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Nasopharyngeal Embryonal Rhabdomyosarcoma- A Rare Case Report

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Abstract

Embryonal rhabdomyosarcoma is a rare neoplasm that arises from skeletal muscle cells. Most common site for rhabdomyosarcoma is head and neck region. This malignant mesenchymal neoplasm occurs most commonly in boys between the age group of two to six years. It is difficult to diagnose a case of rhabdomyosarcoma solely on the basis of histopathological examination. We present a case of a 7 year old male child presenting with pain and epistaxis and was diagnosed as embryonal rhabdomyosarcoma.

Keywords: NIL

Introduction

Rhabdomyosarcoma is a malignant mesenchymal neoplasm that arise from skeletal muscle cells[1]. Most common site for rhabdomyosarcoma are head and neck region, genitourinary tract, retroperitoneum and rarely extremities [2]. Amongst the head and neck region most common sites include orbit, paranasal sinus and soft tissue in cheek and neck[3]. Embryonal rhabdomyosarcoma is most common subtype of rhabdomyosarcoma[1] and occurs in children between birth and 15 years of age. Histologically embryonal rhabdomyosarcoma contains mixture of spindle cells and undifferentiated round cells and immature striated muscle like cells (rhabdomyoblasts) with abundant eosinophillic cytoplasm.

It is hypothesized that rhabdomyosarcoma arises from proliferation of embryonal mesenchymal cells that have a tendancy to differentate towards muscle differentiation[4-6].

We present a case of 7 year old male child who presented with a nasopharyngeal mass.

Case Report: A 7 year old male patient presented with complaints of pain in neck, loss of appetite, pain while swallowing food and epistaxis along with loss of weight .On general physical examination patient had pallor. On further examination a CT scan was planned and revealed an enhancing mass centered at the sphenopalatine fossa causing its widened appearance with its extension into the nasopharynx and posterior nasal cavity. A small biopsy was taken and on histopathological examination revealed nasopharyngeal angiofibroma. An MRI was planned to know the extent of mass lesion which revealed a homogenously enhancing nasopharyngeal mass extending anteriorly into posterior nasal cavity and in bilateral ethmoidal air cells and soft palate along with lymph nodal masses in the right posterior triangle and radiologically a possibility of malignant transformation of nasopharyngeal angiofibroma was given. Surgical excision of the mass was done. Histopathological examination of the mass revealed a polypoidal mass lined by respiratory epithelium. The sub-epithelium tissue revealed thin grenz zone beneath which a condensed layer of round to spindle cells having hyperchromatic nuclei, fine chromatin, inconspicuous nucleoli and scant eosinophillic cytoplasm (cambium layer)was seen. Beneath this layer nests of atypical plump spindled to round cells with fine chromatin, prominent nucleoli and abundant vacuolated cytoplasm with dense lympho-plasmacytic infiltrate was seen. Occasional mitosis was present. A final diagnosis of embryonal rhabdomyosarcoma was made and IHC was advised. IHC revealed positivity for desmin, myogenin and high Ki-67 index.

Discussion:

Rhabdomyosarcoma is a rare tumor more commonly encountered in young boys with a peak age of two to six years[7-8]. Our patient was a 7 year old male child. Most common site for rhabdomyosarcoma is head and neck region but nasopharynx is a rare site for rhabdomyosarcoma.

Anatomically head and neck rhabdomyosarcoma is divided in two categories: parameningeal (including nose, nasopharynx, paranasal sinuses, mastoid region, infra-temporal and pterygopalatine fossae and medium ear) and non-parameningeal (which include scalp, orbit, parotid gland, oral cavity, oropharynx and larynx)[9]. Patients who present with parameningeal tumors usually have symptoms like pain, voice alterations, airway obstruction, epistaxis and cranial nerve paralysis[7].In our patient an enhancing mass was seen centered at the sphenopalatine fossa and he presented with complaints of pain and epistaxis.

For diagnosis of rhabdomyosarcoma, a biopsy is performed initially followed by confirmation by immunohistochemistry. Our patient was diagnosed clinically and on a previous biopsy as angiofibroma. However, on histopathological examination of the surgically excised mass, a diagnosis of embryonal rhabdomyosarcoma was made which was later confirmed by immunohistochemistry.

On histopathological examination we also found some dilated congested vascular channels and blood vessels along with stellate cells and perivascular inflammation suggestive of angiofibroma. This morphology created a dilemma, however the presence of cambium layer, nests of spindle to round cells (rhabdomyoblasts) were diagnostic findings of an embryonal rhabdomyosarcoma. The final report of IHC confirmed the diagnosis.

Figures:

Hence to conclude we recommend a proper clinico-radiologic evaluation, thorough histopathological examination followed by immunohistochemistry for diagnosis of embryonal rhabdomyosarcoma especially at a rare site.

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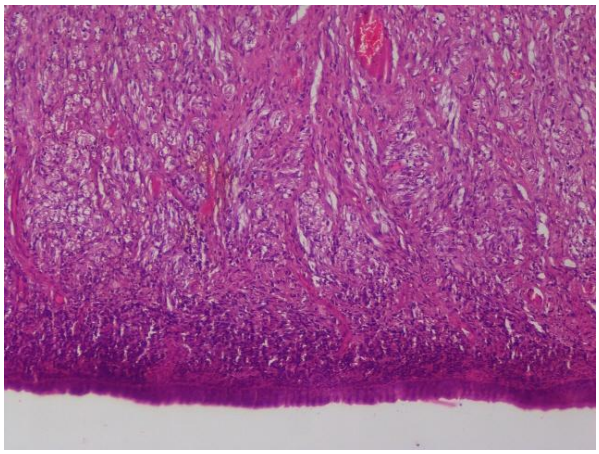
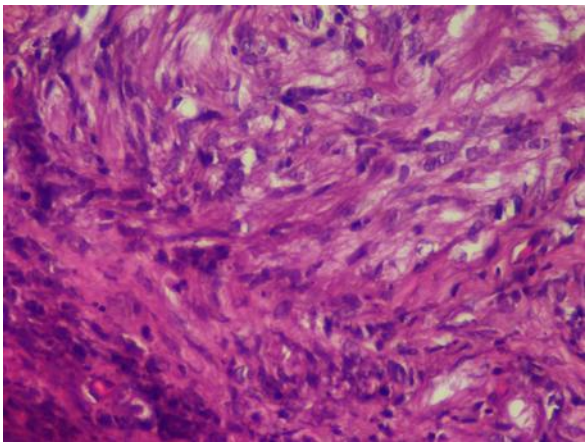
Volume 5, Issue 4; July-August 2022; Page No 1012-1014

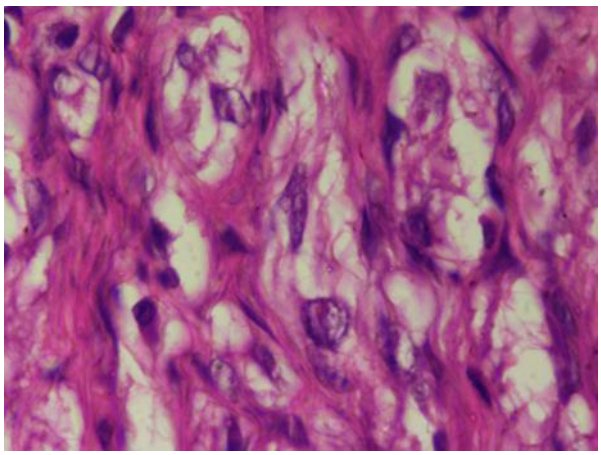
a) section shows respiratory lining epithelium with underlying thin grenz zone beneath which cambium layer is seen(H &E ,40X)



b) atypical plump spindle to round cells(H&E 400X)



c) atypical cells with prominent nucleoli and abundant vacuolated cytoplasm (H&E 400X)



Volume 5, Issue 4; July-August 2022; Page No 1012-1014