Case Report

Retroperitoneal pleomorphic rhabdomyosarcoma metastasizing to inguinal lymph node

Shagufta Qadri, Kiran Alam, Feroz Alam

Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, Uttar Pradesh, India

ABSTRACT

Rhabdomyosarcoma (RMS) is a highly aggressive, malignant tumor of skeletal muscle cell, associated with an early and a wide spread metastasis. Although a commonly occurring soft tissue sarcoma in the pediatric population, it is seldom encountered in adults. Outcome for adult RMS is poorly documented due to its rarity. We report a case of pleomorphic RMS (PRMS) in a 50 years male presenting with an intra-abdominal mass along with a swelling in the right inguinal region measuring 4 cm × 3 cm. Computed tomography revealed an ill-defined intra-abdominal mass arising from the peritoneum. Abdominal mass was resected along with the dissection of inguinal lymph node. Histopathological examination of these masses coupled with the immunohistochemistry, confirmed the diagnosis of PRMS metastasizing into inguinal lymph node. Despite of adjuvant radiotherapy and chemotherapy, the patient couldn't survive >3 months and died of widespread lung metastasis.

Key words: Metastasis, pleomorphic sarcomas, retro-peritoneum, rhabdomyosarcoma

INTRODUCTION

Soft tissue sarcomas makes up <1% of all adult malignancies, of which rhabdomyosarcoma (RMS) accounts for 3% of all soft tissue sarcomas.[1] Stout was first to introduce pleomorphic RMS (PRMS) into the medical literature in 1946,[2] as "classical" RMS. Due to the infrequency with which RMSs are encountered, only tenuous cases of PRMS has been reported in English literature till date.

We hereby report a rare case of PRMS of retro-peritoneum metastasizing to inguinal lymph node, with a comprehensive literature review. Succinctly, describing the tumor and its management, with an aim to provide a cumulative contribution to the medical literature.

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CASE REPORT

A 50-years-old male presented with an intra-abdominal mass and swelling in the right inguinal region. He also complained of dragging sensation and constipation. On per-abdominal examination, a mass was felt in right lumbar region. Abdominal Computed tomography revealed an ill-defined intra-abdominal mass filling right retroperitoneal space [Figure 1]. On local examination of the inguinal region, a large globular swelling of about 4 cm × 3 cm was felt. It was firm in consistency, which neither fixed to the underlying tissue nor the overlying skin. Abdominal mass was resected by open laparotomy, followed by inguinal lymph node dissection. Both the specimens were sent for histopathological examination. Grossly the abdominal mass was a huge, irregular growth measuring 18 cm × 12 cm × 10 cm, firm to soft in consistency with glistening, gelatinous areas. On cut section, it showed gray-white mass with some hemorrhagic and necrotic areas. The dissected lymph node was firm in consistency with central necrosis on cut section [Figure 2]. Microscopic examination revealed the tumor composed of highly pleomorphic, spindle-shaped cells arranged in a fascicular pattern [Figure 3]. The individual cells were showing high N: C ratio, variable

Address for correspondence: Dr. Shagufta Qadri, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh - 202 002, Uttar Pradesh, India. E-mail: qadridrshagufta@gmail.com

amount of the fibrillary cytoplasm, many bizarre polygonal rhabdomyoblasts with prominent nucleoli and abundant eosinophilic cytoplasm [Figure 4]. High mitotic count and necrosis were also noticed. On immunohistochemistry, the tumor cells showed strong cytoplasmic desmin positivity [Figure 5] and focal nuclear positivity for myogenin [Figure 6]. Histomorphologic findings complemented with immunohistochemistry, prompted to the diagnosis of PRMS metastasizing

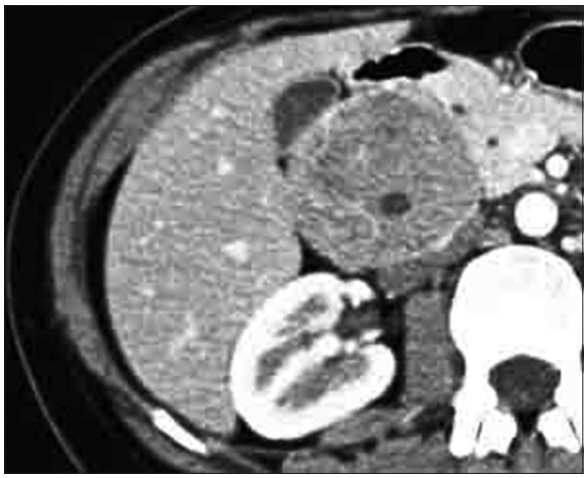

Figure 1: Abdominal computed tomography showing an ill-defined intra-abdominal mass filling right retroperitoneal space

Figure 2: Resected lymph node showing central necrosis on cut section

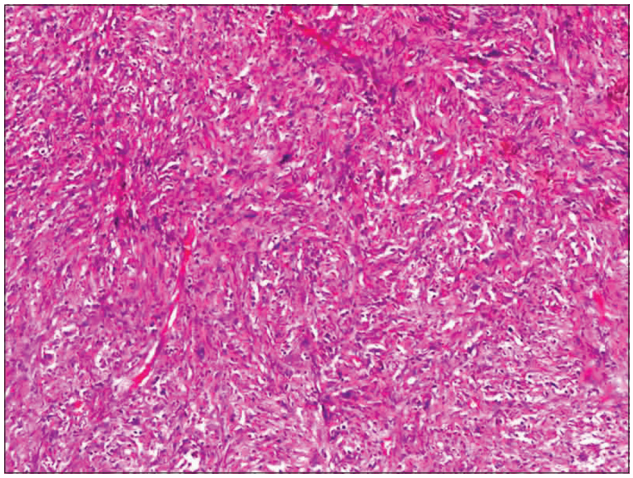

Figure 3: Section showing the tumor composed of highly pleomorphic, spindle-shaped cells arranged in a fascicular pattern (H and E, ×100)

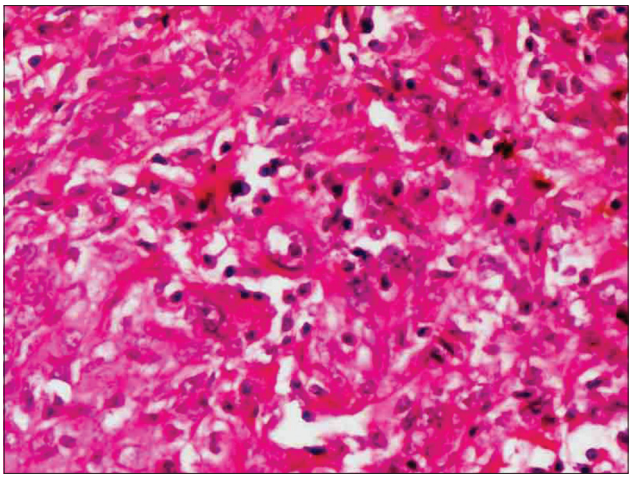

Figure 4: Polygonal rhabdomyoblasts with prominent nucleoli and the abundant eosinophilic fibrillary cytoplasm (H and E, ×400)

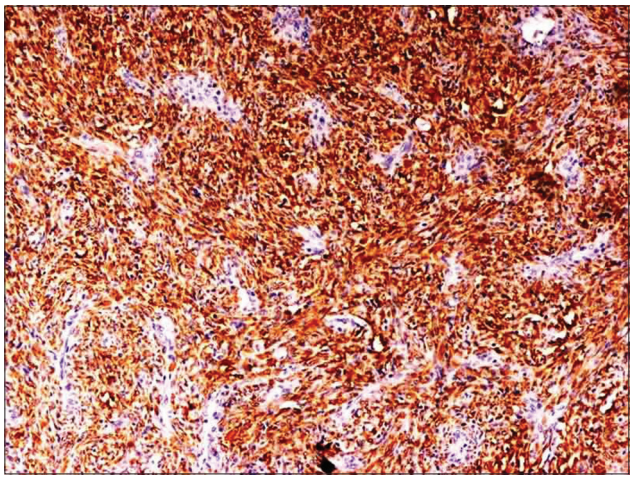

Figure 5: Tumor cells showing strong cytoplasmic desmin (H and E, ×100)

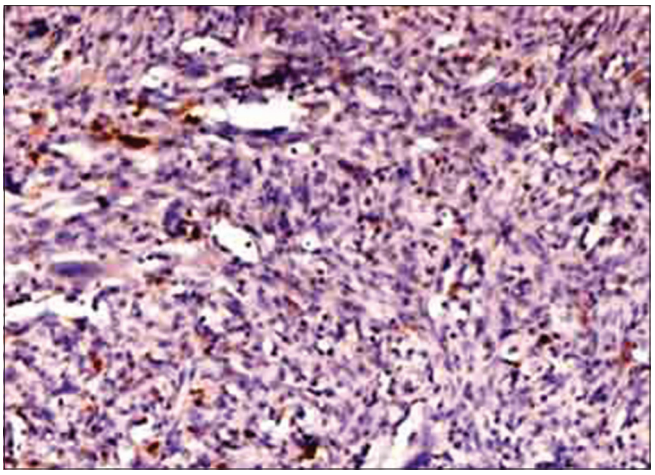

Figure 6: Tumor cells showing focal nuclear positivity for myogenin (H and E, ×100)

into inguinal lymph node. Although the surgery was accompanied with adjuvant radiotherapy and chemotherapy, the patient couldn't survive >3 months and died of widespread lung metastasis.

DISCUSSION

Rhabdomyosarcoma 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.[2] In a study performed by Furlong *et al*., PRMS is mentioned as a rare variant of RMS that almost always occurs in adults, with mean age of 49 years and has a very poor prognosis,[2] our case also corresponded to these notable finding. The tumor is often large (>10 cm), and mostly fleshy, well-circumscribed, intramuscular masses with focal hemorrhage and extensive necrosis. It is noticed that, in general tumors growing into the body cavities, such as those involving naso-pharynx, and urinary bladder, are fairly well circumscribed, multi-nodular, or distinctly polypoid with a glistening, gelatinous, gray-white surface that on cross-section often shows patchy areas of hemorrhage or cyst formation. However, the deep-seated tumors arising in the musculature are not well-defined and frequently infiltrate into the surrounding tissues. PRMS can be distinguished from embryonal and alveolar RMS on the basis of histomorphology. Normally, on microscopic examination the tumor cells are seen to be arranged in sheets and lobules. Cells are pleomorphic, with round to elongated nuclei and abundant eosinophilic cytoplasm. Areas of necrosis are common phenomena. The diagnosis of RMS can be difficult with conventional histological techniques, and it could easily be confused with undifferentiated high-grade pleomorphic sarcoma (malignant fibrous histiocytoma).[3] Immunohistochemical analysis should, therefore, address specific skeletal muscle markers and nonspecific myoid markers for achieving the diagnosis of PRMS. There may be a spectrum of differentiation in PRMS, substantiated by variable myoregulatory protein expression, which goes uncharted if relied purely upon morphologic appearance. PRMS should be differentiated from other pleomorphic sarcoma subtypes including differentiated liposarcomas, malignant peripheral nerve sheath tumors, pleomorphic leiomyosarcoma and metastasis from metaplastic carcinoma.[4] The presence of pleomorphic polygonal rhabdomyoblasts on routine hematoxylin and eosin stain coupled with positive immunohistochemical staining with anti-myoglobin antibody and anti-sarcomeric actin antibody, characteristic of striated muscle are very helpful in the diagnosing RMSs.[5] Several studies have reported a high sensitivity and specificity for MyoD1 and/or myogenin in recognizing PRMS and its distinction from other adult pleomorphic soft tissue sarcomas.[6] We confirmed the histomorphologic findings in our case by the positive immunoreactivity to desmin and myogenin antibodies, which was also implemented by Ceylan *et al*.[7] in diagnosing a case of PRMS involving the urinary bladder. However, Tallini *et al*. reported sensitivities of MyoD1 and myogenin estimated to be only 53% and 56%, respectively, in cases of PRMS.[8] According to intergroup RMS study classification, patients with metastases at diagnosis are classified as stage IV. Major metastatic sites include the lung, lymph nodes, and bone marrow, followed by the heart, brain, meninges, pancreas, liver, and kidney. The lungs are involved in at least two-thirds of patients with metastasis and comprises of 14% case at the time of initial diagnosis,[9] even the patient presented in this report died of widespread lung metastasis, in spite of receiving adjuvant chemo and radiotherapy. The incidence of lymph node metastasis largely depends on the location of the tumor. Although in our case the nodal metastasis is from retroperitoneal RMS, it is more often associated with RMSs of the prostate, para-testicular region, and extremities than with those of the orbit and head and neck.[10] As the disease is preponderant in pediatric and adolescents population, there is a paucity of literature inscribing its management in adults. The optimal treatment of this malignancy is not yet determined, but multimodality approaches combining surgical excision, followed by chemotherapy with or without radiotherapy is the most recommended mode of managing PRMS. Ogilvie *et al*. in their study found that combining surgery and radiation therapy with chemotherapy using doxorubicin, ifosfamide, and vincristine yielded 55% overall and 64% disease-free survival at 2 years.[11,12]

CONCLUSION

- Outcome of RMS in adults appears to be worse than those occurring in children
- Diagnosis of PRMS is achieved by combining histomorphological, immunohistochemical and ultra-structural findings that identifies the skeletal muscle phenotype
- The multipronged treatment approach is combining surgical resection, radiation and systemic chemotherapy, recommended for managing PRMS, still needs further evaluation.

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