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EMBRYONAL RHABDOMYOSARCOMA ARISING FROM NASOPHARYNX : A RARE CASE REPORT .

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Abstract

Background - Nasopharyngeal cancer is realtively rare world wide especially in the Indian subcontinent .Among Nasopharyngeal malignancies ,nasopharyngeal Rhabdomyosarcoma is an uncommon soft tissue sarcoma , particularly prevalent in children. Rhabdomyosarcoma is a highly invasive tumor originating from mesenchymal tissue ,mainly affecting pediatric populations.

Case presentation - We report a case of Embryonal Rhabdomyosarcoma in a 8 yr old female presenting with chief complaints of nasal obstruction and snoring since last 15 days which was sudden in onset and progressive in nature.

Conclusion - This case underscores the importance of considering rare malignancies like embryonal rhabdomyosarcoma in the differential diagnosis of nasopharyngeal tumors, especially in pediatric patients . further research and reporting of such cases are essential for better understanding and management of this rare condition.

keywords - Nasopharyngeal rhabdomyosarcoma , embryonal , parameningeal , pediatric oncology , rare malignancy.

INTRODUCTION:

Nasaopharyngeal cancer is a rare malignancy in most part of the world and it is one of the most confusing, commonly misdiagnosed and poorly understood disease. In spite of high incidence of cancer of oral cavity and other parts of pharynx, nasopharyngeal cancer is uncommon in Indian subcontinent.(1)

Nasopharyngeal rhabdomyosarcoma is a common soft tissue sarcoma in children.

Rhabdomyosarcoma is a highly invasive malignant tumor arising from mesenchymal tissue and mainly affects children.. A small number of cases have been reported where the tumor arise from palate, nasopharynx or middle ear. Its presentation in nasopharynx is a rare entity and only 5 cases have been reported in the literature.(2)

One of the earliest reference of rhabdomyosarcoma of the nasopharynx is that given by Bergmann, bruns and mikulics (1904) who quoted a case, where the tumor has probably originated from the superior maxilla or the epipharynx.

Rhabdomyosarcoma are generally divided by meningeal and parameningeal sites.

Para-meningeal sites include- middle ear, nasal cavity, paranasal sinuses, nasopharynx and infratemporal fossa. Because of the risk of intracranial extension via the para-meningeal locations sinonasal rhabdomyosarcomas often have a poor prognosis.

Rhabdpmyosarcoma can be subdivided into 4 types-

1) Embryonal and embryonal botryoid

2) Alveolar

3) Pleomorphic

4) Mixed.

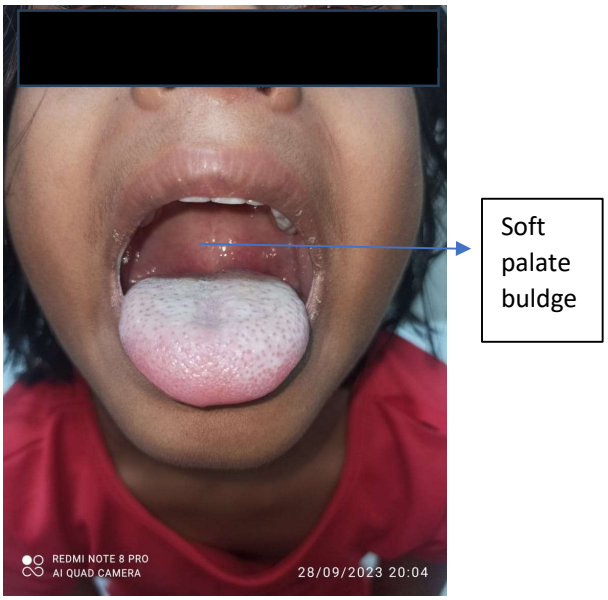
Embryonal is the most commonly encountered rhabdomyosarcoma of children.[3]

Here we are presenting a case of embryonal rhabdomyosarcoma arising from nasopharynx.

CASE REPORT:

A 8 year old female patient presented to ENT OPD with chief complaint of nasal obstruction and snoring since last 15 days which was sudden in onset and progressive in nature. Nasal obstruction was aggravated in the past 5 days which is aggravating on lying down and has no relieving factors. There is no history of fever, no history of weight loss and no history of significant nasal bleeding.

On examination of nose bilateral polypoidal mass was seen which doesn't bleed on touch and is soft in consistency. Patient had a soft palate bulge. On ear examination buldged out TM was seen in left ear.



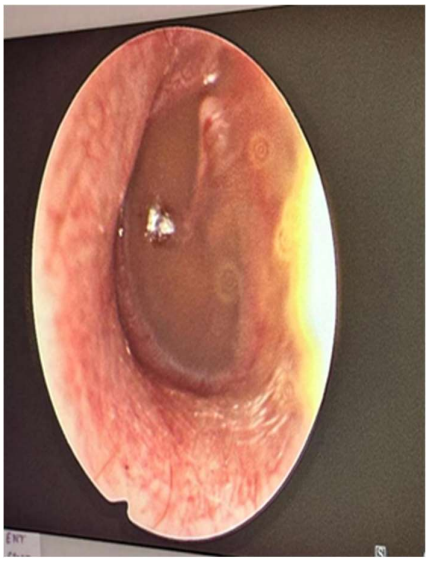
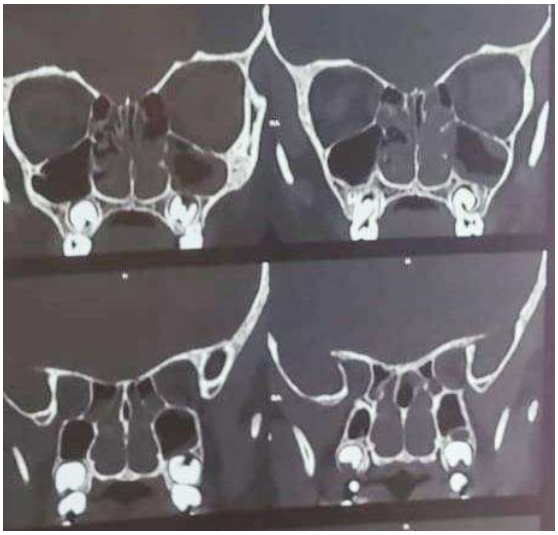


Fig 1- soft palate buldge **Fig 2- Buldged out TM**

On posterior rhinoscopy there is a pinkish mass hanging from nasopharynx into the oropharynx.

On CECT face and neck revealed a mass of around 54x45x26mm in left lateral pharyngeal wall causing near complete obliteration of nasopharyngeal and oropharyngeal airway. Anteriorly mass is seen indenting on soft palate, posteriorly extending upto nasopharyngeal wall, laterally into left parapharyngeal space. Mass is displacing left ICA and indenting upon the pterygoid muscles. There is widening of petroclival fissure. The mass is suggestive of neoplastic etiology likely schwannoma.



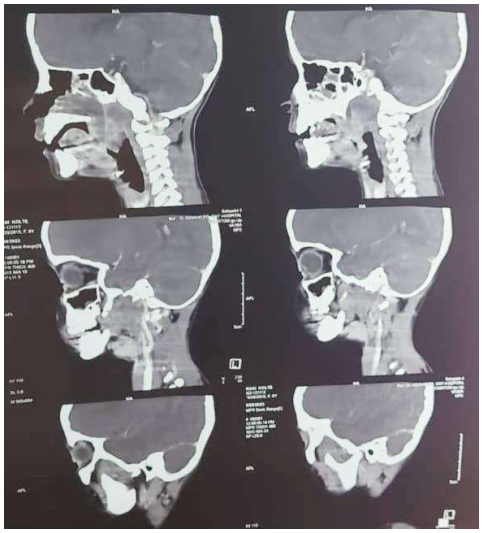


Fig3- coronal view of CT-PNS **Fig 4- saggital view of CT-PNS**

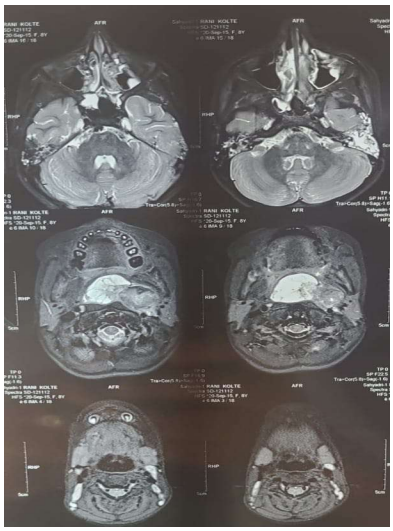


Fig 5- axial view of CT-PNS

Patient was posted for debulking immediately as difficulty in breathing increased. Grossly the tumor was white, soft to firm in consistency and friable. The tumor was around 10x5cm. The mass was removed in toto and send for frozen section: report showed: rhabdomyosarcoma or nasopharyngeal carcinoma. Left ear myringotomy was also done as the patient has severe ear pain due to left serous otitis media.

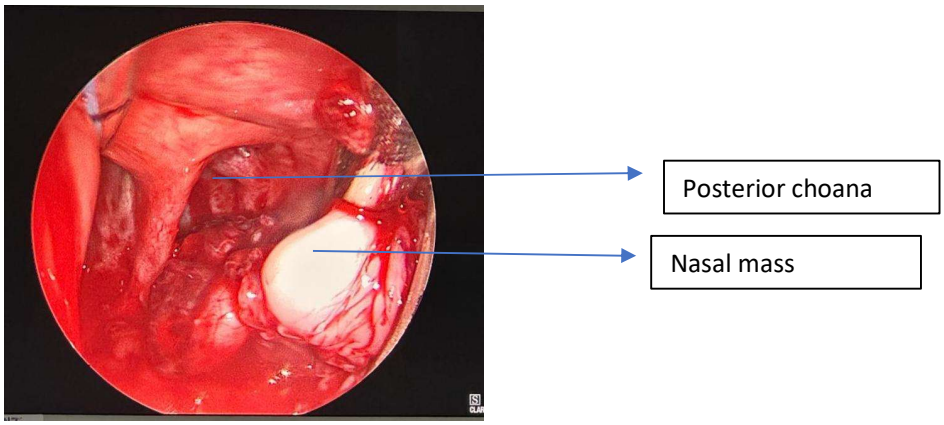


fig 6- endoscopic image of nasopharyngeal mass

The specimen was sentt for histopath: which showed most probable diagnosis to be embroyal nasopharyngeal rhabdomyosarcoma and was send for IHC for confirmation: which proved the diagnosis to be nasopharyngeal rhabdomyosarcoma and the patient was send for further management of radiotherapy and chemotherapy. And there is no recurrence till the time of reporting.

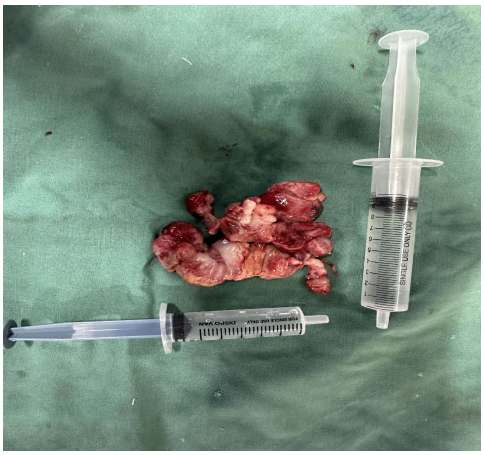


Fig 7- gross image of mass

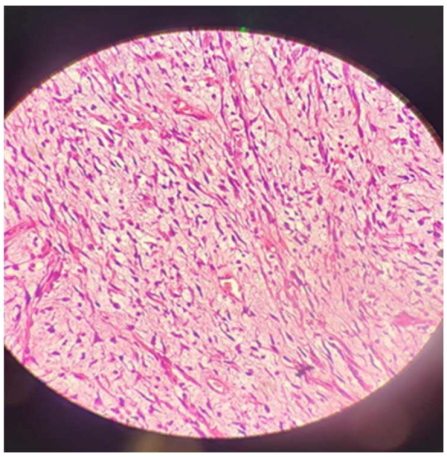


fig 8- Histopathological image of the mass

DISCUSSION:

Nasopharynx represents the most superior portion of the pharynx, bounded superiorly by skull base and inferiorly by the soft palate, anteriorly by nasal cavity, posteriorly posterior pharyngeal wall and laterally by medial pterygoid plates and superior constrictor muscle. The nasopharynx connects the nasal cavity to the oropharynx and contain Eustachian tube and adenoids.

Benign tumors of nasopharynx are rare and the commonest is angiofibroma .

Nasopharyngeal cancers comprises 2% of all malignant tumors of head and neck in children and adult below the age of 30yrs but its incidence in Chinese is high as 18% of all malignant tumors. It occurs more frequently than any other tumors of upper respiratory tract.

Soft tissue sarcoma accounts for 7% of cancers in children and 1% of cancers in adults. Approximately half of the population of pediatric patients with soft tissue sarcoma have rhabdomyosarcoma, which is a high grade, malignant neoplasm in which cancer cells have a propensity for myogenic differentiation.

The most common sites of rhabdomyosarcoma are head and neck, extremities and urogenital tract. The etiology of rhabdomyosarcoma remains unclear. Most cases of rhabdomyosarcoma re sporadic. However the disease is associated with familial syndromes.

In our case the patient presented with severe nasal obstruction and hearing loss, and there was bilateral polypoidal mass. The patient was immediately posted for endoscopic debulking as we suspected the mass could be malignant and there was alarming increase in breathing difficulty and soft palate buldge.

The whole mass was removed successfully in toto endoscopically and three mass was detached from all its attachments. The parapharyngeal space is also checked for any extension. We

observed that there was no intracranial extension and bony destruction. In post op the child has immediate relief of her symptoms.

The histopath was found to be embryonal rhabdomyosarcoma so the patient was further advised for radiotherapy and chemotherapy.

Unfortunately, rhabdomyosarcoma produces distant mets which is associated with poor survival. The effect of radiation and chemotherapy is debatable on sarcoma.

SUMMARY

Rhabdomyosarcoma usually runs a rapid course unless diagnosed and treated promptly, clinically it may be mistaken as polyp or angiofibroma. It has very poor prognosis and tend to reoccur. The tendency to lymph node metastasis is also more. They also tend to be less responsive to only radiotherapy and chemotherapy. So multi-modality treatment is the mainstay management of rhabdomyosarcoma. This includes surgical management along with radiotherapy and chemotherapy.

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