Primary sclerosing liposarcoma of the ovary: a case report and review of the literature

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Abstract

Primary sarcomas of the ovary are rare tumours, comprising under 4% of all histologically diagnosed ovarian malignancies. Most reports in literature are either in the format of a single case report or a small case series.

Primary liposarcoma of the ovary represents an extremely rare tumour, with only three cases of myxoid variant being reported in the literature so far.
Here we report a case of a primary sclerosing liposarcoma of the ovary arising in a 67-year old lady. To the best of our knowledge, this is the first reported example of a sclerosing variant of liposarcoma arising in the ovary.

Introduction

Ovarian sarcomas represent rare pathologic rarities with only a few cases reported in the literature. These include fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, angiosarcoma, rhabdomyosarcoma and osteosarcoma {1}.

We report a case of a primary sclerosing liposarcoma of the ovary arising in a 67-year old lady, which showed confirmatory MDM2 amplification on fluorescent in-situ hybridisation (FISH) analysis. Only three other cases of ovarian liposarcoma have been reported in the literature {1, 2, 3}, all of which have been of the myxoid subtype. Being in an uncommon location, it may be difficult to differentiate these lesions from other primary ovarian tumours with an adipocytic component and overlapping sclerosing features. Hence, the review of reported case will be given along with differential diagnosis, diagnostics and current therapy treatments.

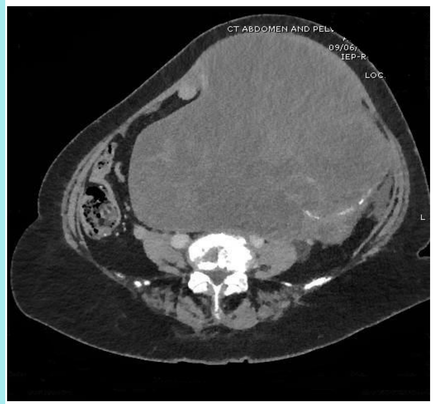

Figure 1. Last CT scan shows large multinodular mass occupying large area of abdomen and extending to the retroperitoneal space. Previous CT scans showed mass originated from the ovary.

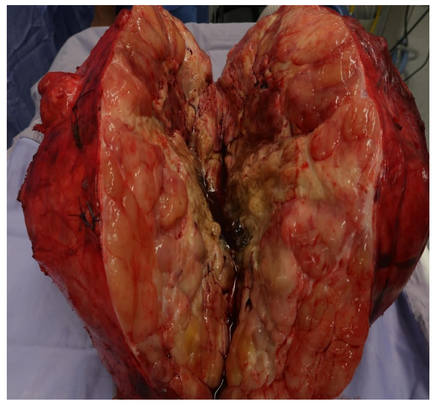

Figure 2. The tumour at the time of surgery as a voluminous multinodular mass, apparently encapsulated with a prominent vascular network, central haemorrhage and ischemic necrosis

Case presentation

The patient was a 67-year old female who presented as an emergency admission with abdominal discomfort, constipation and urinary incontinence. Her serum CA125 was unremarkable at 23.4 U/mL. CT scan demonstrated a complex left pelvic mass with multiple septations and calcifications, arising from the adnexa and compressing the left ureter (Figure 1). No areas suspicious for pelvic metastases were seen. MRI suggested the possibility of teratoma, although the appearances were not entirely specific. Following multidisciplinary team discussion she underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy. By the time of surgery, the mass had increased in size to her xiphisternum, and had grown retroperitoneally with attachment to the large abdominal vessels.

Macroscopically, the uterine cavity was deformed and displaced by an ill-defined, nodular white tumour with budding into the surrounding tissue. The cut surface was soft, grey and haemorrhagic (Figure 2).

Histologically the tumour had a heterogeneous appearance with both hypercellular and low-cellularity areas, divided into lobules by thick fibrous septa. The lesion comprised atypical pleomorphic cells with spindled, oval and stellate-like nuclear forms. The cells showed a compact, nested arrangement with storiform patterns, set in abundant and focally myxoid stroma with less prominent areas of hyalinisation. There was a prominent arborizing vascular network throughout the lesion, and interspersed foci of mature adipose tissue including occasional lipoblasts. Widespread coagulative necrosis was present (Figure 3).

With immunostaining the tumour cells showed diffuse strong positivity for p16 and S-100, while CD34 and CDK4 showed patchy strong staining in around 80% of cells. MDM2 demonstrated nuclear staining of variable intensity in 30-40% of cells. MUC4 and STAT6 were negative (Figure 4).

The case was sent to a tertiary/referral centre for confirmation of diagnosis and molecular analysis. Positive MDM2 gene amplification was detected by interphase FISH analysis, confirming the diagnosis of well-differentiated sclerosing liposarcoma with areas of low grade de-differentiation.

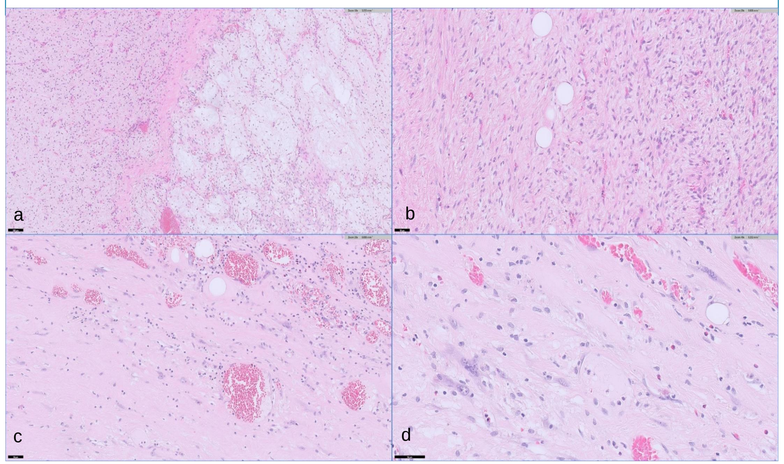

Figure 3. Microscopically tumour is characterized by multilobular proliferation of atypical adipocytes with focally myxoid stromal component (Figure 3a); mature looking adipocytes are focally present (Figure 3b); prominent arborizing and focally ectatic capillaries are seen throughout the lesion, surrounded by abundant hyalinised/sclerosing stromal component (Figure 3c and 3d)

Discussion and conclusions

We report the first confirmed case of primary ovarian sclerosing liposarcoma, and only the fourth published case of liposarcoma of the ovary. Primary pure soft tissue sarcomas of the ovary are rare lesions. In such cases, metastasis from a primary soft tissue tumour is far more likely and should always be considered in such cases with clinical and radiological correlation. Extensive clinical and radiologic investigation in our case largely excluded the fact that ovarian tumour represented metastatic disease. Similarly, a retroperitoneal/intra-abdominal liposarcoma has been excluded on the basis that the tumor was a well-circumscribed mass initially contained within the ovary. The myxoid subtype is the most common type of liposarcoma known to arise in children and adolescents. Liposarcomas can also arise from elsewhere in the female pelvic organs, but are most commonly ovarian in origin as in this case. There is also a report of a retroperitoneal de-differentiated liposarcoma mimicking a primary ovarian tumour {4}.

On account of the characteristic features of liposarcomas, the differential diagnosis is relatively narrow for a tumour showing this morphology. However, it can also show more heterogenous appearances in the examples seen from other sites. Differential diagnoses include myxoid proliferations such as myxomas, followed by other ovarian tumors, such as germ cell and sex cord-stromal tumours. In particular, Sertoli-Leydig cell tumor, with an extensive myxoid change, as well as sclerosing stromal tumor of the ovary should be considered. Immunohistochemistry can be used to distinguish between these entities. In this case, FISH molecular analysis was used as a confirmatory diagnostic adjunct due to the well-recognised MDM2 amplification that is commonly associated with liposarcomas {5}.

Due to the rarity of ovarian sarcomas, there is limited data on the clinical behaviour and the optimal management of these tumours. In the case of our patient, the clinical course was rather dramatic as she presented with an enormous mass, which had grown significantly since her CT of a few months earlier and was densely attached to the big abdominal vessels, resulting in profuse intraoperative bleeding.

Complete en-bloc resection with a margin of uninvolved tissue is the current mainstay of treatment. Despite surgical resection with clear margins, the risk of developing relapsed disease in abdominal liposarcomas remains high; however, outcomes are improved when treatment is delivered at specialist centres with experience in managing this rare disease. {6}

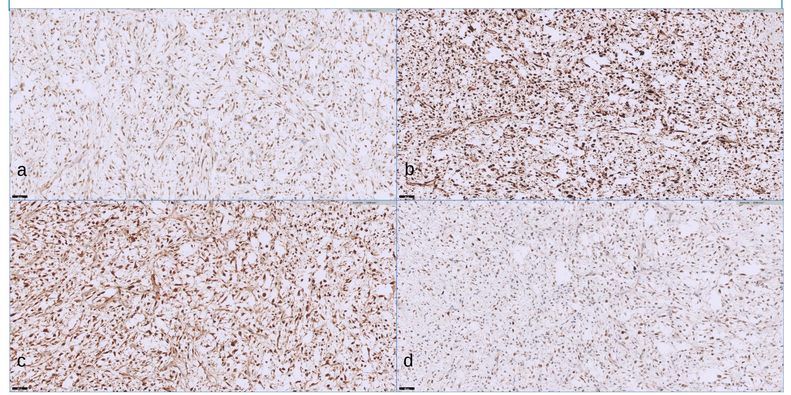

Figure 4. The immunohistochemistry shows expression of CDK4 (Figure 4a), p16 (Figure 4b), S-100 (Figure 4c) and MDM2 (Figure 4d) in neoplastic cells

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Declaration of conflict of interest

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Full informed patient consent was obtained for this case report.