Case Report

A Case of Ovarian Fibromatosis as an Incidental Finding in an Acute Ovarian Torsion: Differential Diagnosis and Management

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Abstract

Ovarian fibromatosis (OF) is a rare non-neoplastic condition, more frequent in young females and characterised by ovarian enlargement with proliferation of collagen-producing spindle cells in the stroma. It usually presents with abdominal pain, menstrual disorders or as a solid mass. Hirsutism and virilisation are possible. The relevance of this subject remains in the usual misdiagnosis as a malignant tumour, which may lead to unnecessary oophorectomies. We report the case of a 17-year-old female who presented with acute intense pain in the right iliac fossa and vomits. On examination, she complained of intense pain without any signs of peritonism. An abdominal ultrasound was performed, finding an enlarged right adnexal containing a heterogenic cyst without flow to the ovary. A Pfannenstiel laparotomy showed a right ovarian torsion. The haemorrhagic cyst was drained and the ovary was detorsed. Bilateral ovarian biopsies were performed as both ovaries showed an indurated and cerebroid surface, suggestive of malignancy. Tumoural markers were negative. Histological examination confirmed OF. After 3 months, the magnetic resonance imaging showed an enlarged right ovary with a fibrous capsule surrounding both ovaries. In conclusion, OF can also be found incidentally in the context of an ovarian torsion. Since its appearance may be dismissed as malignant, it is important to recognise it and remain conservative. Biopsies can be taken to make the differential diagnosis.

Keywords: Ovarian fibromatosis, ovarian torsion, paediatrics

Introduction

Ovarian fibromatosis (OF) is a rare non-neoplastic condition predominantly found in young females. It usually presents with abdominal pain, menstrual irregularities and sometimes virilisation.[1,2] It can also be found as a solid pelvic mass with ascites.[1] It was first described by Young and Scully in 1984, and since then, only around 30 cases have been reported.[2] Due to its low incidence and its atypical, non-specific way of presentation, there is lack of information in the literature.

In this article, we would like to report the first case of concomitant OF in a young female with acute ovarian torsion. The relevance of this subject remains in the usual misdiagnosis from its appearance as a malignant tumour. This may lead to an aggressive treatment with unnecessary oophorectomies.

Case Report

A 17-year-old female presented to our emergency department with acute and intense pain in the right iliac fossa. She compared the pain to her menstrual periods but more intense and associated with two emetic episodes. Her menarche was at 12 years old, with regular menstrual cycles. We expected the next menstrual cycle in 6 days forward. On physical examination, she appeared prostrated by the pain and pale, presenting with intense pain at the palpation of the right iliac fossa without any sign of peritonitis. An abdominal ultrasound was performed, showing a right adnexal enlargement associated with a heterogeneous cyst that measured 56 mm × 49 mm × 52 mm [Figure 1]. Blood flow could not be identified in either the ovary or the lesion.

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A laparotomy with Pfannenstiel incision was undertaken, confirming a torsion of the right ovary. The sero-haemorrhagic cystic content was drained and the ovary was detorsed. Our attention shifted to the ovary as it was enlarged, indurated, pale and irregular, with a cerebroid surface and exophytic lesions. Suspecting malignancy, we looked for the other ovary, whose appearance was identical but smaller, since it was not oedematous [Figure 2]. Although the aspect of both ovaries was suggestive of malignancy at sight, taking into account the low incidence of malignant tumours at the patient's age,[3] we decided to take bilateral ovarian wedge biopsies and a sample of the ascetic liquid for further examination. No peritoneal implants were observed. Histological examination demonstrated fibromatoid proliferation of collagen-producing spindle cells entrapping follicular structures, consistent with OF. Pre-operative follicle-stimulating hormone, beta-human chorionic gonadotropin (HCG), alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA) and CA-125 were within the normal range. The patient was discharged 2 days later. A follow-up pelvic ultrasound 2 months later showed a persistently enlarged but well vascularised right ovary. The central cystic lesion was nearly completely reabsorbed (19 mm). The left ovary appeared normal. In the absence of new torsion episodes, a 3-month post-operative magnetic resonance imaging (MRI) showed a persistently enlarged right ovary and fibrous capsule surrounding both ovaries.

Discussion

OF is a rare benign disorder. It is more common in young females with a mean age of 25 years old.[2] Nevertheless, its real incidence is unknown because it is infrequent and does not follow a typical clinical debut, being asymptomatic in many cases. When symptomatic, it can present with menstrual abnormalities,[2] abdominal or pelvic pain and a solid mass with or without ascites.[1] Hirsutism and virilisation have also been described, sometimes related to a slight increase of serum CA-125 levels.[1] Our case is the first to report ovarian torsion as a way of presentation, highlighting the importance of keeping this pathology in mind for the differential diagnosis.

Macroscopically, we can find enlarged ovaries with a lobulated and firm external surface because the fibromatous process involves the whole ovarian parenchyma.[4] It is bilateral in 20% of cases. Microscopically, it is characterised by the proliferation of collagen-producing spindle cells in the ovarian stroma surrounding normal follicles with collagenous thickening of the ovarian cortex.[5]

The pathogenesis of OF remains poorly understood. It has been compared to massive oedema of the ovary, for which a histological overlap has been observed.[2,6] Some studies have postulated that the oedema caused by intermittent torsions of the ovary could stimulate a reactive proliferation of fibroblasts,[2,5,6] whereas others have suggested that the enlargement of the ovaries due to fibromatosis could cause torsions and lead to oedema.[1] The latter hypothesis would be reinforced by the fact that fibromatosis has been shown to be a clonal process, postulating as a neoplasm rather than a reactive inflammatory process.[7]

Due to its frequency in presentation as a solid mass, pre-operative differential diagnosis with other ovarian tumours must be performed. Serum markers such as CEA, beta-HCG, AFP and CA-125 must be checked. MRI findings can help to distinguish OF from ovarian fibromas, Brenner and Krukenberg tumour.[8] In the case of OF, we can observe a homogeneously, well-limited, often unilateral mass. On T2, we can observe the entrapment of normal cystic follicles in the fibrous cortex, which is known as the 'black garland' sign. This is considered pathognomonic but not always found.[8]

Due to its low incidence, there are no guidelines on the way to proceed with this condition. As the 'black garland' sign is not always observed and the resemblance to many malignant diseases, it is often necessary to perform an explorative laparotomy or laparoscopy biopsy. In our case, OF was diagnosed after an ovarian torsion that was probably caused by the presence of a cyst. There is no literature relating ovarian torsion to OF. If found incidentally, an intraoperative wedge biopsy to exclude malignancy is an option.[1,4,6] There are some

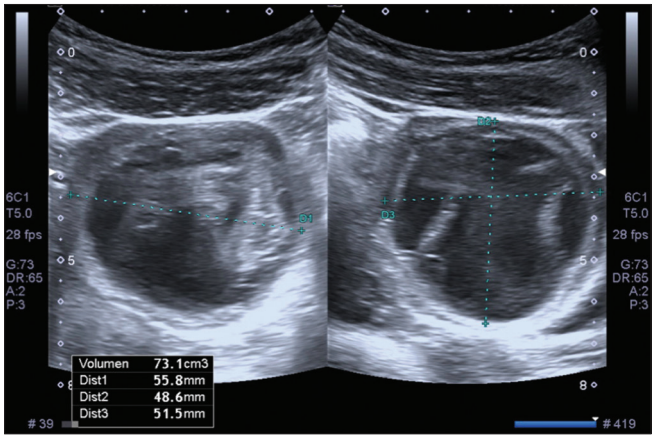

Figure 1: Ultrasound appearance of the torsed ovary. In the images, we can observe a heterogenic cyst of 56 mm × 49 mm × 52 mm with septums

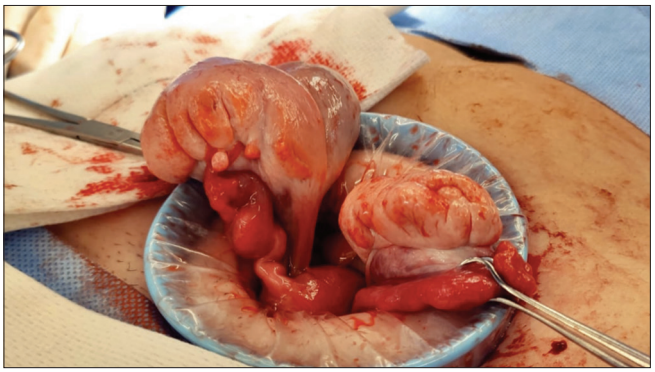

Figure 2: Intraoperative aspect of the ovaries. On the left, an enlarged right ovary. Both with a cerebroid surface and exophytic lesions

reports of peritoneal fibrous proliferation in OF, which shows that it can have a locally aggressive behaviour.[8,9]

Conclusion

OF is a rare condition, but it is more frequent in young females. Even though it is more often presented as a pelvic mass or menstrual disorder, we can find it incidentally in the context of an ovarian torsion. The pathological appearance of the ovaries in surgery may be misleading. In this case, an ovary-preserving procedure should be performed, confirming the benignity of the lesions with intraoperative biopsies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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