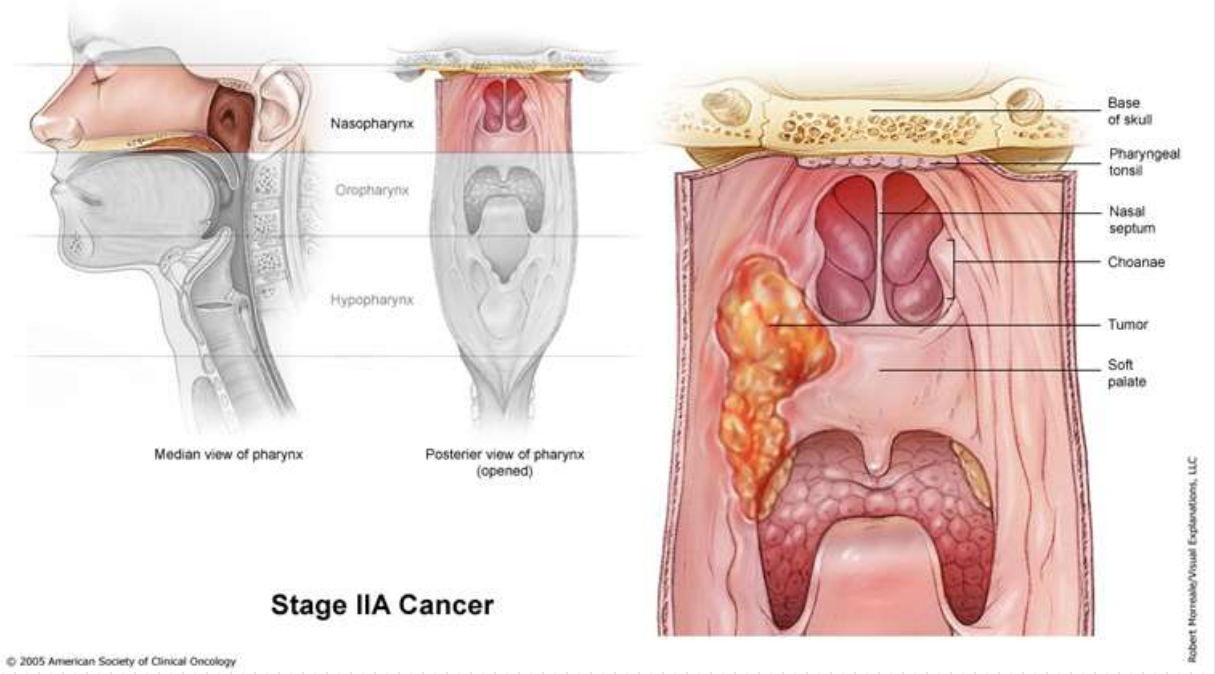
Nasopharyngeal Carcinoma



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Nasopharyngeal carcinoma (NPC)

- A tumour arising from the epithelial cells that cover the surface and line the nasopharynx.
- Approximately one third of nasopharyngeal carcinomas of the undifferentiated type are diagnosed in adolescents or young adults.
- Although rare, NPC accounts for one third of childhood nasopharyngeal neoplasms

Etiology-1

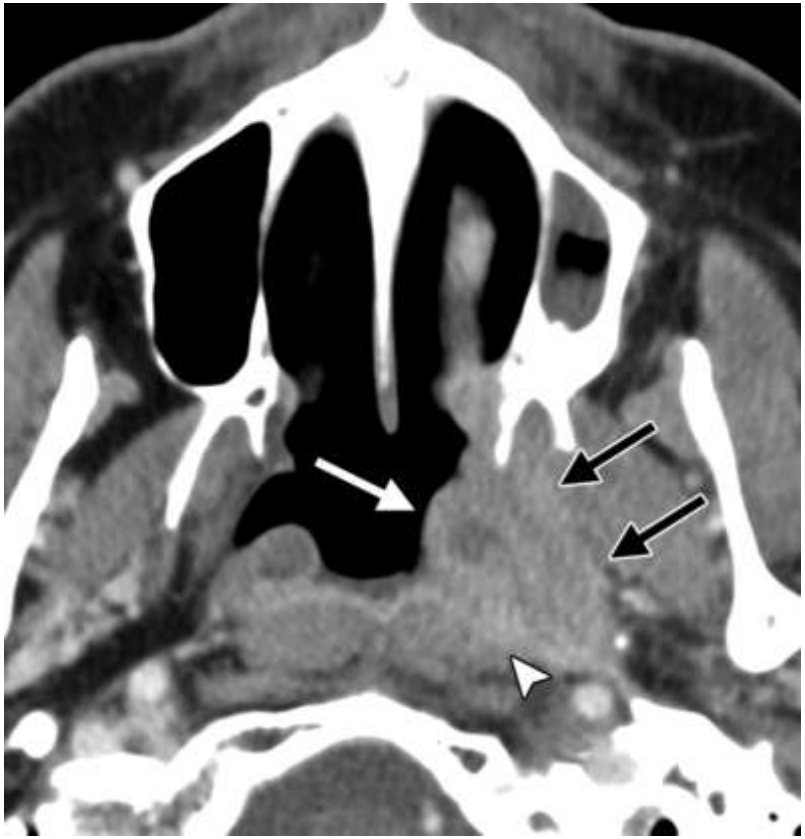
- NPC is the commonest epithelial cancer in adults.
- The detection of nuclear antigen associated with Epstein-Barr virus (EBNA) and viral DNA in NPC type 2 and 3, has revealed that EBV can infect epithelial cells and is associated with their transformation.

Etiology-2

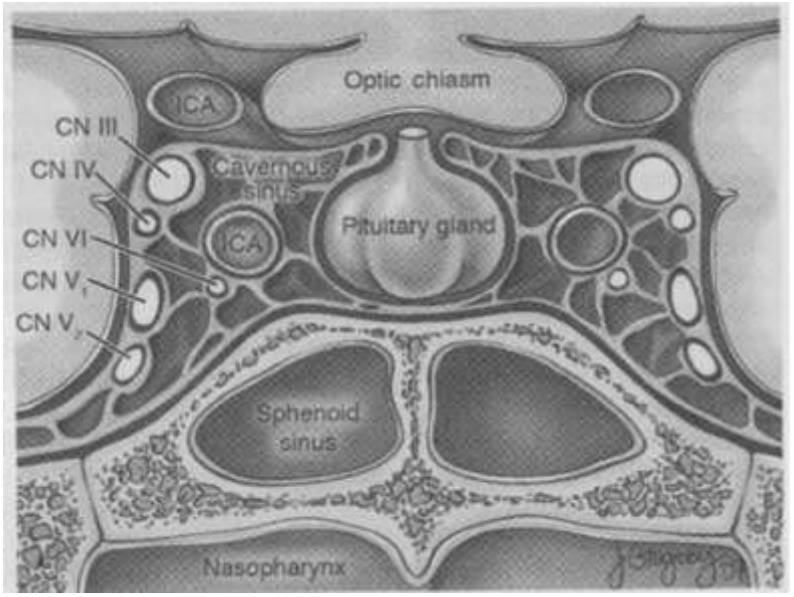
- The aetiology of NPC (particularly the endemic form) seems to follow a multi-step process, in which EBV, ethnic background, and environmental carcinogens all seem to play an important role.
- In adults, other likely etiological factors include genetic susceptibility, consumption of food (in particular salted fish) containing carcinogenic volatile nitrosamines.

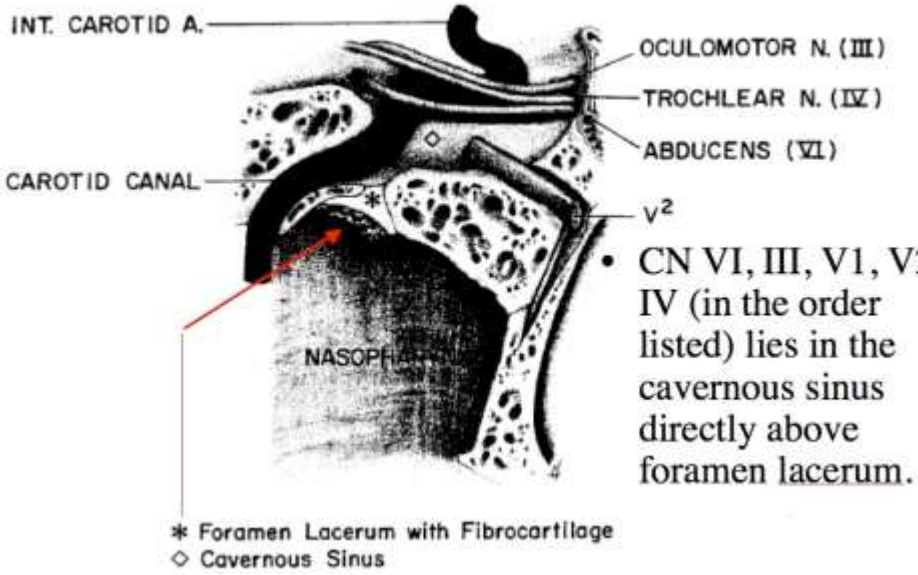
Clinical presentation

NPC usually originates in the lateral wall of the nasopharynx, which includes the fossa of Rosenmuller.



It can then extend within or out of the nasopharynx to the other lateral wall and/or postero-superiorly to the base of the skull or the palate, nasal cavity or oropharynx.





- CN VI, III, V1, V2, IV (in the order listed) lies in the cavernous sinus directly above foramen lacerum.

- It then typically metastasises to cervical lymph nodes.
- Distant metastases may occur in bone, lung, mediastinum and, more rarely, the liver

CLINICAL COURSE

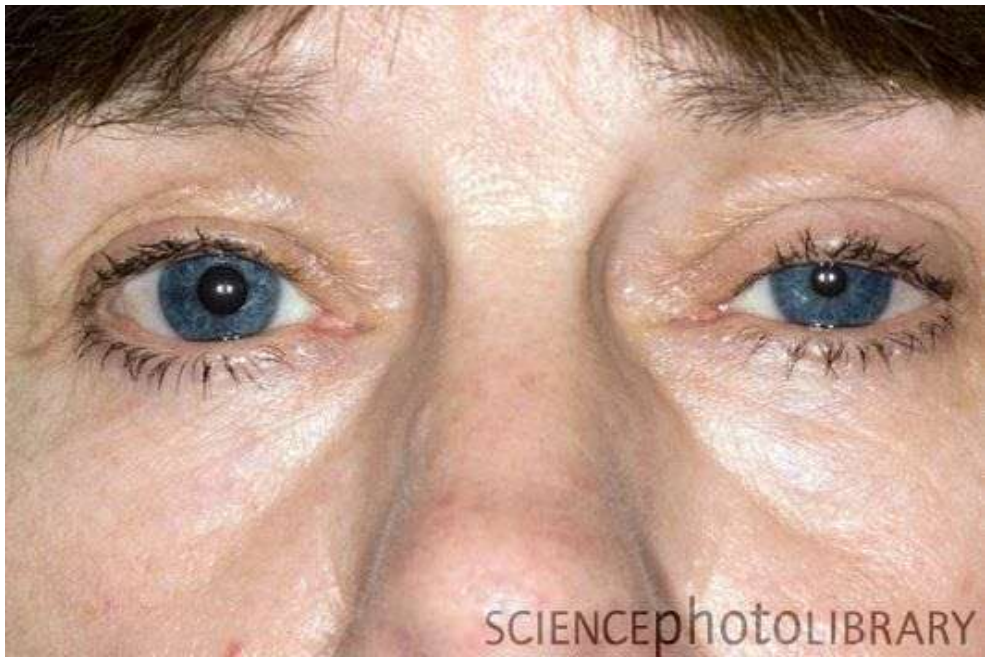
- Unilateral hearing loss from a middle ear effusion is the most common finding.
- Another common presenting complaint is a neck mass resulting from regional spread.
- Large or exophytic lesions may cause nasal obstruction or epistaxis.
- As the tumour enlarges, adjacent cranial nerves may become involved.

- **Xerophthalmia** may result from involvement of the greater superficial petrosal nerve at the foramen lacerum.
- **Facial pain** may indicate Trigeminal nerve involvement.
- **Diplopia** may occur with isolated Abducens nerve injury.

- **Ophthalmoplegia** indicates involvement of cranial nerves III, IV and VI, usually in the cavernous sinus or the superior orbital fissure.
- **Horner's syndrome** occurs with injury to the cervical sympathetic chain and more extensive skull base involvement produces deficits of the lower cranial nerves (IX, X, XI, XII)

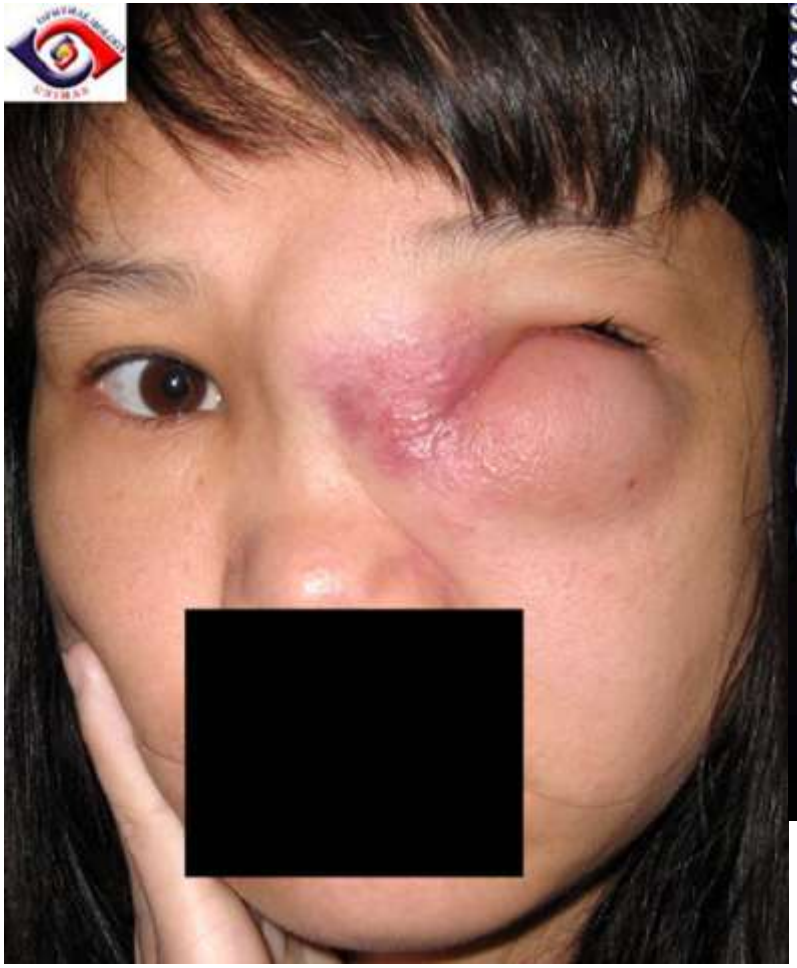
Horner's syndrome

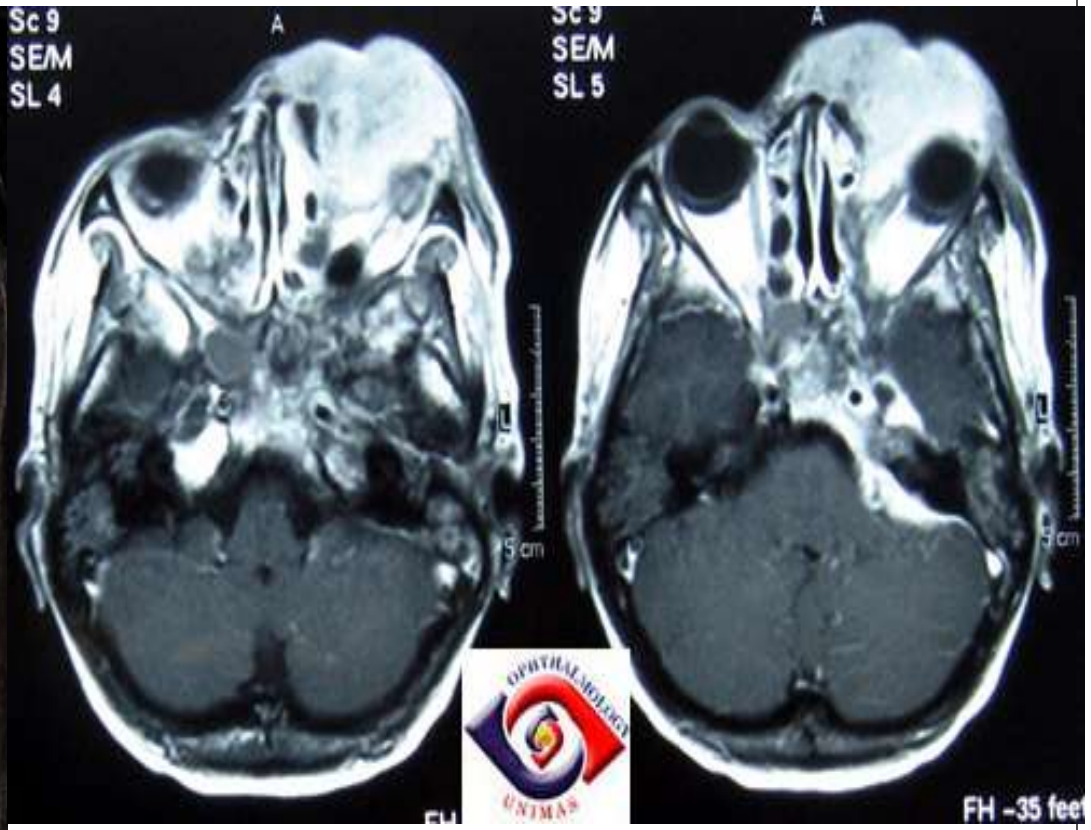
- Partial ptosis (drooping of the upper eyelid from loss of sympathetic innervation to the superior tarsal muscle)
- Upside-down ptosis (slight elevation of the lower lid)
- Aanhidrosis (decreased sweating on the affected side of the face)
- Miosis (small pupils)
- Enophthalmos (the impression that the eye is sunk in)



Clinical presentation

- Metastatic spread may result in bone pain or organ dysfunction.
- Rarely, a paraneoplastic syndrome of osteoarthropathy may occur with widespread disease.

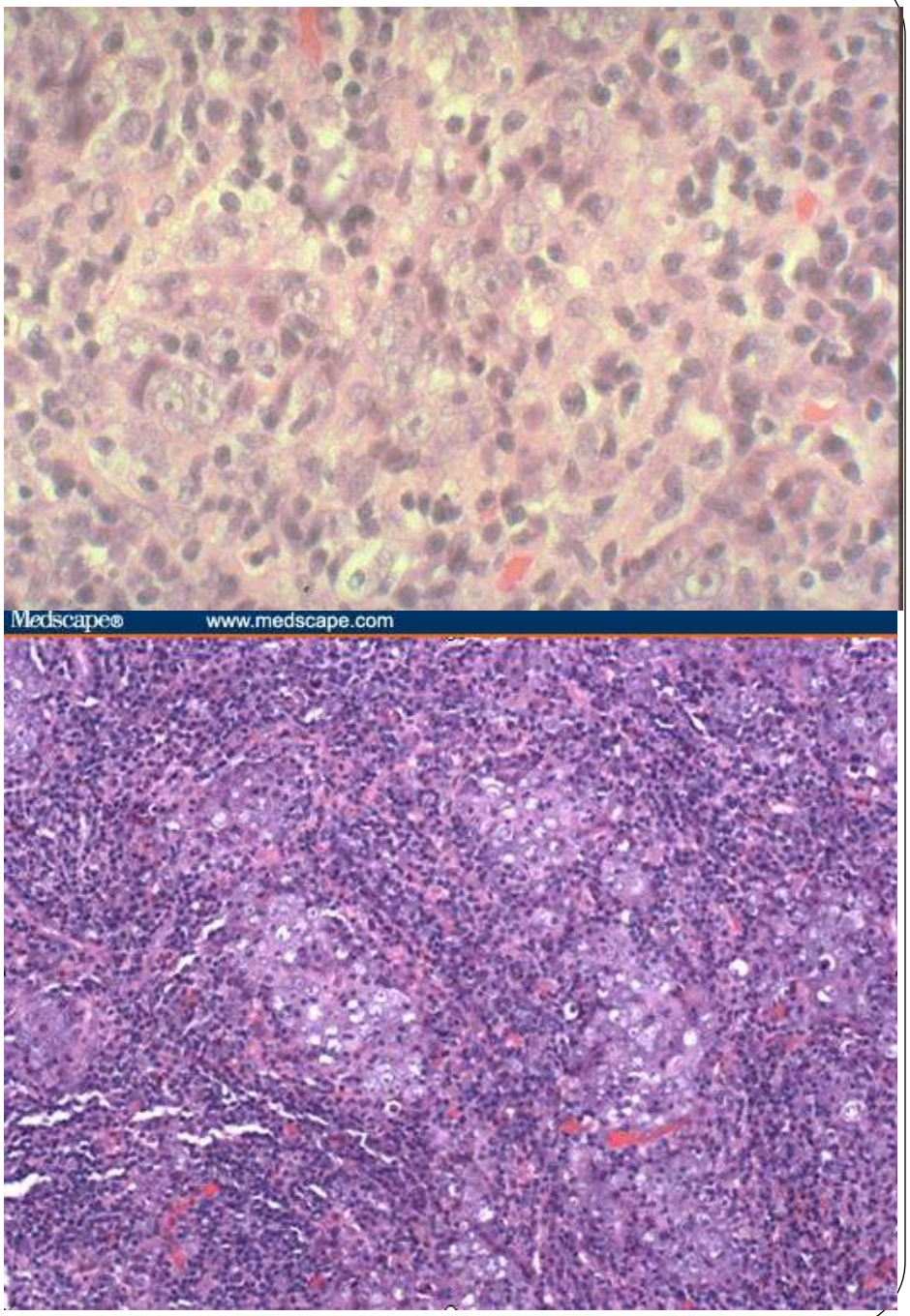



Sc 9
SE/M
SL 4
A
5 cm
Sc 9
SE/M
SL 5
A
5 cm
FH -35 feet

Histopathology

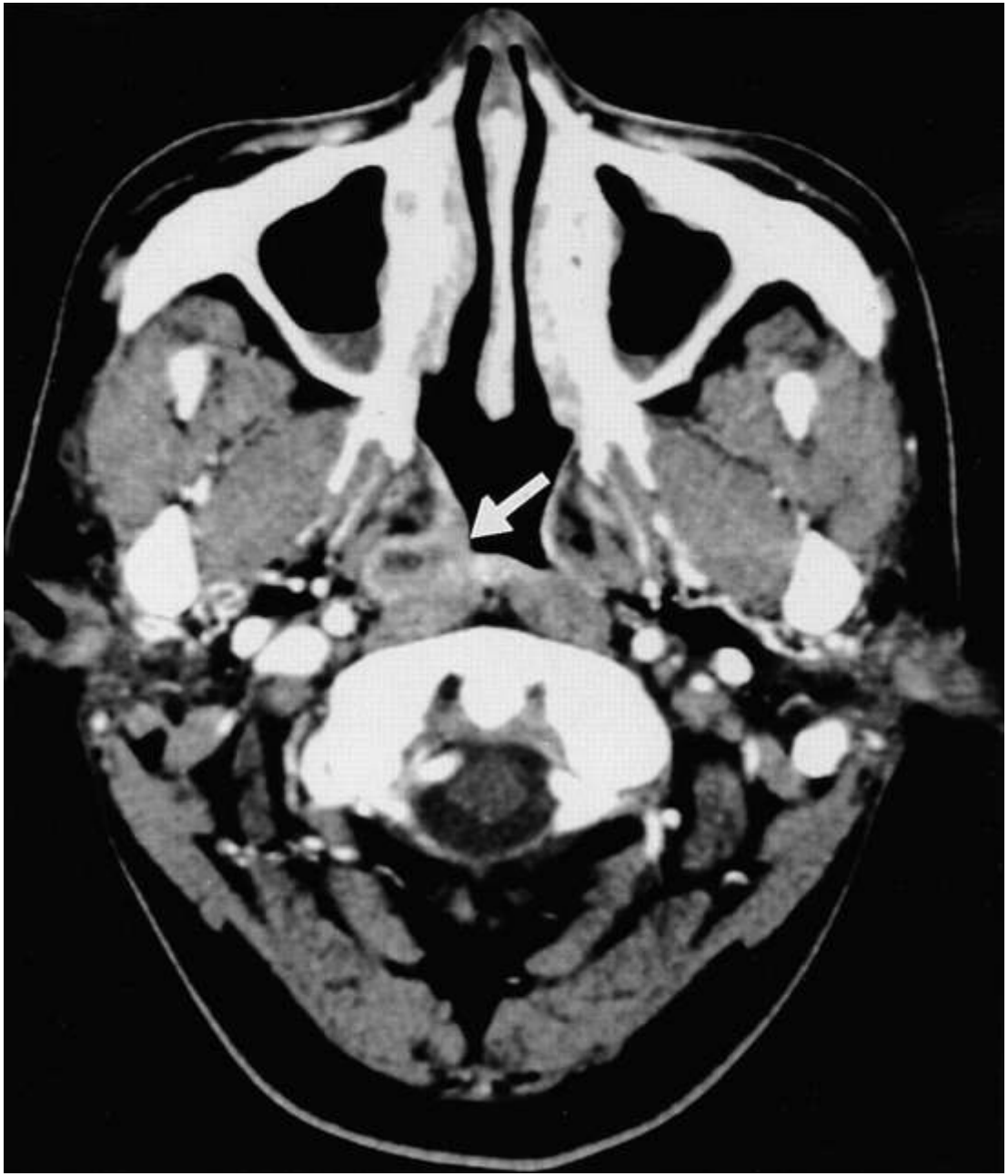
Three subtypes of NPC are recognized in the World Health Organisation (WHO) classification:

- Type 1: squamous cell carcinoma, typically found in the older adult population
- Type 2: non-keratinizing carcinoma
- Type 3: undifferentiated carcinoma



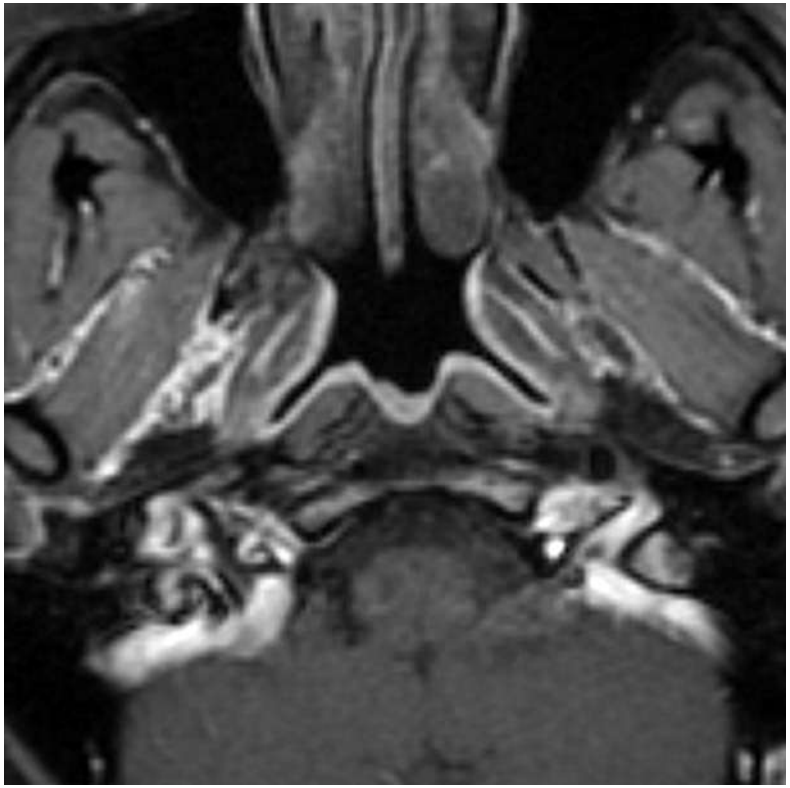
Diagnostic methods

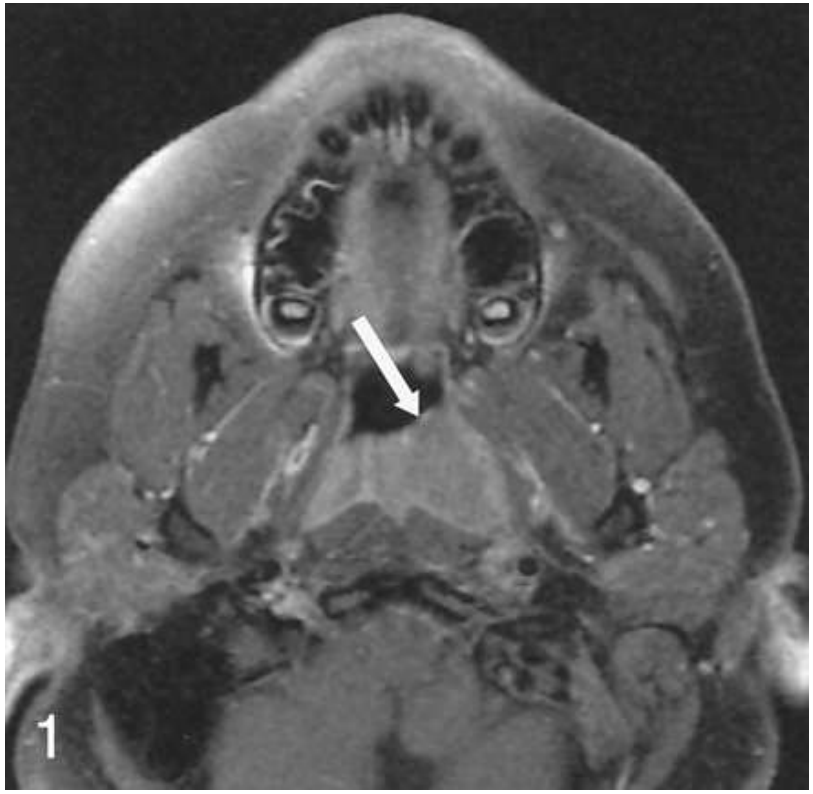
- Clinical evaluation of the size and location of cervical lymph nodes.
- Indirect nasopharyngoscopy to assess the primary tumor.
- Neurological examination of cranial nerves.



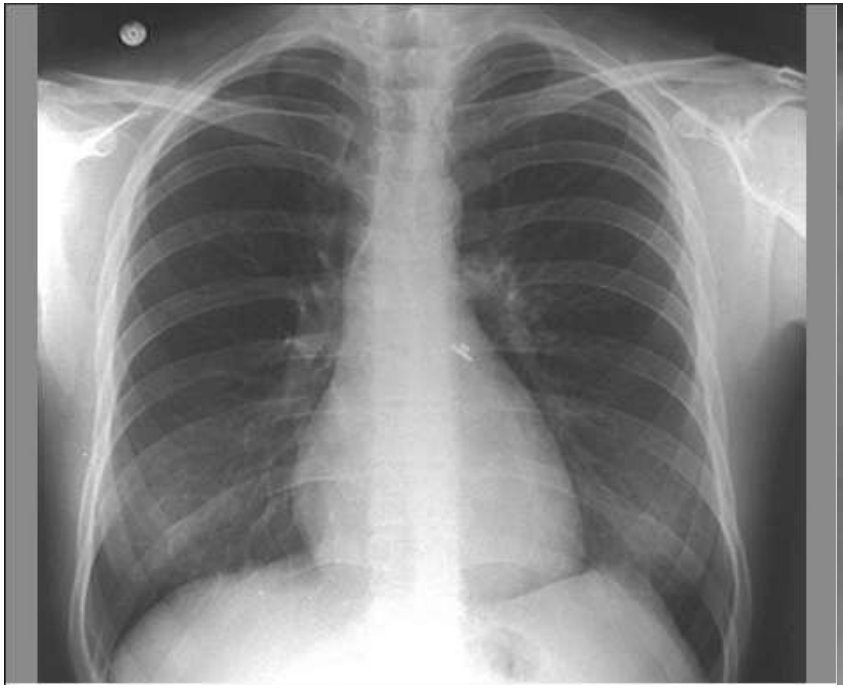
Contrast enhanced axial CT showing recurrence of nasopharyngeal carcinoma in the right nasopharynx (arrow) invading posterolaterally.

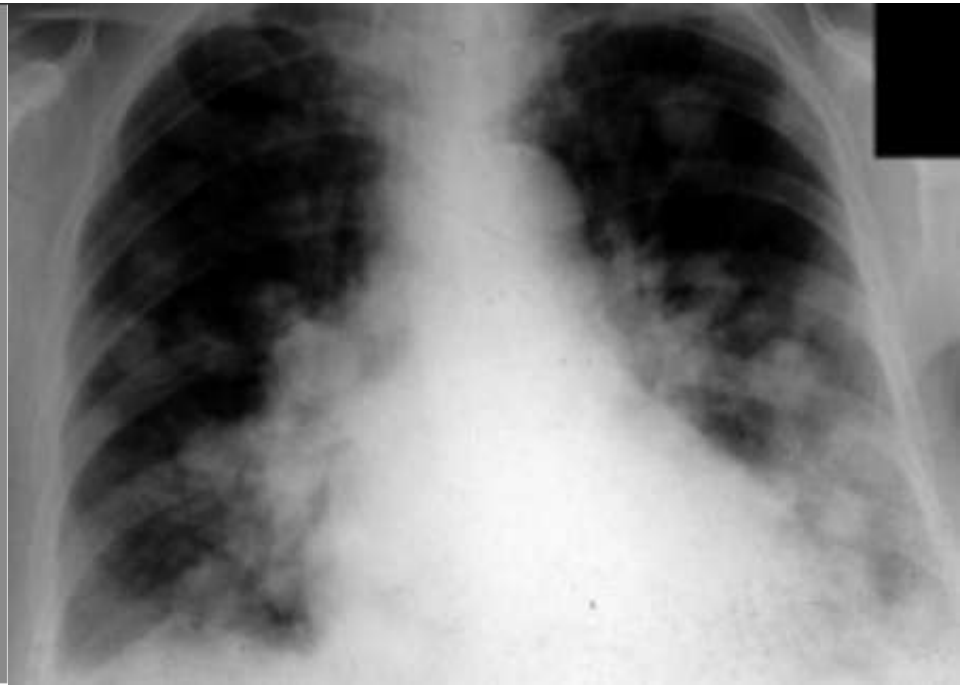
MRI

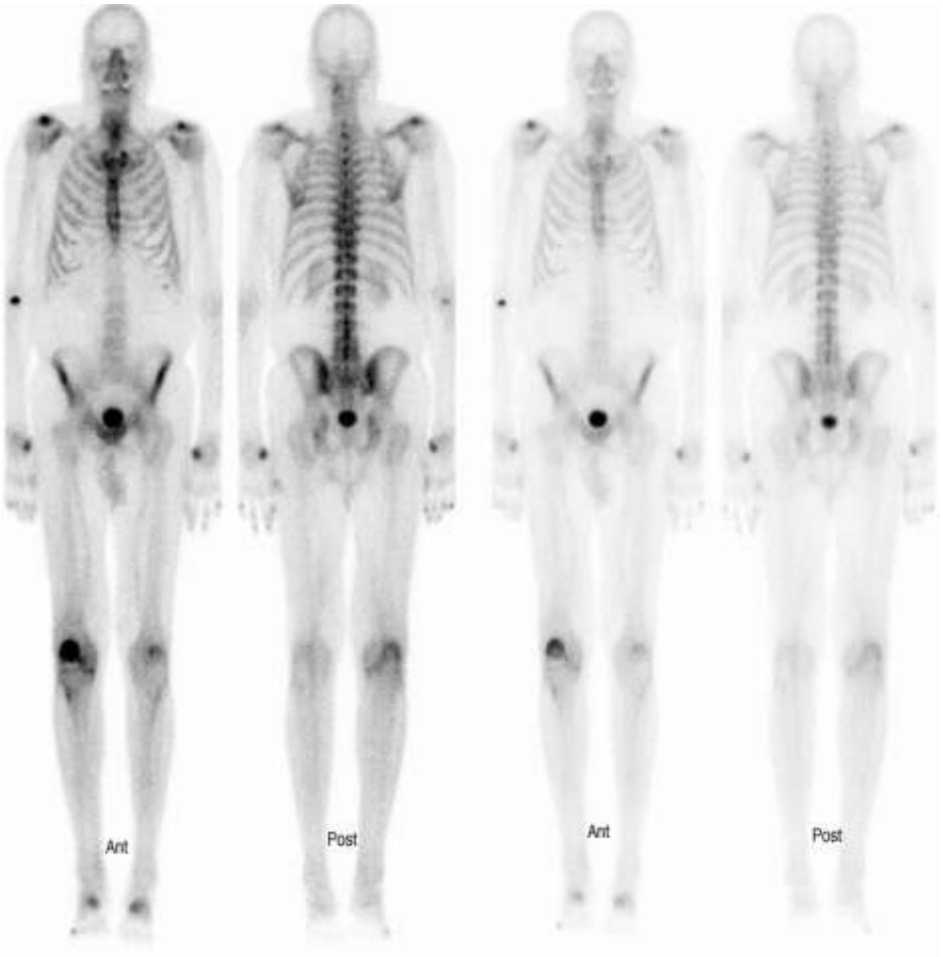




Chest radiography to see if NPC has spread to the lungs.



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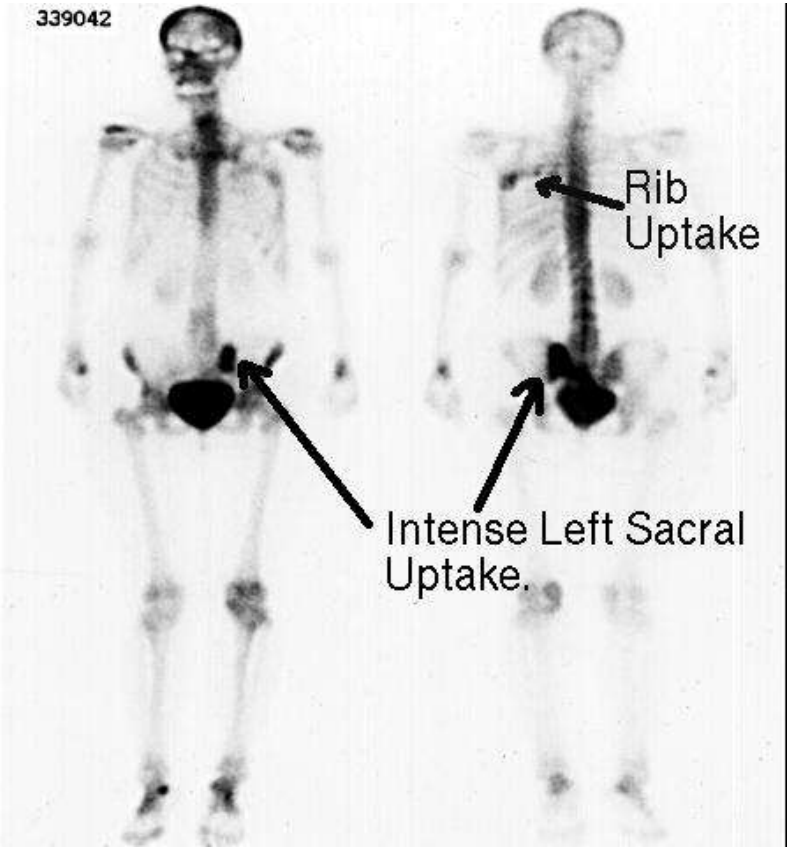
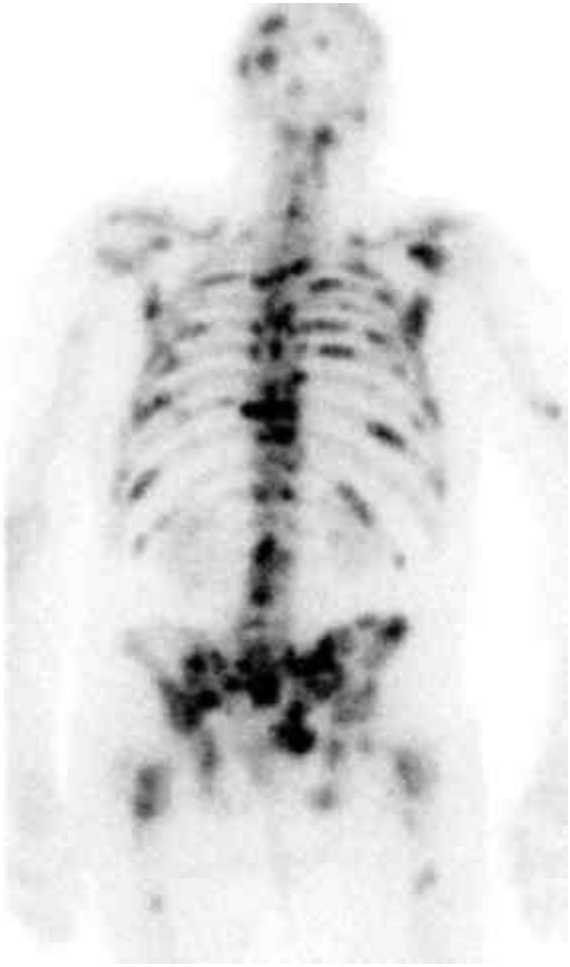

339042
Rib
Uptake
Intense Left Sacral
Uptake.

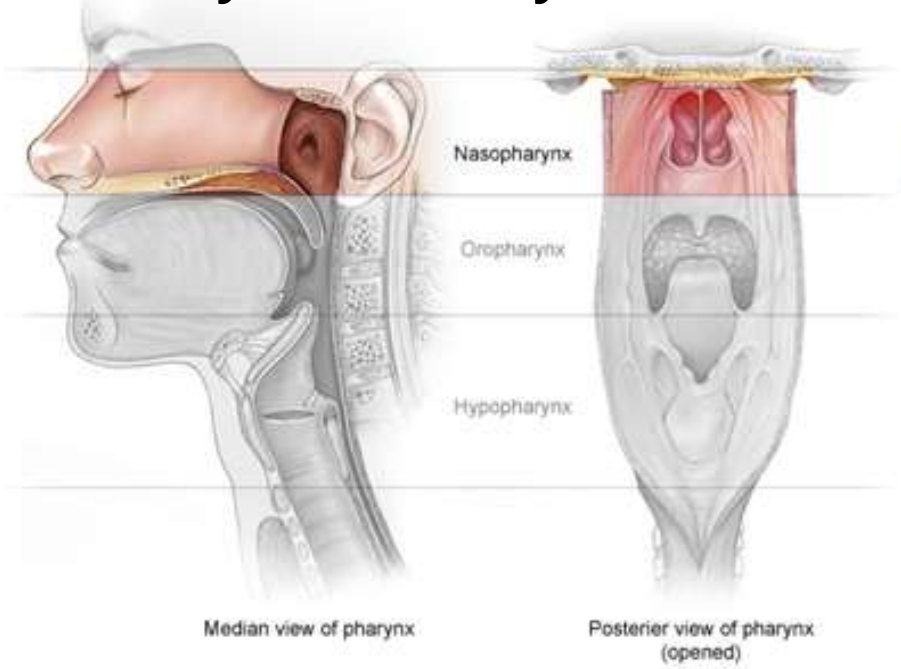
Bone scintigraphy by Tc 99 diphosphonate to show whether cancer has spread to the bones.

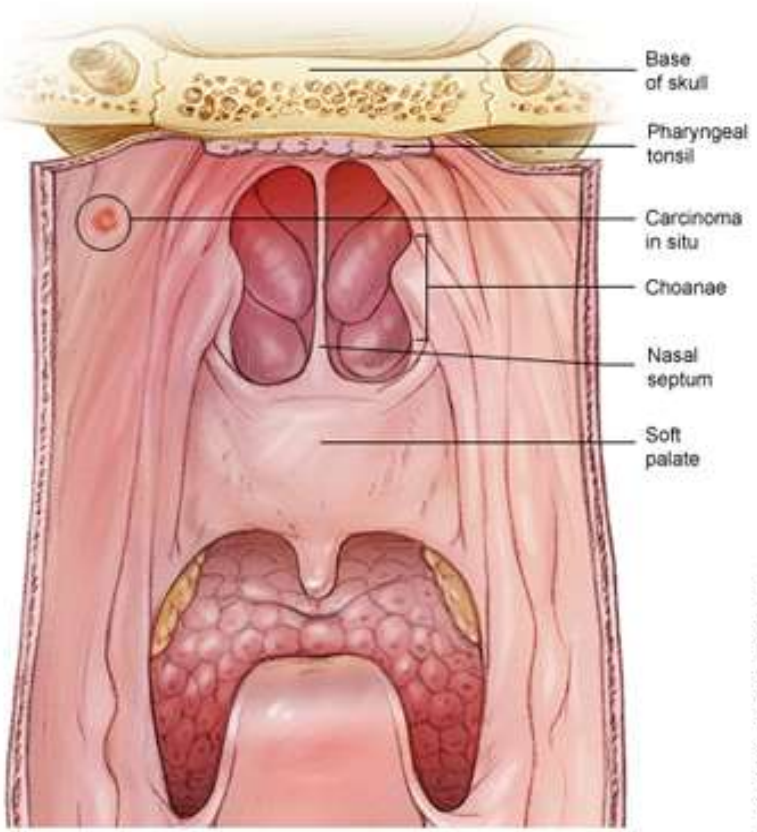


- Full blood count.
- Urea, electrolyte, creatinine, liver function, Ca, PO4, alkaline phosphate.
- EBV viral capsid antigen and EBV DNA.
- Biopsy of either the lymph nodes or primary tumor for histological examination.

A carcinoma in situ (Tis) with no spread to lymph nodes (N0) or distant metastasis (M0).

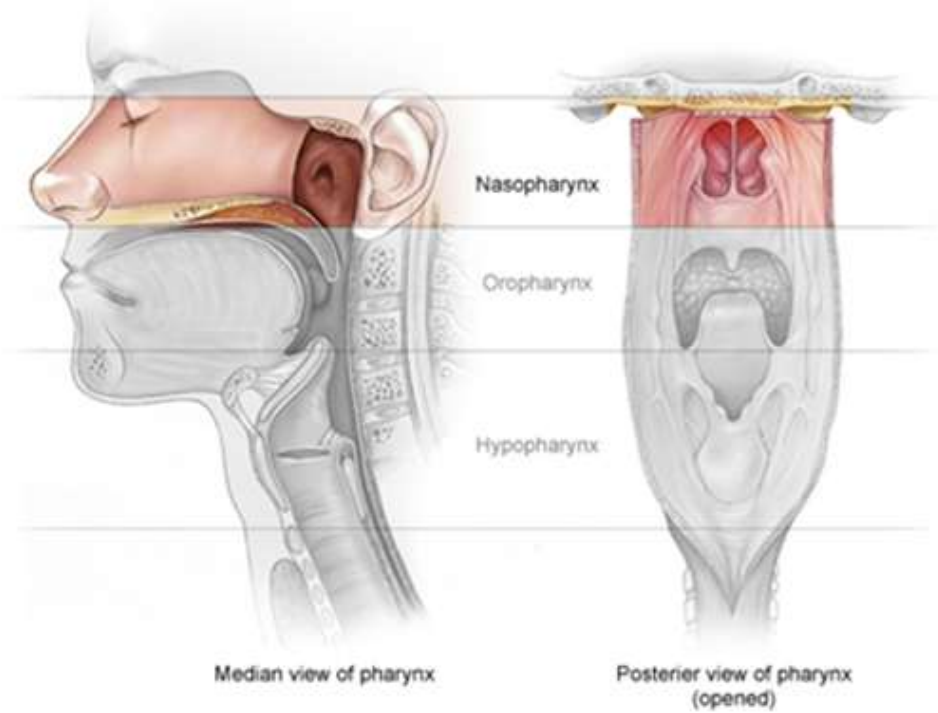
Tis: This describes a stage called carcinoma (cancer) in situ. This is a very early cancer where cancer cells are found only in one layer of tissue.

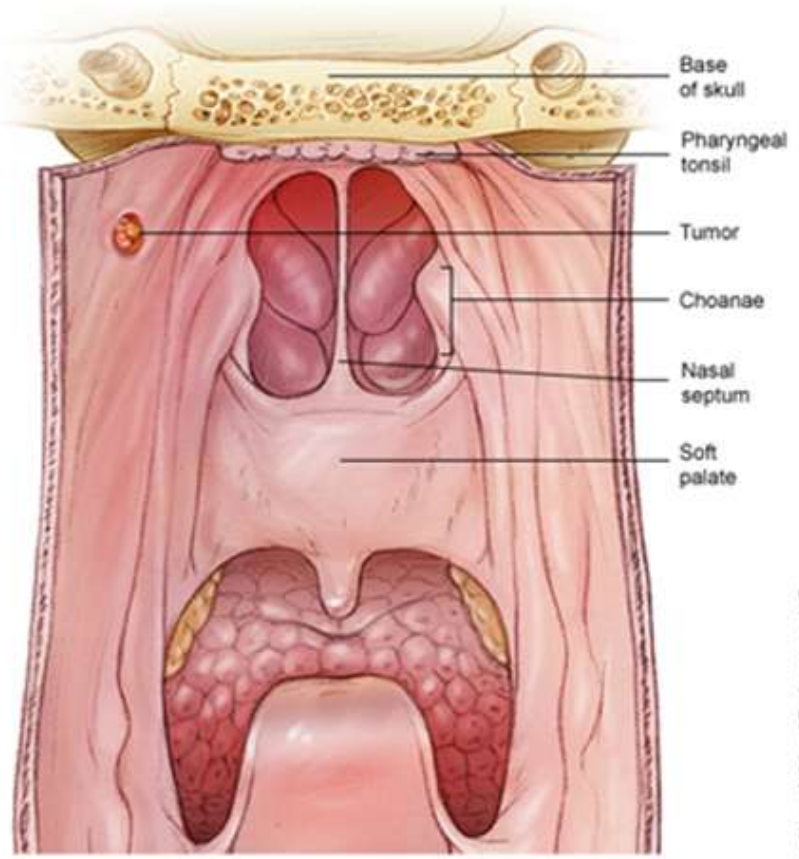




Stage 0 Cancer

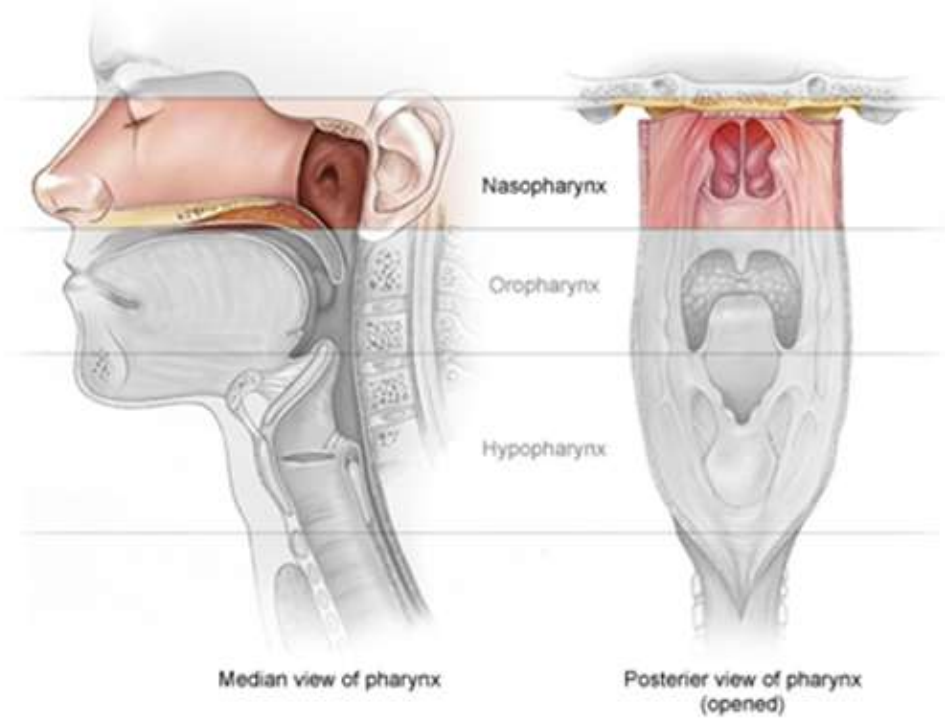
Stage I: A small tumor (T1) with no spread to lymph nodes (N0) and no distant metastasis (M0)

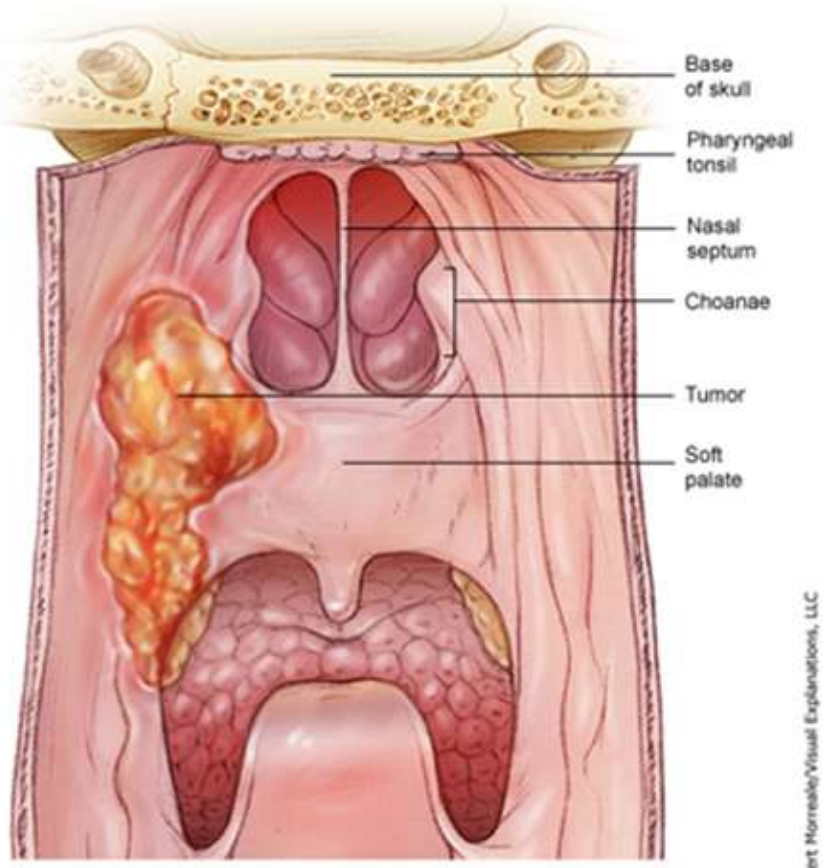




Stage I Cancer

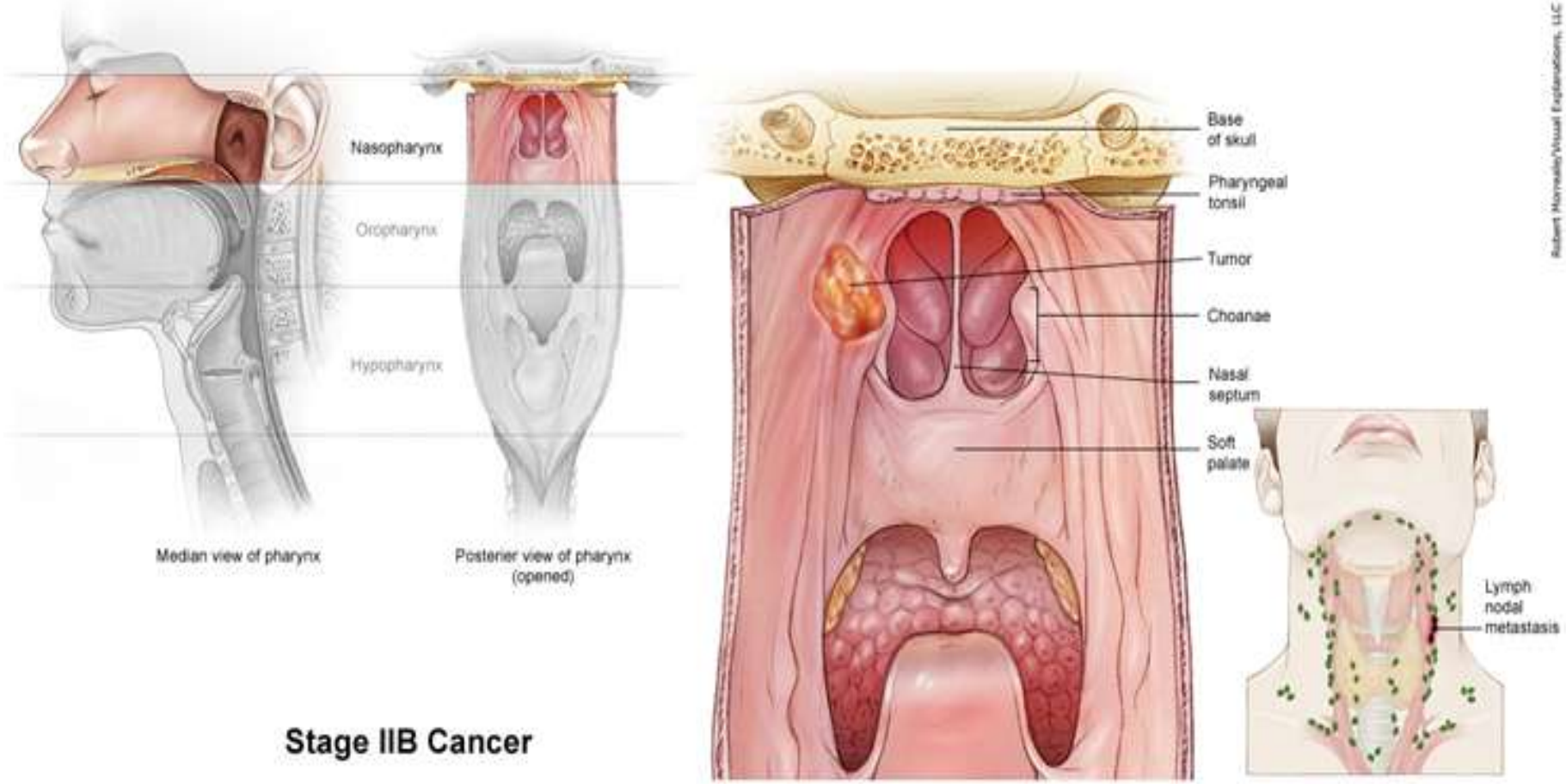
A tumour that has extended beyond the nasopharynx (T2) but has not spread to lymph nodes (N0) or to distant parts of the body (M0).



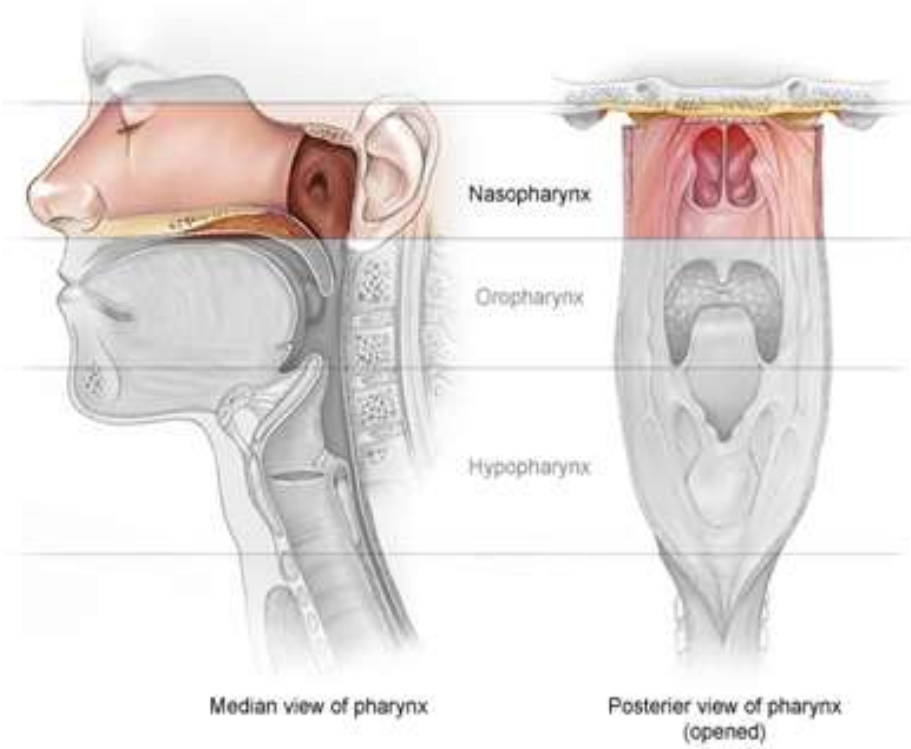


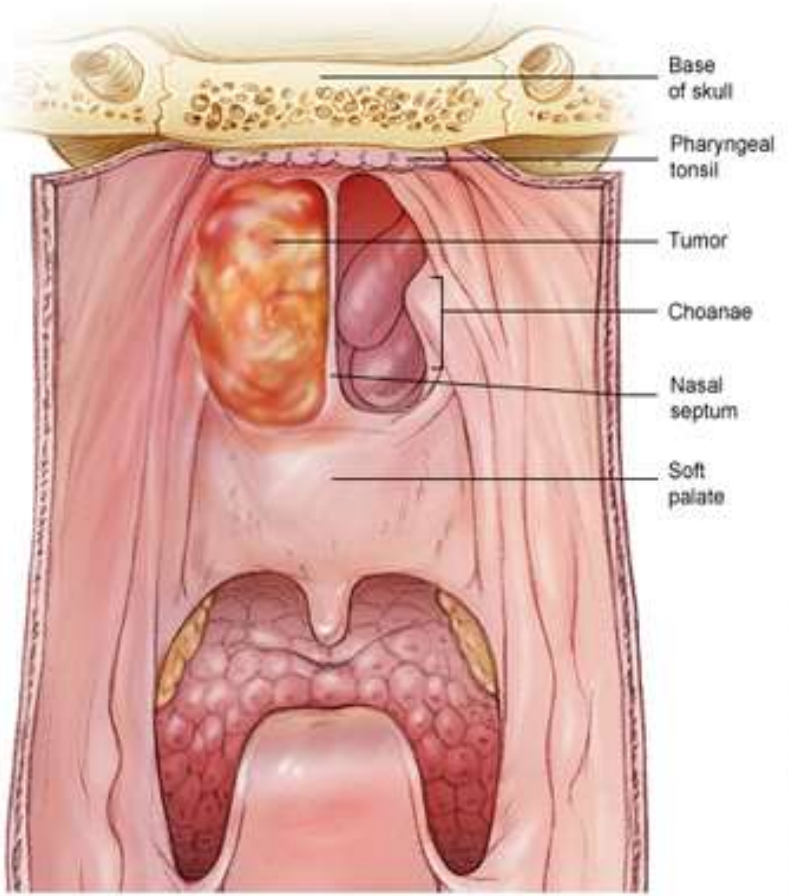
Stage IIA Cancer

Stage IIB: A tumour (T1 or T2) that has spread to lymph nodes (N1) but has not metastasized (M0)



Stage III: This describes a non-invasive and invasive tumour (T1 or T2) that have spread to lymph nodes (N1 or N2) but have not metastasized (M0), or it describes a larger tumour (T3) with or without nodal involvement (N0, N1, or N2) and no metastasis (M0).





Stage III Cancer

MANAGEMENT

- External beam radiation therapy continues to be the mainstay of treatment for this lesion.
- Doses of 6500 to 7000 cGy are directed at the primary lesion and the upper echelon lymph nodes.
- If clinically positive, lower cervical nodes are included in the field.
- **Brachytherapy** is occasionally used as an adjuvant to external beam radiation or in cases of recurrent/residual tumor.

Surgical management

- Primarily used to obtain tissue for histologic examination and for EBV testing.
- If an obvious tumor is present in the nasopharynx, biopsy under local anesthesia in the clinic may be practical if the patient is cooperative.
- If the tumor is not obvious or if sufficient tissue cannot be obtained in clinic, the patient should be taken to the operative room for formal endoscopy and biopsy under general anesthesia.

- Chemotherapy as an adjuvant to radiation therapy has yet to demonstrate a significant improvement in long-term outcome and therefore continues to be used mainly as a palliative measure.
- While immunotherapy has also not shown any clear improvement in survival to date, the close association of certain anti-EBV antibodies with an improved prognosis offers the hope of effective immunologically based approach in the future.
- Also, a vaccine to protect against EBV related disease may one day be reality.