CASE REPORT

Primary ovarian rhabdomyosarcoma

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SUMMARY

Sarcomas represent a diverse and extremely complex group of malignancies that can arise from mesenchymal tissue at any site in the body. It is believed rhabdomyosarcoma (RMS) arises from immature cells destined to form skeletal muscles. There are, however, instances where these tumours can occur in locations where skeletal muscles are not typically present. RMS of the ovaries is an extremely rare malignant ovarian tumour. We present a case of 45-year-old woman with progressive abdominal distension and significant weight loss. A fixed pelvic mass and palpable left supraclavicular lymph nodes were discovered during clinical examination. Computed Tomography scans revealed bilateral adnexal masses as well as pelvic and para-aortic lymphadenopathy. A diagnosis of RMS of the ovary was obtained after excisional biopsy of the left supraclavicular lymph node and ultrasound-guided biopsy of the ovarian tumour. We also discuss the difficulties encountered throughout the patient's diagnosis and treatment. She underwent neoadjuvant chemotherapy before cytoreductive surgery after being diagnosed with advanced ovarian RMS. Unfortunately, she died after the first course of chemotherapy due to cancer progression.

INTRODUCTION

The most common soft-tissue sarcoma in childhood is rhabdomyosarcoma (RMS), which usually affects the genitourinary tract, extremities, and head and neck region. The genitourinary tract includes the urinary bladder, prostate, and paratesticular soft tissues.[1] In contrast, primary ovarian RMS is an extremely rare tumour, which can cause diagnostic dilemmas for pathologists and clinicians alike. An accurate diagnosis is crucial, because RMS treatment is different from other tumours that happen in the ovary.[2] Predicting the biological behaviour of primary ovarian RMS is difficult due to a scarcity of evidence.[1] Here, we provide one such case from both a clinical and pathological standpoint.

CASE REPORT

A 45-year-old nulliparous woman with no previous comorbidities presented with 2-month-old lower abdomen pain and distension, as well as considerable weight loss. There had been no changes in bowel or urine patterns in the past.

On general examination, there was a palpable 2×2 cm left supraclavicular lymph node. A fixed pelvic mass equivalent to the size of a gravid uterus at 24 weeks was discovered during an abdominal examination. There were no signs of ascites. Upon pelvic examination, a fixed mass was felt at the right adnexa.

A computed tomography (CT) scan revealed a solid adnexal tumour emerging from the right ovary, measuring 9×11×18 cm (Figure 1a and 1c). There were numerous enlarged lymph nodes (up to 2.5 cm) in both iliac regions, as well as an enlarged left supraclavicular lymph node (Figure 1b) and matted para-aortic lymph nodes from the aortic bifurcation to the renal level (Figure 1c). There was minimal ascites present, but no metastatic lesions in the lungs or liver. CA125, CA19-9, and alpha-fetoprotein (AFP) laboratory tests all came back with increased levels (411U/ml (normal 35), 58U/ml (normal 34), and 21.6IU/ml (normal 5.8), respectively. At this point, we were considering advanced epithelial ovarian malignancy.

An excisional biopsy of the left supraclavicular lymph node was performed, as well as an ultrasound-guided biopsy of the ovarian tumour. Microscopic examination of both specimens showed tissue infiltrated by sheets and clusters of malignant cells with minimal fibrous stroma. The malignant cells display monotonous small to medium size nuclei, inconspicuous nucleoli, and scanty cytoplasm. Mitosis and apoptotic bodies were brisk. The tumour cells were immunopositive for desmin, myogenin (nuclear), synaptophysin, INSM-1, WT-1 (cytoplasmic), and INI1/SMARCB1 (nuclear) while immunonegative for pancytokeratin, epithelial membrane antigen, calretinin, CK20, chromoganin, OCT3/4, CD45, and GATA-3.

Small cell carcinoma of the ovary of hypercalcemic type (SCCOHT), small cell carcinoma of the ovary of pulmonary type (SCCOPT), neuroendocrine carcinoma, and other entities, including epithelial, germ cell, lymphoproliferative disorders, desmotic small round cell tumours, and nephroblastomas have been ruled out by immunochemistry. After a thorough examination of the patient, radiologically, and pathologically, we concluded that it was a primary ovarian rhabdomyosarcoma (RMS).

In view of the advanced disease, she underwent the first cycle of chemotherapy VAC regime (vincristine, actinomycin-D,

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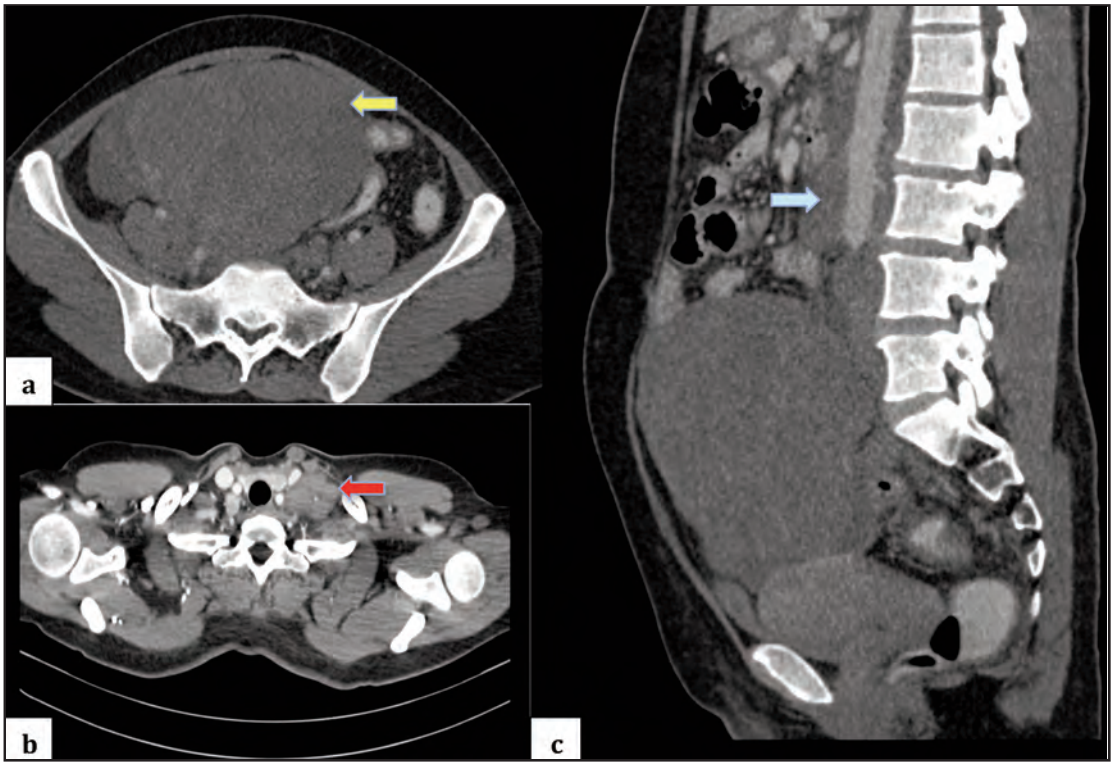

Fig. 1: Computed tomography (CT) imaging
a) Axial CT image showing adnexal mass (yellow arrow) insinuating into the right hemipelvis
b) Axial CT image showing enlarged left supraclavicular node (red arrow)
c) Sagittal CT image showing adnexal mass in relation to the uterus inferiorly and the matted paraaortic nodes (blue arrow)

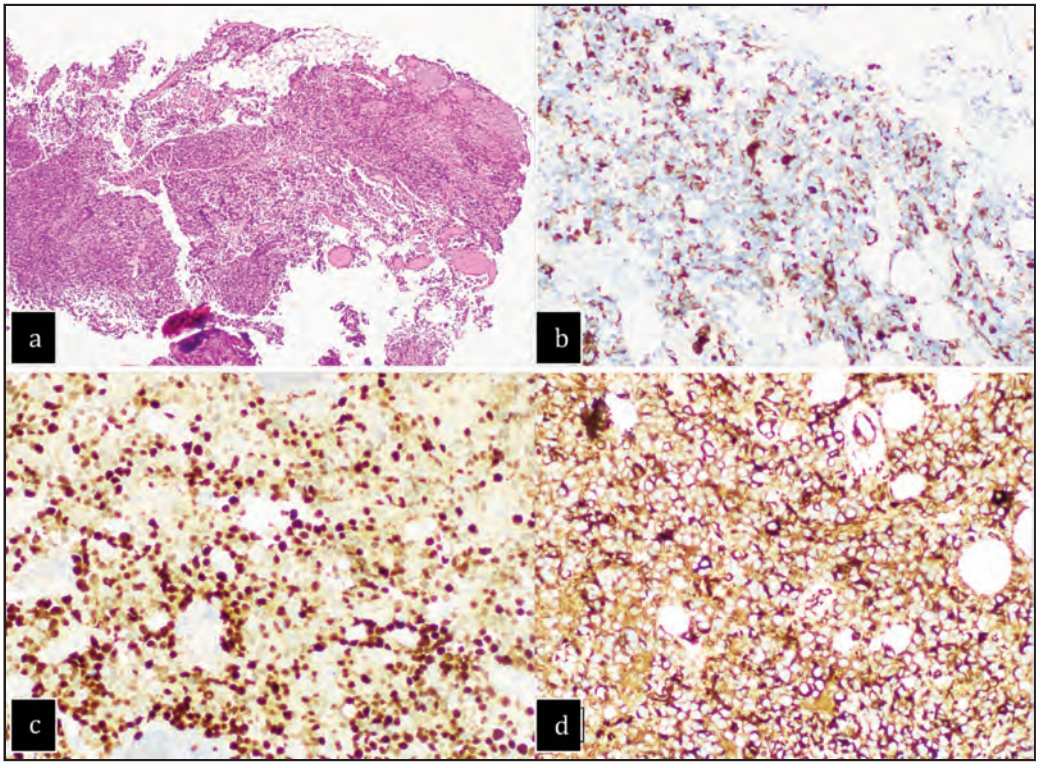

Fig. 2: Microscopic examination findings
a) Diffuse sheets of undifferentiated tumour cells with extensive tumour necrosis (H&E ×40 magnification)
b) Desmin; mesenchymal stain for myogenic differentiation (×200 magnification)
c) Myogenin nuclear positivity. Highlights skeletal muscle differentiation (×200 magnification)
d) WT-1 cytoplasmic positivity (×200 magnification)

and cyclophosphamide). However, her condition deteriorated, and she developed obstructive uropathy and upper gastrointestinal bleeding. Subsequently, she succumbed to death.

DISCUSSION

The most common soft tissue sarcoma in children, accounting for more than half of all soft tissue sarcomas, is RMS. RMS, on the other hand, is extremely uncommon in adults. Soft tissue sarcomas account for less than 1% of all malignancies in adults, while RMS accounts for 3% of these.[3,4] RMS is sporadic in nature, with no known cause or risk factors. The disease, however, is linked to a number of familial syndromes, including Noonan, neurofibromatosis, Beckwith-Wiedemann, Li-Fraumeni, and Costello.[4]

A variety of factors contribute to the different clinical manifestations of RMS, including the location of the origin, the patient's age, and whether or not distant metastases are present. In the course of an initial evaluation of a patient with suspected RMS, it is important to determine the anatomic boundaries of the tumour. All these factors will influence the stage of disease, risk stratification, and eventually the course of treatment.

In conjunction with a thorough physical examination and pelvic ultrasound, a CT scan is useful in assisting the diagnosis and detection of tumours as well as the presence of metastases. Prior to the diagnosis of primary ovarian RMS, a thorough evaluation of the tumour is essential to exclude metastases from other sites or mixed components, such as teratoma, mixed mesodermal tumours, or Sertoli-Leydig cells.[1,2] In our case, clinical and radiological findings did not show any other primary sites. Furthermore, histopathological examination found no evidence of germ cell tumours. Therefore, a final diagnosis of primary ovarian RMS was made.

Guérard et al.,[5] reported that ovarian RMSs are extremely rare and fatal. They documented 14 cases of ovarian RMS ranging in age from 13 months to 86 years, with the majority being over the age of 40. Approximately half of the cases had cancer that had spread beyond the ovary, and survival ranged from 18 days to 15 months after diagnosis. Patient survival appears to be improved through aggressive chemotherapy, radiation, and surgery.

Nielsen et al., subsequently reviewed 13 cases, of which 11 had embryonal histology and 2 had alveolar histology.[6] A follow-up of these patients revealed a poor outcome as 7 of them died within 10 days to 26 months. Cribbs et al., described two paediatric patients with advanced primary ovarian RMS; both underwent complete resection of their primary tumours and were treated with vincristine, doxorubicin, and cyclophosphamide, which resulted in a favourable outcome.[7]

Qureshi et al. reported a case of a 21-year-old woman with pleomorphic ovarian RMS who underwent two surgeries but refused chemotherapy or radiotherapy and was well through the last follow-up.[1] In an article by Ezem et al., a 13-year-old girl was diagnosed with primary ovarian RMS after laparotomy bilateral ovariectomy.[8] Postoperatively, the patient presented with metastatic deposits to the spine. However, she could not afford chemotherapy or radiotherapy. She passed away 93 days after the presentation.

Vanidassane et al. recently described a case of ovarian RMS in a young girl.[2] When an ovarian epithelial tumour was suspected, washings, an omentectomy, and a left salpingo-oophorectomy were performed. Histopathology initially suggested a low-grade myxoid fibrosarcoma. The patient developed a recurrence after a month, and subsequent morphological and immunohistochemical findings confirmed that it was embryonal RMS. A final diagnosis of primary ovarian RMS stage 4 was reached after a thorough pathological, clinical, and radiographic examination. The patient was treated with a high-risk VAC regimen that included vincristine, actinomycin D, and cyclophosphamide. As stated in the case report, the patient is in the midst of treatment.

RMS risk stratification includes histologic classification, pre-surgical stage, and post-surgical clinical group. Surgery, chemotherapy, and radiotherapy are all treatment options. VAC regimen (vincristine, actinomycin D, and cyclophosphamide) and the IVA regimen (ifosfamide, vincristine, and actinomycin D) are the two main chemotherapy regimens used to treat RMS. Ionizing radiation (IR) therapy has been found to be an effective tool in reducing disease recurrence rates, and it is frequently used in patients with advanced stages of cancer. When total resection is not achievable, IR therapy is typically utilised to supplement patient management.[3,4]

CONCLUSION

Primary ovarian RMS is an incredibly rare and lethal form of malignancy. It occurs in a wide range of age groups, usually presents in advanced stages with delayed presentation, and has poor outcomes. Thorough evaluation of the tumour is crucial to rule out metastasis from other sites, as treatment of RMS is different from that of other types of malignant ovarian tumour.

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CONSENT

Written informed consent was taken from the patient from the start of managing the case for publication.

CONFLICT OF INTEREST

There was no conflict of interest.

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