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Case Report

Nasopharyngeal carcinoma in a young adolescent with unilateral palatal palsy and neck mass – A rare case report

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ABSTRACT

Less than 1% of all head and neck neoplasms are juvenile nasopharyngeal angiofibromas, a cancer almost exclusively found in adolescent males. Here we present a case report of 14 year old male presented with neck swelling, nasal discharge mixed with blood, right sided proptosis and right sided palatal palsy where we performed CECT scan of the patient which showed homogenous opacity over nasopharynx extending to soft palate and posteriorly to C1 vertebra and DNE was performed. Later patient was planned for DNE along with biopsy which came out to be undifferentiated non-keratinized squamous cell carcinoma grade III. Further patient received external beam intensity modulated radiation therapy with complete response to treatment.

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1. Introduction

Nasopharyngeal carcinoma (NPC) is a tumor of nasopharynx originating from its epithelial cell lining. It is a tumor of the head and neck region in adults, with varying incidences in different region of the world. High risk areas for NPC include places like southern China, Hong Kong, and Taiwan¹ while in India it is high in Nagaland, Mizoram and Manipur with incidence rates of 19.4, 6 and 5 per 100,000 person/year respectively.² Certain human leukocyte antigen (HLA) subtypes, smoking, eating salted seafood, and occupational exposures are risk factors for NPC.³ NPC in paediatric is different from its adult counterpart. It is rare for such tumor in paediatric age group with only about 3% of all NPC found in patients younger than 18 years.⁴ The head and neck region is where about 5% of primary malignant neoplasms in children develop.⁵ NPC, on the other hand, makes for about 2% of head and

neck malignant neoplasms in children.⁶ Paediatric NPC typically manifests as an advanced locoregional illness and is biologically more aggressive (WHO grade III).⁷

2. Case Presentation

A 14 year old male patient, without any previous medical history presented with his mother at PGIMS, Rohtak ENT outpatient clinic with right sided neck swelling for 5 months of duration as chief complaint along with nasal discharge mixed with blood, right sided proptosis and change in voice. On local examination of the patient he was found to have right side palatal palsy with right sided facial neuralgia. Right side jugulodigastric node was enlarged to around 4x3 cm in size. We undertook further examination by performing head and neck CECT scan which revealed homogenous opacity in nasopharyngeal area anteriorly pressing against hard and soft palate and posteriorly against the C1 vertebra, basisphenoid and occupying the posterior choana.

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Under the Karl Storz telescope, friable lobulated mass was noted over the posterolateral aspect of nasopharynx which was soft in consistency, reddish pink in color, doesn't bleed on touch. Diagnostic nasal endoscopy along with biopsy under local anaesthesia was performed which confirmed the diagnosis of nasopharyngeal carcinoma with histological report suggestive of undifferentiated non-keratinizing WHO grade III. H and E stain was used showing lymphocytes and tumor cells having moderate eosinophilic to amphophilic cytoplasm, round nuclei, prominent nucleoli and vesicular chromatin.



Figure 1: Clinical picture showing right sided palatal palsy



Figure 2: Computed tomography photo showing homogenous opacity anteriorly pressing against hard and soft palate and posteriorly to C1 vertebra

The patient underwent external beam intensity modulated radiation therapy with a preauricular field

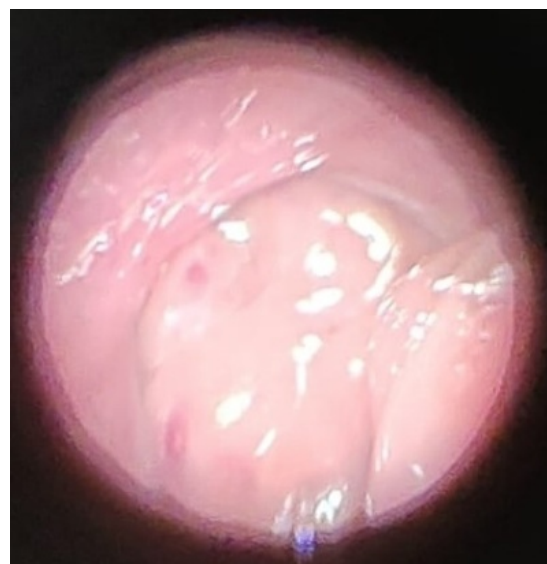


Figure 3: Diagnostic nasal endoscopic photo showing reddish pink, fleshy lobulated nasopharyngeal mass

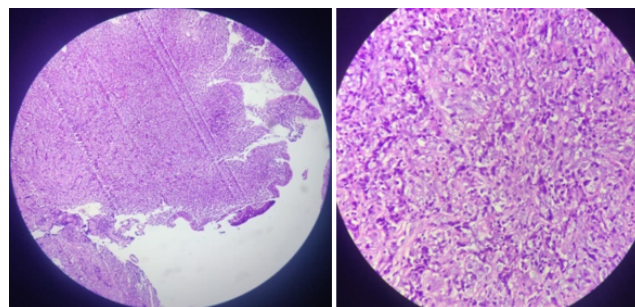


Figure 4: Histopathological photo showing undifferentiated non-keratinized squamous cell carcinoma.

to a total dose of 70 Gy concurrent with Cisplatin 100mg/m² three weekly followed by Cisplatin and 5FU (adjuvant). Evaluation was done on weekly basis during the course of radiotherapy and one month after the completion of radiotherapy and every 3 months for one year and then 6 monthly. The patient had a complete response to treatment without relapse with no late post-radiation complication.

3. Discussion

A typical malignant tumour found in the nasopharynx is nasopharyngeal carcinoma arising mostly from Fossa of Rosenmuller. The prevalence of NPC rises with age and peaks between the ages of 50 and 59, with a lesser peak occurring in late infancy.³

From endemic to non-endemic areas, paediatric NPC incidence varies. Around 0.5 per million children (ages 10 to 14) and 1.08 million teenagers (ages 15 to 17) are affected by NPC each year.⁸ NPC frequently manifests as otitis media, nasal congestion, and obstruction. Bilateral

cervical lymphadenopathies are frequently the initial illness symptom because of the extensive lymphatic outflow of the nasopharynx. The World Health Organisation (WHO) recognises three histological subtypes: Types of squamous cell carcinoma include I) keratinizing squamous cell carcinoma, II) non-keratinizing squamous cell carcinoma, and III) undifferentiated squamous cell carcinoma. The type III nasopharyngeal carcinoma has a high rate of distant metastasis, typically involving the bones, liver, and lung. It is strongly related with Epstein-Barr virus infection of the malignant cells. However, bone marrow metastasis is still an uncommon illness that can materially shorten patient survival times. Retrospective reviews of 23 patients with recurrent nasopharyngeal cancer were conducted by Zen et al. After receiving rigorous treatment for 2-4 months, bone marrow metastases was discovered in five of the individuals. Anaemia, leukopenia, thrombocytopenia, sepsis, soreness of the sternum, and fever were the clinical features of such metastasis. All five patients had severe local and regional illness, and their conditions rapidly deteriorated once bone marrow metastases was identified.⁹

The gold standard for initial diagnosis of NPC is with nasopharyngeal endoscopy with biopsy and radiological imaging to aid in the staging and extent of disease. CT and MRI are often performed with MRI being the preferred imaging modality.^{10,11}

Because NPC is radiosensitive, radiation has been the cornerstone of its management for many years.¹² The use of doses greater than 65 Gy in the past to control tumours has had severe short- and long-term consequences, including secondary cancers and growth abnormalities as well as stomatitis, dental caries, hypothyroidism and other endocrine abnormalities, xerostomia, and hearing loss (related to both radiotherapy and cisplatin treatment).¹³

4. Conclusion

In this case, the main course of treatment was intensity modulated radiation therapy with adjuvant chemotherapy and is widely used in the treatment of nasopharyngeal carcinoma. Paediatric NPC is rare and usually presents in advanced stage. However, it responds well to radiotherapy alone or with concomitant use of chemotherapy.

5. Source of Funding

None.

6. Conflict of Interest

None.

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