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CASE REPORT

Ovarian Fibromatosis

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Clinical Summary

A 23-year-old married female presented with complaints of lower abdominal pain and fullness of 4 months duration. The patient was in lactational amenorrhea following a full-term normal vaginal delivery 18 months ago.

Physical examination revealed left adnexal mass and ascites. Abdominal ultrasound confirmed left ovarian mass and significant ascites. CA-125 was within normal range.

An exploratory laparotomy with resection of left ovarian mass and drainage of about 3 l of ascitic fluid was done.

On gross examination of the specimen, a 26 × 16 × 8 cm globular, solid mass with lobulated white external surface weighing 4.5 kg was seen. The mass was attached to a 2-cm-long pedicle. The cut surface was firm, white to gray and solid with a few cystic areas Fig. 1.

Histopathological Examination

Revealed fibromatoid proliferation of collagen producing spindle cells separated by dense collagen surrounding normal follicular derivatives (corpus albicans and cystic follicles). Moderately cellular fascicles of spindle cells were interspersed with relatively acellular bands of dense collagen. Small foci of stromal edema and foci of uninvolved ovarian stroma were also seen.

With these classical histological features, a diagnosis of ovarian fibromatosis was made (Figs. 2, 3, 4).

Discussion

Fibromatosis describes a group of fibrous tissue proliferations that, although benign, can be locally invasive, but do not metastasize [1]. X-chromosome inactivation molecular analyses have confirmed these tumors to be clonal, thus identifying them as neoplasms and not products of an inflammatory response.

Fibromatosis of the ovary was first described by Young and Scully in 1984 as a tumor-like enlargement of one or both ovaries due to proliferation of collagen producing ovarian stroma [2]. It is a rare clinicopathological entity and just over 30 cases have been described in the literature [3]. It is seen predominantly in young patients with an average age of 25 years. The clinical manifestations include menstrual abnormalities or amenorrhea, recurrent abdominal pain, and rarely, virilization. A majority of the patients have a palpable pelvic mass. It has also been described as an incidental finding during pregnancy and the postpartum period. Preoperative MRI and intraoperative frozen section may help to define the benign nature of the disease.

Microscopically, ovarian fibromatosis is characterized by proliferation of collagen producing spindle cells that typically surround normal follicular structures. Foci of

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Fig. 1 Gross specimen of the tumor with lobulated white to gray external surface

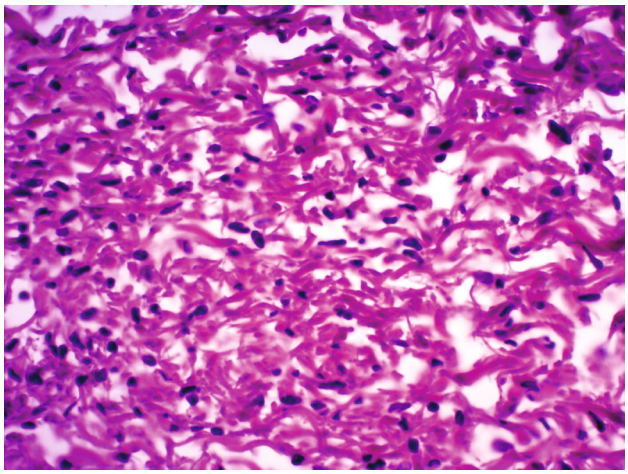

Fig. 2 Proliferation of collagen producing spindle cells (Hematoxylin and eosin, ×400)

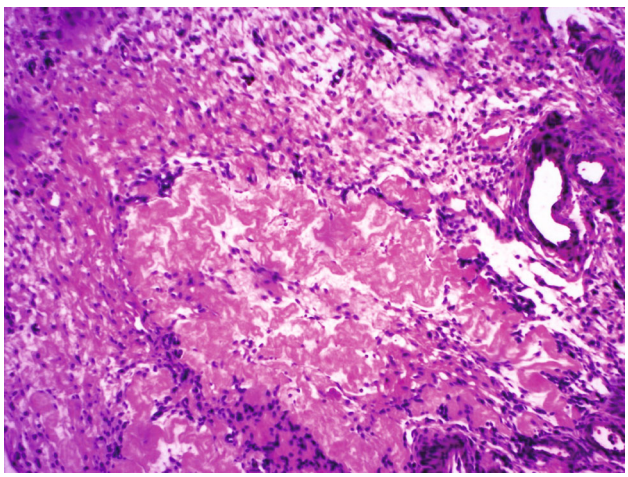

Fig. 3 Spindle cell proliferation surrounding follicle derivatives (Hematoxylin and eosin, ×100)

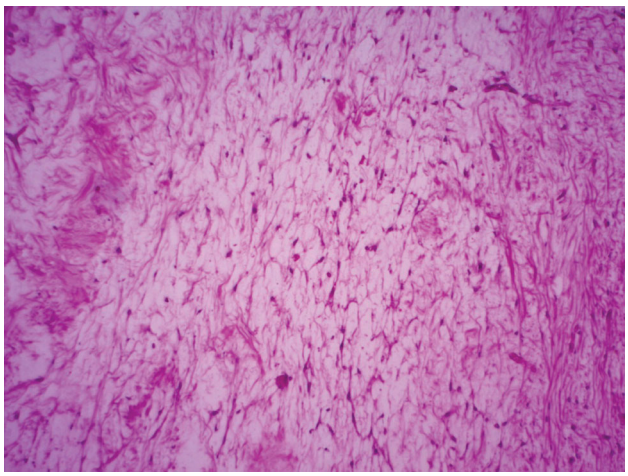

Fig. 4 Foci of stromal edema (Hematoxylin and eosin, ×100)

stromal edema, luteinized stromal cells, and focal proliferation of sex cord elements may be encountered. Ovarian fibroma resembles fibromatosis histologically, however, it usually appears in older age groups, is typically nonfunctioning, and normal follicular structures are not seen. The few small aggregates of sex cord cells in fibromatosis are distinguished from those of a Brenner tumor in their number, size, and shape. The absence of signet ring cells and the typically unilateral mass help to distinguish fibromatosis of the ovary from the Krukenberg tumor.

Due to rarity of the lesion, its pathogenesis is not well known. It has been suggested that fibromatosis represents the "burned out" stage of a reactive fibroblastic proliferation that at one end of the spectrum is represented by massive edema and at the other by a variety of highly cellular fibroblastic tumor-like lesions [4]. It is observed that fibromatosis of the ovary is not a well-recognized entity and many clinicians are not aware of its existence. It is, therefore, often misdiagnosed as a malignant ovarian tumor, and more extensive treatment than is necessary is undertaken. The important clinical message is that this entity should be considered in young women presenting with an ovarian mass as a simple surgical resection is curative.

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