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Case Report Open Access

Retrorectal Myxoid Fibrosarcoma: A New Entity

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Abstract

We report the case of a 40-year-old man who admitted to our department because of progressive increasing abdominal girth and weight loss. CT scan showed a huge retrorectal tumor. The mass was totally excised by laparotomy. At five years, the patient was doing well with disease free. Tumors occurring in the retrorectal space are rare; we report herein the first case of a myxoid fibrosarcoma of retrorectal space.

Keywords: Retrorectal tumor; Fibrosarcoma; Surgery

Introduction

The presacral space, which contains different types of embryonic tissue, is a potential site for several tumors. Primary fibrosarcoma is an exceptional retrorectal tumor. We report the case of a 40-year-old man with a huge retrorectal tumor treated successfully by surgery alone. This is the first report of a case of a myxoid fibrosarcoma of retrorectal space.

Case Report

A 40-year-old man was admitted to our department because of progressive increasing abdominal girth, asthenia and weight loss. On physical examination, the abdomen was distended owing to a huge, non-tender, palpable mass with no clear borders. Laboratory findings and tumor markers were within normal limits. A computed tomography scan revealed a bulky inhomogeneously enhancing retrorectal mass displacing completely the abdominal organs, with no clear signs of infiltration. T1- and T2-weighted magnetic resonance imaging showed a well-defined soft-tissue mass measuring 24 × 17 × 9.0 cm located in retrorectal space (Figure 1). At laparotomy, a giant mixte tumor that filled completely the retrorectal space was found. The mass was totally excised and the main difficulty was to guarantee clear margins sparing the rectum and the sacrum (Figure 2). The rectum was spared and no intestinal resection was necessary to have clear margins. No lymph node dissection was done.

The mass weighed 18 kg and appeared grossly multinodular, partially confluent with solid areas. The histopathological examination showed fibroblasts with myxoid stroma and a rich capillary network (Figure 3).

On immunohistochemical study, the tumor cells expressed vimentin, SMA and MUC 4 (Figure 3). There was no expression of desmin, CD34 and EMA. The diagnosis of a low grad fibromyxoid sarcoma was established. The postoperative course was uneventful. The patient was discharged one week after surgery. Five years later, the patient was free from recurrence.

Discussion

The true incidence of tumors occurring in the retrorectal (presacral) space is unknown, yet several retrospective series suggest that between one and six patients will be diagnosed annually in major referral centers [1]. The retrorectal space contains multiple embryologic remnants derived from a variety of tissues. Tumors that develop in this space are both macroscopically and histologically heterogeneous. Most lesions are benign, but malignant neoplasms are not uncommon. Solid lesions are more likely to be malignant than are cystic lesions. Neurogenic lesions typically arise from peripheral nerves and represent about 10% of retrorectal tumors [2]. These tumors include neurofibromas and sarcomas, neurilemomas, ependymomas, and ganglioneuromas. Fibromyxoid sarcoma is a rare soft tissue sarcoma usually located in the deep soft tissue in the groin or lower extremities. No case located in retrorectal space has been reported in literature review [2]. Thus, it is important to differentiate this tumor from other soft tissue tumors [3].

Fibrosarcoma occurs more commonly in men than in women. It can be diagnosed in patients of any age, but it is diagnosed more frequently in patients in the fourth or the fifth decade of life, as in our patient.

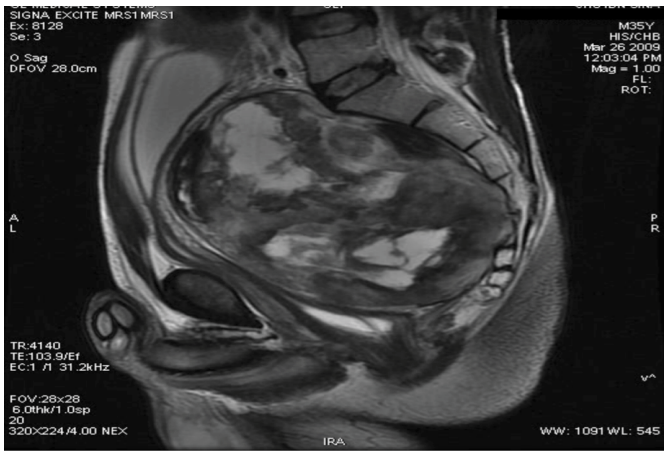

Figure 1: Magnetic resonance imaging showing a well-defined soft-tissue mass measuring 24 × 17 × 9.0 cm located in the retrorectal space.

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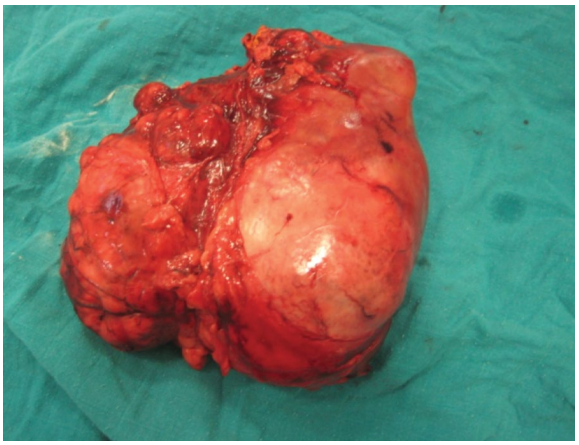

Figure 2: Resection Specimen.

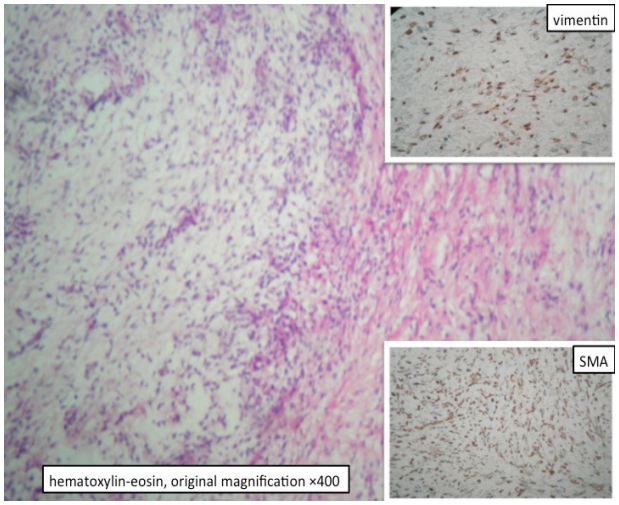

Figure 3: Histopathological examination showing fibroblasts with myxoid stroma and a rich capillary network (hematoxylin-eosin, original magnification ×400).

Symptoms of retrorectal tumors are often nonspecific and are related to the location and to the size of the lesion. The majority of benign cystic lesions are asymptomatic and usually discovered on routine rectal examination. Pain and neurologic dysfunction might be the presenting symptoms and are related to the route of the involved nerve.

Pelvic MRI is emerging as the most sensitive and specific imaging study of these tumors [4,5].

Almost all retrorectal tumors require surgical management [4]. The case described herein suggests that the dimension alone should not be considered as a contraindication for an aggressive surgical approach. From a technical point of view, clear margins of resection can be difficult to obtain for these tumors because of their proximity, attachment to, or often invasion of major anatomic structures. When a major volume tumor cannot be removed, aggressive resection of other organs must be done [6,7].

Histologically, these neoplasms demonstrated contrasting fibrous and myxoid areas, a swirling, whorled growth pattern (at least in part), and bland, deceptively benign-appearing fibroblastic spindle cells. The cellularity of these tumors is low with a rich capillary vascular network visible in myxoid areas [3]. In some cases, fibrosarcoma might be difficult to distinguish from other tumors such as a dedifferentiated liposarcoma or malignant fibrous histiocytoma. The presence of a storiform pattern and epithelioid type cells would support the diagnosis of a malignant fibrous histiocytoma.

Non-surgical treatment, such as radiation treatment and chemotherapy, might improve local control making the appearance of clinically evident metastatic disease less likely. Although adjuvant therapy has enhanced the chance of cure for retroperitoneal sarcomas, there are no studies for tumors of retrorectal space. In fact, chemotherapy for retrorectal sarcomas seems to be ineffective. Thus, further studies are necessary to clarify the role of adjuvant treatment for local control of these tumors [3].

Overall survival appears to be good if the resection is complete [7]. Indeed, our patient was disease free five years after surgery.

Conclusion

Myxoid fibrosarcoma of retrorectal space is rare. Surgery remains the key of treatment and might be of an aggressive approach in managing huge tumors.

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Conflict of interest:

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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