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CASE REPORT

Laparoscopic resection of cystic adenomyosis in a teenager with arcurate uterus

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Abstract Whereas diffuse adenomyosis is a common finding in parous women, cystic adenomyosis causing severe dysmenorrhoea is very rarely reported and possibly under-diagnosed, particularly in adolescents. We report a case of juvenile cystic adenomyosis in a 19-year-old nulliparous patient presenting with secondary dysmenorrhoea and non-cyclical pain. A 20-mm adenomyotic cyst in the fundal myometrium was successfully excised laparoscopically by modified myomectomy. It is mandatory to take persistent primary and early secondary dysmenorrhoea seriously, especially after poor response to medical treatment and to have a low threshold for further investigations, because a diagnosis of cystic adenomyosis requires targeted therapeutic intervention.

Keywords Juvenile cystic adenomyosis · Adenomyoma · Uterine cyst · Laparoscopic surgery · Arcuate uterus · Modified myomectomy

Introduction

Diffuse adenomyosis is typically present in up to 20% of hysterectomy specimens of parous women over the age of 30, but circumscribed forms, such as solid or cystic adenomyoma, also termed cystic adenomyosis, are less frequent. Cystic adenomyosis can be divided into an adult form, which is present in 5–7% of hysterectomy fibroid uteri [1] and often associated with the diffuse subtype, and a juvenile form seen in nulliparous women between 13 and 20 years of age, for which the true incidence is not known; less than ten cases have been reported to date [2–9], most of them in Japan. Although the aetiology is still a cause for debate, the adult form appears to result from traumatic breech of the interface between the endometrium and the myometrium, for instance after uterine instrumentation followed by bleeding into dispersed endometrial glands, which become dilated and turn into cystic spaces of typically less than 5 mm in diameter. In contrast, the juvenile form is thought to result from a congenital defect of the development of the Müllerian duct, which appears to lead to larger cystic spaces lined with endometrium between 1 and 3 cm in diameter [6]. The early presentation of pain soon after menarche may mislead clinicians into making the diagnosis of functional, primary dysmenorrhoea; further differential diagnoses include ovarian endometriomata, degenerative uterine fibroids and bicornuate uterus with a non-communicating uterine horn.

Case

A 19-year-old nulliparous woman presented with an 18 months' history of severe and worsening dysmenorrhoea with cramps, which were insufficiently relieved by non-steroidal anti-inflammatory medication, oral narcotics and the combined oral contraceptive pill. The worst pain occurred on the first day of her menses. In addition, she also complained of non-cyclical left-sided pelvic pain. She went through the menarche when she was 13 years old.

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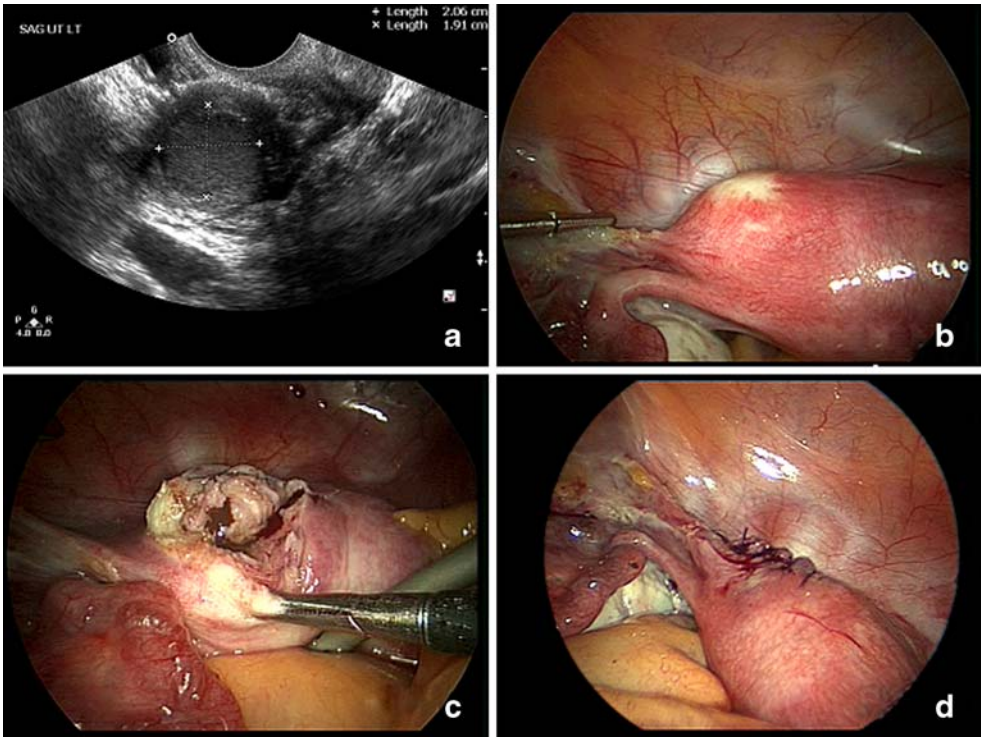

Fig. 1 **a** Trans-vaginal ultrasound scan of uterus showing a spherical 2-cm lesion with mid-level echoes suggestive of a longstanding blood collection. **b** Adenomyoma in situ prior to excision. Apparent blanching can be seen secondary to vasopressin injection. There is no external discoloration as typically seen in prominent endometriotic lesions. **c** Intra-operative image of uterus showing the enucleation using a modified myomectomy approach. Altered blood, similar to endometrioma fluid, can be seen draining from cystic lesion. **d** Repair of the site of the resection site with interrupted 2/0 PDS sutures

Pelvic examination revealed a normal vagina, vulva and adnexae and a normal-sized anteverted uterus containing a 2-cm firm fundal mass.

Trans-vaginal ultrasound performed immediately before the operation demonstrated a 2-cm myometrial lesion in the left lateral fundal area (Fig. 1a). Interestingly, a trans-vaginal ultrasound scan 3 months before showed a fluid level within the lesion. In fact, changing morphology with intermittent visualisation of a fluid level in cystic adenomyoma has been reported before [10]. Both ovaries appeared normal.

Hysteroscopy and operative laparoscopy were performed on day 12 of her menstrual cycle under general anaesthesia. Laparoscopy was carried out via direct entry. A 10-mm Excel Optiview® port was introduced intra-umbilically and a further three 5-mm lateral ports were placed under vision, one on the left side and two on the right with 6-cm distance in between. The lower ports were placed 1 cm medial to the anterior iliac spines. Operative findings included an arcurate uterus with clearly visible ostia and bilaterally patent fallopian tubes, thus ruling out a non-communicating uterine horn. A 20-mm nodular lesion with an external appearance similar to that of a fibroid (Fig. 1b) was embedded in the uterine myometrium and located in the fundus towards the left side just caudally to the round ligament. The rest of the pelvis appeared entirely normal. After vasopressin (Abraxis, Schaumburg, IL, USA) at a dilution of 1 U in 50 ml of normal saline was injected superficially into the lesion, a Harmonic Ace® scalpel was used to incise the visceral peritoneum overlying the lesion and the adjacent parametrium transversely. During enucleation of the lesion, it was difficult to establish a clear plane and thus the tumour was divided and drained of brown fluid reminiscent of an ovarian endometrioma (Fig. 1c). There was

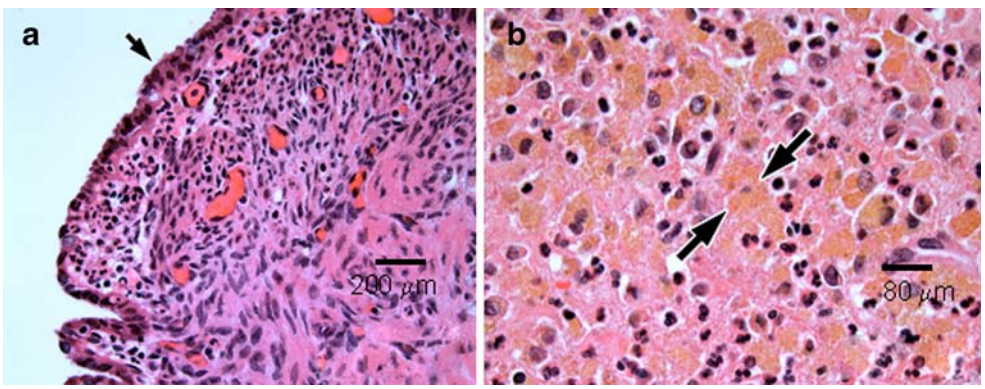

Fig. 2 **a** Section of adenomyotic cyst wall stained with haematoxylin and eosin. A layer of inactive endothelium (*arrows*) lines the cystic space (×100 magnification). **b** Haematoxylin and eosin-stained section of myometrium (×400 magnification) showing abundant haemosiderin-laden histiocytes (*arrows*)

no continuity with the pelvic cavity. Further, careful excision with the Harmonic scalpel was employed to remove the cyst and the surrounding fibrosis. Haemostasis was achieved with bipolar diathermy and the cavity was closed with a layer of deep intramural and parametrial intracorporeal continuous 2/0 PDS sutures on a CT1 needle and a superficial layer including the superficial muscularis and serosa using 3/0 PDS sutures (Fig. 1d). The bisected specimen was removed through the umbilical port. Surgery lasted 90 min and the operative blood loss was less than 20 ml.

The patient was discharged home after 24 h. Eighteen months following the procedure, the patient remained pain free and does not require any medication. Histopathology showed a cystic cavity with endometrial and stromal lining, confirming the presumptive diagnosis of cystic adenomyosis. In the surrounding tissue, there were no other foci of adenomyosis (Fig. 2a) and abundant haemosiderin within histiocytes was visualised (Fig. 2b)

Discussion

Since the first case report of juvenile cystic adenomyosis in 1996 [2], several other cases have recently drawn attention to this rarely diagnosed condition. However, increased awareness of this condition and improved access to transvaginal ultrasound scanning, MRI and laparoscopy may lead to better recognition of juvenile cystic adenomyosis in the future. This case emphasises the importance of linking the clinical findings with repeated ultrasound imaging and to explore the pelvis laparoscopically. None of the preoperative investigations in isolation, including MRI, would have helped to reach a definitive diagnosis; moreover, one-off ultrasound imaging alone may have led to the misdiagnosis of ovarian endometrioma or intramural fibroid. In fact, diagnostic laparoscopy revealed a lesion that was identical in external appearance with a uterine fibroid, and without the ultrasound scan that had demonstrated a fluid level, it could have been labelled as a fibroid and would probably not have been removed at the time of surgery. In this context, it is important to remember that fibroids rarely cause dysmenorrhoea [11], thus a painful 'fibroid' may reveal itself as an adenomyoma only during the operation or on histological examination.

It is important to state that this young woman was not operated on, on the basis of a radiological image, but after 18 months of incapacitating pain, unresponsive to medication. This pain is similar to pain caused by cryptomenorrhoea associated with a non-communicating uterine horn. Postoperatively, the patient is completely pain free and successfully pursuing her career. This patient derived optimal benefit from surgery, which she would not have had from medical treatment.

It can be tempting to label dysmenorrhoea in adolescents as 'functional' and commence medical treatment. However, apart from a single patient, who responded well to combined oral contraceptives [5], all patients, including the present, responded poorly to medical treatment such as non-steroidal anti-inflammatory medication or gonadotropin-releasing-hormone analogues. In juvenile cystic adenomyosis, pelvic examination may be unremarkable, and thus lack of response to medical treatment should trigger further investigations, including ultrasound scanning, laparoscopy and hysteroscopy.

In contrast to the present patient, who complained of secondary dysmenorrhoea, most investigators reported primary dysmenorrhoea. The present case illustrates that a pre-existing developmental cyst within the myometrium may require a period of repetitive bleeding in order to become symptomatic as evidenced by the histological examination revealing numerous haemosiderin-laden histiocytes.

It is of particular interest that the present patient was suffering from juvenile cystic adenomyosis in addition to having an arcuate uterus, supporting developmental aetiology of juvenile cystic adenomyosis.

If there is no response to medical treatment, the optimal treatment for juvenile cystic adenomyosis appears to be resection. The laparoscopic route is preferable to laparotomy, if technically feasible, in order to avoid a negative impact on future fertility through adhesions.

In conclusion, this case draws attention to a little known but not uncommon cause for both primary and secondary dysmenorrhoea, which can affect young nulliparous women and should be included in the differential diagnosis of dysmenorrhoea in young women, which have been reviewed by Drosdzol et al. [12]. It is best investigated with ultrasound scanning and MRI and can be treated laparoscopically with the same approach as used for myomectomy.

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Conflict of interest There is no actual or potential conflict of interest in relation to this article.

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