Case report

Synovial sarcoma in an unusual site

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Introduction

Synovial sarcoma (SS) arises in tissue that contains synovium, usually in the extremities, and only infrequently in other parts of the body. The retroperitoneal space is an unusual site for SS, with only 16 cases described in the world literature (1). We report a case of retroperitoneal SS and discuss the differential diagnosis.

Case report

A 38-year old woman with 3 children was admitted to the Colombo North Teaching Hospital in May 1998. She had abdominal distension, pain in the right iliac fossa and loss of appetite for 3 months. She also complained of frequent vomiting. General examination revealed pallor. Her abdomen was distended, with a scar in the right iliac fossa. This area was tender. Percussion showed flank dullness. Rectal examination did not show any abnormality. She had undergone surgery while working as a housemaid in a Middle East country in 1996. A mass in the abdomen along with a part of the small intestine to which it was adherent had been resected. The diagnosis was benign spindle cell tumour.

On admission to our unit an ultrasound scan was performed which showed an infiltrating mass in the retroperitoneal space. At surgery a large tumour filling the retroperitoneal space was found. Several adjacent organs, including the intestines, were adherent to the tumour, and complete resection was difficult. It was removed piecemeal and a portion was sent for histology.

The patient was transferred to Cancer Institute, Maharamagama, where she received chemotherapy and radiotherapy, and was discharged with advice to attend the outpatient clinic for follow up. However, the patient could not bear the cost of travelling, and sought Ayurvedic treatment. According to relatives she developed generalised swelling and shortness of breath, and died in April 1999.

Pathology

The cut surface of the tumour showed haemorrhagic

and cystic areas. A major portion of the tumour was yellow. It was a cellular tumour composed of spindle cells of monotonous appearance with plump nuclei (Figure). The cells were arranged in fascicles with a focal whorled appearance, and a haemangiopericytoma pattern. Mitotic activity was brisk. There was little collagen present, but focal myxoid changes were seen. Strong focal cytokeratin (CK) and epithelial membrane antigen (EMA) positivity was seen in the spindle cells. However CD34, S100, desmin and actin staining were negative.

Figure. Fascicular appearance with focal haemangiopericytoma pattern. (H&E $\times 100$)

Discussion

Synovial sarcoma accounts for 5 to 10% (2) of all the soft tissue sarcomas, and may arise in any synovial tissue. About 90% of these lesions occur in the extremities (2). However, this tumour can arise in regions lacking synovial tissue, such as the head and neck region (4), heart (5), abdominal wall (6), oesophagus (7), and other rare sites such as retroperitoneum (1,3). This strengthens the theory that SS does not arise from synovial cells but from undefined mesenchymal cells (8).

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Of the 16 cases documented, the clinicopathological features of 10 cases have been reviewed (1). Most of these patients presented with abdominal pain, right iliac fossa mass and vomiting. There were equal numbers of biphasic and monophasic SS in this series. Only one patient was alive 5 years after complete removal of the tumour with no distant metastases being reported.

The differential diagnosis of monophasic synovial sarcoma (MSS) at this site includes leiomyosarcoma, malignant peripheral nerve sheath tumour and fibrosarcoma. MSS was not considered in our initial differential diagnosis as the retroperitoneum is an unusual site. However, the immuno-phenotype was compatible with MSS, approximately 80% being CK and EMA positive (9). In tumours where the immunophenotyping is not clearly indicative of SS, cytogenetic studies will reveal a consistent, specific, balanced translocation in over 90% of the tumour (10). SS is an important differential diagnosis in spindle cell tumours of the retroperitoneum, but often overlooked because this is a rare site of occurrence.

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Maintaining morale during wartime

In these dark days the Prime Minister would be grateful if all his colleagues in the Government, as well as high officials, would maintain a high morale in their circles; not minimising the gravity of events, but showing confidence in our ability and inflexible resolve to continue the war until we have broken the will of the enemy to bring all of Europe under his domination.

A memo from Winston Churchill to cabinet members and all senior officials, 1941.