Retroperitoneal synovial sarcoma

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Abstract: The case report is of a female patient with a retroperitoneal synovial sarcoma. Her clinical history, imaging examinations and management are discussed. Retroperitoneal synovial sarcoma is a difficult pathology to diagnose simply from the clinical presentation of the patient because some patients may have the disease for a long period of time before any symptoms occur and it might be completely asymptomatic. The most common manifestation of the disease is swelling and pain [1].
Keywords: Soft tissue mass, psoas muscle mass.

Case report

A 32 year old female who works as a ballet teacher was referred by her general medical practitioner to an academic hospital for investigation. During her initial consultation she presented with lower back pain and right groin pain which had severely hampered her normal activities as a ballet teacher. The pain in her groin was considerably more severe than it had been over the last few months. The pain was worse at night and was aggravated by any form of movement. Her previous medical history was unremarkable. Clinical examination revealed she had lower back pain, right groin pain, loss of weight of 15kg in the past 4 months, and loss of appetite. She was apyrexial but had tachycardia. Her blood pressure was within normal range. She complained of generalised right lower abdominal pain.

A lumber spine examination was requested to exclude tuberculosis and was reported as normal; all structures appeared intact. She then had an ultrasound scan of the abdomen which revealed a right sided groin mass. This was followed by magnetic resonance imaging of the lumbar spine. A right sided psoas mass was noted in the lower abdomen and it was partly solid and cystic (Figure 1). The right kidney was malrotated and displaced superiorly by the mass (Figure 2). The

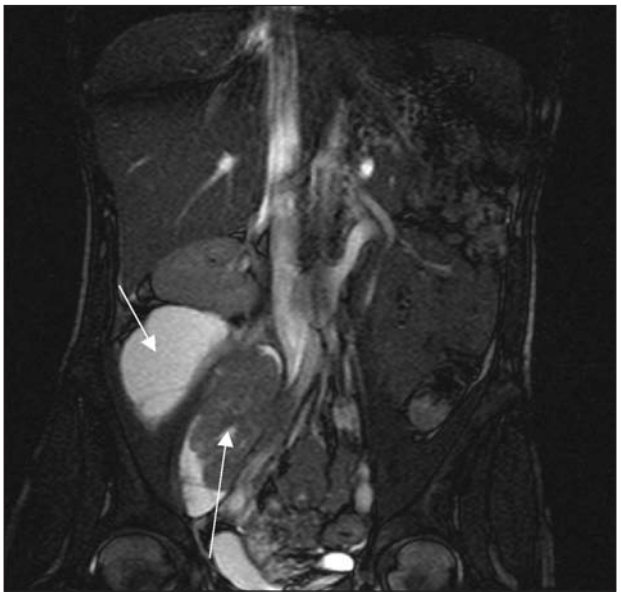

Figure 1: Coronal MR image demonstrating a cystic (long arrow) and solid (short arrow) mass of the right psoas muscle pressing on the right iliac vessels while displacing the right kidney superiorly. The mass measured 10x10cm transverse and 14 cm craniocaudally.

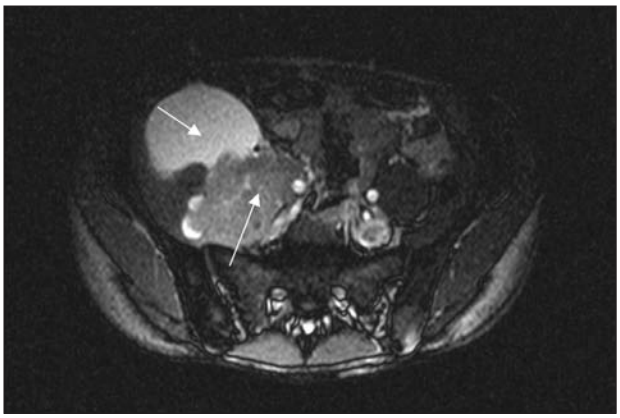

Figure 2: Axial T2 weighted MR image demonstrating the cystic (long arrow) and solid (short arrow) mass.

inferior and soft tissue component of the mass abutted the right sacrum without extension into the neural foramina. The uterus and right ovary appeared normal.The left ovary was not visualized. An ultrasound guided fine needle aspiration biopsy revealed a complex cystic/solid mass lesion noted in the right iliac fossa. The features were suggestive of a psoas mass. Histology and cytology results revealed a necrotic and poorly differentiated sarcoma, favouring a soft tissue tumour. It was noted that the specimen also included pus.

The patient underwent surgery and invasion of the right iliac vessels and compression on the right kidney by the mass was seen. Complete resection of the mass was performed with surgical repair of the right iliac vessels. In her post operative period she received adjuvant chemotherapy to prevent micro-metastases.

Discussion

Synovial sarcoma is usually a fully malignant spindle cell sarcoma of soft tissue origin. Unlike most other spindle cell sarcomas of soft tissue origin, the synovial sarcoma has been noted to metastasise to regional lymph nodes. In general retroperitoneal soft-tissue sarcomas are relatively rare and the radiological diagnosis is often difficult. Most of these tumours are malignant and frequently invade contiguous retroperitoneal organs. Surgical resection is often difficult or impossible [1]. Fine needle aspiration of a sarcoma is not a good idea because it is difficult to obtain a representative tissue sample, and the sarcoma may seed along the biopsy tract. An open biopsy, after a magnetic resonance scan to delineate the true extent of the sarcoma, as well as a of computed tomography scan of the lungs to exclude pulmonary metastases, would be a better option.

Synovial sarcoma is a well-recognized soft-tissue malignancy that typically arises in young adults [2]. It is a relatively common primary soft tissue sarcoma and accounts for approximately 5-10 % of all malignant mesenchymal neoplasms [2]. The majority (80-90%) of synovial sarcomas occur in the extremities, with approximately 60-70% in the lower limbs. Other rare sites of involvement include the neck, pharynx, larynx, thoracic spine and abdominal wall. The retroperitoneal region is the most uncommon place of occurrence [2]. Histologically there are two separate types: monophasic and biphasic. One is derived from the primitive synovial cells and the other from a malignant fibroblastic stroma [3]. These sarcomas occur with an approximately equal sex distribution [2]. Since the retroperitoneal space is highly expandable and deeply hidden, early detection of retroperitoneal synovial sarcoma is difficult [1]. Resection remains the main method of treatment for a primary soft tissue sarcoma but the resectability of a sarcoma depends on the vascularity and tenacity of adherence of the tumour mass to its surrounding structures, as well as the size and location of the mass [1]. It is difficult to do a resection of the mass because of its location near vital organs which can not be surgically removed.

The tumour has no special imaging features that distinguish it from other abdominal tumours. Although the diagnosis of a retroperitoneal sarcoma is based on the pathological characteristics of the specimen obtained by needle or surgical resection, radiographic studies suggest pre-operative diagnosis of synovial sarcoma [2]. Synovial sarcoma is an aggressive tumour and metastases or local recurrence is seen in approximately 50-70% of patients [4]. Primary retroperitoneal sarcoma is a very rare malignant tumour with a high mortality and recurrence rate. Due to a high fatality rate, physicians should always be alerted to the possibility of this pathology in the differential diagnosis of an abdominal mass.

Conclusion

Soft tissue sarcomas in adults are fairly uncommon, which partly accounts for some delays in diagnosis and complicates determination of optimal treatment. Retroperitoneal sarcoma is a rare disease but when appropriately managed the success rates may even approach that of an extremity soft tissue sarcoma which has a reduced fatality or local recurrence rate [4].

References

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Editorial comment: A reviewer stated that the role of ultrasound, in the investigation of a patient who presents with clinical signs and symptoms described in the case report, is important as it allows for a differential diagnosis of query a psoas abscess or a retroperitoneal haematoma.

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