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Pleomorphic Rhabdomyosarcoma of Gluteal Region – A Case Report

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Abstract: Rhabdomyosarcoma (RMS) is the most common form of soft tissue sarcoma in the first 2 decades of life, with a peak age incidence in very young children. Due to the rarity of adult RMS, information regarding its clinical and biologic characteristics is very limited. Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children. It is seen rarely in adults. There are three variants of RMS which include embryonal and alveolar variants which are commonly seen in children and pleomorphic variant seen in adults. The prognosis of the tumor is worse in adults when compared to children, with an overall 5-year survival of less than 30%.
Keywords: Rhabdomyosarcoma, pleomorphic rhabdomyosarcoma, Gluteal tumor

INTRODUCTION

Rhabdomyosarcoma (RMS) is a highly malignant mesenchymal tumor thought to originate from immature striated muscle. It is characterized by the presence of cells having an identifiable striated muscular differentiation with rhabdomyoblasts. The pleomorphic form is typically more frequent in adults and has a poor prognosis [1].

RMSs are solid tumors that are common in children, representing 5% of all childhood cancers [2]. In contrast, RMSs are rare in adults, with soft-tissue sarcomas making up fewer than 1% of malignancies in adults and RMSs accounting for 3% of all soft tissue sarcomas [3].

CASE REPORT

A 52 year old female came with complaint of a swelling in right gluteal region for few months with sudden increase in size. Incisional biopsy was done before specimen excision.

We received a skin covered soft tissue mass measuring 20 x 19 x 10 cm (Fig.1). Skin measuring 19 x 5 cm. Centre of skin showed a fresh scar measuring 3.3 cm with intact sutures. Cut surface showed a grey white tumor measuring 16.5 x 14 x 6 cm extending from subcutaneous area reaching upto the underlying muscle at a distance of 4cm from the superior margin, 3cm from the inferior margin, 3 cm from the lateral margin, 0.3cm from the medial margin and extending upto the deep resected margin, reaching at a distance of 0.5cm from the skin (Fig. 2). Necrosis occupied less than 10% of area. Cut surface of tumor showed grey white lobulation with multiple yellowish chalky white areas and foci of necrosis. Firm with well defined margins and foci of infiltrating margins.

Fig-1: External surface of the specimen

Fig-2: Cut surface showing grey white tumor

Sections from incisional biopsy showed a cellular neoplasm composed of oval to polygonal and spindle shaped cells with abundant eosinophilic cytoplasm and enlarged pleomorphic nuclei with prominent nucleoli (Figs.3,4,) Cells were arranged in diffuse sheets. Tumor giant cells, multinucleated giant cells and mitotic figures (>10/10hpf) were seen. The impression from incisional biopsy was given as high grade pleomorphic spindle cell sarcoma.

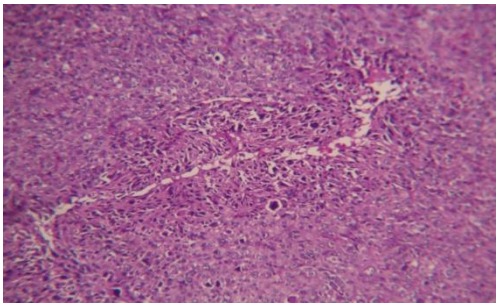

Fig-3: 10x. Cellular neoplasm composed of spindle cells and tumor giant cells

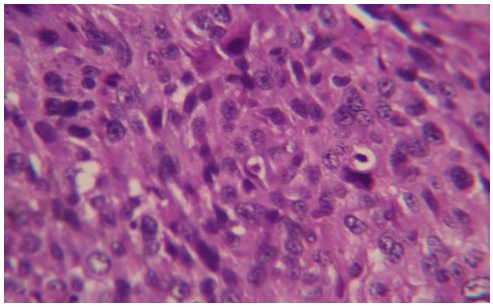

Fig-4: 40x. Ovoid to elongated cells with eosinophilic cytoplasm, and pleomorphic nuclei and mitotic figures

Sections from the excised specimen of mass in the gluteal region showed a cellular neoplasm composed of pleomorphic spindle shaped cells, and anaplastic tumour cells with enlarged hyperchromatic nuclei with some of them showing prominent nucleoli. Multinucleated tumor giant cells were also seen. Atypical mitotic figures were evident (Figs.3,4,7,8) Tumour cells were arranged in interlacing bundles, fascicles and focally in a storiform pattern (Figure 6,7). Areas of necrosis, calcification and hyalinisation were seen. All resected margins and skin were free of tumor except the deep margin, where focal infiltration was seen. With the above histomorphological features, the following two possibilities to be considered - Pleomorphic undifferentiated sarcoma (previously referred to as Malignant Fibrous Histiocytoma and Pleomorphic Rhabdomyosarcoma.

Immunohistochemistry with the marker Myogenin, was performed to choose between the two, and the tumor cells showed positivity for the marker, clinching the diagnosis of Pleomorphic rhabdomyosarcoma (Fig.9).

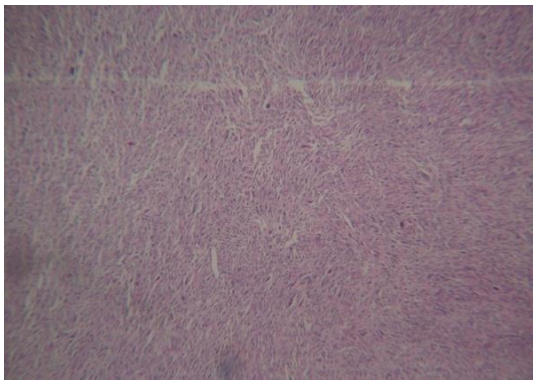

Fig-5: H & E. 4x: Tumor cells in sheets & bundles

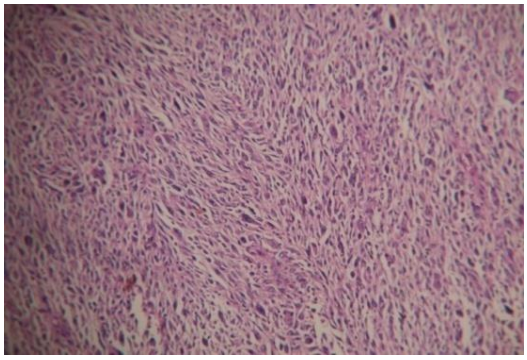

Fig-6: H & E. 10x. Tumor cells in intersecting bundles with pleomorphic giant cells

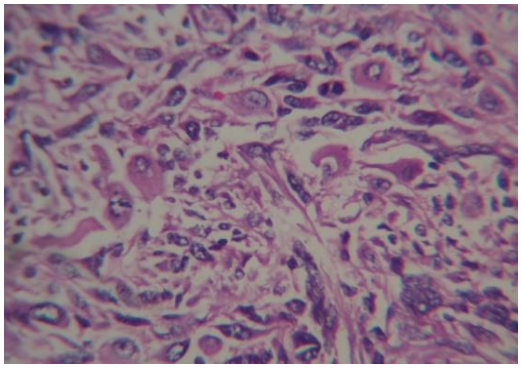

Fig-7: H & E. 45x. Strap cells, rhabdomyoblasts & tumor giant cells

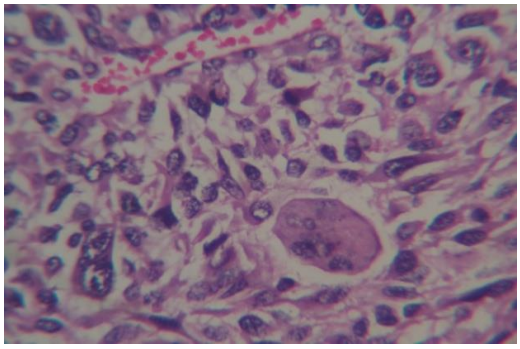

Fig-8: H & E. 45x. Shows tumor giant cells

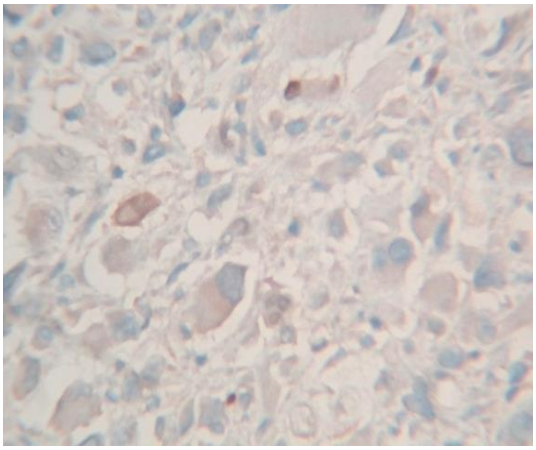

Fig-9:IHC – Myogenin. 45x. Cytoplasmic positivity

DISCUSSION

The different subtypes of RMS are embryonal, alveolar, and pleomorphic. Overall, the embryonal subtypes is the most common subtype, accounting for up to 49% of all RMSs [4]. The alveolar subtype accounts for approximately 30% of all RMSs and most commonly affects adolescents. It is mostly an intramuscular tumor in the soft tissues of the extremities [3].

The pleomorphic subtype is the least common subtype. Pleomorphic RMS is a rare variant of rhabdomyosarcoma (arises in adults > 45 years). It mostly arises in adults with a peak incidence in the fifth decade of life with a predilection for males. Most commonly it occurs in the skeletal muscle of the extremities, particularly the thigh. Less commonly, in the abdomen/ retro-peritoneum, chest/ abdominal wall, spermatic cord/testis and upper extremities. It presents with a rapidly growing, painless mass of several months' duration. Grossly, tumor is usually large (> 10 cm), fleshy, well-circumscribed. They are intramuscular masses with focal hemorrhage and extensive necrosis. Histologically, pleomorphic rhabdomyosarcoma can be distinguished from embryonal and alveolar rhabdomyosarcoma by the association of loosely arranged, haphazardly oriented, large, round or pleomorphic cells with hyperchromatic nuclei and deeply eosinophilic cytoplasm. Tumor cells may be arranged in a haphazard pattern. The most helpful light microscopic feature suggesting this diagnosis is the presence of large bizarre tumor cells with deeply eosinophilic cytoplasm with some cell-to-cell moulding. Other features include phagocytosis by tumor cells, presence of intracytoplasmic glycogen, and a moderately dense lymphohistiocytic infiltrate. It is histologically similar to a malignant fibrous histiocytoma; in fact, many previously diagnosed, pleomorphic RMSs have been reclassified as fibrous histiocytomas, making the diagnosis of pleomorphic RMS an even rarer entity [3].

In adults with RMS, lymphadenopathy appears to be more common. Lungs have been reported to be the most frequent site of distant disease [6].

The presence of metastases at presentation along with tumor size, tumor resectability, and patient age at presentation are also prognostic factors. Histologic subtype also has a bearing on prognosis. Embryonal tumors are usually associated with the most favorable outcome and pleomorphic, the least favorable. In our case there was no metastasis.

Treatment of patients with RMS is usually primarily surgery if excision is attainable, followed by radiation therapy and chemotherapy. Survival rates for patients with non-metastatic disease have improved substantially throughout the last 30 years [7, 8]. Although surgical resection, either as a primary procedure or a secondary strategy (after initial chemotherapy), is important for local control, complete resection is not feasible at some sites of disease. Radiotherapy has therefore assumed a major role in the management of many patients, though late sequelae can be significant.

The prognosis for patients with RMS has been typically very poor, with a 5-year survival rate of less than 50% even in the most recent series [5, 9]. However, this survival rate is improved compared with the rates reported in older series, which were as low as 21% [10].

CONCLUSION

Though the treatment for adult RMS is surgery, radiotherapy and chemotherapy or in combination, careful long-term follow-up is necessary due to high risk of recurrence. Gross removal of these sarcomas appears to provide better disease control. Resection of the tumor followed by radiotherapy remains an important factor in prognosis

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