CASE REPORT / Olgu Sunumu

Intraabdominal Synovial Sarcoma, A Rare Tumour and Rare Localisation

İntraabdominal Sinovyal Sarkom, Nadir Lokalizasyonda Nadir Bir Tümör

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Abstract

Synovial sarcomas are often seen in the soft tissues, intra-abdominal synovial sarcomas are extremely rare. An intra-abdominal mass starting from the umbilicus and extending into the pelvis was detected in a 53-year-old man admitted with abdominal pain and a palpable mass. Total mass excision surgery was performed and the mass was found to be an intra-abdominal synovial sarcoma arise from small bowel wall histopathologically. Unfortunately, this is a malignancy with a poor prognosis and a short survival despite all therapeutic efforts. **(Sakarya Med J, 2018, 8(2):669-673)**

Keywords intra-abdominal mass; sarcoma; small bowel wall; synovial sarcoma

Öz

Sinovyal sarkomlar sıklıkla yumuşak dokuda gözlenirler, intra abdominal sinovyal sarkomlar oldukça nadirdir. Karın ağrısı ve palpabl kitle ile başvuran 53 yaşında erkek bir hastada umbilikustan başlayıp, pelvise kadar uzanım gösteren karın içi kitle saptandı. Total kitle eksizyonu yapılan hastada, histopatolojik olarak ince barsak duvarından kaynaklanan intraabdominal sinovyal sarkom bulundu. bu malignite zayıf prognoza sahiptir ve yapılan tüm tedavilere rağmen sağkalımı kısadır.**(Sakarya Tıp Dergisi, 2018, 8(2):669-673).**

Anahtar kelimeler ince barsak duvarı; intraabdominal kitle; sarkom; sinovyal sarkom

Introduction

Synovial sarcoma is a rare malignancy accounting for 5~10% of all soft-tissue sarcomas. It has an aggressive course and poor prognosis, with a reported mean 5-year survival of 66%.[1] It is typically encountered in the third to fifth decades of life with a male preponderance. In 95% of the cases, the tumour is localised to the extremities, most commonly around the knee.[2] Rarely, tumours originating from the retroperitoneum, tongue, neck, oesophagus, and abdominal wall have been reported. The recommended treatment is surgical excision followed by chemotherapy and radiation therapy. Here, we report a 53-year-old man who was admitted with a palpable intra-abdominal mass and was ultimately diagnosed with an intra-abdominal synovial sarcoma.

Case report

A 53-year-old man presented with abdominal pain for 4 months and a palpable abdominal mass (Fig. 1).

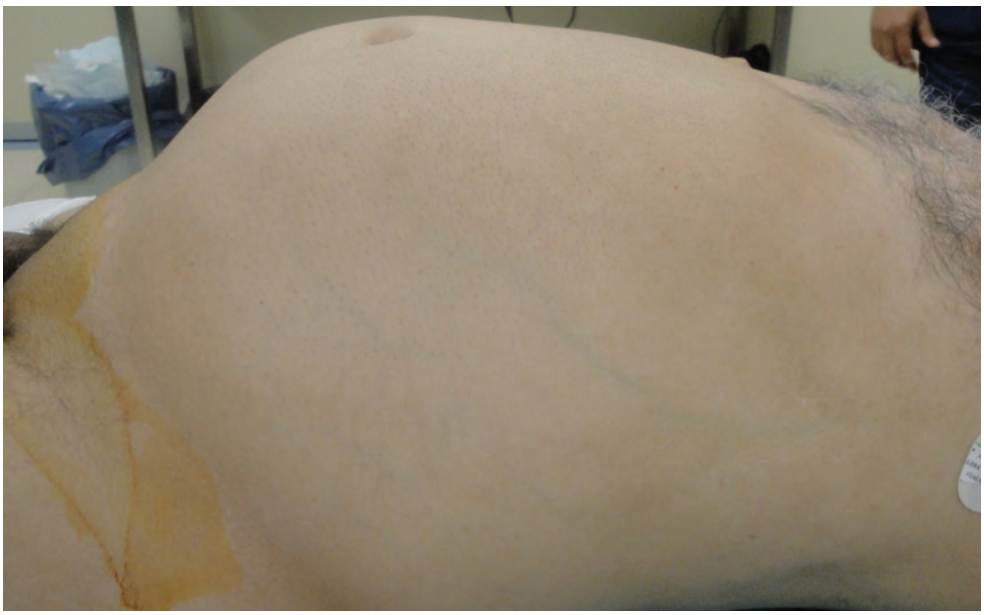
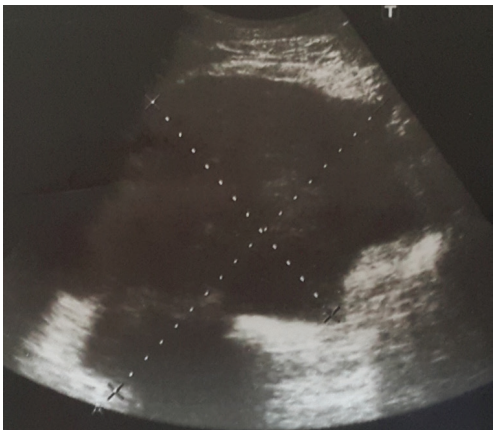


Figure 1. Preoperative patient's view; apparent palpable intraabdominal mass

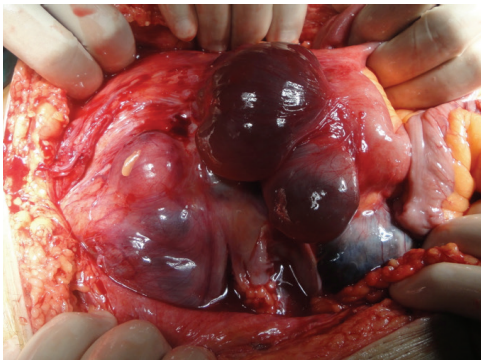
Physical examination detected a mass starting from the periumbilical region with indistinct borders. Abdominal ultrasonography (US) revealed a mass extending from the umbilical level to the pelvis, with heterogeneous cystic and degenerated regions and irregular margins. The mass filled the entire lower abdominal region (Fig. 2). The symptoms of an intestinal obstruction emerged and we performed surgery. After obtaining written informed consent, surgical exploration showed a 280 × 180 mm mass with lobulated contours and some cystic components, although it was mainly solid in nature, extending from the umbilical level to the pelvis, surrounding a 20-cm ileal segment (Fig. 3A-B). The mass was excised totally and a partial ileal resection and terminal ileostomy were performed. An iatrogenic injury to the urinary bladder during the surgery was repaired primarily. The immunohistochemical staining showed that the major cell types were spindle and epithelial cells revealed a synovial sarcoma with CD99 and EMA positive, but were negative for S-100, protein, desmin, CD34. (Fig. 4,5)

In the postoperative course, the patient had acute renal failure, aspiration pneumonia. The overall



condition of the patient worsened in the 8th postoperative day and he died of multi-organ failure on day 10.

Figure 2. Ultrasound image; a mass extending from the umbilical level to the pelvis, with heterogeneous cystic and degenerated regions and irregular margins



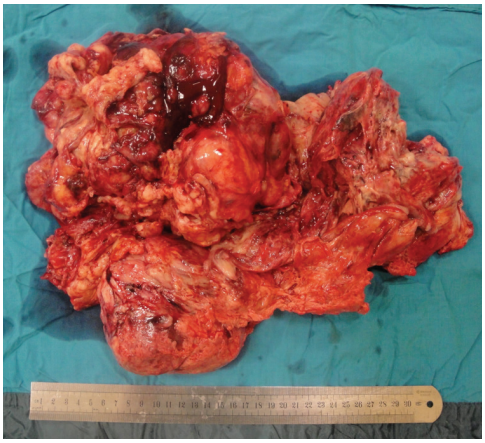


Figure 3A. Operative view; intraabdominal mass is huge, lobulated contour and solid-cystic in nature.
Figure 3B. Operative view; intraabdominal mass is huge, lobulated contour and solid-cystic in nature.

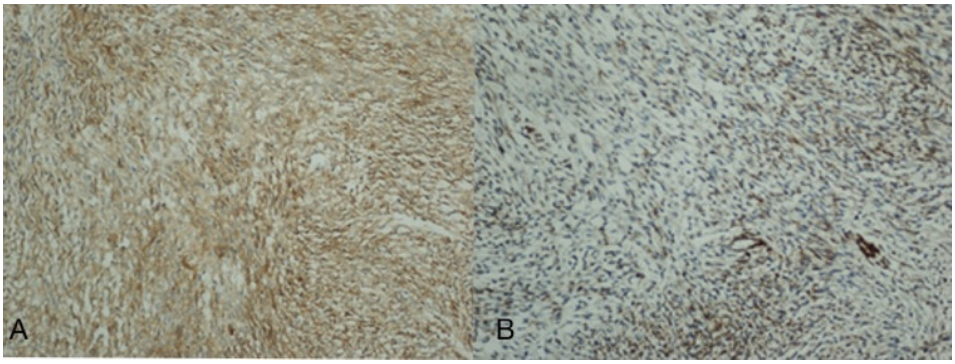


Figure 4. Histological microphotograph of synovial sarcoma, stained with CD99 x20

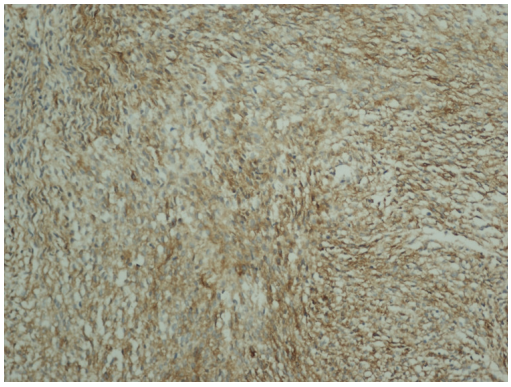


Figure 5. Histological microphotograph of synovial sarcoma, stained with EMA x20

Discussion

Synovial sarcomas are rare aggressive tumours that typically arise in the periarticular areas[2]. Other rare anatomic locations of this tumour are the thorax, neck, parathyroid, tongue, larynx, mediastinum, oesophagus, heart, lung, abdominal wall, gastrocolic ligament, small bowel mesentery, prostate, kidney, and retroperitoneum.[3,4] In this case the tumor arose from small bowel wall.

Clinically, an intra-abdominal synovial sarcoma often presents as a painful, palpable soft mass, as with tumours in other locations. Non-specific gastrointestinal complaints such as abdominal pain, bloating, weight loss, and vomiting may accompany the clinical presentation, depending on the size of the intra-abdominal mass.[3] In this case, the abdominal pain had been present for 4 months, but was overlooked by the patient, until a palpable mass was detected.

Intra-abdominal localisation is an extremely rare presentation of synovial sarcomas. In a retrospective study of 300 cases, Fisher et al. reported that only 11 (3.6%) of the patients had an intra-abdominal mass.[5] Those 11 cases had a mean age of 49 (range 25–75) years and a mean mass size of 6 × 47 cm. In our patient, the mass measured 28 × 18 cm and the patient's age was within the reported age range for synovial sarcomas.

Histopathologically, a synovial sarcoma is a soft-tissue tumour that shows signs of mesenchymal and epithelial differentiation, which can be detected under a light microscope, immunohistochemically, and by electron microscopy.[6] Although the diagnosis of biphasic synovial sarcomas is often straightforward, a monophasic fibrous synovial sarcoma should be differentiated morphologically from fibrosarcoma, malignant peripheral nerve sheet tumour, and solitary fibrous tumour.[7] Routine use of immunohistochemical methods may facilitate the differential diagnosis of these conditions. Changchien et al. reported primary or metastatic intra-abdominal synovial sarcomas stained with SMA, EMA, BCL-2, CD99, and S100, but not with desmin.8 The monophasic and biphasic types were differentiated by theFluorescence In Situ Hybridization (FISH) method using SYT-SSX-1. In a study of 121 cases with a synovial sarcoma, Bergh et al. reported that 60% of the cases were monophasic and 40% the biphasic type.[9] Guillou et al. reported that the monophasic type had a more aggressive course.[10] In our case, staining was positive for CD99 and EMA, histopathological study confirmed the diagnosis of monophasic synovial sarcoma.

The major determinant of survival is the histological grade of the tumour, and the dimensions and depth of the tumour are other prognostic factors.[8] Wide local excision and radiation therapy are mainstays in the treatment of synovial sarcomas. Chemotherapy has been also suggested in the treatment of some histological subtypes.[9] Nevertheless, half of the cases die from distant metastasis despite all treatment modalities.

In conclusion, although synovial sarcomas are rarely seen outside the extremities, they can also present as an intra-abdominal mass, albeit extremely rarely. Wide excision is the first step in the treatment. Unfortunately, this is a malignancy with a poor prognosis and a short survival despite all therapeutic efforts.

Conflicts Of Interest: No potential conflict of interest relevant to this article was reported.

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