CASE NOTE

Management of a small incidentally discovered retroperitoneal synovial sarcoma

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Synovial sarcoma is a rare aggressive soft tissue tumour accounting for 7%–8% of all cancerous sarcomas. It usually affects the extremities (85%–95%), not including rare sites of involvement, with the retroperitoneal region being the least common (0.8%–8.3%)[1].

CASE REPORT

A 33-year-old man presented to the emergency department of our clinic after an accidental fall. Physical examination revealed a healthy-appearing asymptomatic man. Immediately after the reported accident, his liver biochemistry results revealed increased aspartate aminotransferase (AST) and alanine aminotransferase (ALT) values (5.36 and 6.65 μkat/L, respectively); the other laboratory values were within normal limits.

The abdominal ultrasound revealed an hypoechoic mass in the right lower quadrant. Although repeat AST and ALT measurements were normal, we decided to perform further imaging studies on an outpatient basis. Abdominal computed tomography (CT) and magnetic resonance imaging (MRI) scans confirmed the presence of a 4-cm low-density retroperitoneal mass located in the right lower quadrant adjacent to the right ureter without invading it, occluding it or altering its course (Fig. 1). We identified no lymphadenopathy. A CT scan of the chest and bone scintigraphy were negative for metastases. The iliac artery and vein as well as their respective branches were not affected by the lesion.

We scheduled a diagnostic laparoscopy, which revealed a retroperitoneal mass in the right lower quadrant, closely adhered to the right ureter. We converted the procedure to open surgery and resected the mass en-block with the right ureter, which we immediately reconstructed in an end-to-end fashion after the intraoperative placement of the pigtail. Surgical margins were macroscopically clear.

The tumour was a solid, sharply demarcated and circumscribed elastic mass measuring 4 cm in diameter. We assessed slides for tumour type (monophasic or biphasic), percentage of necrosis, presence of poorly differentiated areas, presence of myxoid or aemangiopericytic areas and mitotic counts per 10 high-power fields. We performed immunohistochemistry for cytokeratins AE1/AE3 and epithelial membrane antigen (EMA), S100 protein, CD34 and vimentin.

The tumour consisted of both spindle-cell and glandular structures of cuboidal or columnar epithelium (Fig. 2). Myxoid change, focal calcification and hyalinization were apparent. Squamous metaplasia and necrosis were rarely observed. Mitotic activity was less than 15 mitoses/10 high-power field (HPC). Some tumour cells were positive for cytokeratins AE1/AE3 (7 and 20), EMA and vimentin. The staining was negative for CD34. The microscopic margins were without tumour involvement. Based on the histological and immunohistochemical features of the lesion, we diagnosed a grade 2 synovial

sarcoma of biphasic type without aggressive biologic behaviour. We referred the patient to a specialized centre where he received adjuvant chemotherapy with cisplatin and doxorubicin. At 2 years postoperatively, he showed no evidence of residual disease or distant metastases.

Discussion

The first retroperitoneal synovial sarcoma was described in 1955 by Pack and Tabah.[2] Since then, only 21 cases have been reported.[3] Synovial sarcoma usually occurs in middle age, attains a large size, is difficult to resect and recurs locally. This, however, was not the case with our patient, in whom a respectable synovial sarcoma was incidentally diagnosed, the smallest reported in the international literature to our knowledge.[1] Juxtaureteral location also represents a rare primary site for synovial sarcoma, contributing to the unique characteristics of the reported case.[1] The absence of screening criteria for synovial sarcoma results in late diagnosis of the respective tumour, which is characterized by disappointing histopathologic features and dismal prognosis. Presence of metastases, tumour diameter greater than 5 cm, local invasiveness, poor histological differentiation and positive margins have been reported to have a negative prognostic impact.[4] In our patient, clinical and histopathological characteristics corroborate a favourable prognosis, as evidenced by his clinical course.

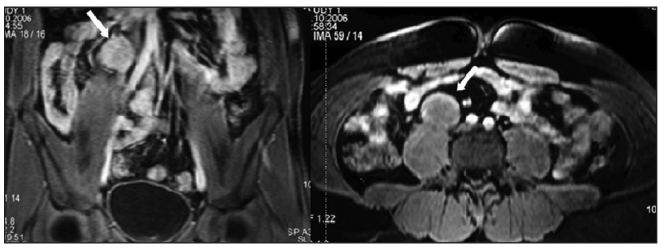

Fig. 1. Magnetic resonance imaging scans demonstrating a soft-tissue mass (white arrows) in the right lower quadrant lying on the psoas muscle.

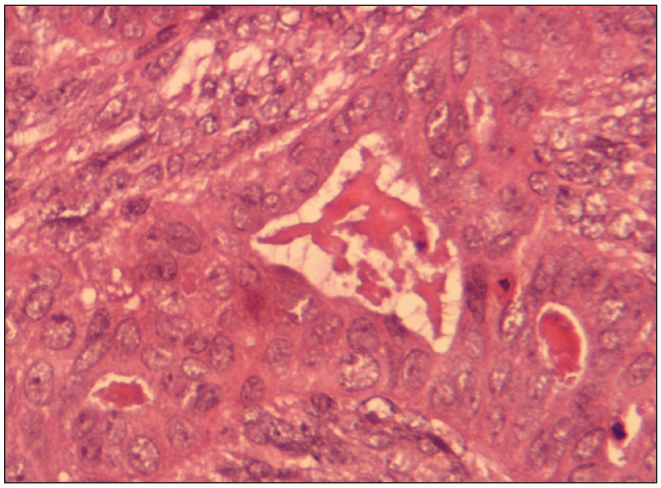

Fig. 2. Micrograph showing spindle-shaped tumour cells and apparent cleft-like spaces (hematoxylin and eosin stain, original magnification ×10).

Early diagnosis followed by definitive surgical removal of the entire tumour offers the only opportunity for cure. Synovial sarcoma has been associated with a poor outcome, and the role of adjuvant chemotherapy remains controversial.[5]

Ultrasonography, although far from helpful in assessing the retroperitoneum, provided us with a finding requiring further imaging evaluation. On the other hand, CT and MRI appear to be more helpful in defining the extent of the disease as well as tumour response to chemotherapy.[2,5]

Incidental diagnosis of synovial sarcoma while it is still asymptomatic accounts for successful surgical removal of the tumour and the favourable clinical course of the respective patient.[1] Synovial sarcoma should be always included in the differential diagnosis of retroperitoneal soft tissue tumours.

Competing interests: None declared.

References

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