Case Report:

Embryonal Rhabdomyosarcoma

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Abstract:
The rhabdomyosarcoma is a sarcoma of soft parts which is more common in children. It is considered a malignant neoplasm, derivative of the striated skeletal muscle and which can originate in any part of the body. In the head and neck region orbit, cranial base, nasal cavity and nasopharynx are most common site of origin. We describe uncommon presentation of the embryonal rhabdomyosarcoma in three pediatric patient of age 4 to 9 years male having lesion in nasopharynx, middle ear and orbit respectively. First patient initially presented with mass in both nostril as well in oral cavity and second patient presented with unilateral facial palsy, mass in right external auditory canal, nasal regurgitation with change in voice and third patient presented with unilateral proptosis of right eye. First patient was operated 3 times due to local recurrence and later received chemotherapy and second and third child received chemotherapy and radiotherapy. Histopathological evaluation of biopsy found to be embryonal rhabdomyosarcoma.

Keywords: Facial paralysis; Rhabdomyosarcoma; Malignancy, Chemotherapy

Introduction:
Rhabdomyosarcoma is an aggressive tumour accounting for 5-8% of all pediatric malignancies and over 50% of all soft tissue sarcomas in children [1-5]. It is considered a malignant neoplasia derived from striated muscle arising in any part of the body; however, it is more common in regions of the head and neck, genitourinary tract, retroperitoneum and extremities [3-5]. Approximately 30% of all pediatric rhabdomyosarcoma occur in the head and neck region and common sites of origin are the orbits, base of the skull, nasal cavity and nasopharynx. In nasopharynx usually it involves the middle ear; paranasal sinus and masticatory space [5]. Rhabdomyosarcoma are classified based on histopathological features, which have distinct clinical characteristics out of which embryonal is the most common type in infants and young children and other are alveolar and anaplastic variants (Pleomorphic variants) [6]. The purpose of this study is to report three case of nasopharyngeal, middle ear and temporal bone and orbital embryonal rhabdomyosarcoma associated with distinct clinical features.

Case series:
In our case series of three patients who were diagnosed and treated for rhabdomyosarcoma between 2009 to 2012. The age of patient ranged from 4 to 9 years. Out of these first patient had fleshy mass in both nostrils (fig:1) and difficulty in breathing since 2 months, similar mass in oral cavity and second patient presented with unilateral facial palsy, mass in right external auditory canal, nasal regurgitation with change in voice and third patient presented with unilateral proptosis of right eye. First patient was operated 3 times due to local recurrence and later received chemotherapy and second and third child received chemotherapy and radiotherapy. Histopathological evaluation of biopsy found to be embryonal rhabdomyosarcoma.
histological diagnosis of Embryonal Rhabdomyosarcoma was made (fig4). Hence cases diagnosed as embryonal rhabdomyosarcoma of nasopharynx, middle ear and temporal bone and orbit respectively. Extension confirmed with computer tomographic scan. Out of three patient first patient underwent surgery as well as chemoradiotherapy and had recurrence twice during the course of treatment and for which debulking surgery was done. Other two patients solely treated with four to eight cycles of chemotherapy consisting of Vincristine (1.4 mg/m²), Cyclophosphamide (750mg/m²) and Actinomycin D (50mg/m²) and localized radiotherapy (5000 rads) after 6 weeks of chemotherapy.

### Table 1:

<table>
<thead>
<tr>
<th>Age</th>
<th>Sex</th>
<th>Site</th>
<th>Type (Histopathology)</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>4 yrs</td>
<td>male</td>
<td>nasopharynx</td>
<td>Embryonal rhabdomyosarcoma</td>
<td>Nasal mass, oral mass, dysphagia and dyspnoea</td>
</tr>
<tr>
<td>7 yrs</td>
<td>male</td>
<td>Middle ear and temporal bone</td>
<td>Embryonal rhabdomyosarcoma</td>
<td>Right external auditory canal mass, right lower motor neuron facial palsy, Change in voice, and nasal regurgitation</td>
</tr>
<tr>
<td>9 yrs</td>
<td>male</td>
<td>orbit</td>
<td>Embryonal rhabdomyosarcoma</td>
<td>Right eye proptosis, diminish of vision</td>
</tr>
</tbody>
</table>

### Discussion:

Three histopathological variants of rhabdomyosarcoma have been described: embryonal, alveolar and anaplastic (pleomorphic [7]. Botryoids and spindle cell variants are subtype of embryonal rhabdomyosarcoma. It is believed that the embryonal, alveolar and botryoides variants are of mesenchymal origin and that the pleomorphic variant is derived from mature skeletal muscle predominantly occur in adults. Both the botryoid and the spindle cell subtypes are associated with very favourable outcomes [7,8]. The alveolar and pleomorphic type has been shown to carry the worst prognosis. Embryonal is the most common histologic type of rhabdomyosarcomas. It is responsible for 70 to 80% of the presentations and its origin is supposed to be triggered by a disorder in mesenchymal primitive differentiation of the musculoskeletal cells in their first or earliest stages of embryogenesis [3,9]. Rhabdomyosarcoma may have their origin in any anatomical localization occurring predominantly in head and neck regions orbits, skull base, nasal cavity, and nasopharynx where there is little or no musculoskeletal tissue [1]. In pediatric cases about 30 to 40% of rhabdomyosarcoma occur in the head and neck regions (10), however the ear and the temporal bone are uncommon sites of involvement [9]. Middle ear rhabdomyosarcomas were reported in the literature with a range of symptoms that clinically simulates chronic otitis media. The most commons presentation are facial nerve palsy, headache, hearing impairment, and bleeding [5]. However, the findings of facial palsy with involvement of nerve roots and lesions of the perineural space and parameningeal spaces are more suggestive of malignancy. In rhabdomyosarcoma involving orbit proptosis is most common presenting sign. Jones et al., have reported that 100% of the cases in their study had
proptosis. [10] However, Schinter et al., have documented 71% of cases with proptosis. [7,8. 11]. Embryonal rhabdomyosarcoma occurs most frequently, accounting for about 71% and 67% cases of orbital rhabdomyosarcoma. [12,13]. Earlier orbital rhabdomyosarcoma was treated by orbital exenteration. In 1979, Abraham et al. Demonstrated irradiation alone or in combination with chemotherapy to be more effective than exenteration for both control and long-term survival. [14] Reports of the efficacy of combined radiotherapy and chemotherapy were confirmed by the Intergroup Rhabdomyosarcoma Study, which showed a three-year survival rate of 93% in a total of 127 patients with localized orbital rhabdomyosarcoma. [15] Radiotherapy and chemotherapy have also been enlisted in treatment of local recurrences and metastatic disease.

In head and neck region embryonal rhabdomyosarcoma of nasopharynx is second most frequent situation orbit being first other site in order of frequency are cheek, external ear, maxillary antrum and retrotonsilar area. In study of Fu & Perzini age at presentation is about 7 years in 16 cases of rhabdomyosarcoma involving nasal cavity or nasopharynx and age ranges from 10 to 29 years out of which 12 patient in first decade and average age was 7 years. Most series suggest slight preponderance to male. Most neoplasm in this category consists of loosely arranged primitive cells situated beneath the epithelium of nasopharynx. Gross appearance 5 out of 16 cases of Fu & Perzini composed of multiple grape like polypoidal masses. Enlarged lymphonodes are more likely associated with distant metastasis [5,9].Metastasis can be present in up to 30% of the cases. The most common affected sites are the lungs, liver, bones and extremities [5]. Rhabdomyosarcomas affecting the perineural and parameningeal spaces, nasopharynx, paranasal sinuses and temporal bone generally are not amenable to surgical resection. In these areas, there is increased risk of serious damage to cranial nerves and deformation of the facial skeleton by radical surgery and it is unlikely to do complete resection. The treatment of choice for these tumors is chemotherapy and radiotherapy, with surgery limited to diagnostic biopsy [9,16]. Plexiform neurofibromas, juvenile angiofibroma, non-Hodgkin lymphoma, nasopharyngeal carcinoma, histiocytosis and cholesteatoma must be included among the differential diagnosis [18]. The diagnosis of middle ear and the temporal bone rhabdomyosarcoma is difficult when associated with inflammatory-infectious diseases, such as exudative otitis media that may mask the base disease and delay the definite diagnosis. In these cases, early correlation with imaging screenings is essential. In our case series of embryonal rhabdomyosarcoma of three different site nasopharynx ,middle ear and temporal bone and orbit only patient with nasopharyngeal embryonal rhabdomyosarcoma initially treated with debulking surgery trans-orally two times 9 month apart to alleviate complaint of dysphagia during course of chemoradiotherapy but patient succumb during course of follow up. Next two patient of embryonal rhabdomyosarcoma of middle ear and temporal bone and orbit solely received VAC regimen and subsequent local Localized Radiotherapy (5000 rads) after 6 weeks of Chemotherapy and patient in 2 year follow up neither had local recurrence nor distant metastasis.

References:
