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RESEARCH ARTICLE

NASO PHARYNGEAL ADENO CARCINOMA: A CASE REPORT

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Abstract

Nasopharyngeal adeno carcinoma (NAC) is a rare malignancy, accounting for lessthan 0.5% of nasopharyngealtumors. We report the case of a 51-year-old man presentingwith right ptosis and a frontal infiltrative lesion, ultimatelydiagnosed as moderatelydifferentiated NAC withbonemetastasis. He wastreatedwith palliative chemotherapy and whole-brainradiotherapy. NAC exhibits distinct clinicalbehavior, with a lower rate of nodal involvement but a tendency for cranial nerve invasion and local recurrence. Due to itsrarity, treatmentis not standardized, but a multimodal approachremains the most effective strategy. Furtherresearchisneeded to optimize management and improveoutcomes.

Introduction:-

Nasopharyngealadenocarcinoma (NAC) is a rare entity, accounting for approximately 0.48% to 0.5% of nasopharyngealneoplasms [1]. Unlikenasopharyngealsquamouscellcarcinoma (NSC), whichrepresents up to 95–98% of nasopharyngeal cancers, NAC ischaracterized by its distinct histologicalorigin and unique clinicalbehavior [2,4,6].

NAC is a histologically distinct formfrom NSC, and the involvement of Epstein-Barr virus (EBV) in itscarcinogenesisappears to benegligible. The rate of cervical lymphnodemetastasesislower in NAC compared to NSC. Clinicalfeatures of NAC include slow growth, a tendency to invadecranial nerves, a high risk of local recurrence, and a low rate of distant metastases.

Management strategies for NAC remain non-standardized due to itslow incidence and the lack of robust data [2,6].

Case Report Patient S.L., a 51-year-old male with no significantmedicalhistory, presentedwith a chronic smoking historyestimated at 20 pack-years, havingquitthreemonthsprior to consultation. No otherknownoncologicalriskfactorswereidentified.

Symptomsbegantenmonthsbefore consultation, characterized by the progressive onset of right-sided ptosis, withoutvisualacuityloss, diplopia, or otherassociatedneurologicalsigns. The condition graduallyworsened, prompting an etiologicalworkup.

Brain MRI (Figure 1) revealed a right frontal infiltrative lesion suggestive of metastasis, measuring 18 mm in thickness and extending over 49 mm. The lesioninvaded the lateral and superior right orbital muscles, with direct contact with the optic nerve. On CT scan (Figure 2), thislesioncausedbone destruction involving the frontal

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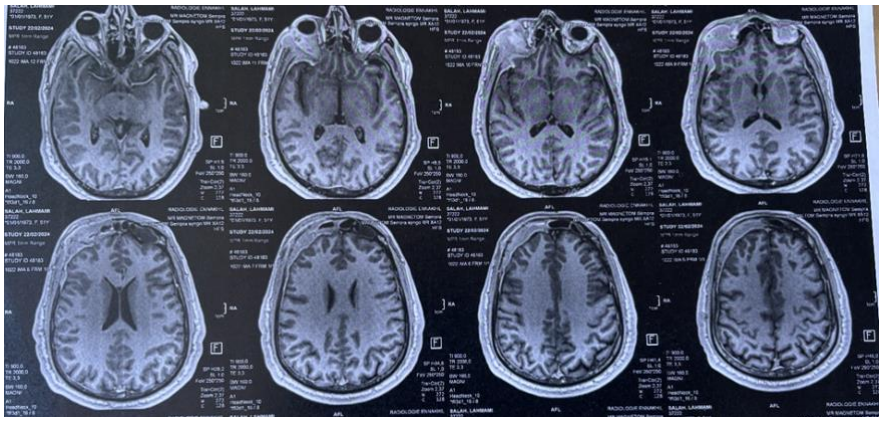


squama, the lesserwing of the sphenoid, and the lateral orbital wall.

Figure 1: Axial brain MRI slices showing a right frontal infiltrative lesion. The lesioninvades the lateral and superior right orbital muscles and is in direct contact with the optic nerve.

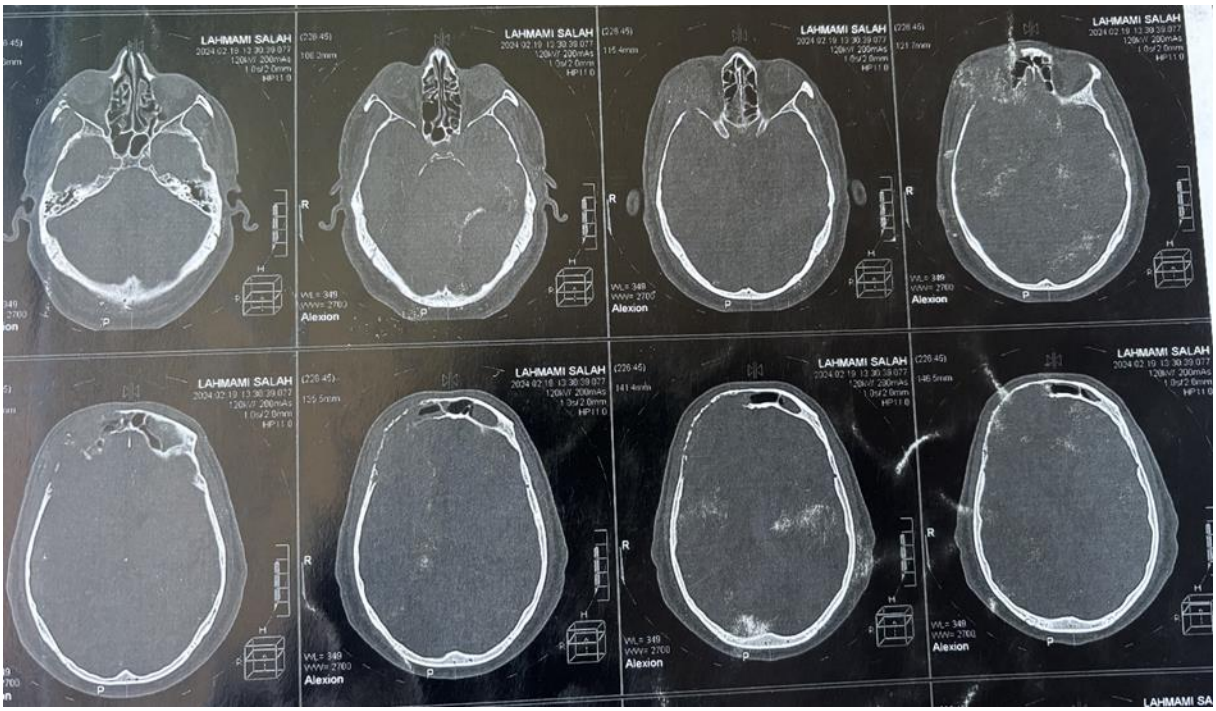


Figure 2: Axial brain CT scans (bonewindow) showing a lesioncausingbonelysis of the frontal squama, the lesserwing of the sphenoid, and the lateralwall of the orbit.

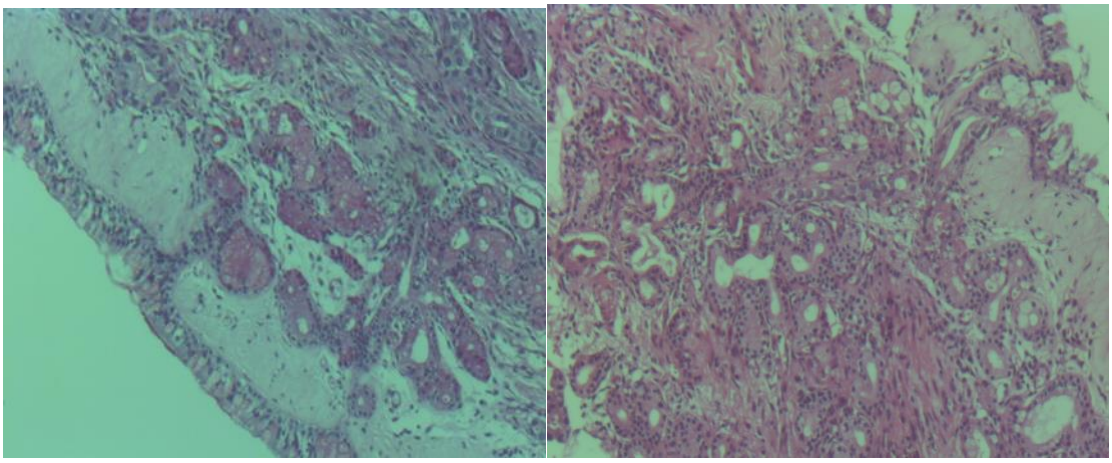
StagingWorkup

- Cervico-thoraco-abdomino-pelvic CT scan (TAP):revealed right jugulocarotid and parotidlymphadenopathymeasuring 10 mm in short axis, with no secondarythoracic, abdominal, or pelviclesions.

- **PET-CT scan:**
  - Presence of a hypermetabolic tissue process centered on the ascendingbranch of the right mandible, infiltrating the ipsilateralmasticatory muscles.
  - Complete hypermetabolicfilling of the right maxillary sinus with infiltration of the temporal process.

- Partial filling of the leftmaxillary sinus, with a hypermetabolic focus in the right nasopharynx extending to the ipsilateralpterygoid muscles, choana, and vomer.
- Hypermetabolism of the frontal sinuses, right sphenoidwing, the floor and lateralwall of the right orbit, with orbital muscle infiltration.

Giventhesefindings, a nasofibroscopywithbiopsywasperformed, confirming the presence of a moderatelydifferentiatednasopharyngealadenocarcinoma (Figures 3, 4).



Figures 3 and 4:Respiratoryepitheliumwith PAS-negativestaining, showing chorion infiltration by a moderatelywell-differentiatedmalignantadenocarcinomatousproliferation.

The patient alsopresentedwith a pathological fracture of the right femoraldiaphysis, requiringsurgical management withintramedullarynailing. Histologicalanalysis of the bonebiopsyconfirmedmetastaticlocalization of adenocarcinoma (Figures 5, 6).

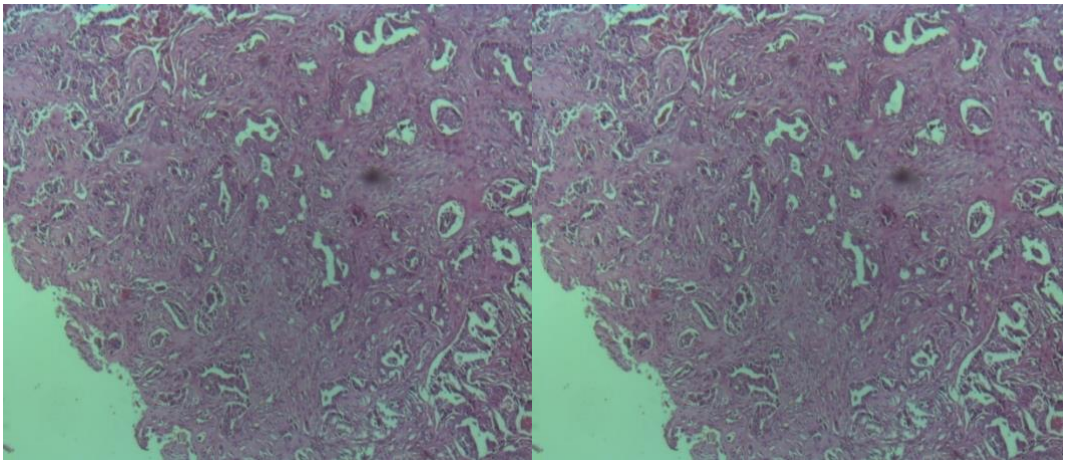


Figure5: Bone metastatictumor tissue from a moderatelywell-differentiatedadenocarcinoma.

Figure 6 :Osteoid tissue withglandular lumen consistent withadenocarcinoma.

After discussion in a multidisciplinarytumorboard (MDT), the followingtherapeuticdecisionwasmade:

- Palliative chemotherapybased on paclitaxel and carboplatin
- Whole-brainradiotherapywith a total dose of 30 Gy in 10 fractions (Figure 7)

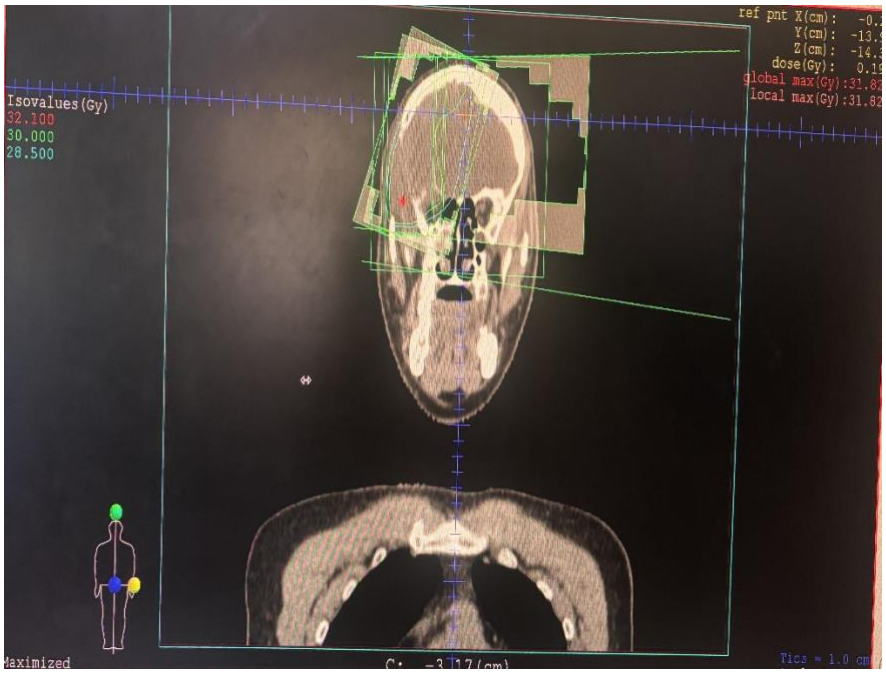


Figure 7:Dosimetric planning image in palliative cerebralradiotherapy.

Discussion:-

NAC is one of the nasopharyngealmalignancieswith a cellular structure characteristic of adenocarcinoma. It mayoriginatefrom the surface mucosalepithelium of the nasopharynx or from the submucosa (arisingfrom minor salivary glands). The World HealthOrganization (WHO) has classifiednasopharyngealadenocarcinomasintopapillaryadenocarcinomas, mucoepidermoidcarcinomas (MEC), adenoidcysticcarcinomas (ACC), and polymorphouslow-grade adenocarcinomas (PLGA) [7].
Recentstudiesclassify NAC intotwosubtypes:mucosal (surface-derived) and submucosal (from minor salivary glands). MucosalNACs are generallylow-grade, affect younger patients—oftenwomen—and have a favorable prognosiswhensurgicallymanaged. In contrast, submucosalNACs are high-grade, mainly affect older men, and have a poorerprognosis, requiring multimodal treatment [5].

The relationshipbetween the virus and nasopharyngealadenocarcinoma (NAC) remainsunclear. Somestudies have detected EBV in salivary gland-type NACs [8,9], whileothersfound no direct link [10]. Methods usedinclude EBV RNA in situ hybridization and PCR analysis of the LMP-1 gene. The results are inconsistent, showingonly partial presence of EBV and no clearrelationshipwith NAC carcinogenesis.

The male-to-female ratio in NAC has been reported as 1.18:1, compared to 2:1 in non-squamouscarcinomas (NSC) [17]. Gender distribution varies depending on histologicalsubtype. The medianage of patients isbetween 40 and 50 years. The slow progression of NAC couldexplain the long duration of symptomsbefore patients seekmedical attention. Schramm et al. reported an averagedelay of 19.3 monthsbeforediagnosis [13,15].

Cervical masses are rare in NAC due to the low rate of cervical lymphnodemetastasis. However, cranial neuropathies are more common in NAC than in NSC, especiallyinvolving the trigeminal nerve, which causes intense facial pain. Trigeminalneuralgiapresents as severe paroxysmal pain due to dysfunction of the fifthcranial nerve [3].

The staging system for nasopharyngealadenocarcinomas (NAC) shouldtakeintoaccountseveralprognosticfactors, such as cranial nerve invasion, skull base erosion, and the presence of positive cervical nodes. Schramm et al. [11] expresseddisagreementwith the ideathatmost patients withcranialneuropathyshouldbeclassified as stage T4 and thereforeproposed a new staging system for salivary gland-type malignancies of the nasopharynx.

The generallypoorerprognosis [12] mainlydepends on tissue type, histologic grade, and TNM staging. Due to the rarity of NAC, no standardizedtreatmentprotocolcurrentlyexists. In high-incidence endemicregions, especially in southern China, NAC treatmentoftenfollowsprotocolsused for non-squamouscarcinomas (NSC). Interestingly, NAC can bewell-controlledwithradiotherapy, with or withoutchemotherapy.

Radiotherapyplays a crucial role in the management of NAC, especially in inoperable patients. Wang et al. [13] reported 5-year survival rates of 86% and 10-year survival rates of 50% for patients treatedwithradiotherapy.

In the future, large-scalemulticenterstudiesshouldbeconducted to furtherevaluate NAC treatmentstrategies. In particular, high-dose radiotherapyusingprecise techniques mayberecommended.

The anatomical structure of the nasopharynx iscomplex. It issurrounded by manycritical tissues and organs, making total or near-total resectiontechnicallydifficult. Nasopharyngealsurgeryistechnicallydemanding [14]. Recentsurgicalapproaches, such as the endonasalendoscopic route [3], have shownpromisingresults, allowingcompleteresectionwithnegativemargins [16]. However, the complexanatomical location of the nasopharynx, near vital structures such as the carotidartery and skull base, presentssignificanttechnical challenges for surgeons.

There are no previous reports specifically on chemotherapy for nasopharyngealadenocarcinoma (NAC), but studies have investigatedchemotherapy for head and neck adenocarcinomas. Althoughthesetumorswere once consideredresistant to cytotoxicdrugs [17,18], severalstudies have confirmedtheirsensitivity to chemotherapy [19,20], particularlywith agents such as platinum compounds, 5-fluorouracil, and anthracyclines [17,20]. Gemcitabine, however, has not shownefficacy in salivary gland adenocarcinomas of the head and neck, according to the EORTC 24982 study [15].

Conclusion:-

In conclusion, although NAC is a rare form of cancer, multimodal management combiningradiotherapy, chemotherapy, and possiblyendoscopicsurgerycurrentlyrepresents the best therapeutic option. Nevertheless, large-scalemulticenterstudies are needed to establishstandardizedtreatmentprotocols and more preciselyevaluate the long-term impact of thesetherapies on patient survival.

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