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Case Report

Embryonal Rhabdomyosarcoma: A Rare Case Report

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ABSTRACT

Rhabdomyosarcoma (RMS) is an aggressive malignant neoplasm of skeletal muscle origin that represents 50% of all soft tissue sarcomas in childhood. It is the most common soft tissue sarcoma of the children, adolescents and young adults. About 35% of RMS arises in the head and neck. The other sites of occurrence are the genitourinary tract, retroperitoneum and to a lesser extent, the extremities. In the head and neck region, the most commonly affected sites are the orbit, paranasal sinuses, soft tissues of the cheek, and the neck. Here we report a case of a 16-year old male suffering from embryonal type of RMS in the pharynx and maxillary sinus and posterior part of oral cavity.
Keywords: Rhabdomyosarcoma (RMS), embryonal, aggressive malignant neoplasm

INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant, soft tissue neoplasm consisting of cells derived from the primitive mesenchyme that exhibit a profound tendency to myogenesis.[1] Rhabdomyosarcoma may be defined as a malignant tumor of the rhabdomyoblasts with a microscopic picture simulating that of striated muscle cells. [2] It was first described by Weber in 1854. [3] The most common sites of occurrence are the head and neck (40%), genitourinary tract (25%), and extremities (20%).[4]

Rhabdomyosarcoma are anatomically divided into 2 categories: parameningeal and nonparameningeal. [4] Parameningeal includes RMS of the nose, nasopharynx, paranasal sinuses, middle ear, mastoid, infratemporal fossa, and pterygopalatine fossa and nonparameningeal includes RMS of the scalp, orbit, parotid gland, oral cavity, oropharynx, and larynx. [3]

Histologically, the RMSs can be divided in embryonal, alveolar and pleomorphic. The embryonal type presents the subtypes: classic, spindle cell and botryoid. [5] The embryonal type represents more than 70% of all cases. [6] The embryonal RMS contains a mixture of spindle and undifferentiated round cells and immature striated muscle like cells (rhabdomyoblasts) with abundant eosinophilic cytoplasm either tightly or loosely packed in a myxoid stroma. The botryoid tumor is a morphologic variety or subtype of the embryonal sort and since there is gross resemblance to cluster of grapes, has been named botryoid. Histologically a multilayered band of cells just below the surface epithelium called the cambium layer is a special finding of this variant. [4]

The treatment of RMS of the head and neck has still some controversy. In the past, the prognosis of RMSs of the head and neck was very poor. However, with the introduction of multidrug chemotherapy, the survival in this group of patients has improved. Afterwards better survival has been obtained with combination of multidrug chemotherapy, external beam radiotherapy, and/or surgery. But still the treatment and survival of these patients remains a challenge. [5]

In this report, we present a case of embryonal Rhabdomyosarcoma in a 16 year old male, and demonstrate the clinical, radiological, histological, and imaginological features of this neoplasm.

CASE REPORT

A 16 year old male reported to us with a chief complaint of painless swelling over right side of face since 1.5 months. The swelling was initially small size and quickly increased (progressive growth). He also had recurrent coryza and decreased hearing capacity from right ear. Patient also complained of difficulty in deglutition and increased snoring. He had history of weight loss, anorexia and generalized weakness. There was no history of any trauma or any systemic disease. There was also no history of any bleeding or pus discharge.

A single right submandibular lymph node was palpable, firm, mobile and non-tender.

A paranasal sinus radiograph (water's view) was taken which showed ground glass haziness in the right maxillary and ethmoidal sinus suggestive of a soft tissue mass like polyp or mucocele in right maxillary sinus. The nasal septum was also deviated towards left side.

The Computed Tomography (CT) scan revealed a heterogeneously enhancing mass of size 4.6 x 5.2 x 9.5 cms in the right nasal cavity. The mass extended into the right pterygopalatine fossa, right ethmoid, maxillary and sphenoid sinuses and through the right choana into the nasopharynx and oropharynx. The findings were suggestive of juvenile nasopharyngeal angiofibroma.

Ultrasonography of neck region showed multiple enlarged hypoechoic lymph nodes in the right submandibular lymph node region and FNAC of the nodes revealed adenocarcinomatous metastasis.

The lesion was subjected for histopathological examination via incisional biopsy (Fig. 1). Histopathological analysis of the hematoxylin/eosin-stained material created an impression of round cell tumour. The histogenesis was also suggestive of non-Hodgkin's lymphoma and undifferentiated carcinoma of nasopharynx. Since no definitive diagnosis was obtained, immunohistochemical staining was done which finally gave the diagnosis as embryonal rhabdomyosarcoma.

Then the treatment was planned as per the stage III clinical staging (T3N1M0). A multi-drug, multi-cycle chemotherapy regimen was planned with a fair prognosis.

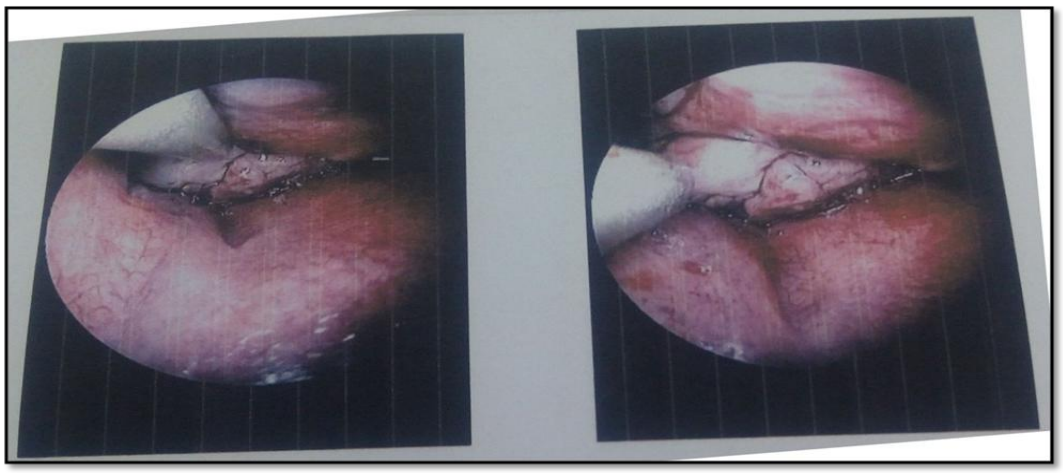

Figure 1. Endoscopic Biops

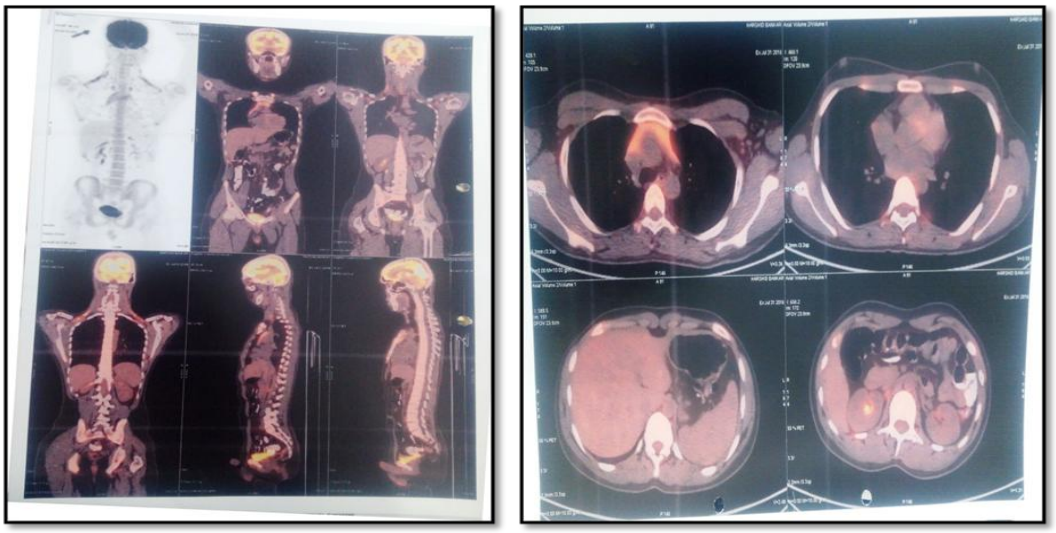

Figure 2. Post-operative PET-CT scan showing no metabolically active disease

After complete treatment again a PET-CT scan (Fig. 2) was performed using 18F Fluoro Deoxy Glucose. It confirmed that there was no metabolically active disease in the treated site. There was also no lymphadenopathy or distant metastasis seen. The patient was then kept on regular follow-up.

Unfortunately after 1 year there was recurrence of the disease with distant metastasis and the patient expired.

DISCUSSION

Rhabdomyosarcoma is the third most common extracranial malignant tumor in children after neuroblastoma and Wilms tumor. [4] The incidence of RMS is highest in children aged 1–4 years, lower in children aged 10–14 years and lowest in those aged 15–19 years.[3] There is found to be a male predilection with 58-73% of cases reported being males. [7,8] Majority of the cases occur in the first two decades of life. Our patient was 16 years old male and did not fall in the very high-risk category. [4]

In head and neck region the RMSs are divided in parameningeal and nonparameningeal as per the anatomy. [5] In general, the patients with oral RMS often have an advanced lesion at the early stage, because of a rapidly enlarging painless mass. [4] In our patient the lesion was of parameningeal type involving nasopharynx, paranasal sinuses and pterygopalatine fossa. However exact primary site could not be determined due to the large extension of the lesion.

Histologically, the embryonal variety shows a mixture of spindle and un-differentiated round cells and immature striated muscle-like cells (rhabdomyoblasts) with abundant eosinophilic cytoplasm either tightly or loosely packed in a myxoid background. In the differential diagnosis with embryonal RMS, the main tumours to be considered are in the category of the so-called '*small round blue cell tumors*'. [1] This was the same in our patient where the histopathological diagnosis was not confirmatory between round cell tumour, non-Hodgkin's lymphoma and carcinoma.

Diagnosis is more accurate with newer techniques such as cytogenetic and immunohistochemical analysis. Histological classification has prognostic significance. For instance, the botryoid and spindle cell variants have a favourable prognosis, while embryonal rhabdomyosarcomas tend to have a better prognosis than alveolar rhabdomyosarcoma. It is therefore important to classify these tumours correctly. [9]
Hence immunohistochemistry was done and the case was found out to be embryonal type of rhabdomyosarcoma.

For the treatment of RMS, intensive doses of chemotherapy in multi-drugs and multi-cycle regimens are used, so that

survival rates of children that develop head and neck RMS can increase. [6] Our patient was treated with vincristine, etoposide and ifosfamide.

The Intergroup Rhabdomyosarcoma Study (IRS) classifies RMS into four distinctive groups according to biological behavior. Group I tumors have higher prognosis whereas groups III and IV have the worst. Studies show that around 50% of RMS patients die following chemotherapy or one year after emission of histopathological diagnosis. [10,11] As a result of their aggressive neoplastic behavior characterized by immature and highly invasive cells, RMS are associated with high rates of recurrence and generalized metastases through the hematogenic and/or lymphatic routes. [5] In our patient also there was recurrence and distant metastasis after one year and unfortunately the patient expired.

Although a marked improvement of the survival rates has been reported in patients with RMS in recent years, a poor prognosis in patients >10 years old, delay diagnosis, and lack of cooperation from the patient and the family are reported to have a negative impact on the prognosis. [3]

CONCLUSION

Dental practitioners frequently encounter cases of children with facial swelling and pain; in most of these cases, dental abscess is considered as the cause and treatment is prescribed accordingly. Taken together, we conclude that in children, any swelling should be carefully examined and treatment outcomes should be regularly followed up. High degree of suspicion, early diagnosing, and a multidisciplinary treatment approach would be of great importance in such cases.

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