Rhabdomyosarcoma of nose, nasopharynx and paranasal sinuses

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Abstract Rhabdomyosarcoma is an aggressive malignant soft tissue tumor that arises from primitive striated muscle cells called rhabdomyoblasts. It is the most common soft tissue malignant tumor in children and accounts for about 7–8% of childhood cancers. It can arise from almost anywhere in the body but it is most common in the head and neck region. This article presents six cases of rhabdomyosarcoma involving the nose, nasopharynx and the paranasal sinuses four of which were treated with a combination of radiotherapy and chemotherapy and two were treated with medial maxillectomy combined with chemotherapy and radiotherapy.

Keywords Rhabdomyosarcoma · Nose · Nasopharynx · Paranasal sinuses

Introduction

Rhabdomyosarcoma is a malignant tumor with skeletal muscle differentiation. It predominantly affects children and accounts for 5–8% of pediatric tumors. In adults it usually affects the extremities, mostly sparing the head and neck. This article presents six cases of rhabdomyosarcoma involving the nose, nasopharynx and the paranasal sinuses, treated in the Department of ENT, Medical College Hospital, Kolkata, West Bengal, India in the period between June 1999 and February 2002.

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Case reports

Six cases of rhabdomyosarcoma are reported in this article. Their ages ranged from 5 to 35 years. Two patients were females and four were males. Table 1 shows that the most common age group for rhabdomyosarcoma is before the second decade (83.3%). A male preponderance was noted in our series (66.6%). Tables 2 and 3 show the clinical presentations of the patients. Nasal obstruction, nasal discharge and cheek swelling were the symptoms common among all the patients. Upper jaw or maxillary swelling and a nasal mass were the commonest signs. As all our cases were primarily maxillary tumors, the radiology (Table 4) revealed haziness of maxillary antrum or erosion of the bony

Table 1 Age-wise distribution of cases

Age (years)	Male	Female	No. of cases
0–10	1	1	2
11–20	2	1	3
21–30	–	–	–
31–40	1	–	1
Total	4	2	6

Table 2 Presenting clinical symptoms

Symptoms	No. of cases
Nasal obstruction	6
Nasal discharge	6
Epistaxis	4
Swelling of cheeks	6
Blurring of vision	3
Ear ache	1

Table 3 Presenting clinical signs

Signs	No. of cases
Nasal mass	6
Upper jaw swelling	4
Palatal bulge	2
Proptosis	3
Cranial nerve palsy	0
Palpable neck nodes	2
Signs of distant metastasis	0

Table 4 Radiological finding

Radiological findings	No. of cases
X-ray PNS	
Haziness of maxillary antrum	6
Occiputo-mental view	
Erosion of bony maxillary wall	4
CT scan findings:	
Involvement of maxillary antrum only	2
Involvement of maxillary antrum, nasal cavity anterior ethmoid and orbit	4
Involvement of all the paranasal sinuses, including the nasal cavity and nasopharynx	1

wall of the maxilla. Histopathology revealed the embryonal variety to be the commonest type.

Four patients were treated with combined radiotherapy and chemotherapy. Out of these three are still surviving with no recurrence. The lone patient with alveolar type was lost on follow-up. Two patients were treated with surgery that is medial maxillectomy with curettage from the remaining portion of the maxillary antrum, followed by chemotherapy and radiotherapy. Both survived for 5 years with no recurrence.

Discussion

Rhabdomyosarcoma arises from primitive undifferentiated mesenchymal tissue. Accounting for 5–8% of pediatric tumors, rhabdomyosarcoma is the third most common pediatric extracranial solid tumor, following neuroblastoma and Wilms tumor [1]. Rhabdomyosarcoma is the most common malignancy to arise within the nasal cavity and paranasal sinuses of children [2].

In childhood, there is a bimodal distribution pattern with

Table 5 Histopathological types

Embryonal	5
Alveolar	1
Pleomorphic	0
Botryoid	0

one peak occurring during the first decade of life and the other peak during adolescence. Dito and Batsakis reviewed 170 cases and found 77.7% patients to be under 12 years of age [3]. Mason and Soule (1965) reviewed 88 cases of rhabdomyosarcoma of head and neck and found 72% of the tumors to be occurring in children [4]. Its incidence in adults in the head and neck in the region is uncommon. Alborus-Saavedra et al. in a study of 35 cases could find only two cases in the fifth decade [5]. In the present study, most of the cases were below 20 years and only one patient was aged in the fourth decade.

In terms of site of involvement most of the available literature has shown the commonest site to be the orbit. Dito and Batsakis in their series found that one-third of the cases had orbital involvement and in fewer than 2% cases only was the maxillary antrum involved [3]. Alborus-Saavedra found only four cases involving the maxillary antrum out of the 35 cases in their series [5].

There are no specific signs or symptoms helping in early diagnosis and treatment of rhabdomyosarcoma. Early tumors typically produce localized nasal symptoms. Rhabdomyosarcoma is no exception; nasal obstruction, rhinorrhea, and recurrent otitis media are the most common presenting symptoms [6]. Persistent pain, sero-sanguineous discharge or cranial nerve palsies eventually indicate the serious cause of the symptoms, the tumor often attaining a fairly large size at initial diagnosis. In the present series most of the cases presented in an advanced stage.

Composed of small, round, blue cells, rhabdomyosarcoma (RMS) resembles fetal skeletal muscle before innervation. Definitive diagnosis of this poorly differentiated tumor requires combined use of histologic analysis, transmission electron microscopy, and assays for muscle specific transcription factors like the MyoD family [7]. The presence of sarcomeres, actin, myosin, and thick filaments lined by ribosomes on electron microscopy are diagnostic for RMS [8].

The Intergroup Rhabdomyosarcoma Studies (IRS) Committee and World Health Organization recognize the embryonal (with botryoid variant), alveolar and pleomorphic subtypes of RMS [9,10]. Accounting for 60% of all cases, embryonal rhabdomyosarcoma is the most frequent form to originate in the head and neck. In our case series also we found out over 80% of cases belong to embryonal variety (Table 5).

On microscopic examination, the embryonal type shows:

- Marked cellularity containing many undifferentiated mesenchymal cells
- Focal areas of myxomatous changes
- Elongated strap shaped cells or small rounded “tadpole” cells with abundant eosinophilic cytoplasm
- Cells containing iron striations.

The alveolar type shows:

- Small round rhabdomyoblasts arranged in nests or cords separated by connective tissue trabeculae.
- Focal areas of alveolar architecture.

Tumors from mucosal surfaces are almost exclusively of the alveolar subtype. The botryoid variety is actually an embryonal tumor arising in the submucosal location giving it a grossly grape-like appearance. In most of the reported series the embryonal variant was the commonest type of rhabdomyosarcoma arising in the nose, nasopharynx and paranasal sinuses. Younger children tend to have the embryonal type of rhabdomyosarcoma; while adolescents experience the alveolar type of rhabdomyosarcoma. In the series of Sutton and Montague of the 30 cases in the head and neck region, 20 were of the embryonal variant, 10 were of the alveolar variety and none were of the pleomorphic type [11]. In the series of Fu and Perzin in their series of 16 cases involving the nose, paranasal sinuses and nasopharynx, 10 tumors had a predominantly embryonal pattern, five showed features of botryoid sarcoma and only one showed an alveolar architecture [12]. In the present study of the six cases, five were of the embryonal variant and only one case showed features of the alveolar type.

The treatment of any malignant tumor requires excision with a wide margin of surrounding normal tissue, with or without chemotherapy and or radiotherapy. In the case of rhabdomyosarcoma of the head and neck the region it is extremely difficult to achieve complete microscopic excision. Andersen et al. (1990) reported that the complete microscopic excision was achieved in only four out of 60 cases. Unless there is a good chance of achieving clear surgical margins with minimal morbidity it was best to err on the side of caution and aim only to do a surgical biopsy of the primary tumor at the outset. In the present series most of the tumors presented at an advanced stage and so were treated with a combination of chemotherapy and radiotherapy, except in two cases involving the maxillary antrum where the mass was removed by medial maxillectomy, followed by chemotherapy and radiotherapy. Current protocols use chemotherapy with radiation therapy. Vincristine, actinomycin D, cyclophosphamide and adriamycin are the most commonly used agents [13].

Conclusion

Rhabdomyosarcoma of the nose and paranasal sinuses is an important malignant tumor affecting different age groups. It is an uncommon though not a rare tumor. The embryonal variant is the commonest according to most of the reported studies and our conclusions are no different. It is an aggressively malignant tumor and frequently presents at an advanced stage. However timely and judicious treatment ensures good results with increased survivability. Surgery should not be delayed where the tumor has been assessed to be surgically resectable. In our series both the patients who underwent surgery had good results. However, considering the fact that most of the tumors of this class in the head and neck region are non-resectable, the mainstay of treatment is a combination of radiotherapy and chemotherapy.

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