

JOURNAL OF CLINICAL AND DIAGNOSTIC RESEARCH

How to cite this article:

BHAT M,DEEPALAKSHMI, VAIDYANATHAN V.MALIGNANT NASAL POLYP IN A CHILD: A CASE REPORT.Journal of Clinical and Diagnostic Research [serial online] 2009 June [cited: 2009 June 1]; 3:1583-1586.

Available from

http://www.jcdr.net/back_issues.asp?issn=0973-709x&year=2009&month=June &volume=3&issue=3&page=1583-1586&id=373

CASE REPORT

Malignant Nasal Polyp in a Child: A Case Report

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ABSTRACT

A one year and two months old male child was brought to our hospital by his parents. He presented with a history of breathing through the mouth and snoring since two months. The clinical examination was suggestive of nasal polyp. Computerised tomography (CT) scan of the nose and paranasal sinuses (PNS) revealed soft tissue density in the left maxillary sinus extending into the left nasal cavity, widening the ostium and infundibulum soft tissue density in ethmoidal sinuses. Endoscopic resection of the mass was done after a histopathological diagnosis of nasal polyp. The patient (pt) presented with recurrence of symptoms, with breathing and feeding difficulty within 15 days. Clinically, a polypoidal mass was seen filling the left nasal cavity, extending into the oropharynx. A repeat endoscopic resection was done and histopathological examination of the mass revealed embryonal rhabdomyosarcoma (parameningeal type). The patient was treated with 32 cycles of chemotherapy and regular follow up was done for two years. The patient is asymptomatic at the end of two years.

Key Words: nasal polyp, rhabdomyosarcoma, embryonal, parameningeal type

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Introduction

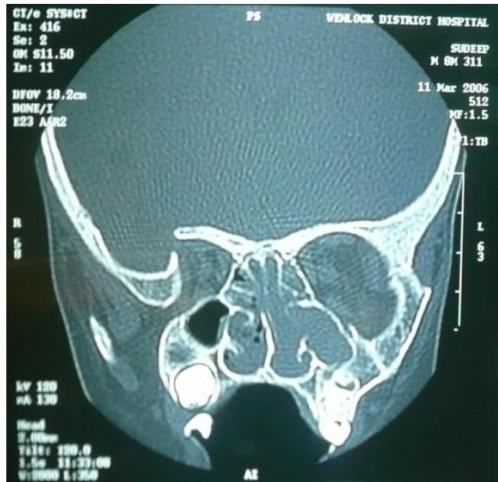
Rhabdomyosarcoma is a cancerous tumour that originates in the soft tissues of the body including muscles, tendons and connective tissues [1]. The most common sites of this tumour are head and neck, bladder, vagina, arms and legs. Embryonal rhabdomyosarcoma occurs in children under six years of age. Alveolar rhabdomyosarcoma occurs in older children. Rhabdomyosarcoma accounts for about 3%

of all childhood tumours [5]. It is the third most common tumour after neuroblastoma and wilm's tumour. The tumour is slightly more common in males than in females [8].

Case Report

A one year and two months old male child was brought to our hospital by his parents with a history of breathing through the mouth, snoring and nasal discharge since two months. On clinical examination, a polypoidal mass was seen filling the left nasal cavity, with mucopus in the middle meatus. The patient was admitted and evaluated and CT scan of the PNS (both coronal and axial scans) found soft tissue density in the left maxillary sinus extending into the nasal cavity, causing widening of the ostium and infundibulum and also soft

tissue density in the left anterior ethmoidal sinuses filling the nasal cavity [Table/Fig 1]. Endoscopic resection of the polypoidal mass was done in toto [Table/Fig 2].The histopathological confirmation of the clinical diagnosis of nasal polyp was made.



(Table/Fig 1) CT scan picture at initial presentation



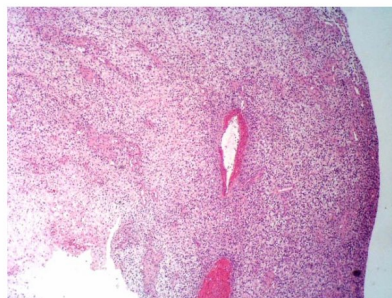
(Table/Fig 2) First resected specimen

feeding, with recurrence of the earlier symptoms. The Ear, Nose and Throat (ENT) examination was suggestive of a mass filling the left nasal cavity and extending into the oropharynx . Endoscopic resection of the mass was done, which was confined to the paranasal sinuses extending into the oropharynx, with no bone erosion or intracranial extension [Table/Fig 3].The histopathological examination was suggestive of embryonal rhabdomyosarcoma of the parameningeal type [Table/Fig 4]. A final diagnosis of embryonal rhabdomyosarcoma of the parameningeal type, stage I was made, based on the Tumour Node Metastasis (TNM) classification. The patient was thereafter treated on the basis of the Intergroup Rhabdomyosarcoma Study (IRS); IV protocol therapy [3]; Vincristine $1.5\text{mg}/\text{m}^2$ weekly, Actinomycin D $0.013\text{mg}/\text{kg}$ day 1 to day 5 and Cyclophosphamide $2.2\text{g}/\text{m}^2$ iv with Mesna every 21 days. A repeat scan after 10 cycles of chemotherapy showed no evidence of recurrence. The patient was followed up regularly for two years, but there was no evidence of recurrence.



(Table/Fig 3)Second resected specimen

After fifteen days, the child presented to the hospital with apnoeic episodes while



(Table/Fig 4) Histopathology of embryonal type of rhabdomyosarcoma

Discussion

Rhabdomyosarcoma comprises of 4% of all childhood tumours [6]. The embryonal type is more common than the alveolar type, and accounts for about 20% of all cases of rhabdomyosarcoma. The incidence of the embryonal type of rhabdomyosarcoma in nose and PNS is 5.4%. The disease is more common in older males and is unlikely in children < 2 years of age, but our patient was 1 year and 2 months old, which makes it an uncommon presentation [8]. A nasal mass resembling a polyp, is the most common presentation of rhabdomyosarcoma of the nose and PNS, as seen in our case, which lead to its early diagnosis. A child with a nasal mass should always be viewed with suspicion, as all swellings in the paediatric age group are not benign. The CT scan demonstrates poorly defined inhomogenous soft tissue masses destroying the adjacent bone [5], unlike the present case which had no obvious bone destruction. This could be because of its early detection. Histopathology revealed undifferentiated round or spindle cells, together with a few scattered eosinophilic rhabdomyoblasts in alternating cellular and myxoid areas. Some elongated rhabdomyoblasts showed angulation of muscle fibres (broken straw sign) which were characteristic of embryonal rhabdomyosarcoma [9]. [Table/Fig 4] Immunohistochemistry was positive for desmin and muscle actin. Myo D₁ and myogenin are useful markers [10]. Stage III and IV tumours are highly aggressive and locally invasive, but in our case, due

to early detection of the tumour in stage I, it did not show any evidence of invasion [2].

Patients Can Be Divided Into:

Low risk [4].

Favourable histology (embryonal)

Stage I and II

Age ranging from 1 to 10 years

Favourable sites are orbit, head and neck including infratemporal fossa.

High Risk [4].

Unfavourable histology (alveolar)

Stage III and IV

Age greater than 10 years

Unfavourable sites as parameningeal extraperitoneal sites, especially those with alveolar histology.

Even though our patient belonged to Stage I, we grouped it under high risk, as the ethmoid sinus was involved with parameningeal extension, with impeding intracranial complication.

Multimodality treatment is advocated for rhabdomyosarcoma, which includes surgical excision and post op chemotherapy. Post op conventional fractionation radiation therapy is advised for advanced cases. The estimated 5 year overall actuarial survival rate ranged from 74% to 79% in various studies [7].

Conclusion

Among rhabdomyosarcomas, embryonal rhabdomyosarcoma of the parameningeal type in the head and neck belongs to the high risk group. Histopathological examination plays a very important role in conclusive diagnosis. Early presentation of head and neck lesions leads to early diagnosis, complete surgical resection and chemotherapy, which helps to reduce mortality and prevent recurrence. Thorough clinical assessment and a suspicious approach to a paediatric nasal mass is the key to diagnosis. This case was reported as the patient had an early diagnosis, prompt surgical intervention

and chemotherapy and had no recurrence at the end of 2 years of follow up.

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