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PAEDIATRIC NASOPHARYNGEAL CARCINOMA: A RARE CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Nasopharyngeal carcinoma (NPC) is a rare malignancy and is one of the commonly misdiagnosed and poorly understood disease. It accounts for 1-5% of paediatric cancers and predominantly seen in adolescent male. Despite multi-factorial pathogenesis, strong association with Epstein Barr Virus has been found in genetically susceptible host. In younger patients more aggressive biological behaviour has been observed, although overall survival is better as compared to adults. As the condition is rare and clinical presentation is non-specific, diagnosis is often delayed and misinterpreted in paediatric patients. EBV-DNA levels are used to monitor the disease in these patients and also have a prognostic values also. Radiotherapy has been used as the main stay of treatment for early disease while concurrent chemo-radiotherapy has been demonstrated to prolong survival in loco-regionally advanced disease. There is paucity of data in Indian paediatric patients. We are presenting such case for its rarity.

INTRODUCTION

NPC is known to have well defined geographical distribution primarily affecting individuals from three different populations; viz. Chinese in South East Asia, Arabs in North Africa and Eskimos in Arctic [1, 2]. The incidence of NPC is low in most parts of the world with an age adjusted incidence of less than 1 per 100,000 people with a male preponderance [3]. In India, Nagaland has the highest incidence with 4.3 per 1,00,000 (4). The pattern of age distribution of NPC varies in different part of the world. Hirayama reported that the incidence in both the sexes starts rising after the age of 20-24 and reaches the plateau between 45-54 years of age [4]. Commune *et al* reported a younger age peak at 10-19 years in addition to the main peak at 50-59 years in Tunisia which is an area of intermediate risk for nasopharyngeal carcinoma [5]. Similar findings were seen in Uganda Kenya and Sudan suggesting that younger age group is affected in areas of intermediate incidence. In India no specific bimodal age peaks were observed in a study from North East region of India. In spite of differences among races and geographical distribution, NPC forms 1%-5% of all paediatric cancers and 20%-50% of all primary malignant nasopharyngeal tumors in children [6-8]. The occurrence of this tumour is rare in children with only 3% of cases occurring in patients with less than 18 years of age [9]. Paediatric NPC is an aggressive tumor with presentation mostly as advanced loco-regional disease. Certain HLA association like HLA-A2, HLA-B17, HLA-Bw26 and EBV has been shown to have increased risk of having nasopharyngeal carcinoma [10]. The detection of EBV nuclear antigen and viral DNA in nasopharyngeal carcinoma has revealed that EBV can infect epithelial cells and can be the reason for their malignant transformation. NPC is also found to be associated with consumption of salted fish which contain nitrosamines, a procarcinogen, recurrent nasal infections and poor hygiene.

CASE PRESENTATION

A 13 year old male child along with his father presented to the department of Otorhinolaryngology with complaints of multiple unilateral (left) neck swellings for last one year, decreased hearing from left ear from last 5 months and decreased mouth opening from last 1 month. Initially the swelling was single, painful and was confined to the left side of the neck which gradually increased in size and number. Patient also complained of pain and decreased hearing from left ear for last 5 months without any history of ear discharge. Patient had intermittent episodes of fever which used to get relieved on taking medications. There was no history of epistaxis. On examination of neck multiple lymph nodes were palpable with varying size, with the largest one being of the size of 3 X 2cm. Lymph nodes were non tender, firm to hard in consistency with restricted mobility and fixity to underlying deep structures. Rigid nasal endoscopy revealed lobulated mass in left nasopharynx arising from Fossa of Rosenmuller and involving superior, lateral and posterior wall of nasopharynx. Oral examination revealed grade II trismus with normal bilateral palatal movements. Otoscopy findings were suggestive of serous otitis media, confirmed with pure tone and impedance audiometry which revealed conductive hearing loss and B type curve respectively.

A Contrast Enhanced CT Scan was done which revealed an ill defined, lobulated, infiltrative mass with epicentre at Fossa of Rosen Muller with size of 51 X 59 X 41mm, predominantly on left side, moderate post contrast enhancement, infiltration of left masticator space with involvement of both medial and lateral pterygoid muscles, involvement of left infratemporal fossa, and destruction of pterygoid bones (figure 2). Chest X ray and ultrasound abdomen were done to rule out distant metastasis.

Endoscopic biopsy from the primary tumor as well as incisional biopsy of neck nodes were taken under general anaesthesia (figure 1). Histopathological examination of the tissue showed poorly differentiated carcinoma with prominent lymphoplasmocytic infiltrate (figure 3) . Staging of the tumour was done according to AJCC 2002 grading system and was found to be Stage IV (T4 N1 M0). Patient was planned for concomittent chemoradiotherapy because of advanced locoregional disease.

Fig 1. Showing cervical lymphadenoapthy with scar mark of incisional biopsy.

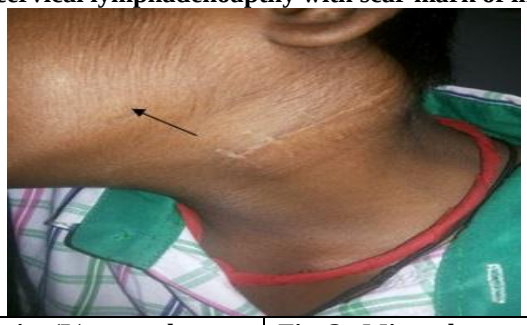


Fig 2. Axial scan showing growth in (L) nasopharynx with extension into pterygopalatine and infratemporal fossa.

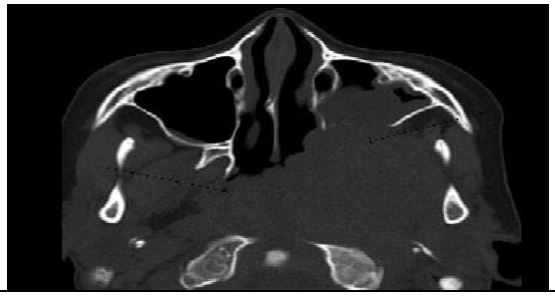
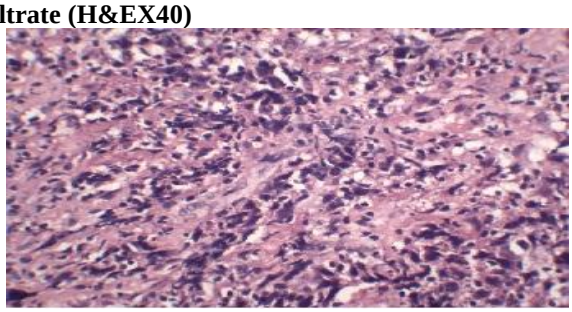


Fig 3. Microphotograph show groups of atypical cells with surrounding desmoplasia and lymphoplasmacytic infiltrate (H&EX40)



DISCUSSION

Nasopharyngeal carcinoma is a rare tumour in children that arises from the epithelial cells of the nasopharynx. Undifferentiated paediatric nasopharyngeal carcinoma is endemic tumour in Southern China, South East Asia, the Mediterranean and Alaska [1]. The age adjusted annual incidence rate varies with age, being 0.1 per million at age 0 to 9 years, 0.8 per million at age 10 to 14 years, 1 to 2 per million at age 15 to 19 years [11]. The median age at diagnosis is 15.3 years [12].In North America and Mediterranean region some places have bimodal age distribution. Paediatric nasopharyngeal carcinoma is more common in black male children. There has been found a strong association between the consumption of salt preserved fish containing nitrosamines and nasopharyngeal carcinoma risk [13]. Most common presentation of nasopharyngeal carcinoma in children is cervical lymphadenopathy which can be attributed to rich

network of lymphatics in the nasopharynx. In paediatric population, neck swelling can be a reactive lymphadenitis, any congenital lesion or a benign neoplasm, but a firm, non tender, neck mass in a child should be considered malignant unless proved otherwise [14]. Patients with nasopharyngeal carcinoma can also present with vague symptoms of influenza like viral respiratory infections, a feature very common in children thus leading to a delayed diagnosis in children. They can also present with trismus, hearing loss, symptom of otitis media as seen in the present case. On involvement of skull base patient can present with cranial nerve palsies [11]. Large tumours can present with epistaxis and loss of nasal tone of voice. Various differential diagnosis kept in mind were Nasopharyngeal rhabdomyosarcoma, Non Hodgkin's Lymphoma, Hodgkin's disease, Juvenile Angiofibroma, Germinal Rhabdomyoma and Haemangioma tumor [1].

WHO has classified nasopharyngeal carcinoma into three categories depending upon features of light microscopy [15].

Type I – Keratinising squamaous cell carcinoma with worst prognosis [10].

Type II- Non keratinizing carcinoma (least common) [10].

Type III- Undifferentiated carcinoma or lymphoepithelioma

Although Type II and III varieties are associated with distant metastasis but have better prognosis because of radio sensitivity. In children the most common histological variant is Type III (lymphoepithelioma) [10]. Distant metastasis is a poor prognostic indicator and usually appears in bones, lungs, mediastinum and less commonly in liver [11]. Optimum treatment for nasopharyngeal carcinoma in children is still not established; however the core treatment for nasopharyngeal carcinoma in paediatric patient in non metastatic disease is radiotherapy [16]. Undifferentiated variety of nasopharyngeal carcinoma is highly sensitive to radiotherapy, though high doses can lead to hypopitutarism and hypothyroidism [17]. Due to anatomical considerations and tendency to have cervical adenomegalies, paediatric patients are not good candidates for surgery. A recent treatment protocol from The Children oncology Group ARAR0331 uses radiotherapy alone for stages I and II using 61.2 Gy and 66.6 Gy respectively [18]. For advanced disease neoadjuvant chemotherapy with Cis-platin and 5 Fluoro-Uracil followed by radiotherapy [18]. 20-50 % of patients experience relapse or metastatic disease usually within 1-2 years of diagnosis [1]. Ozyar *et al* has reported event free survival of 68% at 5 year in a rare cancer network study involving 8 different countries [19].

CONCLUSION

Nasopharyngeal carcinoma is a very rare malignancy of the paediatric age group. It is diagnosed late in its natural course because of its rarity and overlapping of symptoms with many other similar non malignant and frequently occurring conditions in children. An early diagnosis depends upon high index of suspicion. With recent advances in treatment protocols including radiotherapy techniques there are good overall survival of the patients.

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CONFLICT OF INTEREST

No financial interest or any conflict of interest.

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