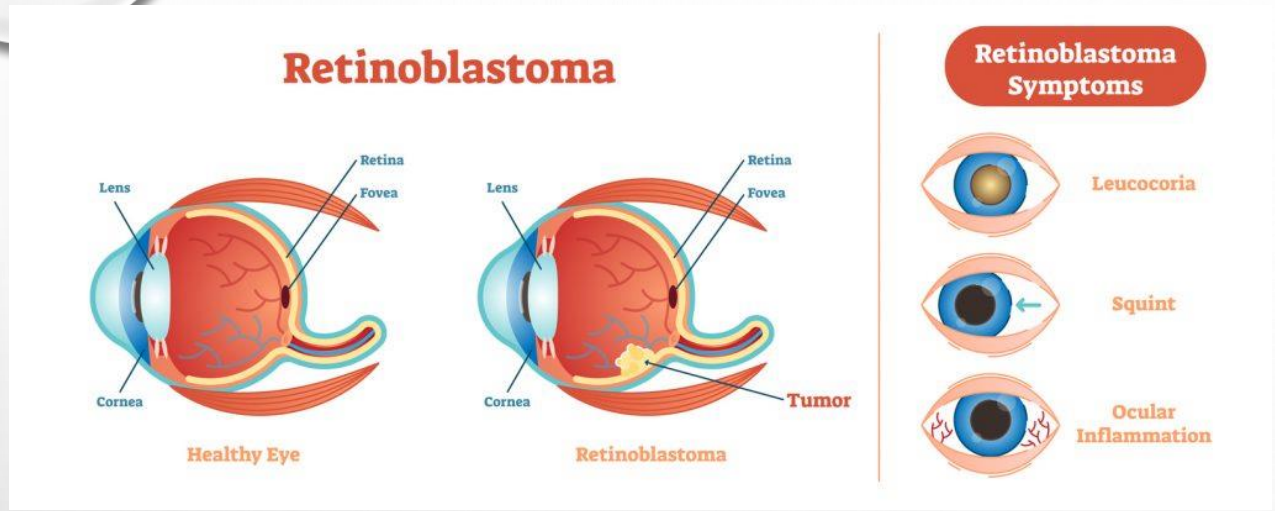
NEOPLASMA PANCA INDERA

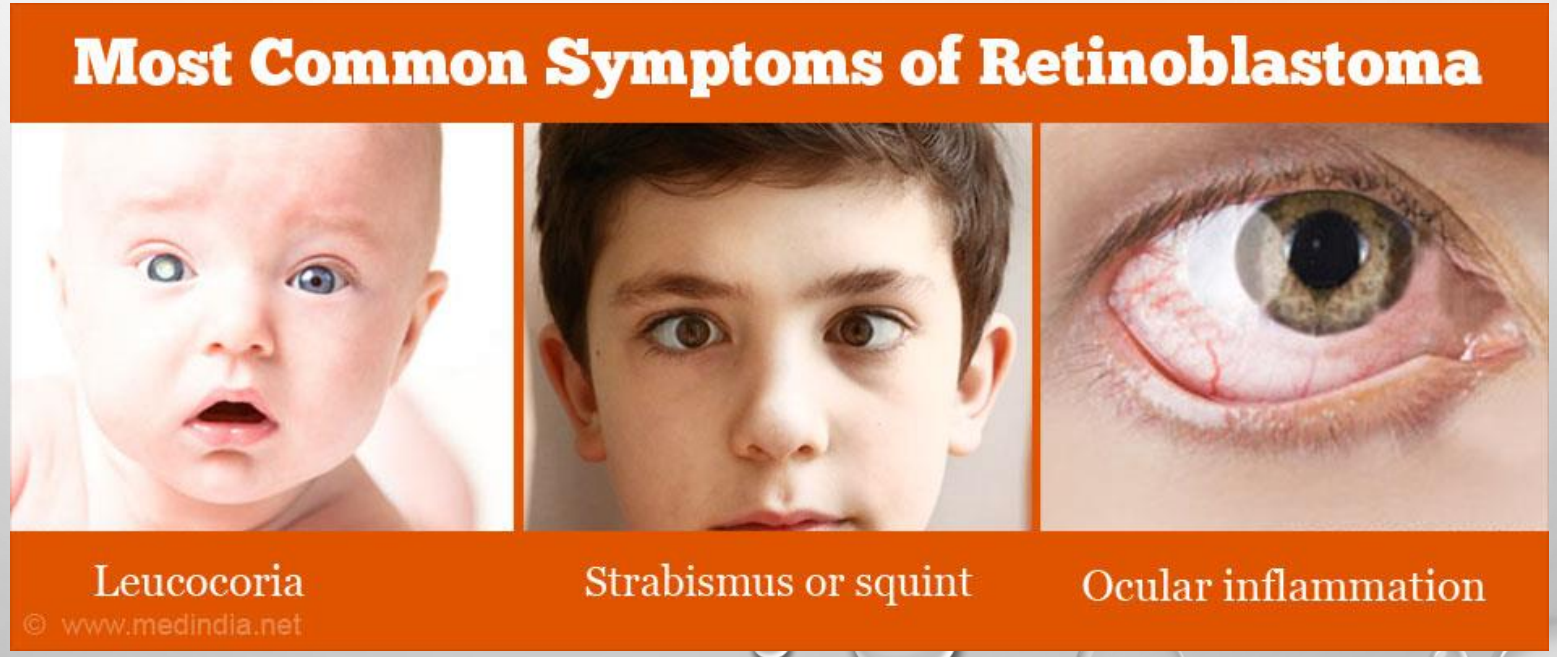
DIAN YULIARTHA LESTARI

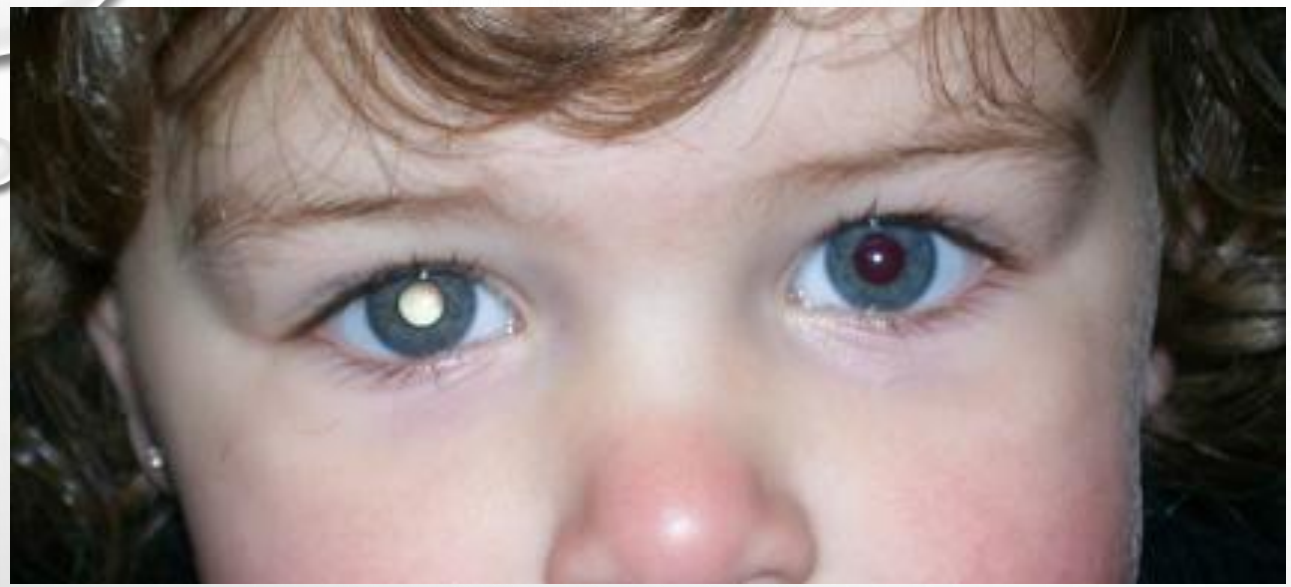
RETINOBLASTOMA

- MOST COMMON INTRAOCULAR TUMOR OF CHILDREN WITH INCIDENCE OF 1 PER 20,000 LIVE BIRTHS
- MAY BE CONGENITAL BUT NOT RECOGNIZED UNTIL AGES 6 MONTHS TO 2 YEARS
- 60% SPORADIC, 40% FAMILIAL (AUTOSOMAL DOMINANT)
- DEVELOPS IN 80 - 90% OF THOSE WITH MUTANT ALLELES IN RETINOBLASTOMA (RB) GENE AT 13Q14
- NEED MUTATIONS IN BOTH ALLELES TO INACTIVATE RB GENE, A NEGATIVE GROWTH REGULATOR
- PATIENTS WITH HEREDITARY RETINOBLASTOMA HAVE A GERMLINE MUTATION IN ONE ALLELE; DEVELOP TUMORS AFTER SOMATIC MUTATION IN SECOND ALLELE ("SECOND HIT"); IN SPORADIC CASES, BOTH ALLELES HAVE SOMATIC MUTATIONS

- BILATERAL IN 30% OF ALL CASES, 90% OF FAMILIAL CASES; SOME PATIENTS WITH BILATERAL TUMORS ALSO HAVE SIMILAR TUMOR OF PINEAL GLAND, TERMED "TRILATERAL" RETINOBLASTOMA, ASSOCIATED WITH POOR PROGNOSIS
- WHITE REFLEX (LEUKOKORIA) PRESENT IN AFFECTED EYE; ALSO RETINAL DETACHMENT
- TENDS TO INVADE OPTIC NERVE (PARTICULARLY LARGE EXOPHYTIC TUMORS WITH SECONDARY GLAUCOMA); CAN INVADE UVEAL TRACT
- DISTANT METASTASES TO CRANIAL VAULT, SKELETAL SYSTEM
- **SECOND PRIMARIES FOR FAMILIAL TUMORS:** 6 - 20% AFTER 10 - 20 YEARS, USUALLY OSTEOSARCOMA (50% OF TUMORS) AND RHABDOMYOSARCOMA, CLOSE TO IRRADIATED FIELDS; ALSO RHABDOID TUMORS


Retinoblastoma
Lens
Retina
Fovea
Cornea
Healthy Eye
Lens
Retina
Fovea
Cornea
Tumor
Retinoblastoma
Retinoblastoma Symptoms
Leucocoria
Squint
Ocular Inflammation


Most Common Symptoms of Retinoblastoma
Leucocoria
Strabismus or squint
Ocular inflammation
© www.medindia.net



- **PROGNOSTIC FACTORS**
  - **5 YEAR SURVIVAL:** 90% IF UNILATERAL, SLIGHTLY LESS IF BILATERAL
  - **POOR PROGNOSTIC FACTORS:**
    - INVASION OF OPTIC NERVE (REPORT AS PRELAMINAR OR RETROLAMINAR INVOLVEMENT, WITH OR WITHOUT RESECTION LINE INVOLVEMENT)
    - INVASION OF UVEAL TRACT OR SCLERA
    - SEEDING OF VITREOUS
    - INVOLVEMENT OF ANTERIOR SEGMENT
    - EXTENSIVE OCULAR TISSUE AND
    - TUMOR NECROSIS IS ASSOCIATED WITH OTHER FACTORS
    - DIFFERENTIATION DOES NOT APPEAR TO HAVE PROGNOSTIC VALUE

- **TREATMENT**
  - **EARLY:** RADIATION THERAPY, CRYOPEXY, XENON ARC PHOTOCOAGULATION
  - **LARGE TUMORS:** ENUCLEATION
  - **INVOLVEMENT OF OPTIC NERVE MARGIN:** RADIATION OF ORBIT AND SYSTEMIC CHEMOTHERAPY
  - **BILATERAL TUMORS:** RADIATION THERAPY TO LESS AFFECTED EYE WITH POSSIBLE CHEMOTHERAPY OR BILATERAL RADIATION
  - **RECURRENCES:** PHOTOCOAGULATION, CRYOTHERAPY OR COBALT DISKS

- **GROSS DESCRIPTION**
  - CREAMY WHITE WITH CHALKY AREAS OF CALCIFICATION AND YELLOW NECROTIC AREAS
  - MAY GROW INWARD (ENDOPHYTIC) OR OUTWARD TOWARD CHOROID (EXOPHYTIC)
  - RARELY ARE DIFFUSELY INFILTRATIVE; TYPICALLY SEEDS INTRAOCULARLY

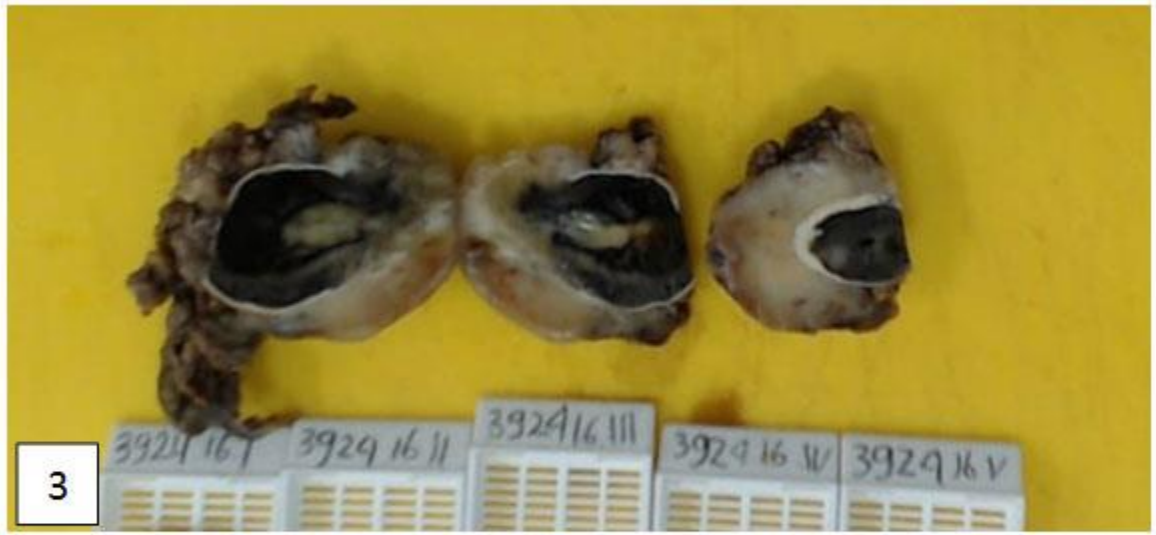


Figure 3: Tumor macroscopy.The tumor covered and infiltrated the eye ball.

- **MICROSCOPIC (HISTOLOGIC) DESCRIPTION**
  - SHEETS, TRABECULAE AND NESTS OF SMALL BLUE CELLS WITH SCANT CYTOPLASM, HYPERCHROMATIC NUCLEI AND SCANTY STROMA
  - FREQUENT NECROSIS OF TUMOR CELLS AWAY FROM VESSELS AND CALCIFICATION
  - ALSO **FLEXNER-WINTERSTEINER ROSETTES** (CELLS LINE UP AROUND EMPTY LUMEN DELINEATED BY A DISTINCT EOSINOPHILIC CIRCLE COMPOSED OF TERMINAL BARS ANALOGOUS TO OUTER LIMITING MEMBRANE OF NORMAL RETINA)
  - ALSO **HOMER-WRIGHT ROSETTES** (NUCLEI ARE DISPLACED AWAY FROM LUMEN),
  - **FLUERETTES** (TUMOR CELLS ARRANGED SIDE BY SIDE WHICH SHOW DIFFERENTIATION TOWARDS PHOTORECEPTORS)
  - FREQUENT **AZZOPARDI PHENOMENA** (BASOPHILIC DEPOSITS AROUND BLOOD VESSELS, ALSO SEEN IN SMALL CELL CARCINOMA); FREQUENT MITOTIC FIGURES; VARIABLE APOPTOTIC CELLS
- **DIFFERENTIATED RETINOBLASTOMA:** BIPOLAR-LIKE CELLS ARE PRESENT
- **UNDIFFERENTIATED RETINOBLASTOMA:** LARGE, ANAPLASTIC CELLS WITHOUT ROSETTE FORMATION
- **RETINOCYTOMA:** MARKED PHOTORECEPTOR DIFFERENTIATION; CELLS HAVE ABUNDANT CYTOPLASM, LESS HYPERCHROMATIC NUCLEI; BENIGN, WITH CALCIFICATION BUT WITHOUT NECROSIS OR MITOTIC ACTIVITY

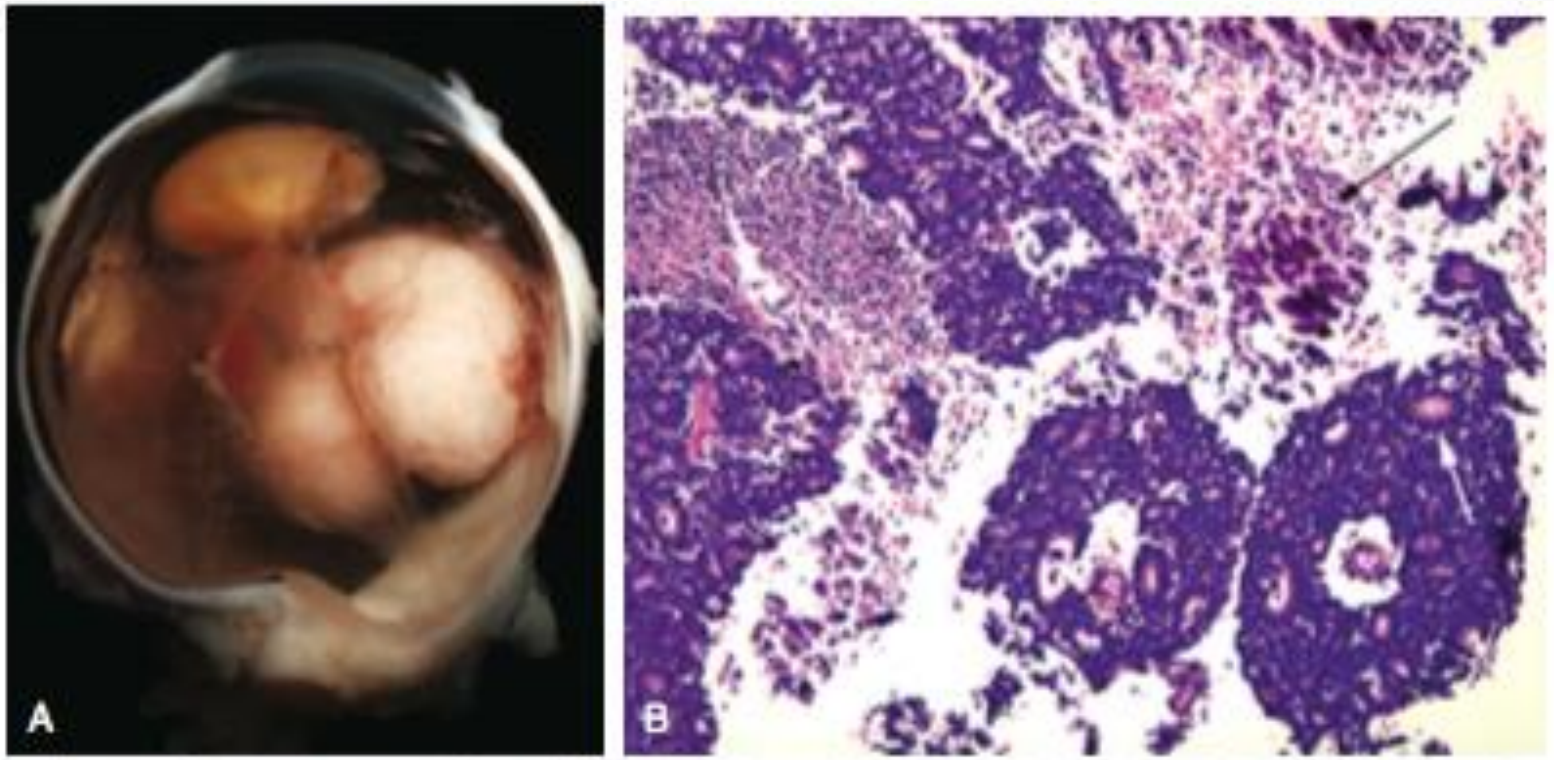
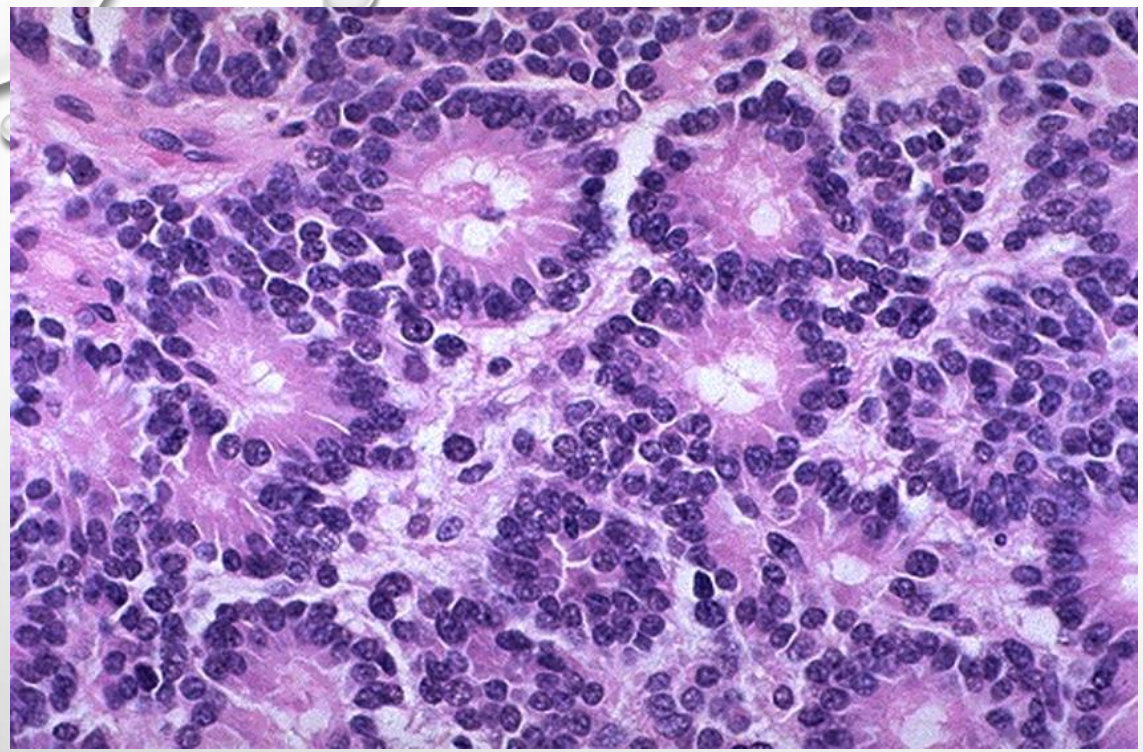
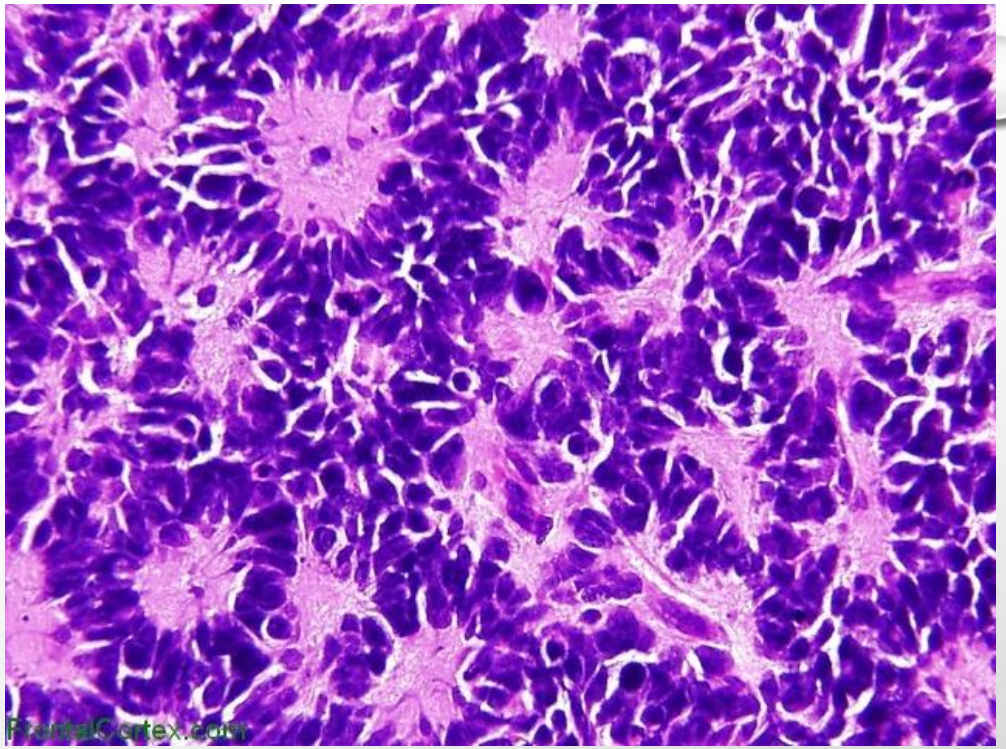


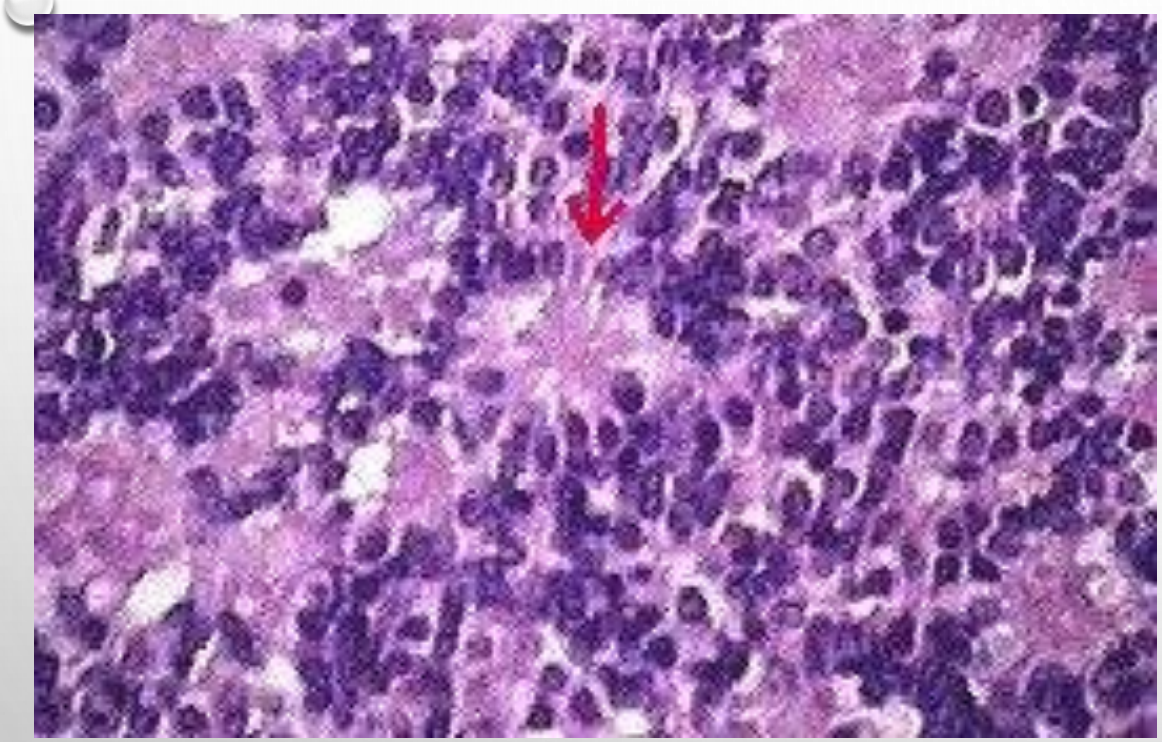
Figure 29-24 Retinoblastoma. **A,** Gross photograph of retinoblastoma. **B,** Tumor cells appear viable when in proximity to blood vessels, but necrosis is seen as the distance from the vessel increases. Dystrophic calcification *(dark arrow)* is present in the zones of tumor necrosis. Flexner-Wintersteiner rosettes—arrangements of a single layer of tumor cells around an apparent "lumen"—are seen throughout the tumor, and one such rosette is indicated by the white arrow.



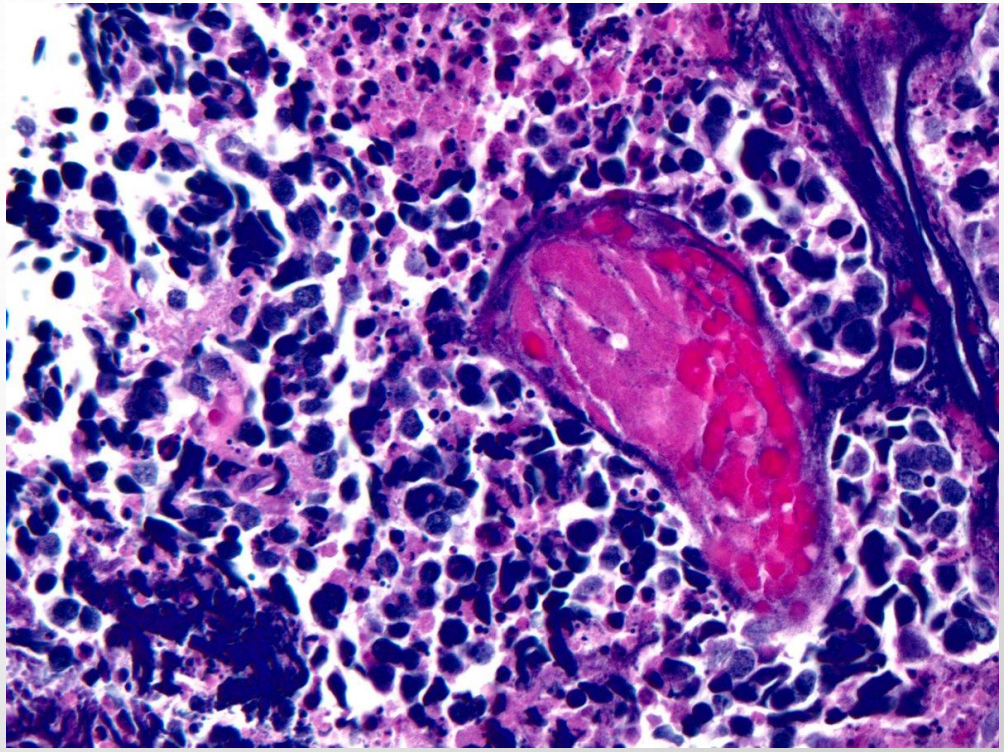
Flexner-Wintersteiner rosettes



Homer-Wright rosettes



fluerettes



Azzopardi phenomena

Pathologic TNM staging of retinoblastoma, AJCC 7th edition

Primary tumor (T)

- **pTX:** primary tumor cannot be assessed
- **pT0:** no evidence of primary tumor
- **pT1:** tumor confined to eye with no optic nerve or choroidal invasion
- **pT2:** tumor with minimal optic nerve or choroidal invasion
- **pT2a:** tumor superficially invades optic nerve head but does not extend past lamina cribrosa OR tumor exhibits focal choroidal invasion, but not both
- **pT2b:** tumor superficially invades optic nerve head but does not extend past lamina cribrosa AND exhibits focal choroidal invasion
- **pT3:** tumor with significant optic nerve or choroidal invasion
- **pT3a:** tumor invades optic nerve past lamina cribrosa but not to surgical resection line OR tumor exhibits massive choroidal invasion, but not both
- **pT3b:** tumor invades optic nerve past lamina cribrosa but not to surgical resection line AND exhibits massive choroidal invasion
- **pT4:** tumor invades optic nerve to resection line OR exhibits extraocular extension elsewhere, but not both
- **pT4a:** tumor invades optic nerve to resection line but no extraocular extension identified
- **pT4b:** tumor invades optic nerve to resection line AND extraocular extension identified

Regional lymph nodes (N)

- **pNX:** regional lymph nodes cannot be assessed
- **pN0:** no regional lymph node involvement
- **pN1:** regional lymph node involvement (preauricular, cervical)
- **N2:** distant lymph node involvement

Metastasis (M)

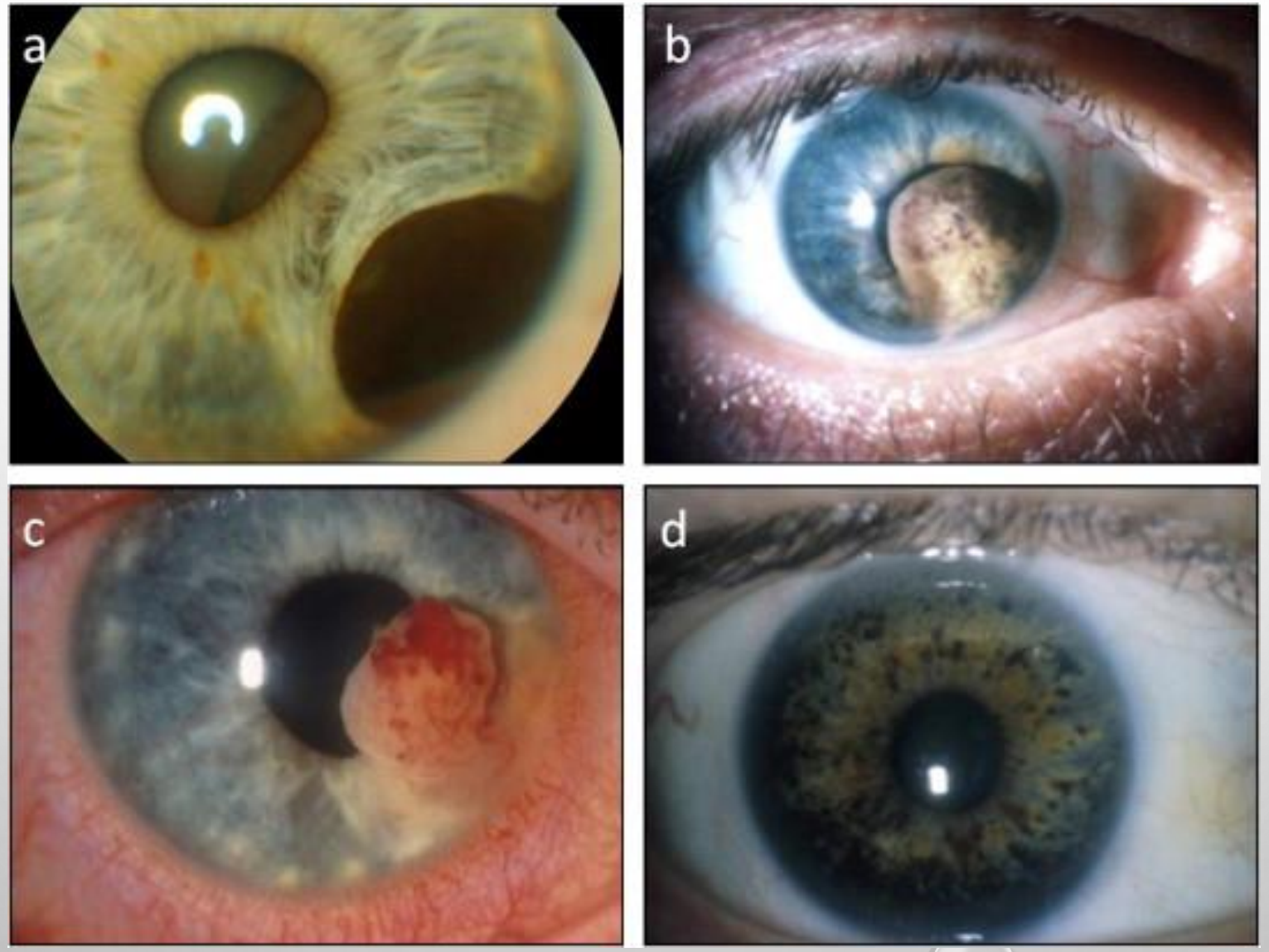
- **cM0:** no metastasis
- **pM1:** metastasis to sites other than CNS
- **pM1a:** single lesion
- **pM1b:** multiple lesions
- **pM1c:** CNS metastasis
- **pM1d:** discrete mass(es) without leptomeningeal or CSF involvement
- **pM1e:** leptomeningeal or CSF involvement

UVEAL MELANOMA

- MALIGNANT NEOPLASM OF MELANOCYTES WITHIN UVEAL TRACT, INCLUDING IRIS, CILIARY BODY AND CHOROID
- TUMOR CENTER LOCATED IN IRIS, CILIARY BODY OR CHOROID
- APPROXIMATELY HALF METASTASIZE, ALMOST EXCLUSIVELY TO LIVER
- TUMOR SIZE, LOCATION AND CELL TYPE ARE THE MOST IMPORTANT HISTOPATHOLOGIC PROGNOSTIC FACTORS BUT MOLECULAR TESTING PLAYS AN INCREASING CLINICAL ROLE IN PROGNOSTICATION

- **EPIDEMIOLOGY**
  - MOST COMMON PRIMARY INTRAOCULAR MALIGNANCY IN ADULTS
  - INCIDENCE IS 5.1 NEW CASES PER MILLION PEOPLE IN UNITED STATES, WHICH IS LOW COMPARED TO OTHER COUNTRIES
  - MEAN AGE AT DIAGNOSIS IS 61 YEARS
  - RISK FACTORS INCLUDE LIGHT SKIN, CAUCASIAN RACE, LIGHT EYE COLOR, INABILITY TO TAN; WEAK ASSOCIATION WITH SUNLIGHT EXPOSURE

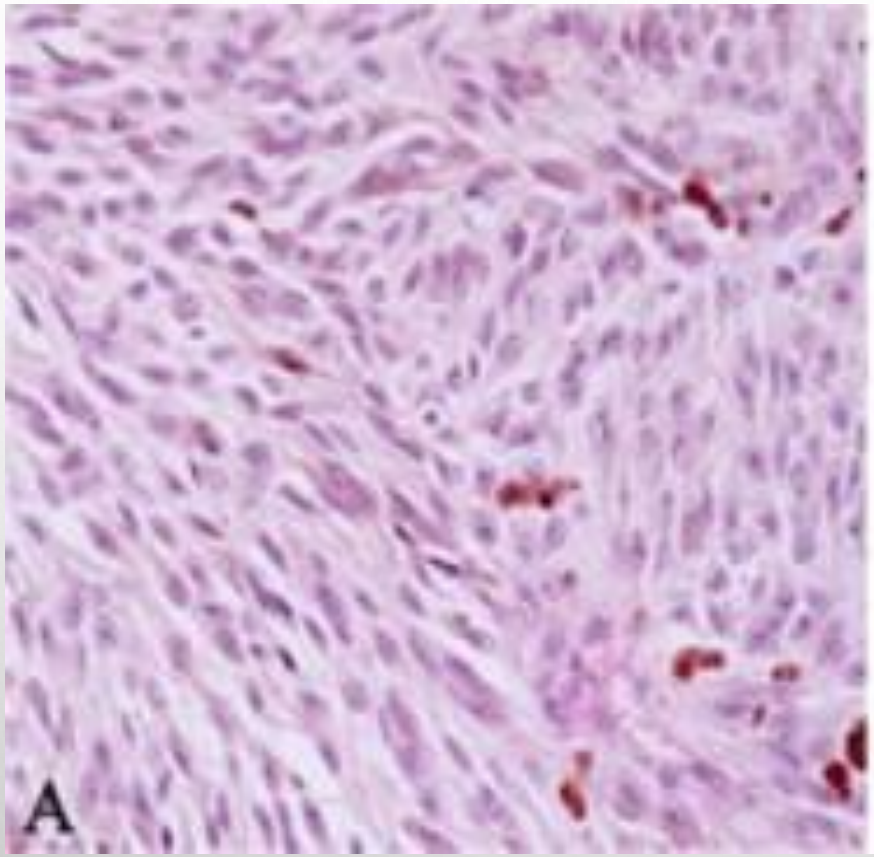
- **CLINICAL FEATURES**
  - HALF OF CASES ARE ASYMPTOMATIC AND DISCOVERED ON EXAM; HALF PRESENT WITH VISUAL DISTURBANCE
  - **MELANOMA OF IRIS:** PRESENTS AS ELEVATED MASS WITH VARIABLE PIGMENTATION, OFTEN WITH DISTORTION OF PUPIL AND PROMINENT VESSELS
  - **MELANOMA OF CHOROID:** IRREGULAR, SLATE GRAY, SOLID, CHOROIDAL TUMOR THAT MAY EXTEND THROUGH BRUCH MEMBRANE INTO RETINA AND VITREOUS PRODUCING RETINAL DETACHMENT, MACULAR EDEMA, CHOROIDAL HEMORRHAGE; OCCASIONALLY SPREADS ALONG SCLERAL CANALS INTO ORBIT, RARELY INVADES OPTIC NERVE
  - **MELANOMA OF CILIARY BODY:** MAY INTERFERE WITH ACCOMMODATION OR CAUSE LOCALIZED CATARACT
  - TENDS TO SPREAD THROUGH SCLERAL EMISSARY CANAL
  - UP TO HALF OF CASES METASTASIZE
  - DISTANT METASTASES NEARLY EXCLUSIVELY TO LIVER (95%), SOMETIMES TO LUNG AND
  - LYMPHATIC SPREAD UNCOMMON DUE TO LACK OF LYMPHATIC CHANNELS IN EYE


a
b
c
d

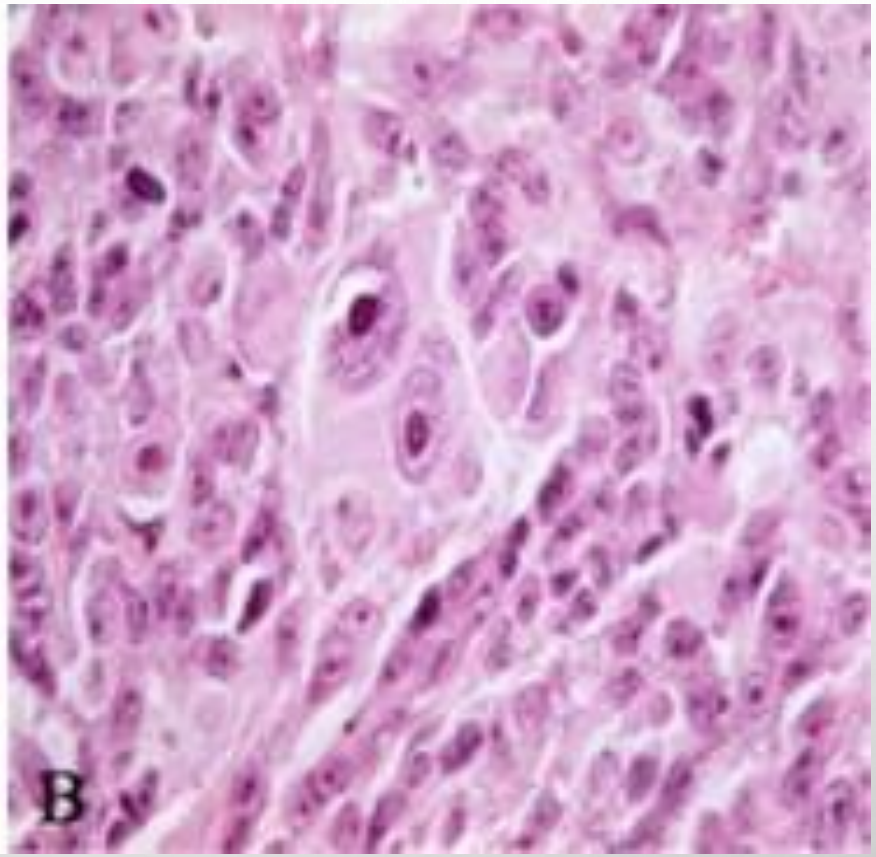
- **GROSS DESCRIPTION**
  - SMALL UVEAL MELANOMAS MAY BE DIFFICULT TO DISTINGUISH FROM A NEVUS CLINICALLY; SIZE CRITERIA ARE USED
  - GROSS FINDINGS SUSPICIOUS FOR MELANOMA INCLUDE:
    - ORANGE PIGMENT
    - SUBRETINAL FLUID
    - TUMOR THICKNESS > 2 MM
    - LOW INTERNAL REFLECTIVITY ON ULTRASOUND EXAMINATION

Gross uveal melanoma

- **MICROSCOPIC (HISTOLOGIC) DESCRIPTION**
  - THERE ARE 2 DISTINCT CELL TYPES AND MOST TUMORS CONTAIN A COMBINATION OF THE 2:
    - **SPINDLE B CELLS:** SPINDLE SHAPED CELLS WITH LARGE OVAL NUCLEI AND DISTINCT NUCLEOLI
    - **EPITHELIOID CELLS:** LARGE POLYGONAL CELLS WITH ATYPICAL NUCLEI, 1 OR MORE PROMINENT NUCLEOLI AND SOMETIMES INTRANUCLEAR PSEUDOINCLUSIONS
  - NOTE THAT SPINDLE A CELLS ALSO EXIST, WHICH ARE SPINDLE SHAPED CELLS WITH SLENDER NUCLEI THAT LACK VISIBLE NUCLEOLI; THESE ARE TYPICALLY BENIGN AND ARE THE MELANOCYTES OF UVEAL NEVI



Spindle cell



Epitheloid

- **PROGNOSTIC FACTORS**
  - SIZE OF TUMOR
  - CILIARY BODY INVOLVEMENT
  - EXTENSION OUTSIDE OF ORBIT, METASTASIS AND RECURRENCE HAVE VERY POOR PROGNOSIS
  - CYTOGENETIC ABNORMALITIES, PARTICULARLY MONOSOMY 3 AND GAIN OF 8Q
  - GENE EXPRESSION PROFILING (CLASS 1A, 1B AND 2)
  - HISTOLOGIC FEATURES: MITOSES, CELL TYPE (SPINDLE B VERSUS EPITHELIOID), EXTRAVASCULAR MATRIX LOOPS, TUMOR INFILTRATING LYMPHOCYTES AND MACROPHAGES

WHO classification of tumours of the nasal cavity, paranasal sinuses and skull base

Carcinomas	
Keratinizing squamous cell carcinoma	8071/3
Non-keratinizing squamous cell carcinoma	8072/3
Spindle cell squamous cell carcinoma	8074/3
Lymphoepithelial carcinoma	8082/3
Sinonasal undifferentiated carcinoma	8020/3
NUT carcinoma	8023/3*
Neuroendocrine carcinomas	
Small cell neuroendocrine carcinoma	8041/3
Large cell neuroendocrine carcinoma	8013/3
Adenocarcinomas	
Intestinal-type adenocarcinoma	8144/3
Non–intestinal-type adenocarcinoma	8140/3
Teratocarcinosarcoma	9081/3
Sinonasal papillomas	
Sinonasal papilloma, inverted type	8121/1
Sinonasal papilloma, oncocytic type	8121/1
Sinonasal papilloma, exophytic type	8121/0
Respiratory epithelial lesions	
Respiratory epithelial adenomatoid hamartoma	
Seromucinous hamartoma	
Salivary gland tumours	
Pleomorphic adenoma	8940/0
Malignant soft tissue tumours	
Fibrosarcoma	8810/3
Undifferentiated pleomorphic sarcoma	8802/3
Leiomyosarcoma	8890/3
Rhabdomyosarcoma, NOS	8900/3
Embryonal rhabdomyosarcoma	8910/3
Alveolar rhabdomyosarcoma	8920/3
Pleomorphic rhabdomyosarcoma, adult type	8901/3
Spindle cell rhabdomyosarcoma	8912/3
Angiosarcoma	9120/3
Malignant peripheral nerve sheath tumour	9540/3
Biphenotypic sinonasal sarcoma	9045/3*
Synovial sarcoma	9040/3

Borderline/low-grade malignant soft tissue tumours	
Desmoid-type fibromatosis	8821/1
Sinonasal glomangiopericytoma	9150/1
Solitary fibrous tumour	8815/1
Epithelioid haemangioendothelioma	9133/3
Benign soft tissue tumours	
Leiomyoma	8890/0
Haemangioma	9120/0
Schwannoma	9560/0
Neurofibroma	9540/0
Other tumours	
Meningioma	9530/0
Sinonasal ameloblastoma	9310/0
Chondromesenchymal hamartoma	
Haematolymphoid tumours	
Extranodal NK/T-cell lymphoma	9719/3
Extraosseous plasmacytoma	9734/3
Neuroectodermal/melanocytic tumours	
Ewing sarcoma/primitive neuroectodermal tumour	9364/3
Olfactory neuroblastoma	9522/3
Mucosal melanoma	8720/3

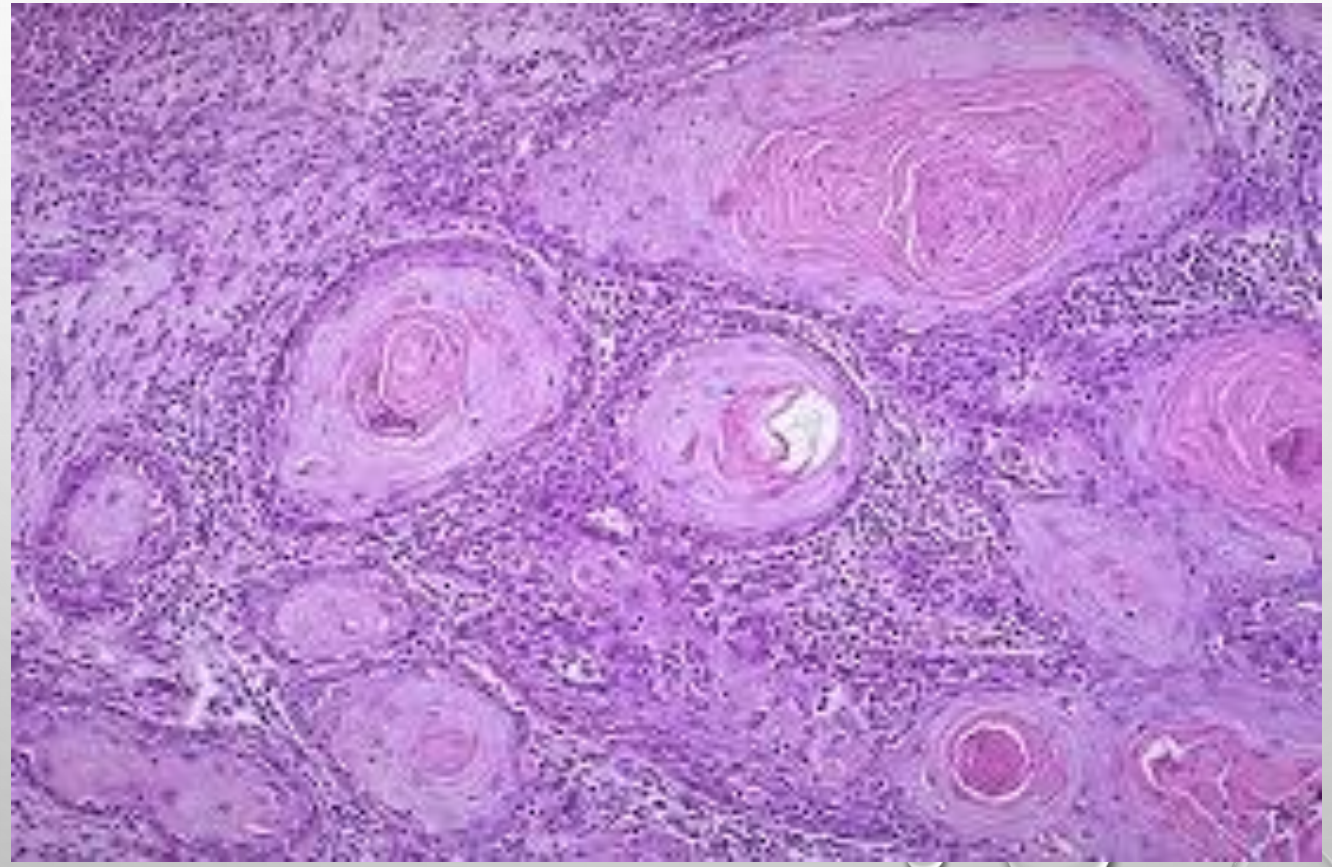
The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) [778A]. Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours. The classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.
*These new codes were approved by the IARC/WHO Committee for ICD-O.

KERATINIZING SCC

- DEFINISI
  - MALIGNANT EPI THELIAL NEOPLASM ARISING FROM THE SURFACE EPITHELIUM LINING THE NASAL CAVITY AND PARANASAL SINUSES AND EXHIBITING SQUA MOUS DIFFERENTIATION.
- SYNONIM : **EPIDERMOID CARCINOMA**
- EPIDEMIOLOGI
  - SINONASAL KSCCS ARE RARE
  - KSCC MOST OFTEN AFFECTS PATIENTS IN THEIR 6^{TH}-7^{TH} DECADES OF LIFE
  - MEN ARE AFFECTED TWICE AS OFTEN AS WOMEN
- ETIOLOGY :
  - CIGARETTE SMOKING INCREASES RISK
  - WOOD DUST, LEATHER DUST, INDUSTRIAL EXPOSURE → KSCC
  - HPV → NON KSCC

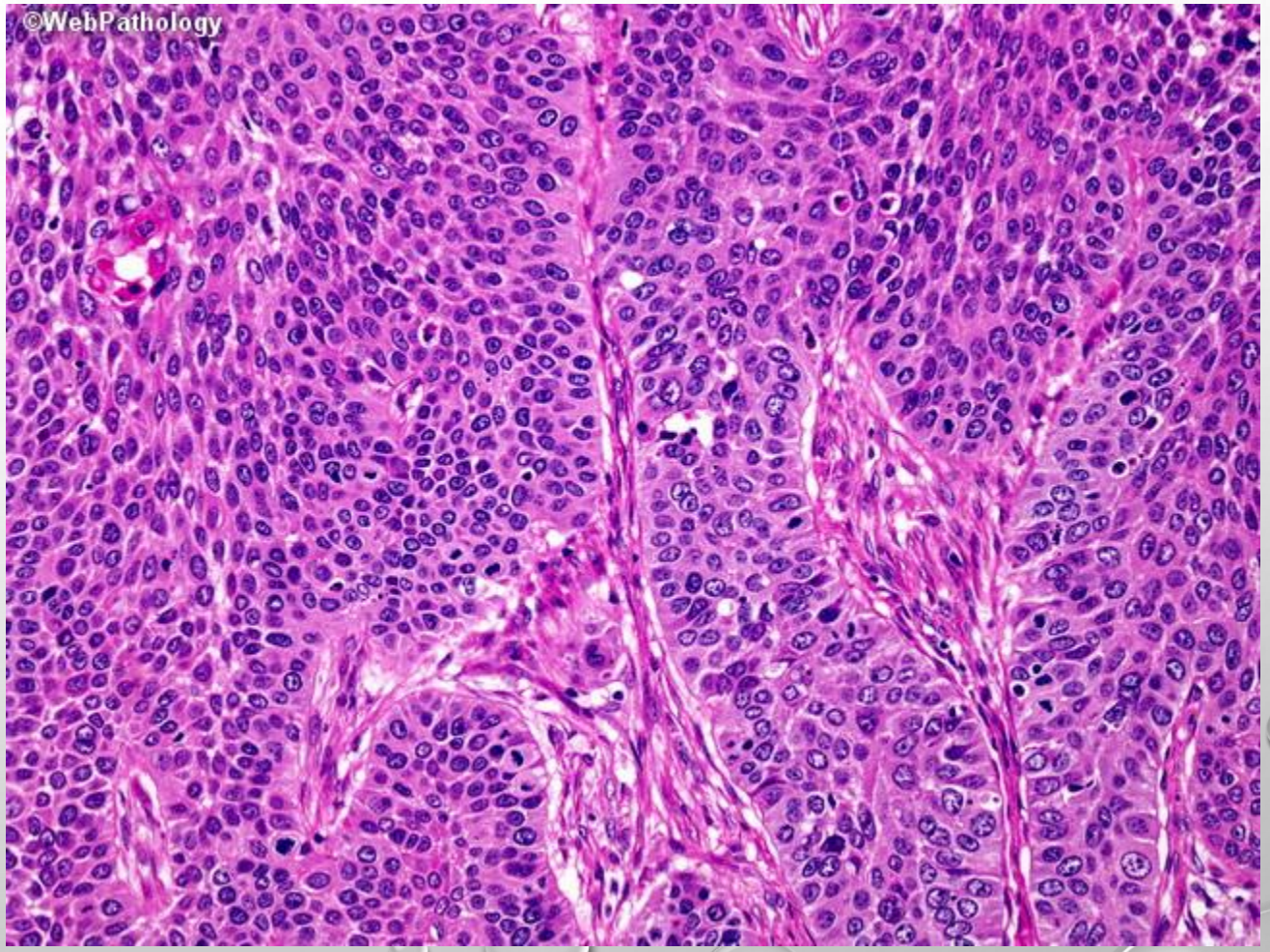
- LOCALIZATION :
  - SINUS MAXILLARIS >>
  - NASAL CAVITY
  - SINUS ETHMOIDALIS
- CLINICAL FEATURES :
  - NASAL OBSTRUCTION
  - EPISTAXIS
  - RHINORRHOEA
  - FACIAL PAIN AND/OR PARALYSIS, DIPLOPIA, AND PROPTOSIS ARE INDICATIVE OF MORE-ADVANCED TUMOUR GROWTH
- MACROSCOPY
  - EXOPHYTIC OR ENDOPHYTIC, WITH VARIOUS DEGREES OF ULCERATION, NE CROSIS, AND HAEMORRHAGE.

- MICROSCOPY
  - CONVENTIONAL SCC
  - GRADING : WELL DIFFERENTIATED, MODERETLY DIFFERENTIATED, POORLY DIFFERENTIATED



NON KERATINIZING SCC

- SYNONIM :
  - SCHNEIDERIAN CARCINOMA
  - TRANSITIONAL CELL CARCINOMA
  - CYLINDRICAL CELL CARCINOMA



TNM classification of carcinomas of the nasal cavity and paranasal sinuses

TNM classification[a,b]

T – Primary tumour

TX Primary tumour cannot be assessed
T0 No evidence of primary tumour
Tis Carcinoma in situ

Maxillary sinus

T1 Tumour limited to the antral mucosa, with no erosion or destruction of bone
T2 Tumour causing bone erosion or destruction, including extension into hard palate and/or middle nasal meatus, except extension to posterior wall of maxillary sinus and pterygoid plates
T3 Tumour invades any of the following: bone of posterior wall of maxillary sinus, subcutaneous tissues, floor or medial wall of orbit, pterygoid fossa, ethmoid sinuses
T4a Tumour invades any of the following: anterior orbital contents, skin of cheek, pterygoid plates, infratemporal fossa, cribriform plate, sphenoid or frontal sinuses
T4b Tumour invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than maxillary division of trigeminal nerve (V2), nasopharynx, clivus

Nasal cavity and ethmoid sinus

T1 Tumour limited to one subsite of nasal cavity or ethmoid sinus, with or without bony invasion
T2 Tumour involves two subsites in a single site or extends to involve an adjacent site within the nasoethmoidal complex, with or without bony invasion
T3 Tumour extends to invade the medial wall or floor of the orbit, maxillary sinus, palate, or cribriform plate
T4a Tumour invades any of the following: anterior orbital contents, skin of nose or cheek, minimal extension to anterior cranial fossa, pterygoid plates, sphenoid or frontal sinuses
T4b Tumour invades any of the following: orbital apex, dura, brain, middle cranial fossa, cranial nerves other than V2, nasopharynx, clivus

N – Regional lymph nodes (i.e. the cervical nodes)

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in a single ipsilateral lymph node, ≤ 3 cm in greatest dimension
N2 Metastasis as specified in N2a, N2b, or N2c below
N2a Metastasis in a single ipsilateral lymph node, > 3 cm but ≤ 6 cm in greatest dimension
N2b Metastasis in multiple ipsilateral lymph nodes, all ≤ 6 cm in greatest dimension
N2c Metastasis in bilateral or contralateral lymph nodes, all ≤ 6 cm in greatest dimension
N3 Metastasis in a lymph node > 6 cm in greatest dimension

Note: Midline nodes are considered ipsilateral nodes.

M – Distant metastasis

M0 No distant metastasis
M1 Distant metastasis

Stage grouping

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T1–2	N1	M0
	T3	N0–1	M0
Stage IVA	T1–3	N2	M0
	T4a	N0–2	M0
Stage IVB	T4b	Any N	M0
	Any T	N3	M0
Stage IVC	Any T	Any N	M1

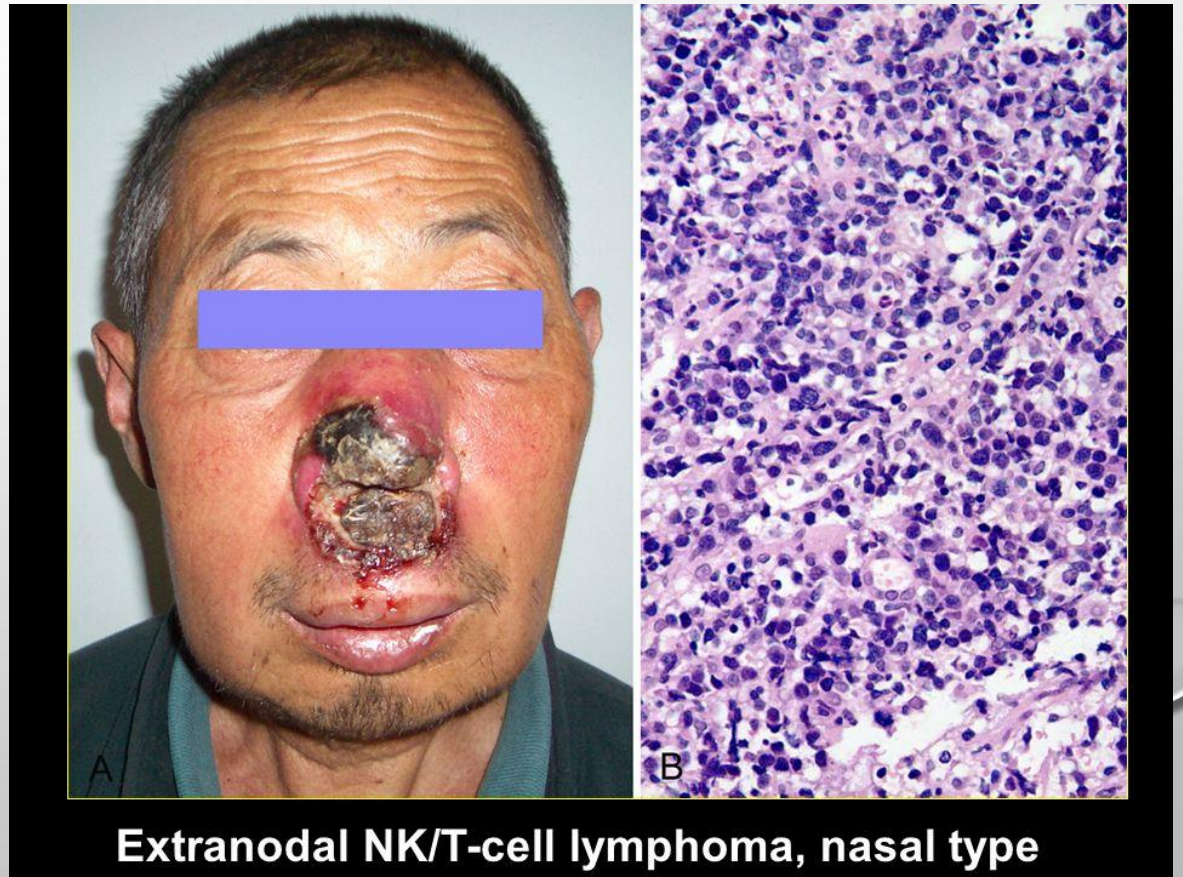
[a]Adapted from Edge et al. [625A] – used with permission of the American Joint Committee on Cancer (AJCC), Chicago, Illinois; the original and primary source for this information is the AJCC Cancer Staging Manual, Seventh Edition (2010) published by Springer Science+Business Media – and Sobin et al. [2228A].

[b]A help desk for specific questions about TNM classification is available at http://www.uicc.org/resources/tnm/helpdesk.

MIDLINE GRANULOMA

- EXTRANODAL NK/T CELL LYMPHOMA
- **EPIDEMIOLOGY**
  - RARE; MORE PREVALENT IN ASIANS AND NATIVE AMERICAN POPULATIONS OF MEXICO, CENTRAL AMERICA AND SOUTH AMERICA
  - ADULTS, OFTEN CHINESE (IN US, OF ASIAN/HISPANIC DESCENT
  - MORE COMMON IN MALES THAN FEMALES
- ETIOLOGY → EBV
- **SITES**
  - MOST COMMON LOCATION IS UPPER AERODIGESTIVE TRACT (NASAL CAVITY, NASOPHARYNX, PARANASAL SINUSES, PALATE)

- **CLINICAL FEATURES**
  - DESTRUCTIVE SINONASAL OR MIDLINE FACIAL TUMORS, MAY DISSEMINATE RAPIDLY, SOMETIMES ASSOCIATED WITH HEMOPHAGOCYTIC SYNDROMES, EBV+
  - SKINNODULAR, ULCERATED LESIONS; INTESTINEPERFORATION; OTHER SITESMASS LESIONS
  - IF SIGNIFICANT BONE MARROW AND PERIPHERAL BLOOD INVOLVEMENT, MAY OVERLAP WITH AGGRESSIVE NK-CELL LEUKEMIA



SINONASAL PAPILOMA

- SINONASAL PAPILLOMA IS A BENIGN EPITHELIAL NEOPLASM OF SINONASAL TRACT
- WHO HAS DIVIDED SINONASAL PAPILLOMA INTO 3 DISTINCT TYPES
  - INVERTED PAPILLOMA
  - EXOPHYTIC PAPILLOMA
  - ONCOCYTIC PAPILLOMA
- **TERMINOLOGY**
  - ALL TYPES: SCHNEIDERIAN PAPILLOMA AND EPITHELIAL PAPILLOMA
  - EXOPHYTIC TYPE: TRANSITIONAL CELL PAPILLOMA, FUNGIFORM PAPILLOMA, RINGERTZ PAPILLOMA AND SEPTAL PAPILLOMA
  - ONCOCYTIC TYPE: CYLINDRICAL CELL PAPILLOMA AND COLUMNAR CELL PAPILLOMA

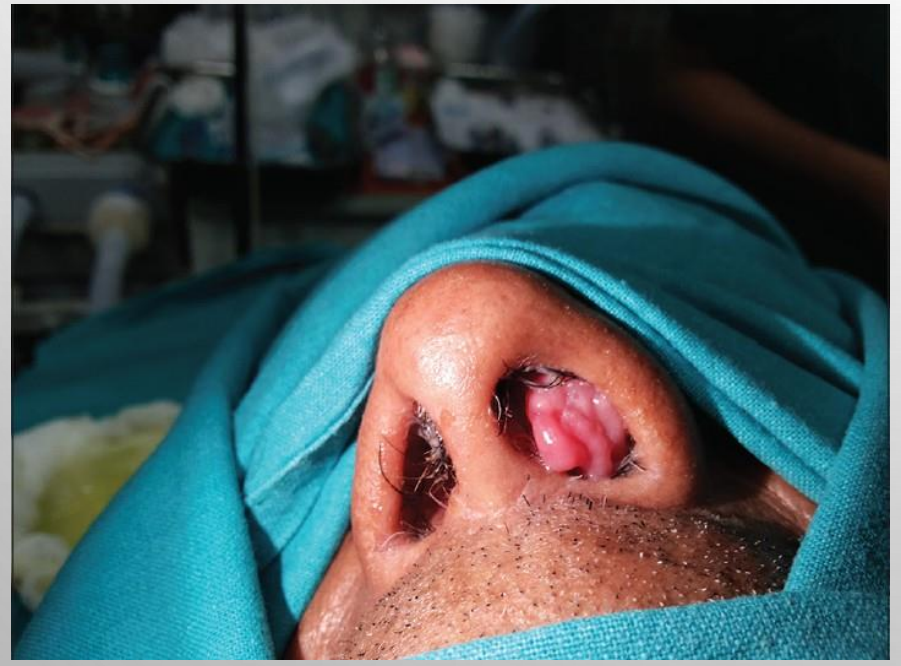
Comparison of essential features of the 3 types of sinonasal papilloma

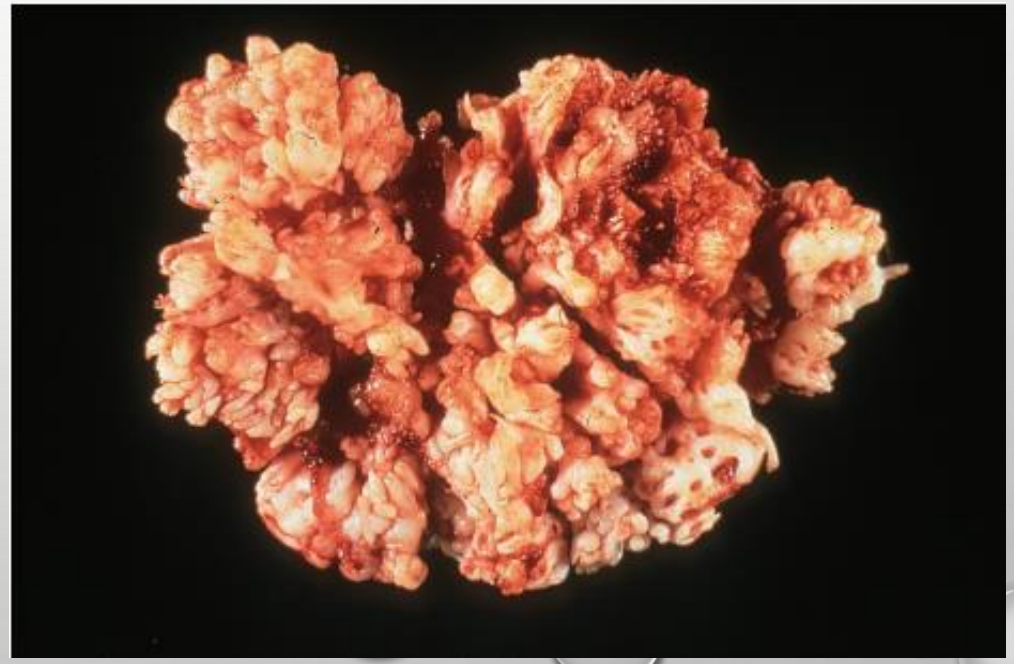
	Inverted papilloma	Exophytic papilloma	Oncocytic papilloma
Frequency	Most common	Second most common	Least common
Location	Lateral nasal wall / paranasal sinus	Nasal septum	Lateral nasal wall / paranasal sinus
Male to female ratio	2 - 3:1	10:1	1:1
Most common age of presentation	5th to 6th decades	3rd to 5th decades	5th to 6th decades
Association with human papillomavirus (HPV)	High risk HPV Low risk HPV	Low risk HPV	No association
Architectural pattern	Endophytic (inverted)	Exophytic (filiform)	Exophytic or endophytic
Epithelial lining	Squamous, transitional or respiratory	Squamous, transitional or respiratory	Oncocytic
Molecular alterations	*EGFR* activating mutation	None reported	*KRAS* mutation
Risk of malignant transformation	5 - 15%	~0%	4 - 17%

- **EPIDEMIOLOGY**
  - ANNUAL INCIDENCE IS 0.74 - 2.3 PER 100,000 POPULATION
  - INVERTED PAPILLOMA IS THE MOST COMMON SUBTYPE, FOLLOWED BY EXOPHYTIC PAPILLOMA; ONCOCYTIC PAPILLOMA IS THE LEAST COMMON
  - INVERTED AND ONCOCYTIC PAPILLOMA MOST COMMONLY AFFECT PATIENTS IN THEIR 5^{TH} TO 6^{TH} DECADES
  - EXOPHYTIC PAPILLOMA OCCURS IN 3^{RD} TO 5^{TH} DECADES
  - INVERTED AND EXOPHYTIC PAPILLOMA OCCUR MORE FREQUENTLY IN MALES, WITH A MALE TO FEMALE RATIO OF 2 - 3:1 AND 10:1
  - ONCOCYTIC PAPILLOMA AFFECTS BOTH GENDERS EQUALLY
- **CLINICAL FEATURES**
  - SYMPTOMS ARE USUALLY NONSPECIFIC : NASAL CONGESTION, NASAL OBSTRUCTION, NASAL DISCHARGE OR EPISTAXIS

- **SITES**
  - SINONASAL PAPILLOMA COMMONLY AFFECTS NASAL CAVITY OR PARANASAL SINUSES
  - EXOPHYTIC PAPILLOMA → THE NASAL SEPTUM
  - INVERTED AND ONCOCYTIC TYPES → LATERAL NASAL WALL OR PARANASAL SINUSES
  - INVERTED PAPILLOMA MAY SECONDARILY EXTEND TO NONSINONASAL SITES, E.G. PHARYNX, EAR, CRANIAL CAVITY
  - SINONASAL PAPILLOMA IS USUALLY UNILATERAL; BILATERAL INVOLVEMENT IS RARE
- **ETIOLOGY**
  - ROLE OF HIGH RISK HUMAN PAPILLOMAVIRUS (HPV) IN INVERTED PAPILLOMA REMAINS CONTROVERSIAL
  - REPORTED RATE OF HIGH RISK HPV IN INVERTED PAPILLOMA AND CARCINOMA EX INVERTED PAPILLOMA RANGES FROM 0 – 100%
  - LOW RISK HPV, ESPECIALLY TYPES 6 AND 11, HAS ALSO BEEN DETECTED IN INVERTED PAPILLOMA AND IS MORE COMMON THAN HIGH RISK HPV
  - OTHER ETIOLOGIC FACTORS IMPLICATED IN INVERTED PAPILLOMA INCLUDE EXPOSURE TO WELDING, ORGANIC SOLVENTS AND SMOKING
  - EXOPHYTIC PAPILLOMA MAY BE RELATED TO LOW RISK HPV, ESPECIALLY TYPES 6 AND 11
  - NO SIGNIFICANT ASSOCIATION BETWEEN ONCOCYTIC PAPILLOMA AND HPV

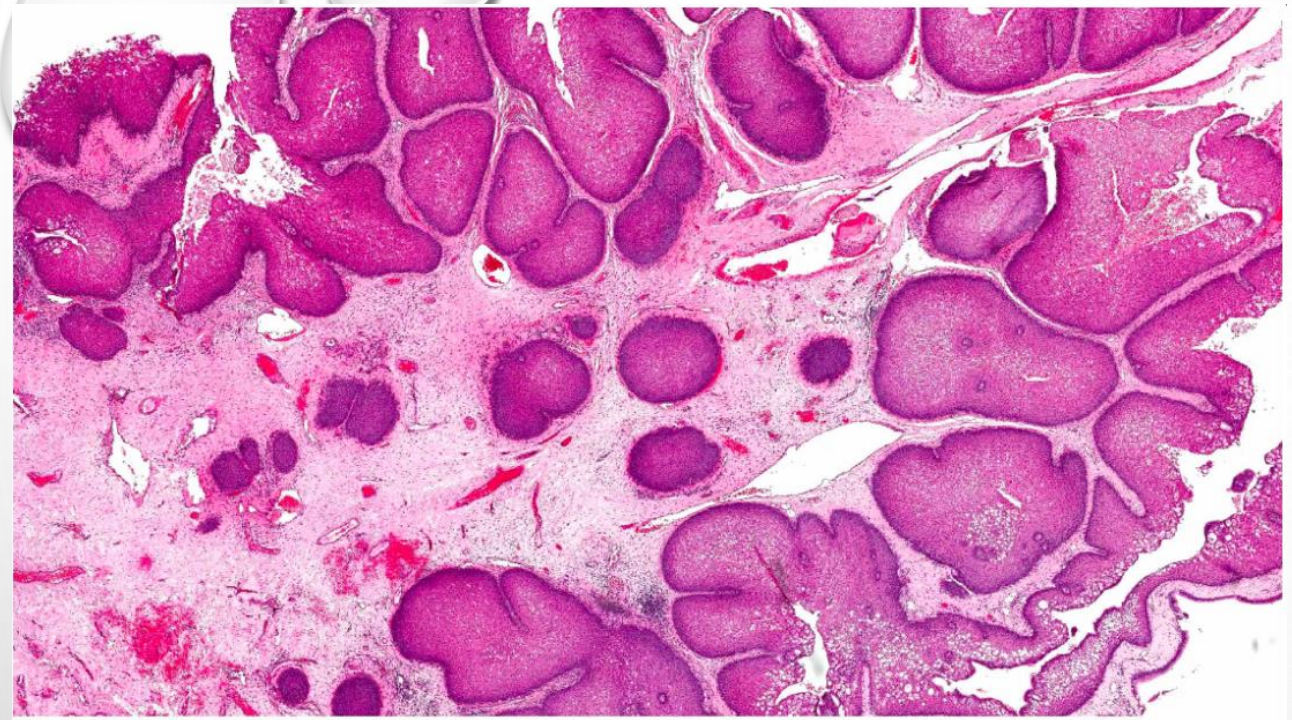
- **GROSS DESCRIPTION**
  - INVERTED PAPILLOMA: PINK-TAN-GRAY, SOFT TO MODERATELY FIRM POLYPOID GROWTH WITH CONVOLUTED OR WRINKLED SURFACE
  - EXOPHYTIC PAPILLOMA: USUALLY MUSHROOM SHAPED AND EXOPHYTIC WITH PAPILLARY APPEARANCE
  - ONCOCYTIC PAPILLOMA: FLESHY POLYPOID GROWTH OF VARIABLE COLOR
  - GENEROUS OR COMPLETE SAMPLING IS ADVISED TO SEARCH FOR FOCUS OF MALIGNANT TRANSFORMATION



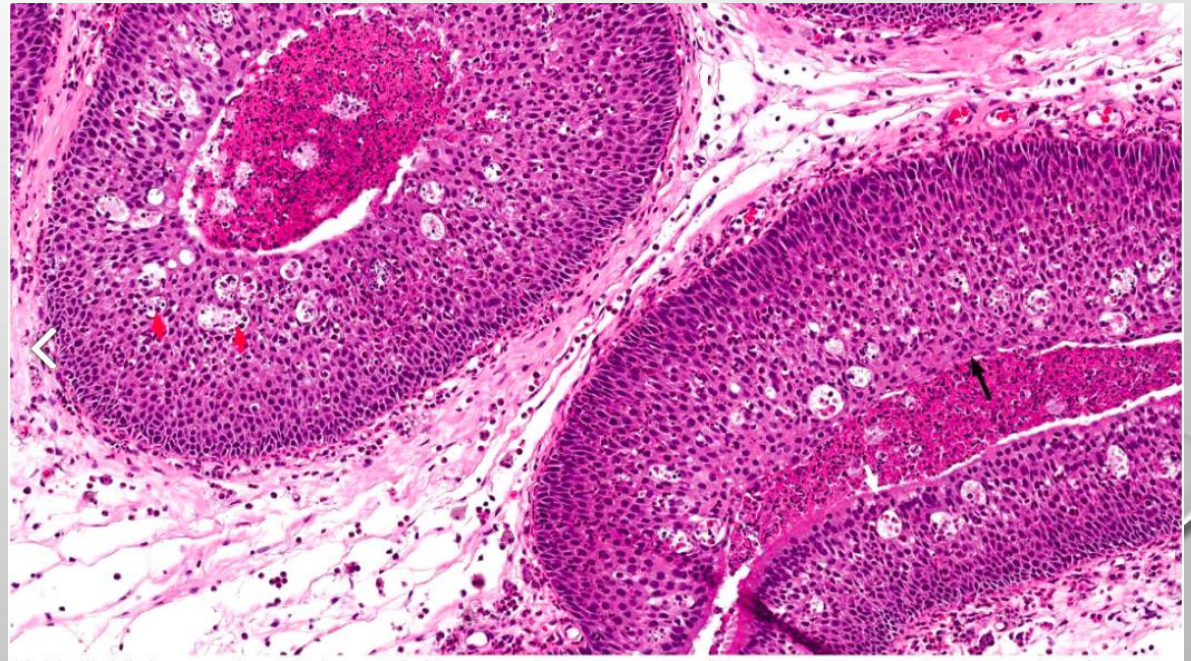


MICROSCOPIC (HISTOLOGIC) DESCRIPTION

- **INVERTED PAPILLOMA:**
  - ARCHITECTURE: PROMINENT DOWNWARD ENDOPHYTIC GROWTH OF ROUND TO ELONGATED INTERCONNECTED EPITHELIAL NESTS WITH SMOOTH OUTER CONTOUR
  - EPITHELIUM IS HYPERPLASTIC (5 - 30 CELL LAYERS IN THICKNESS) AND MAY BE OF SQUAMOUS, TRANSITIONAL OR RESPIRATORY TYPE
  - TRANSMIGRATING NEUTROPHILS AND NEUTROPHILIC MICROABSCESSES MAY BE SEEN
  - STROMA MAY HAVE EDEMA OR CHRONIC INFLAMMATION
  - SEROMUCINOUS GLAND IN THE LAMINA PROPRIA IS COMMONLY DECREASED OR ABSENT

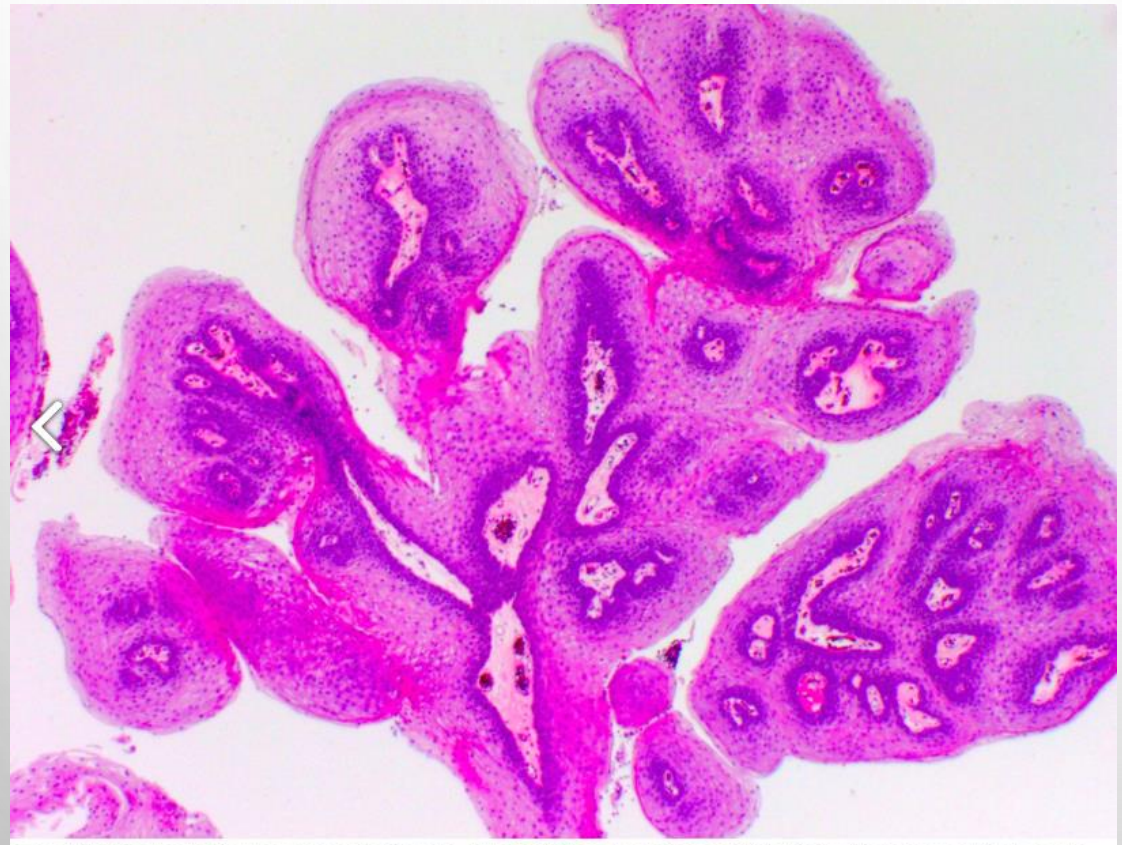


Inverted papilloma is characterized by endophytic growth of epithelial nests with smooth outer contour

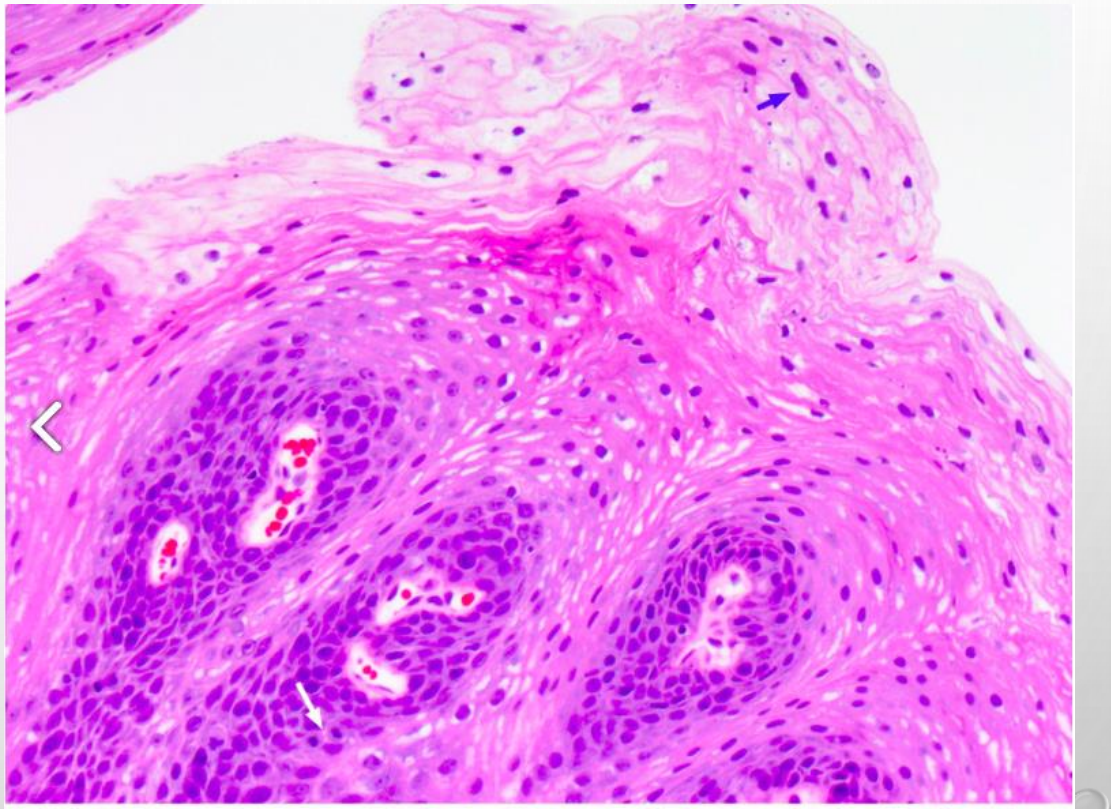


Epithelial lining can be respiratory (white arrow) or squamous (black arrow); note the transmigrating neutrophils and microabscesses (red arrows) in the epithelium

- **EXOPHYTIC PAPILLOMA:**
  - ARCHITECTURE: FILIFORM OR PAPILLARY ARRANGEMENT WITH DELICATE FIBROVASCULAR CORE
  - EPITHELIAL LINING CAN BE EITHER SQUAMOUS, RESPIRATORY OR TRANSITIONAL; IT MAY CONTAIN MUCUS SECRETING CELLS AND GOBLET CELLS
  - VARIABLE KOILOCYTOSIS MAY BE PRESENT
  - USUALLY NO / SCANT SURFACE KERATINIZATION
  - MITOTIC FIGURES ARE ABSENT OR LIMITED TO THE BASAL LAYER
  - MINIMAL INFLAMMATORY CELLS

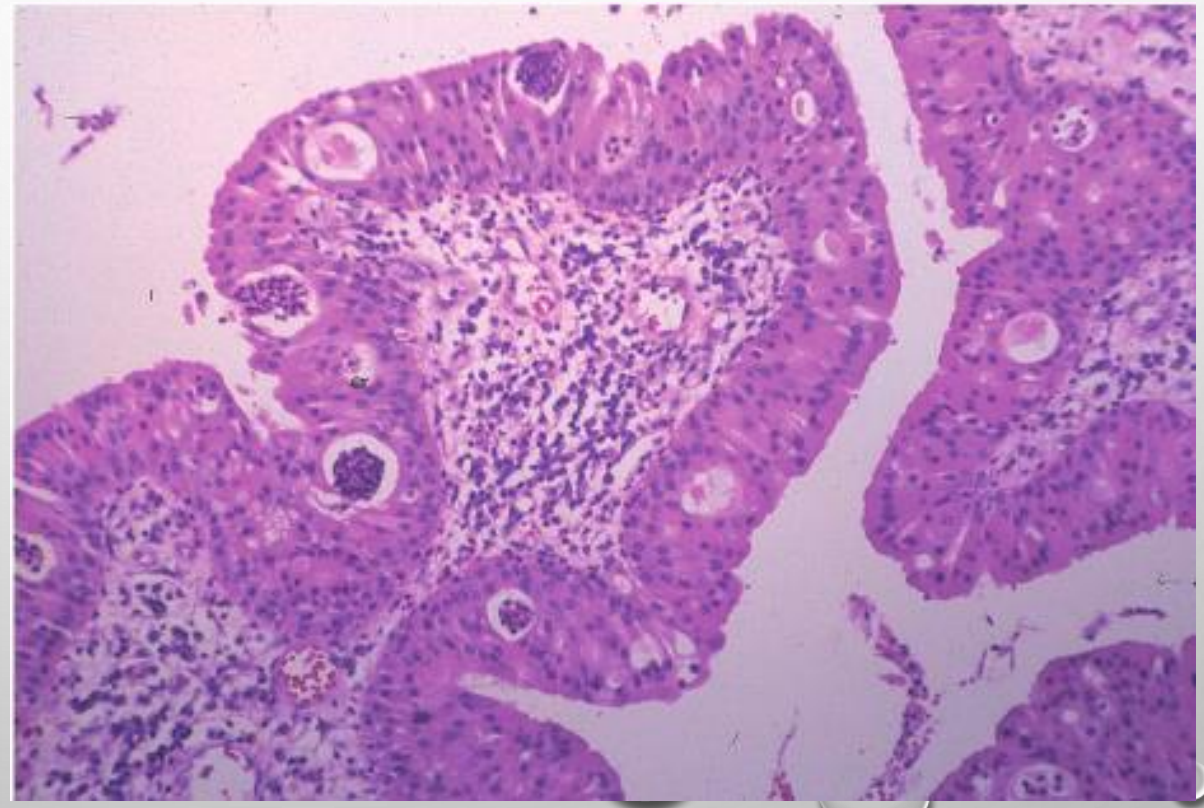


Exophytic papilloma has delicate branching papillae with thin fibrovascular core



It is lined by squamous epithelium without significant atypia; focal koilocytic change (blue arrow) and rare basal mitotic figures (white arrow) may be seen

- **ONCOCYTIC PAPILLOMA:**
  - ARCHITECTURE: MAY HAVE ENDOPHYTIC (INVERTED) OR EXOPHYTIC GROWTH PATTERNS
  - EPITHELIAL LINING IS PSEUDOSTRATIFIED AND COLUMNAR WITH ABUNDANT EOSINOPHILIC GRANULAR CYTOPLASM AND HYPERCHROMATIC UNIFORM NUCLEI
  - INTRAEPITHELIAL MUCIN FILLED CYSTS WITH NEUTROPHILIC MICROABSCESSES MAY BE SEEN



WHO classification of tumours of the nasopharynx

Carcinomas	
Nasopharyngeal carcinoma	
Non-keratinizing squamous cell carcinoma	8072/3
Keratinizing squamous cell carcinoma	8071/3
Basaloid squamous cell carcinoma	8083/3
Nasopharyngeal papillary adenocarcinoma	8260/3
Salivary gland tumours	
Adenoid cystic carcinoma	8200/3
Salivary gland anlage tumour	
Benign and borderline lesions	
Hairy polyp	
Ectopic pituitary adenoma	8272/0
Craniopharyngioma	9350/1
Soft tissue tumours	
Nasopharyngeal angiofibroma	9160/0
Haematolymphoid tumours	
Diffuse large B-cell lymphoma	9680/3
Extraosseous plasmacytoma	9734/3
Extramedullary myeloid sarcoma	9930/3
Notochordal tumours	
Chordoma	9370/3

The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) [776A]. Behaviour is coded /0 for benign tumours, /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours. The classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.

TNM classification of carcinomas of the nasopharynx

TNM classification[a,b]

T – Primary tumour

TX	Primary tumour cannot be assessed
T0	No evidence of primary tumour
Tis	Carcinoma in situ
T1	Tumour confined to nasopharynx, or extends to oropharynx and/or nasal cavity
T2	Tumour with parapharyngeal extension (which denotes posterolateral infiltration of tumour)
T3	Tumour invades bony structures of skull base and/or paranasal sinuses
T4	Tumour with intracranial extension and/or involvement of cranial nerves, infratemporal fossa, hypopharynx, orbit, or masticator space

N – Regional lymph nodes (i.e. the cervical nodes)

NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Unilateral metastasis in cervical lymph node(s), and/or unilateral or bilateral metastasis in retropharyngeal lymph nodes, ≤ 6 cm in greatest dimension, above the supraclavicular fossa
N2	Bilateral metastasis in cervical lymph node(s), ≤ 6 cm in greatest dimension, above the supraclavicular fossa
N3	Metastasis in cervical lymph node(s), > 6 cm and/or in the supraclavicular fossa
N3a	> 6 cm in greatest dimension
N3b	in the supraclavicular fossa[c]

Note: Midline nodes are considered ipsilateral nodes.

M – Distant metastasis

M0	No distant metastasis
M1	Distant metastasis

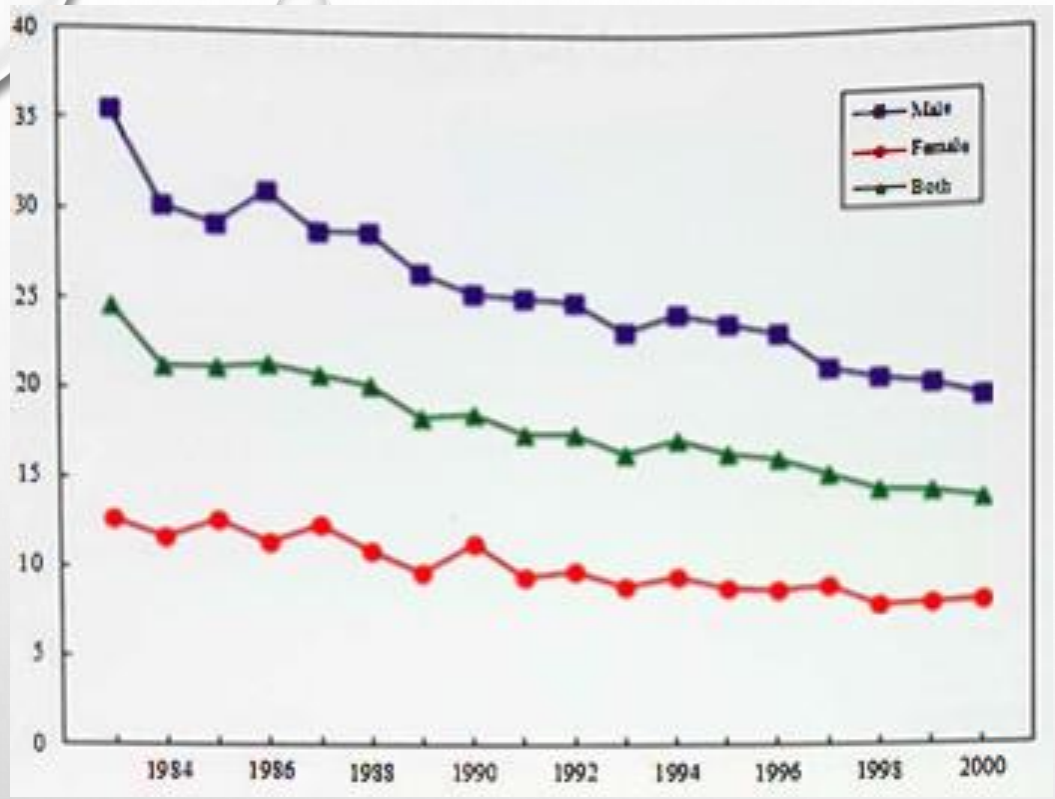
Stage grouping

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T1	N1	M0
	T2	N0–1	M0
Stage III	T1–2	N2	M0
	T3	N0–2	M0
Stage IVA	T4	N0–2	M0
Stage IVB	Any T	N3	M0
Stage IVC	Any T	Any N	M1

[a]Adapted from Edge et al. [625A] – used with permission of the American Joint Committee on Cancer (AJCC), Chicago, Illinois; the original and primary source for the information is the AJCC Cancer Staging Manual, Seventh Edition (2010) published by Springer Science+Business Media – and Sobin et al. [2228A].
[b]A help desk for specific questions about TNM classification is available at http://www.uicc.org/resources/tnm/helpdesk.
[c]The supraclavicular fossa is the triangular region defined by three points: (1) the superior margin of the sternal end of the clavicle, (2) the superior margin of the lateral end of the clavicle, and (3) the point where the neck meets the shoulder; this includes caudal portions of levels IV and V.

NASOPHARYNGEAL CA

- SYNONYMS
  - LYMPHOEPITHELIAL CARCINOMA
  - UNDIFFERENTIATED CARCINOMA WITH LYMPHOID STROMA
  - SQUAMOUS CELL CARCINOMA (WHO GRADE 1)
  - NON-KERATINIZING CARCINOMA (WHO GRADE 2)
  - UNDIFFERENTIATED CARCINOMA (WHO GRADE 3)
- DEMOGRAPHICS VARY GREATLY BY REGION
  - UNITED STATES: RARE, INCIDENCE OF 0.4 PER 100,000 IN WHITES
  - AFRICA: COMMON; #1 CHILDHOOD CANCER; ASSOCIATED WITH EBV
  - SOUTH CHINA: MOST COMMON CANCER IN ADULTS (18% OF CANCERS IN HONG KONG, 21.4 PER 100,000 IN HONG KONG,, RARE IN CHILDREN
- 70% MALE
- STRONGLY ASSOCIATED WITH EBV INFECTION FOR UNDIFFERENTIATED AND NONKERATINIZING SUBTYPES
- OTHER RISK FACTORS: CONSUMPTION OF SALT PRESERVED FISH CONTAINING CARCINOGENIC NITROSAMINES, FAMILY HISTORY, SPECIFIC HLA CLASS I GENOTYPES, TOBACCO SMOKING, CHRONIC RESPIRATORY TRACT CONDITIONS AND LOW CONSUMPTION OF FRESH FRUITS AND VEGETABLES



Insidens berdasar usia

Table 2.01 Structures involved by local infiltration of nasopharyngeal carcinoma; MRI data of 308 patients, Pamela Youde Nethersole Eastern Hospital, Hong Kong.

Structures involved	Frequency
Adjacent soft tissues	
Nasal cavity	87%
Oropharyngeal wall, soft palate	21%
Parapharyngeal space, carotid space	68%
Pterygoid muscle (medial, lateral)	48%
Prevertebral muscle	19%
Bony erosion / paranasal sinus	
Nasal septum	3%
Pterygoid plate(s), pterygomaxillary fissure, pterygopalatine fossa	27%
Maxillary antrum	4%
Ethmoid sinus	6%
Sphenoid sinus; sphenoid bone; foramina lacerum, ovale, and rotundum	38%
Clivus	41%
Petrous bone, petro-occipital fissure	19%
Jugular foramen, hypoglossal canal	4%
Pituitary fossa/gland	3%
Extensive/intracranial extension	
Cavernous sinus	16%
Cerebrum, meninges, cisterns	4%
Infratemporal fossa	9%
Orbit, orbital fissure(s)	4%
Hypopharynx	2%

- DOTEC :
  - DIPLOPIA
  - OTOREA
  - TINITUS
  - EPISTAXIS
  - CEPHALGIA

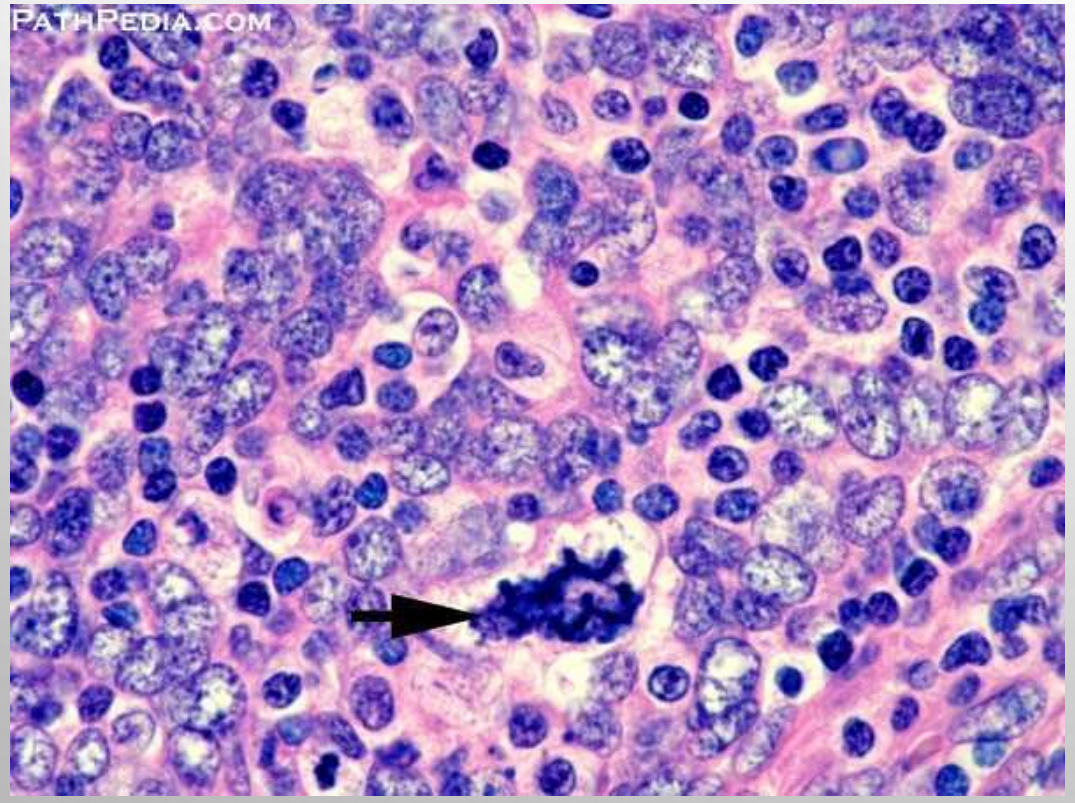


Table 2.02 Common presenting symptoms and signs of nasopharyngeal carcinoma; data from 722 consecutive patients treated at Pamela Youde Nethersole Eastern Hospital, Hong Kong SAR, China, in 1994–2001.

Presenting features	Frequency
Symptoms	
Neck mass	42%
Nasal (postnasal drip, discharge, bleeding, obstruction)	46%
Aural (tinnitus, discharge, earache, deafness)	42%
Headache	16%
Double vision, squint, blindness	6%
Facial numbness	5%
Speech/swallowing problem	2%
Weight loss	4%
Physical signs	
Enlarged neck node(s)	72%
Bilateral neck nodes	35%
Neck nodes extending to supraclavicular fossa	12%
Cranial nerve palsy	10%
Deafness	3%
Dermatomyositis	1%

WHO classification of tumours of the hypopharynx, larynx, trachea and parapharyngeal space

Malignant surface epithelial tumours	
Conventional squamous cell carcinoma	8070/3
Verrucous squamous cell carcinoma	8051/3
Basaloid squamous cell carcinoma	8083/3
Papillary squamous cell carcinoma	8052/3
Spindle cell squamous cell carcinoma	8074/3
Adenosquamous carcinoma	8560/3
Lymphoepithelial carcinoma	8082/3
Precursor lesions	
Dysplasia, low grade	8077/0
Dysplasia, high grade	8077/2
Squamous cell papilloma	8052/0
Squamous cell papillomatosis	8060/0
Neuroendocrine tumours	
Well-differentiated neuroendocrine carcinoma	8240/3
Moderately differentiated neuroendocrine carcinoma	8249/3
Poorly differentiated neuroendocrine carcinoma	
Small cell neuroendocrine carcinoma	8041/3
Large cell neuroendocrine carcinoma	8013/3
Salivary gland tumours	
Adenoid cystic carcinoma	8200/3
Pleomorphic adenoma	8940/0
Oncocytic papillary cystadenoma	8290/0
Soft tissue tumours	
Granular cell tumour	9580/0
Liposarcoma	8850/3
Inflammatory myofibroblastic tumour	8825/1
Cartilage tumours	
Chondroma	9220/0
Chondrosarcoma	9220/3
Chondrosarcoma, grade 1	9222/1
Chondrosarcoma, grade 2/3	9220/3
Haematolymphoid tumours	

The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) [776A]. Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours. The classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.

TNM classification of carcinomas of the larynx

TNM classification*,†

T – Primary tumour
- TX Primary tumour cannot be assessed
- T0 No evidence of primary tumour
- Tis Carcinoma in situ

Supraglottis
- T1 Tumour limited to one subsite of supraglottis, with normal vocal cord mobility
- T2 Tumour invades mucosa of more than one adjacent subsite of supraglottis or glottis or region outside the supraglottis (e.g. mucosa of base of tongue, vallecula, or medial wall of pyriform sinus), without fixation of the larynx
- T3 Tumour limited to larynx with vocal cord fixation and/or invades any of the following: postcricoid area, pre-epiglottic space, paraglottic space, inner cortex of thyroid cartilage
- T4a Tumour invades through the thyroid cartilage and/or invades tissues beyond the larynx; for example, trachea, soft tissues of neck including deep/extrinsic muscle of tongue (genioglossus, hyoglossus, palatoglossus, and styloglossus), strap muscles, thyroid, oesophagus
- T4b Tumour invades prevertebral space or mediastinal structures, or encases carotid artery

Glottis
- T1 Tumour limited to vocal cord(s) (may involve anterior or posterior commissure), with normal vocal cord mobility
  - T1a Tumour limited to one vocal cord
  - T1b Tumour involves both vocal cords
- T2 Tumour extends to supraglottis and/or subglottis, and/or with impaired vocal cord mobility
- T3 Tumour limited to larynx with vocal cord fixation and/or invades paraglottic space and/or inner cortex of the thyroid cartilage
- T4a Tumour invades through the outer cortex of the thyroid cartilage and/or invades tissues beyond the larynx; for example, trachea, soft tissues of neck including deep/extrinsic muscle of tongue (genioglossus, hyoglossus, palatoglossus, and styloglossus), strap muscles, thyroid, oesophagus
- T4b Tumour invades prevertebral space or mediastinal structures, or encases carotid artery

Subglottis
- T1 Tumour limited to subglottis
- T2 Tumour extends to vocal cord(s), with normal or impaired mobility
- T3 Tumour limited to larynx, with vocal cord fixation
- T4a Tumour invades cricoid or thyroid cartilage and/or invades tissues beyond the larynx; for example, trachea, soft tissues of neck including deep/extrinsic muscle of tongue (genioglossus, hyoglossus, palatoglossus, and styloglossus), strap muscles, thyroid, oesophagus
- T4b Tumour invades prevertebral space or mediastinal structures, or encases carotid artery

N – Regional lymph nodes (i.e. the cervical nodes)
- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Metastasis in a single ipsilateral lymph node, ≤ 3 cm in greatest dimension
- N2 Metastasis as specified in N2a, N2b, or N2c below
- N2a Metastasis in a single ipsilateral lymph node, > 3 cm but ≤ 6 cm in greatest dimension
- N2b Metastasis in multiple ipsilateral lymph nodes, all ≤ 6 cm in greatest dimension
- N2c Metastasis in bilateral or contralateral lymph nodes, all ≤ 6 cm in greatest dimension
- N3 Metastasis in a lymph node > 6 cm in greatest dimension

Note: Midline nodes are considered ipsilateral nodes.

M – Distant metastasis
- M0 No distant metastasis
- M1 Distant metastasis

Stage grouping

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T1–2	N1	M0
	T3	N0–1	M0
Stage IVA	T1–3	N2	M0
	T4a	N0–2	M0
Stage IVB	T4b	Any N	M0
	Any T	N3	M0
Stage IVC	Any T	Any N	M1

*Adapted from Edge et al. [625A] – used with permission of the American Joint Committee on Cancer (AJCC), Chicago, Illinois; the original and primary source for this information is the AJCC Cancer Staging Manual, Seventh Edition (2010) published by Springer Science+Business Media – and Sobin et al. [2228A].
†A help desk for specific questions about TNM classification is available at http://www.uicc.org/resources/tnm/helpdesk.

CONVENTIONAL SCC

- EPIDEMIOLOGY :
  - SCC OF THE LARYNX AND HYPOPHARYNX IS THE 2ND MOST COMMON RESPIRATORY TRACT CANCER, AFTER LUNG CANCER
  - IT ACCOUNTS FOR 1 .6-2% OF ALL MALIGNANT TUMOURS IN MEN AND 0.2-0.4% IN WOMEN
  - IT OCCURS MOST FREQUENTLY IN THE 6TH AND 7TH DECADES OF LIFE.
- ETIOLOGY :
  - CIGARETTE SMOOKING
  - ALCOHOL CONSUMPTION
- LOCALIZATION :
  - SUPRAGLOTTIS >>

CLINICAL FEATURES

- HOARSENESS (WITH GLOTTIC AND SUPRAGLOTTIC SCC)
- DYSPNOEA AND STRIDOR (WITH SUBGLOTTIC SCC).
- DYSPHAGIA
- CHANGE IN THE QUALITY OF VOICE
- SENSATION OF A FOREIGN BODY IN THE THROAT
- HAEMOPTYSIS
- ODINOPHAGIA
- WHEEZING OR STRIDOR (TRACHEAL)
- ACUTE RESPIRATORY FAILURE (TRACHEAL)
- COUGH (TRACHEAL)
- MASS ON THE NECK (HYPOPHARING)

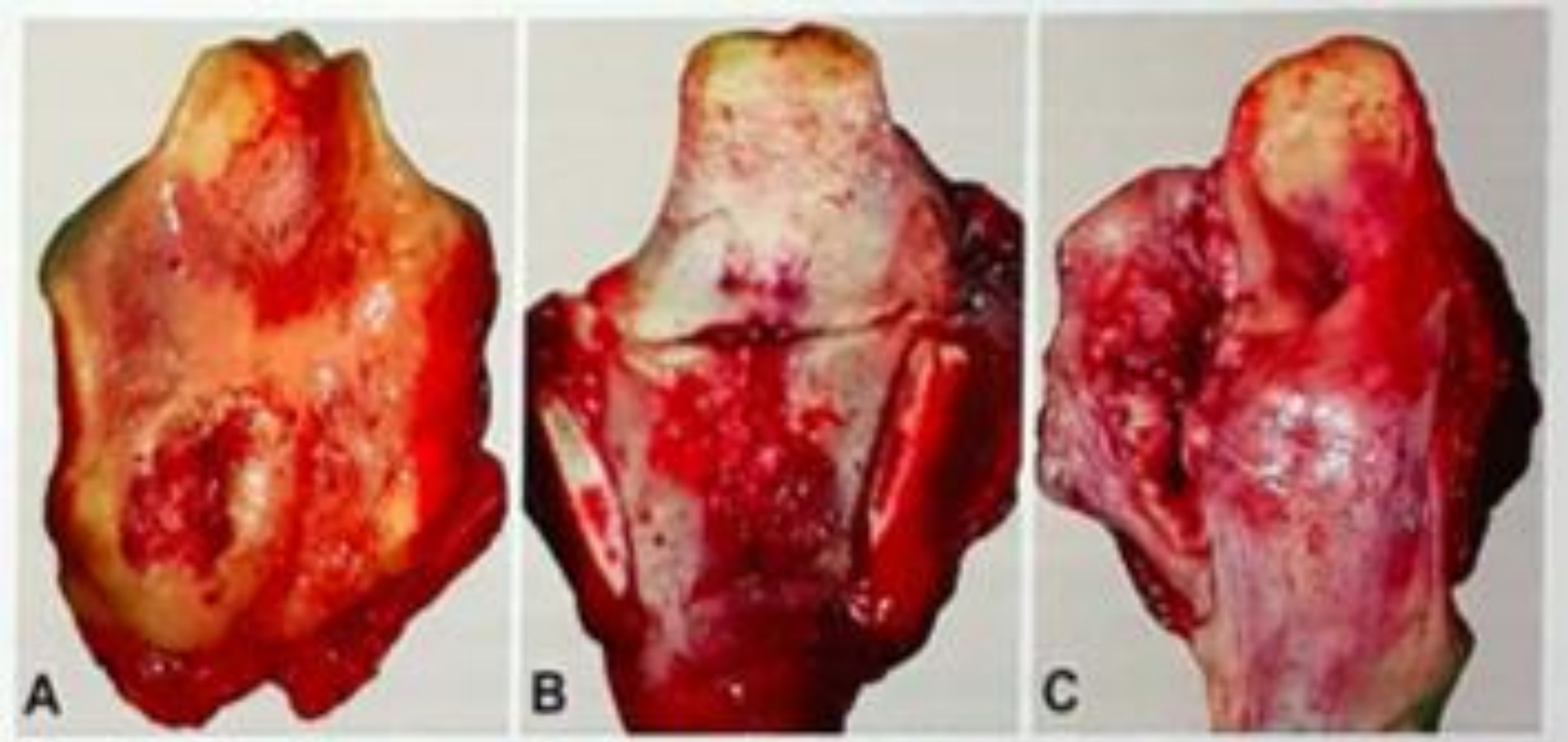


Fig. 3.01 Macroscopic appearance of conventional squamous cell carcinoma. A Supraglottic carcinoma of the larynx: an ulcerated tumour with raised edges at the base of the epiglottis. B Subglottic carcinoma of the larynx: a partially flat and partially exophytic nodular tumour of the subglottis, extending to the anterior commissure. C Hypopharyngeal carcinoma of the piriform sinus: a large, ulcerated tumour with raised edges in the piriform sinus, extending to the aryepiglottic fold.

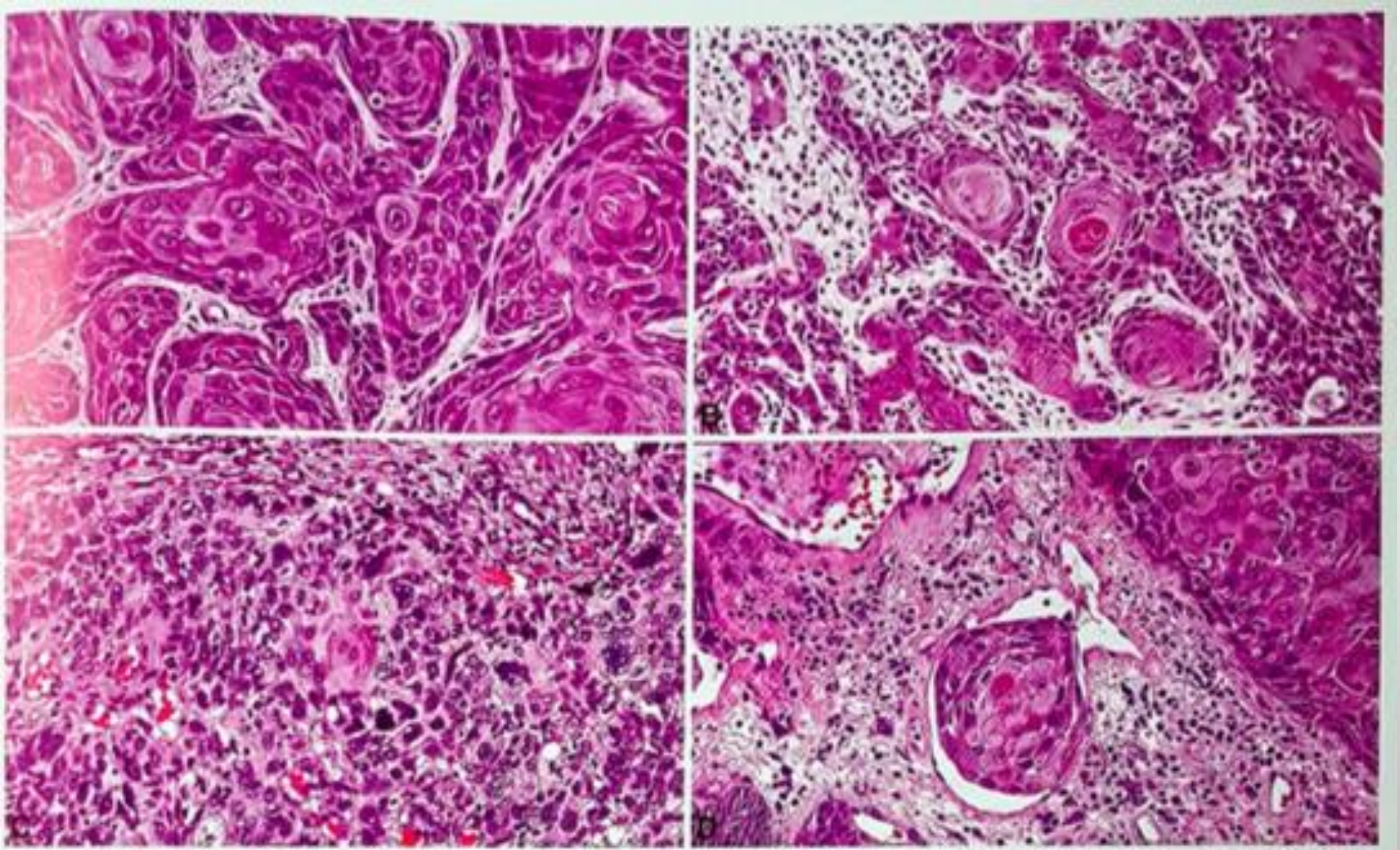


Fig. 3.02 Conventional squamous cell carcinoma. **A** Well-differentiated squamous cell carcinoma: islands of tumour cells with clearly visible squamous differentiation and mild nuclear and cellular pleomorphism. **B** Moderately differentiated squamous cell carcinoma: islands and cords of tumour cells with evident squamous differentiation and moderate nuclear and cellular pleomorphism. **C** Poorly differentiated squamous cell carcinoma: solid growth of tumour cells with marked nuclear and cellular pleomorphism, high mitotic rate, and barely discernible squamous differentiation. **D** Lymphatic invasion by squamous cell carcinoma. A tumour island within a thin-walled lymphatic vessel.

WHO classification of tumours of the ear

Tumours of the external auditory canal	
Squamous cell carcinoma	8070/3
Adenocarcinoma	8420/3
Ceruminous adenocarcinoma	8420/3
Adenoid cystic carcinoma	8200/3
Mucoepidermoid carcinoma	8430/3
Ceruminous adenoma	8420/0
Tumours of the middle and inner ear	
Squamous cell carcinoma	8070/3
Aggressive papillary tumour	8260/1
Endolymphatic sac tumour	8140/3
Otosclerosis	
Cholesteatoma	
Vestibular schwannoma	9560/0
Meningioma	9530/0
Middle ear adenoma	8140/0

The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) [776A]. Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours. The classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.

- EPIDEMIOLOGY :
  - 1 CASE PER 1 MILLION POPULATION
  - 55-65 YEARS OLD
  - PREDOMINANTLY FEMALE
- ETIOLOGY :
  - CHRONIC INFLAMMATION
  - RADIATION
- CLINICAL FEATURES :
  - OTITIS MEDIA AND EXTERNA PAIN
  - HEARING CHANGE
  - CHOLESTEATOME
  - STENOSIS

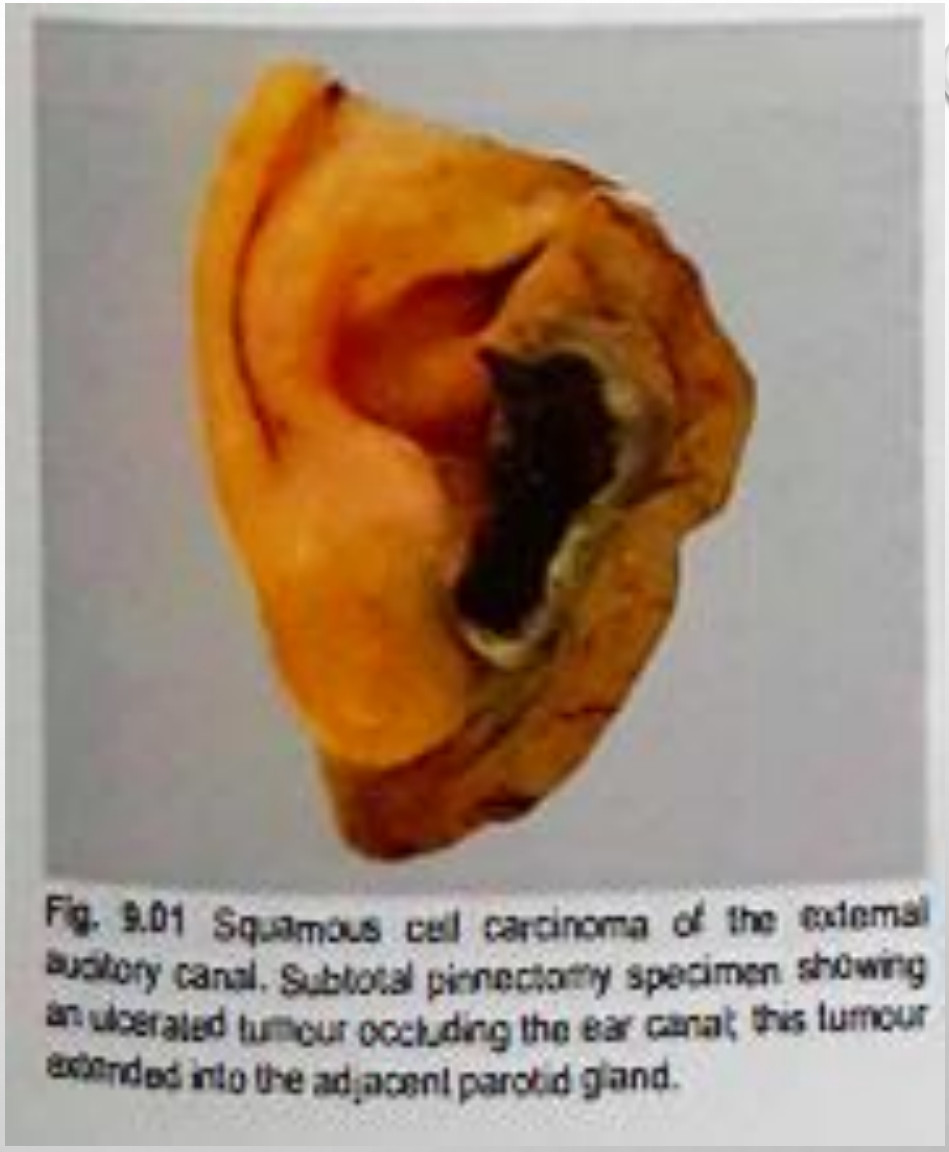


Fig. 9.01 Squamous cell carcinoma of the external auditory canal. Subtotal pinnectomy specimen showing an ulcerated tumour occluding the ear canal; the tumour extended into the adjacent parotid gland.

- MACROSCOPY :
  - WARTY, EXOPHYTIC MASS
- MICROSCOPY :
  - SCC
- PROGNOSTIC :
  - AGGRESSIVE
  - POOR :
    - HIGH CLINICAL STAGE
    - TUMOR DEPTH >8 MM
    - LYMPHOVASCULAR INVASION

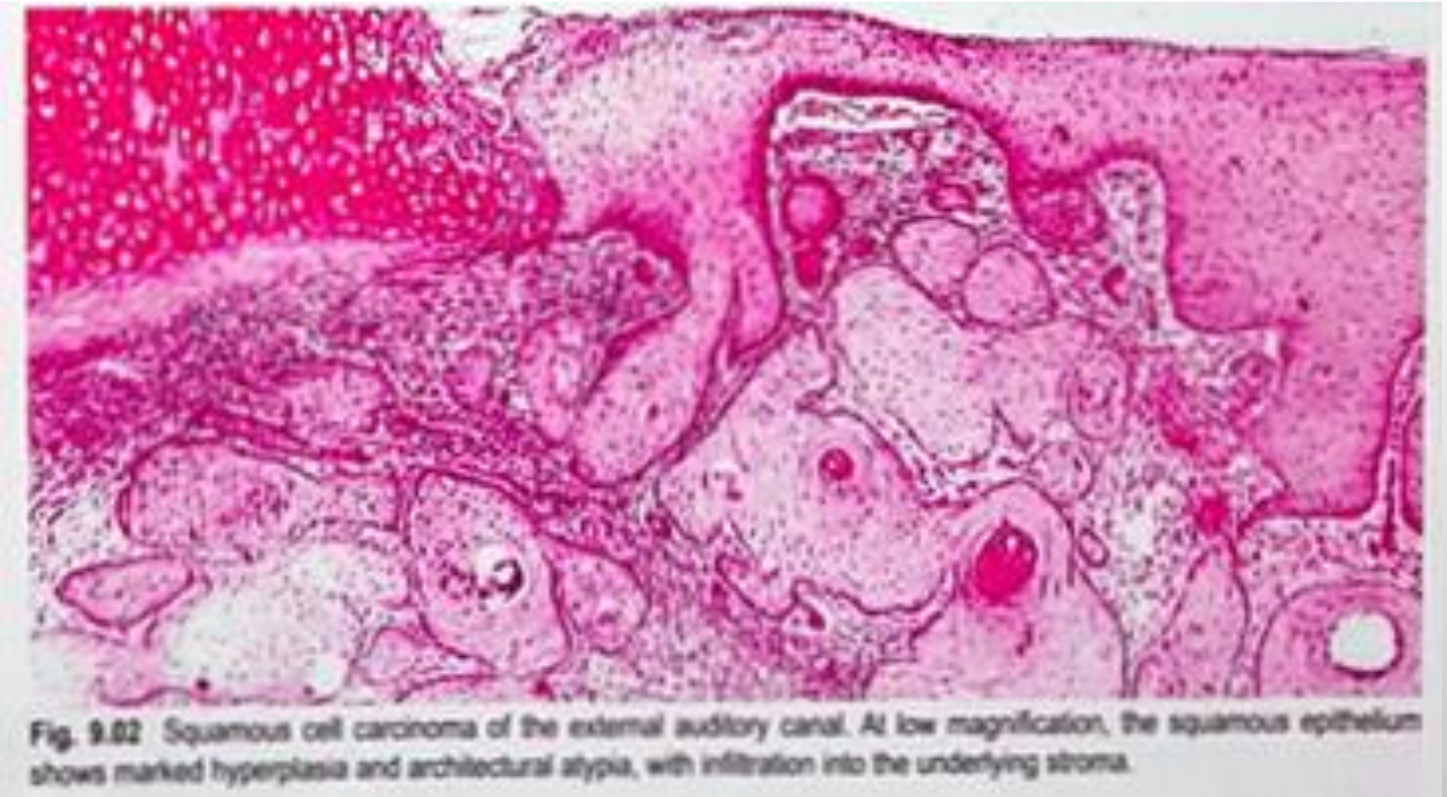
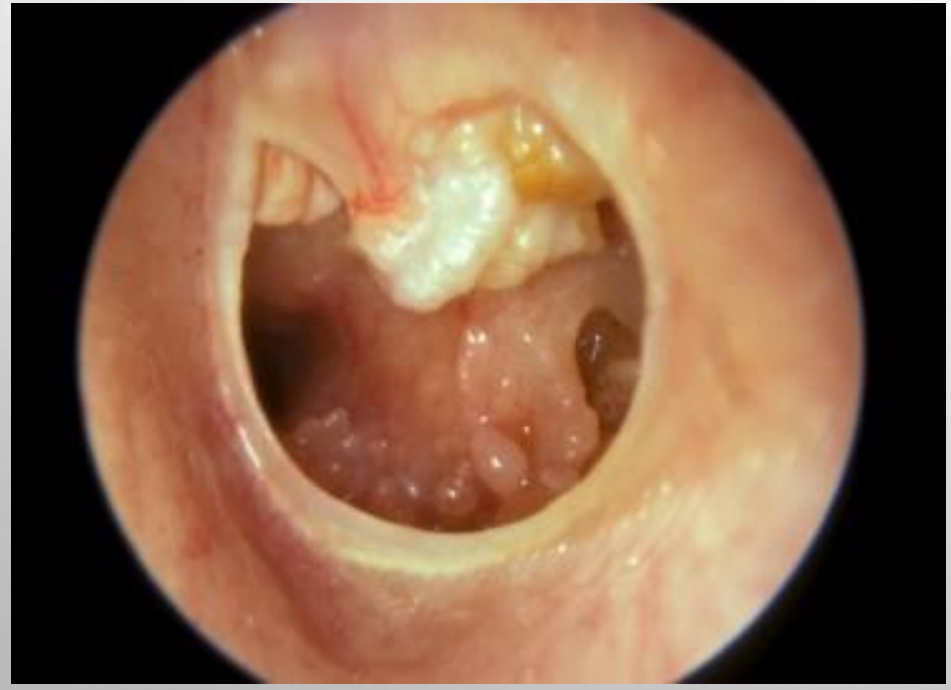


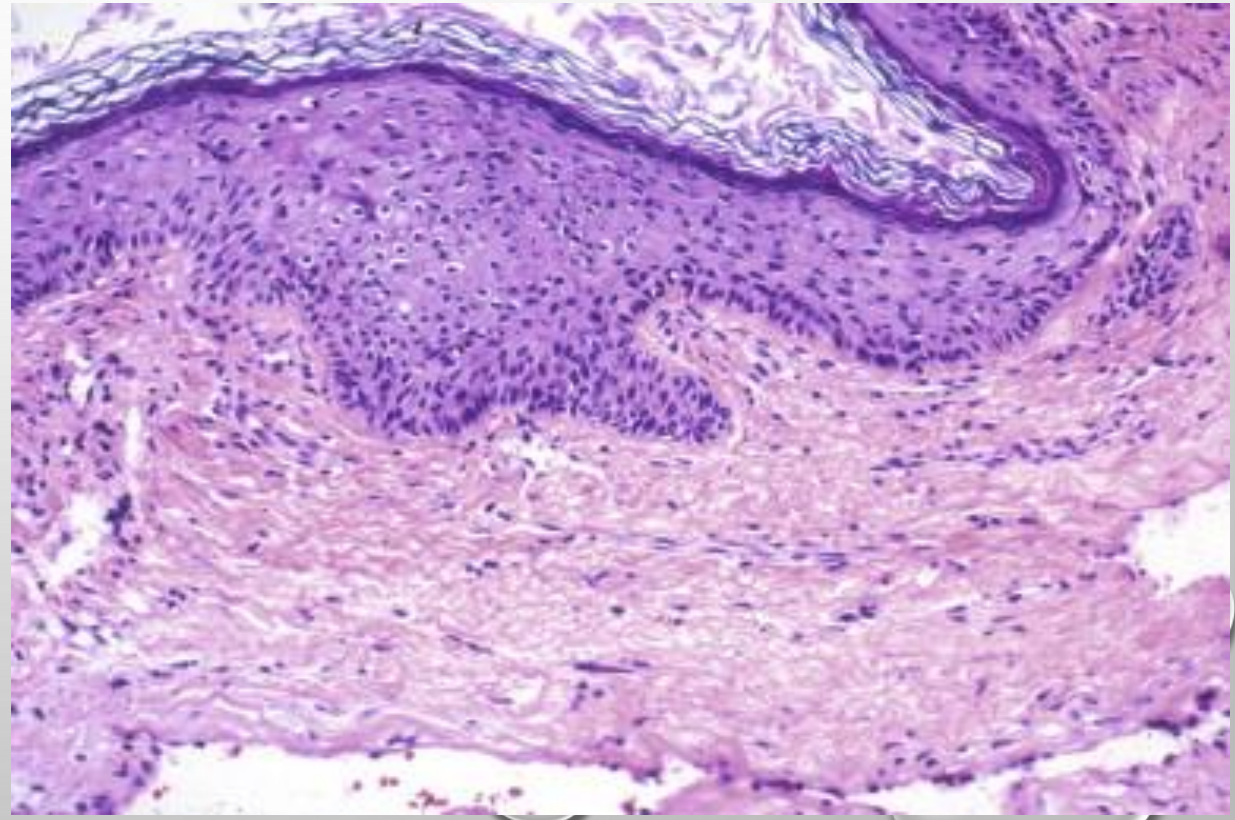
Fig. 9.82 Squamous cell carcinoma of the external auditory canal. At low magnification, the squamous epithelium shows marked hyperplasia and architectural atypia, with infiltration into the underlying stroma.

CHOLESTEATOMA

- CHOLESTEATOMA IS A CYSTIC OR OPEN MASS OF KERATINIZING SQUAMOUS EPITHELIUM IN AIR-FILLED SPACES OF TEMPORAL BONE. ALTHOUGH NOT NEOPLASTIC, IT HAS A PROPENSITY TO ERODE LOCAL STRUCTURES AND TO RECURE AFTER EXICISION
- 3-15 CASES PER 100.000 CHILDREN
- 9-13 CASES PER 100.000 ADULTS
- MALE PREDOMINANCE
- CONGENITAL CHOLESTEATOMA : INFANT AND YOUNG CHILDREN
- ETIOLOGY :
  - CONGENITAL : EMBRYONIC REST
  - ACQUIRED : PERFORATED EARDRUM

- CLINICAL FEATURES :
  - HEARING LOSS
  - FOUL-SMELLING AURAL DISCHARGE
- MACROSCOPY :
  - PEARLY WHITE MASS IN THE MIDDLE EAR CAVITY





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