Radiologic-Pathologic Correlation of Primary Ovarian Leiomyosarcoma: a Case Report and Review of the Literature

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ABSTRACT

Primary ovarian leiomyosarcoma is a rare ovarian sarcoma, primarily affect postmenopausal women. It has a non-specific appearance in the US, CT, and MRI. Here we describe the case of a 71-year-old woman presented with three months of vaginal bleeding, progressive abdominal distention and left pelvi-abdominal mass, with no other complaints. Physical examination, ultrasound, Computed tomography and magnetic resonance imaging revealed a left ovarian tumor with no ascites and no lymphadenopathy. On histology and immunohistochemistry, a diagnosis of leiomyosarcoma was made. Postoperatively the patient did not receive chemotherapy or radiotherapy, and she was doing well after 16 months follow-up and disease free on clinical and radiological examination.

Keywords: Ovarian Sarcoma, Primary ovarian leiomyosarcoma, Radiological studies, magnetic resonance imaging, ultrasound, Computed tomography, Histopathology, Microscopy, Immunohistochemistry, Treatment

1. INTRODUCTION

Primary ovarian sarcomas comprise less than 3% of all ovarian neoplasm primarily affects the post-menopausal women(1). Primary ovarian leiomyosarcoma is a very rare ovarian tumor, with less than 50 cases in the literature(2). Our case reviews the presentation of ovarian leiomyosarcoma, along with the imaging workup and pathological correlation in the post-menopausal patient. Literature of primary ovarian leiomyosarcoma is reviewed.

2. CASE REPORT

The tumor was detected in a 71-year-old nulliparous postmenopausal woman with a history of hypertension who had a three months history of vaginal bleeding, progressive abdominal distention, and large left pelvi-abdominal mass. No abdominal pain or urinary complaints.

On physical examination, the patient was stable, communicating, oriented and not in pain. The abdomen was lax and soft. There was a palpable 10.0-cm left adnexal mass extending from left iliac fossa to umbilicus. Routine investigations were normal. CA-125 and carcinoembryonic antigen levels were normal. CA-125 was 5.8U/ml. Radiological workup as Ultrasound, CT of abdomen and pelvis and MRI pelvis were performed. An abdominal ultrasound showed a large well defined multilobulated heterogeneous predominantly hypoechoic solid mass measures 13 x 9 cm in the left adnexal region with internal vascularity. No posterior acoustic shadowing or enhancement. The uterus and right ovary were normal. (Figure 1A-B).
Fig. 1: A) Greyscale Ultrasound image shows a large heterogeneous hypoechoic solid left adnexal mass. B) Color Doppler Ultrasound shows internal vascularity within the mass.

Enhanced CT scan of abdomen and pelvis showed a large heterogeneously enhanced solid left ovarian mass with intervening necrotic/cystic changes. The mass measures 13.0 x 9.4 x 10.0 cm in the transverse, AP and craniocaudal dimensions respectively. The right ovary is grossly unremarkable. No associated ascites or peritoneal disease. (Figure 2A-C)

Fig. 2. (A, B, C) Enhanced CT scan of abdomen and pelvis (Transverse, Coronal, Sagittal views) shows large heterogeneously enhanced solid left ovarian mass with intervening necrotic/cystic changes

The magnetic resonance imaging (MRI) of the abdomen and pelvis showed a large left pelvic-abdominal mass measuring approximately 13.4 x 9.2 x 9.7 cm in Transverse, AP and Craniocaudal dimensions respectively, arising from the left ovary showing heterogeneous signal intensity on T2-
weighted images and predominant hypointensity signal on T1-weighted images. After IV gadolinium administration the mass showed heterogeneous enhancement. The right ovary is normal. No associated ascites or peritoneal disease. The sigmoid colon is displaced anteriorly by the underlying adnexal mass but no definite invasion identified. No evidence of metastasis. (Figure 3A-E)

Figure 3: (A-E) MRI of abdomen and pelvis images shows large left pelvi-abdominal mass with heterogeneous signal intensity. A, B, C) sagittal, transverse and coronal views of T2 weighted images D) T1 weighted image E) T2 weighted with fat saturation image

The patient underwent total transabdominal hysterectomy with bilateral salpingooophorectomy. Staging biopsies were performed, and samples were sent to his to pathology. Perioperative findings revealed tumor mass with clotted blood was depended on the left ovary, without apparent compromise of uterus and parametria. The patient had a smooth operative period with no complication.

Based on histopathology, the gross description was large multilobulated mass measures 15 x 10.5 x 5 cm. The cut sections show homogeneous, solid white/cream cut surfaces with a lobulated pattern with whitish and necrotic areas in addition to areas of hemorrhage. An attached ovary measures 4 x 1 x 0.5 cm. (Figure 4A-B)

Figure 4: A) Gross specimen demonstrates large multilobulated mass measures 15 x 10.5 x 5 cm. B) Cut a section of mass shows homogenous, solid white/cream cut surfaces with intervening areas of hemorrhage, necrosis and myxoid change.
According to microscopy, the mass is a spindle cell tumor comprising highly cellular proliferation of spindle cells showing elongated, cigar-shaped nuclei, with moderate pleomorphism and abundant mitoses (>20 mitoses /10 HPF). Abundant necrosis also noted. Immunohistochemistry shows the tumor cells to be strongly positive for smooth muscle actin but negative for cytokeratin. Desmin did not work due to some technical fault. The morphological features and immune profile are consistent with a leiomyosarcoma mostly of ovarian origin, considering the clinical findings. Fallopian tubes, right ovary, cervix, and omentum were negative for tumor lesions. Peritoneal lavage cytology was negative for malignancy. (Figure 6A-D)

Post operation, the patient, did not receive chemotherapy or radiotherapy, she is currently asymptomatic and without signs of disease relapse after 18 months of follow-up.

3. DISCUSSION

Leiomyosarcomas (LMS) are classified as malignant mesenchymal tumors accounting for 8% of malignant of tissue tumors\(^3\). Primary ovarian sarcomas...
The rare leiomyosarcomas accounting for less than 0.1% of all ovarian neoplasms, and it represents a heterogeneous group of ovarian tumors primarily affects the post-menopausal patients. The most common primary ovarian sarcoma are rhabdomyosarcoma, fibrosarcoma, stromal cell sarcoma, with the rare leiomyosarcomas accounting for less than 0.1%. Ovarian leiomyosarcoma typically has no significant symptoms. A number of patients were referred to the hospital with abdominal pain or fullness, a few exhibited constipation and one patient exhibited post-menopausal bleeding.

Regarding radiological investigations, CT is the primary imaging modality for the assessment of abdominal leiomyosarcomas as well as for the evaluation of metastatic disease. Ultrasound remains the primary imaging modality for the assessment of adnexal masses. However, the pelvic lesion is optimally evaluated with MRI because of its superior soft tissue contrast to CT. MRI is useful in determining more precisely the site of origin of a mass and relationship between a mass and adjacent structures. The general imaging findings in our case were compatible with those of other reported cases of primary ovarian leiomyosarcoma. According to literature, ovarian leiomyosarcoma usually manifests as large necrotic heterogeneously enhanced soft-tissue masses are usually seen at CT. On MRI it appears as huge mass with an isointense signal to muscle on T1 weighted images and an intermediate or high signal on T2 weighted images. Cystic and necrotic components are often found in the mass.

On gross examination, these tumors are indistinguishable from other sarcomas. Usually, these tumors are solid but cystic degeneration is often seen in large tumors.

Microscopically tumor cells are spindle shaped and uniform, with abundant eosinophilic cytoplasm and elongated nuclei. Cells are organized in whorled, interwoven bundle with an area of pleomorphism. Histopathology with immunohistochemistry is essential tools for the definite diagnosis of leiomyosarcoma. According to Kurian et al. treatment of choice is radical surgery followed by adjuvant chemotherapy or radiotherapy. Although the overall prognosis is poor, early stage disease treated with optimal surgery may offer significant improvements in survival.

4. CONCLUSION

Primary ovarian leiomyosarcoma is a rare tumor with a poor prognosis, primarily affect a postmenopausal woman. It has a non-specific appearance in the US, CT, and MRI. Histopathology with immunohistochemistry is essential tools for definite diagnosis. Optimal debulking surgery remains the cornerstone of ovarian leiomyosarcoma treatment. Although prognosis is poor, the good news that if it diagnosed at an early stage, the treatment will be effective and improve prognosis.

REFERENCES