

A Case of Pleomorphic Variant of Embryonal Rhabdomyosarcoma in Skeletal Muscle - A Rare Case Report

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Abstract

Embryonal Rhabdomyosarcoma (ERMS) is a malignant soft tissue tumour with phenotypical and biological features of embryonic skeletal muscle cells. It is rare in skeletal musculature of extremities and its pleomorphic variant is rare which is associated with worse outcome. Here we are discussing a case of three year old male child who has presented with recurrence of swelling in right gluteal region arising in gluteus maximus muscle who is a known case of Rhabdomyosarcoma (RMS) 1 year back at the same site. Patient was operated with wide local excision and underwent chemotherapy cycles.

Keywords: Embryonal, Pleomorphic, Rhabdomyosarcoma (RMS)

1. Introduction

Rhabdomyosarcoma is malignant mesenchymal tumour with skeletal muscle differentiation. Three subtypes of RMS are recognised- Embryonal, Alveolar and Pleomorphic. Embryonal subtype constitutes about 60% of rhabdomyosarcomas¹. ERMS is the most common soft tissue sarcoma of the childhood comprising 80 to 90% of cases². Common sites are head, neck, genitourinary tract and retroperitoneum¹. Less than 9% of ERMS arise within skeletal musculature of extremities³. Grossly, ERMS form poorly circumscribed, fleshy, pale tan mass that impinge upon neighbouring structures. Microscopically ERMS show alternating areas of cellular condensation and hypercellularity with cells floating in mucoid ground substance. These tumours may exhibit heterogenous degrees of differentiation, matrix and cellularity varying from case to case². Occasional tumours within embryonal category have focally anaplastic features with bizarre nuclear forms are classified as pleomorphic subtype of embryonal rhabdomyosarcoma⁴. However, the presence of anaplastic cells in aggregates of diffuse sheets throughout the tumour portends poor survival for these patients⁵.

2. Case Report

A three year old male child who was a known case of embryonal rhabdomyosarcoma and operated one year back presented with recurrence in right gluteal region. Patient had similar complaints one year back and the mass was excised and histopathologically diagnosed as Embryonal RMS (pleomorphic type). The patient had six cycles of chemotherapy. Now patient presented with similar swelling at the same site. MRI showed multilobulated heterogeneously hyperintense lesion arising from right gluteus maximus muscle. Fine Needle Aspiration Cytology of the swelling was done and reported as High Grade Sarcoma with few multinucleate giant cells. Wide local excision of the tumour was done. Grossly we received a single, poorly circumscribed soft tissue mass measuring 11 x 8 x 5.5 cm with cut section showing fleshy, pale tan appearance. Microscopically, tumour showed cells arranged in diffusely compact pattern. Tumour cells were round to oval with pleomorphic, hyperchromatic nuclei showing prominent nucleoli and scant eosinophilic cytoplasm. Many multinucleate giant cells with few binucleate forms noted. Perivascular pseudorosette

arrangement of tumour cells was noted. Few of tumour cells showed anaplastic features with bizarre nuclei. Occasional stellate shaped cells with elongated nucleus (strap cells) were seen. Immunohistochemically tumor showed positivity for *Myogenin* and *Desmin*.



Figure 1. Gross image showing poorly circumscribed soft tissue mass with cut section showing fleshy pale tan appearance.

Microscopy images showing anaplastic features in embryonal RMS in 3 yrs old child.

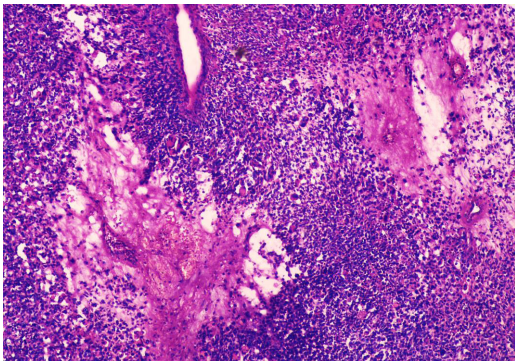


Figure 2. H&E, x 100 low power view showing diffusely compact arrangement of tumor cells with perivascular pseudorosette.

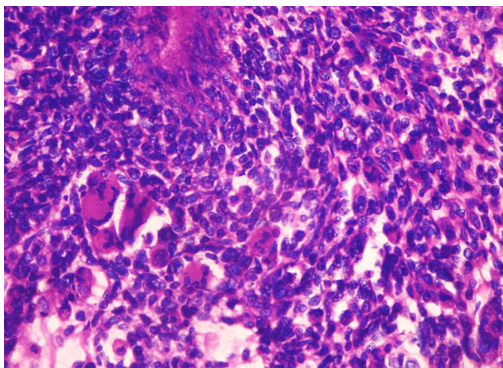


Figure 3. H & E, x 400 high power view showing tumor cells having anaplastic features. Many multinucleate giant cells with binucleate forms noted.

3. Discussion

Children less than 10 years of age are usually affected by ERMS with most cases (36%) occurring in children less than 5 years of age, only 18% arise in adolescence. Common sites of involvement include head, neck, genitourinary tract, prostate, paratesticular soft tissues, oropharynx, parotid, auditory canal, nasopharynx, tongue and cheek. Also occurs in retroperitoneum, abdomen and viscera like kidney and heart. ERMS infrequently involves soft tissues of extremities and trunk³.

Microscopically, ERMS contains primitive mesenchymal cells in varying stages of myogenesis with variable content of rhabdomyoblasts. Stellate cells with sparse amphophilic cytoplasm and central nuclei represent the most primitive end of this spectrum. Rhabdomyoblasts progressively acquire more cytoplasmic eosinophilia and elongation manifested by “tadpole”, strap cell and spider cells. Differentiation becomes more evident after chemotherapy. ERMS form embryoid aggregates of myoblasts in loose myxoid mesenchyme and comprise patternless sheets of spindle and round cells³. Occasional tumours within embryonal category have focally anaplastic features with bizarre nuclear forms, they are classified as pleomorphic subtypes of ERMS. It has been suggested that they are associated with more aggressive clinical course, especially when the pleomorphic features are extensive, conversely well differentiated tumours are associated with excellent response to chemotherapy⁶.

Immunohistochemical markers of skeletal muscle differentiation typify ERMS. These markers correlate with the degree of tumour cell differentiation. Only vimentin is present in the cytoplasm of most primitive cells. Desmin and Actin are acquired by developing rhabdomyoblasts³. Excellent monoclonal antibodies against MyoD and Myogenin have been developed for immunohistochemistry. ERMS stain in more heterogeneous fashion which gives clue to subclassification⁷.

Prognosis of ERMS can be determined by stage, histological classification, age and site of origin. Stage is accomplished by clinical evaluation or surgicopathological evaluation. Age tends to be independent risk factor. Patient aged 1 to 9 years having better outcomes than infants or adolescents. Histologically ERMS with anaplasia have worse outcome than nonanaplastic. The outcome of parameningeal and extremity tumours is poor³.

4. Conclusion

Embryonal rhabdomyosarcomas that occur in extremities are rare. When they are associated with pleomorphism are

rare in children and show more aggressive clinical course, higher relapse rate and lower survival rate⁸.

5. References

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