

Post auricular (extracardiac) fetal rhabdomyoma – a case report

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Abstract: Fetal rhabdomyoma, is a tumor originating in striated muscle, most commonly in the heart. Extracardiac rhabdomyomas most commonly occur in the head and neck region. They warrant a distinctive diagnosis and need to be differentiated from other malignant tumors like rhabdomyosarcoma in view of its good prognosis. These are benign and complete excision of the tumor is usually curative. We report a case of a 5-month-old female infant who presented with a postauricular swelling since birth. Microscopic examination revealed a cellular fetal rhabdomyoma.

Keywords: Rhabdomyoma, postauricular, fetal

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I. Introduction

Rhabdomyoma is a rare tumour originating in striated muscle, most commonly in the heart¹. Extracardiac rhabdomyomas most commonly occur in the head and neck region². Two types of extracardiac rhabdomyomas have been identified in the head and neck region: fetal and adult³. These are benign and complete excision of the tumour with preservation of the surrounding tissues is usually curative.

II. Case Report

A 5-month-old infant was brought to the pediatrics department with complaints of a postauricular swelling since birth. Examination revealed a hard and painless swelling of size 4x3x2 cm. No intracranial communication was noted on sonography. Complete excision of the swelling was done.

Macroscopic features:

We received a partly skin covered lobulated and firm tissue measuring 4x3x2 cm. Cut surface revealed a well circumscribed tumour with a grayish white appearance.

Microscopic features:

On microscopy, a well-circumscribed tumour having variable cellularity in different areas was seen. The tumour was composed of a mixture of undifferentiated spindle shaped cells with indistinct cytoplasm and scattered muscle fibres in varying stages of differentiation. The cells were arranged in a vague fascicular pattern and sheets. Mild cellular pleomorphism was noted however, mitotic activity was rare. Differentiated muscle fibres were more numerous at the periphery than at the centre (Fig. 1 & 2). Special stain for muscle, i.e. Phospho-tungstic acid hematoxylin stain highlighted the cross-striations of the skeletal muscle fibres (Fig.3). Immunohistochemistry confirmed rhabdomyoblastic differentiation in the form of desmin (Fig. 4) and myogenin (Fig.5) which showed cytoplasmic positivity highlighting the cross-striations.

III. Discussion

Rhabdomyomas are rare tumours originating in striated muscle¹ and are generally divided into the following categories: cardiac rhabdomyomas, which are relatively common, and extracardiac rhabdomyomas, which are rare (less than 2% of all tumours having skeletal muscle differentiation)⁴.

Cardiac rhabdomyoma, which is believed to be a hamartoma and not a true neoplasm, is usually diagnosed in children and associated with tuberous sclerosis⁵. Extracardiac rhabdomyomas, rare tumors, are subdivided into adult, fetal and genital histologic subtypes. They are distinct clinically and morphologically⁶. The fetal types are predominantly seen in infants and young children^{2,7} in the head and neck regions. The post-auricular region is the most characteristic site¹. They can be further subdivided into myxoid and cellular types.

On gross pathology, fetal rhabdomyomas are grayish white to tan pink, well circumscribed and soft¹. The cellular fetal rhabdomyomas are unencapsulated but well defined, are more cellular than the myxoid type and contain predominantly primitive mesenchymal cells with few mature muscle cells².

Our case showed the typical features of fetal rhabdomyoma i.e. early inception from birth, post auricular location, slow growth rate and good circumscription. However, the patient was lost for follow-up. The present report adds one more example of a benign undifferentiated mesenchymal tumour which presents all typical features of fetal rhabdomyoma, named by Dehner⁸ who studied nine cases and demonstrated immature mesenchymal cells differentiating into cells showing morphologic features of embryonic muscle.

Cytoplasmic cross-striations are difficult to identify on routine hematoxylin and eosin sections but can be identified with special stains for muscle like Phospho-tungstic acid hematoxylin (PTAH) stain or Masson's trichrome stain in majority of cases. Periodic acid-Schiff (PAS) reaction shows cytoplasmic globules of PAS positive material which is largely digested by diastase pretreatment. The tumour cells are haphazardly arranged in myxoid, often PAS

positive ground substance.

On immunohistochemistry, fetal rhabdomyoma is positive for desmin and vimentin in the primitive cells and desmin and myoglobin in the more mature striated cells². Adult rhabdomyoma is composed of more mature cells which stain for desmin and myoglobin; though immunostaining for vimentin is negative². In our case, immunohistochemistry confirmed rhabdomyoblastic differentiation. (Desmin and MyoD1 were positive).

On electron microscopy, the skeletal muscle origin of both types of rhabdomyoma has been verified by the findings of myofibrils arranged into sarcomeres in various stages of organisation. Focal condensation of myofibrils forms Z bands. Mitochondria, glycogen are found within the cytoplasm inmost of the tumour cells⁴.

Confusion in differentiating benign rhabdomyoma from well-differentiated embryonal rhabdomyosarcoma¹ may occur. Infiltrative margins and obvious histologic features of malignancy such as pleomorphism, a high rate of mitotic activity and rapid growth with early metastasis in embryonal rhabdomyosarcoma contrast sharply with good circumscription, lack of mitotic activity and invariable benign outcome after conservative surgery that are seen in fetal rhabdomyoma.

Generally, the prognosis for fetal rhabdomyomas is good. Recurrence has been reported in a few cases. Metastases are not known. Treatment consists primarily of complete excision with preservation of surrounding tissues. Recurrences are most commonly associated with incomplete excision.

IV. Conclusion:

Extracardiac fetal rhabdomyoma warrants a distinctive diagnosis and needs to be differentiated from other malignant tumors like rhabdomyosarcoma in view of its good prognosis.

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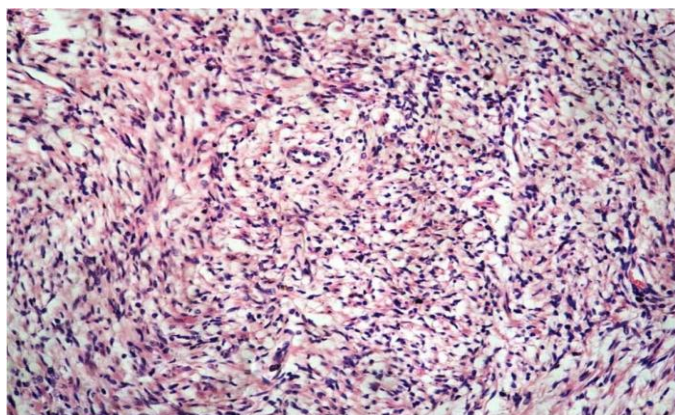


Fig 1 –Prominent spindle cell pattern, cells with eosinophilic cytoplasm and lacking nuclear atypia (H&E 100x)

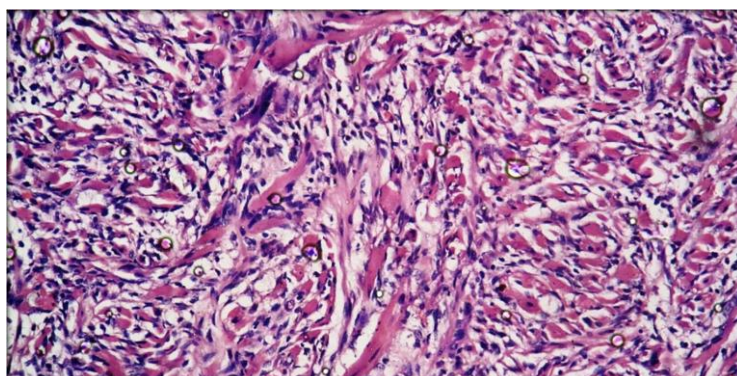


Fig 2- Intersecting bundles of differentiated myofibrils separated by strands of small undifferentiated spindle cells (H&E 400x)

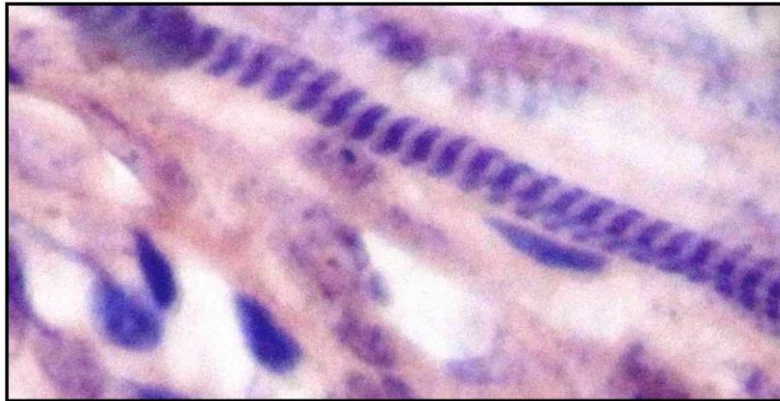


Fig 3- Muscle PTAH stain showing cross striations in differentiated rhabdomyoblastic cells (PTAHx1000)

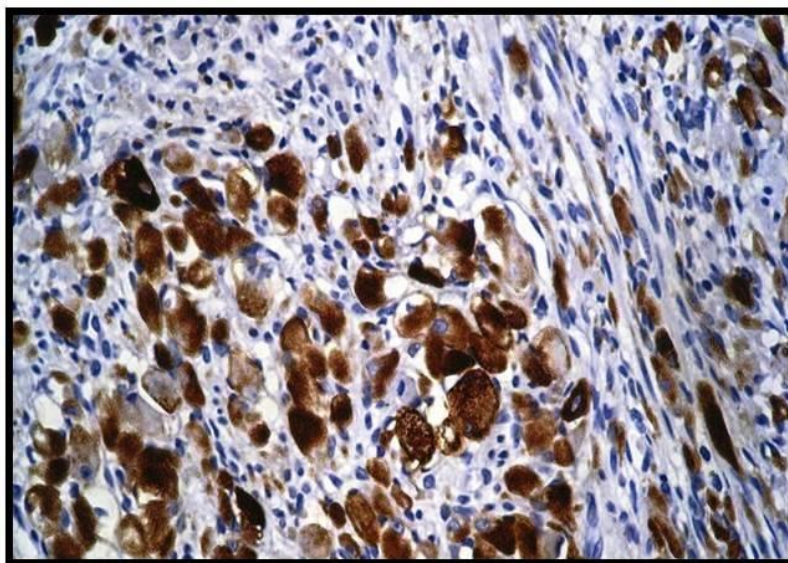


Fig 4- Cytoplasmic staining for desmin (Desmin x400)

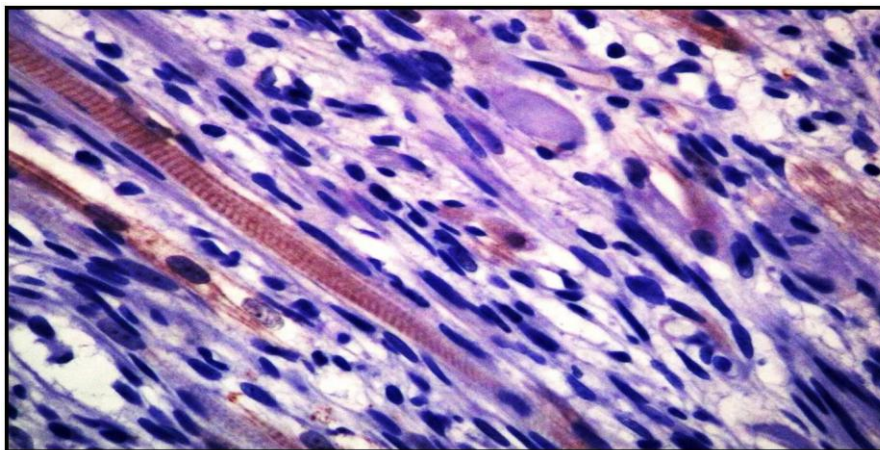


Fig 5- Striated cells showing diffuse cytoplasmic positivity for myogenin (Myogenin x400)

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