Retroperitoneal Fibrosarcoma: A Rare Case Report

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Retroperitoneal sarcomas are rare tumors ranging from only 1% to 2% of all solid malignancies. Among all sarcomas, most of these occur outside the retroperitoneum. Only 10-20% of sarcomas are retroperitoneal sarcomas, and the overall incidence is 0.3-0.4% per 100,000 of the population. The diagnosis and treatment of retroperitoneal sarcomas are challenging because the tumors are quite rare and usually present in later stages of the disease in an anatomically complex location. A rare case of recurrent retroperitoneal fibrosarcoma in a 23-year-old male presented as a mass per abdomen. Retroperitoneal sarcomas are malignant tumors arising from mesenchymal cells, which are usually located in muscle, fat, and connective tissues.

Keywords: Fibrosarcoma, Retroperitoneal, VIMENTIN®, S100 protein

INTRODUCTION

Retroperitoneal sarcomas are rare tumors ranging from only 1% to 2% of all solid malignancies. Among all sarcomas, most of these occur outside the retroperitoneum. Only 10-20% of sarcomas are retroperitoneal sarcomas, and the overall incidence rate is 0.3-0.4% per 100,000 of the population.¹ The highest incidence rates are seen in the 5th decade of life, although they can occur in any age group. Patients with sarcomas present late, because these tumors arise in the large potential spaces of the retroperitoneum and can grow into massive tumors without producing any signs or symptoms.²³

CASE REPORT

A 23-year-old male patient presented with rapidly increasing mass in the right side of the abdomen of 1-month duration. Swelling associated with pricking type of pain, radiating to the right thigh and low-grade fever. A history of significant loss of weight (5 kg in 2 months) present. No history of jaundice and diabetes. A retroperitoneal mass of 28 cm × 15 cm occupying the right lumbosacral region and hypochondrium, epigastrium and umbilical regions. It is not moving with respiration. It is variable in consistency and irregular shape. There is no ascites. Café au leot spots are seen over the abdomen. Patient had a high arched palate (Figures 1-3). He is hypertensive - blood pressure 170/100 mmHg one year back he was operated for a similar complaint and was diagnosed to have retroperitoneal neurofibroma.

Ultrasonography abdomen (Figure 4). A large well defined heterogeneous mass lesion noted in the pelvis, extending up to the right hypochondrium with multiple areas of necrosis. The mass approximately measuring 14.5 cm × 12.8 cm, displacing the ureter and inferior vena cava medially.

Contrast-enhanced computed tomography and magnetic resonance imaging: A large well defined, encapsulated, mixed density mass lesion noted extending from pelvis to right hypochondrium with large areas of necrosis (Figures 5 and 6).

Trucut biopsy: Neurofibroma.

Operative findings: Large retroperitoneal mass with areas of necrosis and calcification, stretching right ureter over it. Mass extending into right iliac fossa and into the pelvis. Upper extent is abutting on to undersurface of the liver. There is increased vascularity. The peritoneum is adherent to tumor. Tumor weighed 5.5 kg (Figures 7-9).

Histopathology: Smear shows cellular tumor consisting of interlacing bundles of spindle cells infiltrating the adipose tissue. They contain scant to moderate amount of fibrillary vacuolated cytoplasm and elongated cigar-shaped nucleus. There are mild nuclear pleomorphism and focal areas of high cellularity and patchy areas of necrosis. Mitotic activity is high (Figure 10).
Impression: Malignant mesenchymal tumor in favor of fibrosarcoma.

Immunohistochemistry-VIMENTIN - strongly positive in the blood vessels and spindle-shaped tumor cells. S-100 - negative. CD-6 - negative.

Final diagnosis: Fibrosarcoma.
DISCUSSIONS

Retroperitoneal sarcomas are rare tumors ranging from only 1% to 2% of all solid malignancies. Among all sarcomas, most of these occur outside the retroperitoneum. Only 10-20% of sarcomas are retroperitoneal sarcomas, and the overall incidence is 0.3-0.4% per 100,000 of the population. Its assumed that sarcomas develop from mesenchymal stem cells residing in muscle, fat, and connective tissues. The origin of these stem cells is not clear, and occasionally even their mesenchymal derivation is doubtful (as for nerve or nerve sheath sarcomas, gastrointestinal stromal tumors, and primitive neuroectodermal tumors). Two prevailing theories suggest that mesenchymal stem cells are found in local tissue pools or arise from the bone marrow. Approximately half of retroperitoneal sarcomas are high-grade tumors. The most common histologic subtypes of retroperitoneal sarcoma are found to be liposarcomas (41%), leiomyosarcoma (28%), malignant fibrous histiocytoma (7%), fibrosarcoma (6%), and malignant peripheral nerve sheath tumor (3%).

CONCLUSIONS

The evaluation and treatment of retroperitoneal soft tissue sarcomas remain challenging. The most important factor in the long-term success in the treatment of primary tumors is complete surgical resection. It is important that patients with these tumors should be evaluated and treated at centers with multidisciplinary treatment planning and expertise in treating these rare tumors. These centers also can invest time and resources into investigations of novel therapies and have access to clinical trials. Local recurrence remains a difficult problem with increased associated morbidity and psychological stress for affected patients.

REFERENCES

Wattamwar, et al.: Retroperitoneal Fibrosarcoma


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