Case Report

Primary ovarian leiomyoma: A rare tumor with a diagnostic dilemma

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Abstract

Primary ovarian leiomyomas are rare tumors accounting for 0.5%–1% of all ovarian tumors. They are seen commonly in the reproductive age group. Morphologically, they resemble leiomyomas occurring at other sites. The definitive preoperative diagnosis is difficult due to the lack of characteristic radiological features. Here, we describe a case of ovarian leiomyoma, which has posed a diagnostic and therapeutic challenge. The final diagnosis was confirmed by immunohistochemistry.

Keywords: Desmin, fibroid, fibroma, leiomyoma, ovary, smooth muscle actin

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INTRODUCTION

Smooth muscle tumors of the female genital tract are commonly seen in the uterus. Ovarian leiomyomas are rare and account for less than 1% of all ovarian tumors. These tumors are usually unilateral, small in size, and generally <3 cm in size. They are found over a wide age range, with a mean age of 43 years. The characteristic histomorphological features will help in identifying the smooth muscle origin of these tumors; however, in some patients, immunohistochemistry may be required to arrive at a final diagnosis. Here, we report a case of an ovarian tumor which was suspected to be a dysgerminoma/fibroma clinically. The definitive diagnosis was made on histopathological and immunohistochemical examination.

CASE REPORT

A 38-year-old woman with previous regular menstrual cycles presented with complaints of amenorrhea of 3

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months' duration followed by spotting for 1 day. She was uniparous with the last childbirth 18 years back. Her past medical history was not significant. Per abdomen examination revealed a 10 cm × 8 cm nontender mass in the suprapubic area extending to the right iliac fossa. Her routine hematological and biochemical investigations were not contributory. Her tumor markers – serum CA-125, alpha-fetoprotein, CEA, and β-HCG – were within normal limits. The ultrasound abdomen revealed an ill-defined lobulated mass in the right ovary, with dense peripheral calcification. The computed tomography abdomen revealed a heterogeneously enhancing abdominopelvic mass arising from the right ovary measuring 12 cm × 12 cm × 7 cm, and the differential diagnosis included dysgerminoma and fibroma. There was also associated right-sided hydroureteronephrosis. Given the clinical suspicion of a malignant tumor, total abdominal hysterectomy, bilateral salpingo-oophorectomy, bilateral pelvic node sampling, and infracolic omentectomy were performed. On gross

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examination, uterus and left adnexa were unremarkable. Right adnexa revealed a solid and hard ovarian mass measuring 12 cm × 11.5 cm × 8 cm. Cut surface was predominantly solid with pale white and peripheral yellow areas. The histopathological examination of the right ovarian tissue showed elongated spindle cells in the fascicles with abundant pale eosinophilic cytoplasm and uniform cigar-shaped nuclei [Figure 1a-c]. Extensive collagenized stroma was noted. Many areas of dystrophic calcification were also seen. As the morphology closely resembled fibroma, immunohistochemistry was done and showed diffuse positivity for vimentin and desmin [Figure 1d and e]; hence, the final diagnosis of ovarian leiomyoma was made. Histopathology of the uterus, left adnexa, lymph nodes, and omentum was unremarkable. The patient was closely followed up, and after 6 months of follow-up, the patient is doing well.

DISCUSSION

Ovarian leiomyoma is accepted as a primary tumor when it originates directly from the ovary. [3] They are rare tumors first reported by Sangalli *et al.* in 1862. [4,5] They are usually asymptomatic or may present with abdominal pain and swelling. Rarely, these cases may present with hydronephrosis, elevated CA-125, hydrothorax, and ascites. [6] Our patient was premenopausal with a history of menstrual irregularities of 3 months' duration. She

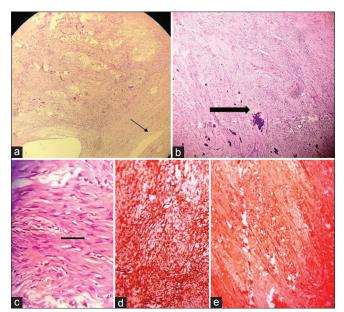


Figure 1: (a) Histopathology of ovarian tumor showing compressed ovarian tissue (shown in arrow) with the spindle cell tumor (H and E, ×100). (b) Histopathology of the tumor showing spindle cell in the fascicles with calcification, shown in arrow (H and E, ×100). (c) Histopathology showing spindle cell tumor with cigar-shaped nuclei (H and E, ×400). (d and e) Immunohistochemistry showing immunoreactive for vimentin and desmin (×100)

presented with a right-sided abdominal mass and was found to have hydroureteronephrosis on the right side. Ovarian leiomyomas are recognized by their characteristic histological features, with a fascicular pattern of growth, abundant eosinophilic cytoplasm, and cigar-shaped uniform nuclei. The tumors that present in the premenopausal age are usually bilateral. In contrast, those in the postmenopausal age are often unilateral. These tumors are thought to arise from the smooth muscle cells of the ovarian hilar blood vessels.[8] Other theories that have been proposed include cells in the ovarian ligament, smooth muscle cells, multipotent cells, or undifferentiated germ cells in the ovarian stroma. A metaplastic change in the stroma in response to endometriotic cysts is also proposed. [8] The tumor is reported to occur more frequently in nulliparous patients. Hence, androgen is thought to play a role in its development. It is also believed to occur more frequently in developmentally abnormal ovaries. [9] In our case, she was uniparous, and the healthy ovarian tissue was visualized. The differential diagnosis includes ovarian fibrothecoma, cellular fibroma, and sclerosing stromal tumor. They have to be distinguished from leiomyomas arising in the broad ligament and uterine leiomyoma becoming parasites on the ovary (wandering leiomyomas).[10] In large tumors, it is essential to distinguish from leiomyosarcomas, spindle cell carcinoma, and metastatic gastrointestinal stromal tumor.[6] The differentiation from leiomyosarcomas is based on the histological features such as mitotic count, cellular atypia, and necrosis.[11] Distinction from fibromas is difficult when there is an excess of collagenization and hyalinization.[12] Immunohistochemistry will help in such cases. Leiomyomas are diffusely positive for desmin/ smooth muscle actin, whereas fibromas tumors are negative or only focally positive. [7,13] In our specimen, we had areas of collagenized stroma due to which it was difficult to differentiate from fibroma, but IHC with vimentin and desmin was helpful in the final diagnosis.

Surgical resection is favored when the tumor is symptomatic or significant in size, and there is a possibility of torsion. In our case, because of the large size and clinical diagnosis of dysgerminoma, radical surgery was performed. The patient was closely followed for 6 months, and there was no recurrence and any fresh complaints.

CONCLUSION

This case of primary ovarian leiomyoma has been described due to its rarity. Leiomyomas should be considered in the differential diagnosis of solid ovarian tumors. Immunohistochemistry is a useful tool in definitive diagnosis and further management.

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Conflicts of interest

There are no conflicts of interest.

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