Monophasic synovial sarcoma of the greater omentum: case report and review of literature

FJ Alcalá Serrano¹, JR Hernández Hernández¹, T Montenegro Dámaso², E López-Tomassetti Fernández¹

¹General Surgery Department, Hospital Universitario Insular de Gran Canaria, Las Palmas, Spain

²Pathology Department, Hospital Universitario Insular de Gran Canaria, Las Palmas, Spain

ABSTRACT

Synovial sarcoma is a malignant spindle cell neoplasm normally arising from tissues around joints, bursa and tendon sheaths. Several reports involving the gastrointestinal tract, mainly the oesophagus and stomach, have been documented; however, the omentum remains an extremely unusual location. Monophasic type is composed exclusively of spindle cells arranged in fascicles. Establishing the correct diagnosis of these tumours could be challenging because of the similarities with gastrointestinal stromal tumours and other mesenchymal tumours with similar histology.

KEYWORDS

Synovial sarcoma – Greater omentum – Pathology – Digestive surgery – Immunohistochemistry

Accepted 3 April 2017

CORRESPONDENCE TO

Francisco Javier Alcalá Serrano, E: fjavi7alcser@hotmail.com

Case Report

A 53-year-old woman was referred to our institution after months of mild abdominal pain associated to hyporexia and weight loss. Her past medical history was unremarkable, only high blood pressure and renal colic were recorded. Physical examination showed a tender, palpable mass in the left upper quadrant. On admission, laboratory tests revealed a $12.800/\text{mm}^3$ white cell count with no other abnormality. Both upper endoscopy and colonoscopy were normal.

Thoracoabdominal computed tomography (CT) showed a multilocular and cystic mass in the upper left quadrant measuring $190 \times 120\text{mm}$ with displacement of the stomach, the transverse colon and small bowel. The right gastroepiploic artery was enlarged surrounding the tumour mass (Figure 1). A suspicious diagnosis of multicystic mesothelioma or peritoneal pseudomyxoma was made and percutaneous ultrasound-guided of the mass was decided, which evidenced necrotic areas and a non-specific low-grade stromal proliferation.

The patient was scheduled for exploratory laparotomy. Surgical findings demonstrated a huge mass arising from the greater omentum and fixed to the left parietal peritoneum with adhesions, surrounded with free serohaemorrhagic fluid from a spontaneous rupture of the capsule. No peritoneal or liver metastases were observed. According to these findings, an omentectomy with en-bloc gastric wedge resection and left lateral peritonectomy were performed to obtain free margins.

Macroscopically, the surgical specimen measured $15 \times 8 \times 4\text{cm}$ in size and consisted of a yellowish soft mass with cystic haemorrhagic and necrotic areas on sectioning. Microscopic examination revealed a sarcomatous hypercellular monomorphic spindle-shaped cell proliferation with haemangiopericytoma-like areas. Several mitotic figures could be seen in the tumour cells. On immunohistochemistry, spindle cells were positive at staining for vimentin, B-cell lymphoma 2 and CD99 antigen and negative for epithelial membrane antigen, smooth muscle actin, desmin, S100 protein, CD34 and creatine kinase. Applying fluorescence in-situ hybridisation (FISH) on paraffin embedded-tissue sections, molecular analysis demonstrated a rearrangement of gene SS18 (SYT) at 18q11 on 95% of the tumour cells, thus confirming the suspected diagnosis of monophasic synovial sarcoma. Margins of the resected tumour and peritoneal washing cytology were negative.

The patient was discharged on postoperative day 12 and received four courses of doxorubicin postoperatively. CT was performed at three months, six months and one year after surgery and evidenced no signs of recurrence.

Discussion

Synovial sarcoma is a rare neoplasm that accounts for around 10% of all soft tissue sarcomas.¹ Synovial sarcoma is usually found in young adults and in the extremities, in relation with synovial structures, joint capsules, bursae and tendon sheaths, but it also occurs in regions lacking synovial

tissue. Three different patterns have been identified: the monophasic type, composed exclusively of spindle-shaped cells; the biphasic type, which consists in both epithelial and spindle cells; and the poorly differentiated type, with worse prognosis and rhabdoid features.

Soft-tissue sarcomas are a heterogeneous group of malignant neoplasms of mesodermal origin. From the greater omentum arise unique cells called human peritoneal mesothelial cells, which display dual mesenchymal and epithelial characteristics and express intermediate filaments typical of both mesoderm (vimentin) and epithelium (cytokeratins). A crucial event is the epithelial to mesenchymal transition, during which epithelial cells convert into fibroblast-like cells. Adult mesothelial cells seem to retain much of this potential for differentiation, which could lead eventually to a sarcomatoid differentiation simulating synovial tissue.

Usually, intra-abdominal synovial sarcoma presents with a very vague and non-specific symptomatology because of the distensibility of the abdominal cavity, which allows them to grow until they compress neighbouring structures. Our search of the English-language literature yielded no more than 50 cases of intra-abdominal synovial sarcoma, most of them documented as case reports. Retroperitoneal sarcomas are more frequently identified than those arising from the gastrointestinal tract, as Fisher *et al.*² noted in a review of 33 cases. Among the cases in the gastrointestinal tract, oesophageal synovial sarcomas were first identified. To our knowledge, only two cases located at the omentum have been published.⁵

Radiographic characteristics of synovial sarcoma are non-specific, normally showing a hypodense soft-tissue mass with no pathognomonic imaging features. Nonetheless, CT is useful determining the extent of the disease and following the response to chemotherapy.⁴

Curative treatment for these tumours consists of wide surgical excision followed by adjuvant chemotherapy. Different schemes of doxorubicin or ifosfamide plus mesna have had encouraging results for SS with acceptable toxicity.

Voluminous masses (greater than 5cm), young age, high mitotic count, retroperitoneal and pelvic location and poorly differentiated component seem to be related to worse prognosis,^{2,5} while vascular and perineural invasion are presumably associated with recurrence and metastatic disease.⁵

Diagnosis of synovial sarcoma is made based on pathological characteristics of the specimen. Both monophasic and biphasic patterns show proliferative sarcomatous cells with scant cytoplasm arranged in fascicles, while the monophasic consists solely of spindle cells. Since gastrointestinal stromal tumour (GIST) is the most frequent intra-abdominal mesenchymal tumour, it is usually the primary differential diagnosis with synovial sarcoma. GISTs have abundant cytoplasm and are positive for c-Kit, CD34 or both, while synovial sarcoma is negative for these factors.

Regardless of the synovial sarcoma subtype and anatomical site, tumour cells show a translocation of chromosomes X and 18, resulting in three alternative fusion products of the SS18 gene (SYT) with either the SSX1, SSX2 or SSX4 gene. Molecular techniques as FISH or reverse transcriptase polymerase chain reaction are accurate in detecting this gene rearrangement, which is specific of synovial sarcoma.

Awareness of this rare neoplasm should alert the physician to further investigate unknown intra-abdominal masses and to highlight the pathological and immunohistochemical study to separate them from other mesenchymal tumours such as c-kit negative GISTs and carcinosarcoma.

A correct diagnosis, with the advent of immunohistochemistry and molecular analysis, would allow a more targeted, effective treatment and better prognosis in patients with synovial sarcoma in the near future.

References

1. Suurmeijer AJH, de Bruijn D, Geurts van Kessel A *et al.* *Synovial Sarcoma*. Lyon: IARC Press; 2013.
2. Fisher C, Volpe AE, Hashimoto C *et al.* Intraabdominal sinovial sarcoma: a clinicopathological study. *Histopathol* 2004; **45**: 245–253.
3. Wang YJ, Wen SC, Chien ST *et al.* Primary intraabdominal sinovial sarcoma. *J Chin Med Assoc* 2006; **69**(10): 492–495.
4. Ko SF, Chou FF, Huang CH *et al.* Primary synovial sarcoma of the gastrocolic ligament. *Br J Radiol* 1998; **71**: 438–440.
5. Kamata K, Wada R, Yajima N *et al.* Primary sinovial sarcoma: molecular diagnosis and prediction of prognosis. *Clin J Gastroenterol* 2013; **6**: 303–308.