Rare disease

Juvenile cystic adenomyoma

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Summary

The purpose of this paper is to describe a case of juvenile cystic adenomyoma in a 17 year-old female patient with severe dysmenorrhoea unresponsive to non-steroidal anti-inflammatory drugs. The patient presents progressively worsening dysmenorrhoea that started 2 years after menarche and a cystic uterine lesion in MRI. The cyclic nature of symptoms, the similarity of the lesion and endometrium in MRI signal intensity and response to hormone suppression are consistent with juvenile cystic adenomyoma. The treatment depends on the age of the patient, severity of her symptoms and size and localisation of the cyst. This is a rare condition in young nulliparous women with a challenging differential diagnosis. This case highlights the relevance of MRI in the patient's study, featuring important characteristics of the lesion that disclosed the final diagnosis.

BACKGROUND

Adenomyosis is a condition characterised by presence of ectopic endometrial tissue within the uterine myometrium, with adjacent smooth muscle hypertrophy.¹ The disease usually occurs in women older than 30.² Diffuse adenomyosis is the most common form, while focal or nodular adenomyosis, particularly the cystic variant, is extremely rare, especially in pediatric population.³

In 2009, Hiroyuki *et al*⁴ defined the diagnostic criteria of juvenile cystic adenomyoma based on age (<30 years), presence of cystic lesion ≥ 1 cm in diameter independent of uterine lumen and covered by hypertrophic myometrium on diagnostic images and associated with severe dysmenorrhea.

There are fewer than 20 cases of juvenile cystic adenomyoma reported in the literature, according to Hiroyuki criteria. Similar disease entities have been reported using different nomenclature, making difficult to estimate the prevalence of the disease in this age group.⁴⁻⁷

In this paper, we used the term Juvenile cystic adenomyoma, according to the diagnostic criteria defined by Hiroyuki *et al*.

Clinical findings are non-specific and include pelvic pain, dysmenorrhea, menorrhagia, uterine enlargement and infertility. In many cases, women may be asymptomatic.⁸

The differential diagnosis includes congenital anomaly with haematometra in a non-communicating horn, fibroid with haemorrhagic or fatty degeneration, congenital uterine cysts and intramietrial hydrosalpinx.² These diagnoses can be distinguished by MRI, that is the primary radiological modality for evaluating cystic adenomyosis and can be crucial for making a correct diagnosis.⁹

Initial therapy for focal adenomyosis involves hormonal suppression of the endometrial tissue with oral contraceptives. If symptoms are severe and refractory to medical management, surgical intervention is recommended.

CASE PRESENTATION

A 17-year-old girl presented to the emergency department of our hospital on day 6 of menstrual cycle with pain felt over the right lower abdomen. She had such complaints

during menses, especially in the last days, for 2 years, with progressively worsening pain. She also had a history of analgesics use, but the pain was refractory to analgesic or non-steroidal anti-inflammatory drugs.

The patient was healthy and had a surgical history of a laparoscopic appendectomy and a laparotomy for left ovary's haemorrhagic cyst.

She had menarche at age 13 and referred regular menstrual cycles lasting 6 days with a normal flow. She denied any history of sexual intercourse.

On examination, the patient was afebrile with stable vital signs. Physical examination revealed diffuse tenderness to palpation with voluntary guarding over the right iliac fossa. Bimanual examination was deferred owing to the presence of narrow introitus. Serum human chorionic gonadotropin, complete blood count, kidney and liver function, pancreatic enzymes, urinalysis and erythrocyte sedimentation rate were within normal limits.

Figure 1 Transabdominal ultrasound performed to evaluate the uterus and adnexa. A transverse plane identifies the lesion at the right margin of the uterus.

Figure 2 T1-weighted axial image demonstrates a cystic lesion contained within the right uterine myometrium with an internal fluid–fluid level, signifying layering of serous and haemorrhagic/proteinaceous fluid.

INVESTIGATIONS

Transabdominal ultrasound was performed which demonstrated a large, thick-walled cystic lesion with a fluid level contiguous with the uterus and adjacent to the right ovary measuring 3.3×2.5 cm (figure 1). Doppler study showed lesion wall vascularity and unremarkable blood flow to the uterus and ovaries.

To confirm the diagnosis, a pelvic MRI was performed, which showed a large cystic structure of 3.3×3.2×2.9 cm contained within the myometrium of right uterus with internal fluid–fluid level. The dependent fluid was hyperintense on T1 and intermediate intensity on T2-weighted images, compatible with haemorrhagic and/or proteinaceous fluid (figure 2). The cyst wall was thickened and demonstrated T2-hypointense signal, reflecting myometrium hypertrophy.

DIFFERENTIAL DIAGNOSIS

The round mass in the right uterine wall, with no apparent communication to the endometrial cavity, correlating with the abnormality seen on ultrasound was considered to be a benign haemorrhagic lesion. The endometrial cavity and adnexal regions were normal. Two normal uterine horns were visualised and the right horn was displaced by the mass (figure 3). The differential diagnosis can be a haemorrhagic leiomyoma, a fibroid with fatty degeneration, an isolated congenital anomaly with haematometra in a non-communicating horn, a congenital uterine cyst and intramyometrial hydrosalpinx.

TREATMENT

Owing to the age of the patient, it was decided not to make the resection of the lesion. The patient was started on continuous oral contraceptive pills and had significant relief of her pelvic pain.

OUTCOME AND FOLLOW-UP

Subsequent follow-up ultrasound and MRI after 6 months and 1 year demonstrated a significant decrease in mass size to less than 2.5 and 1.5 cm, respectively (figure 4). The patient remains asymptomatic.

Figure 3 T2-weighted coronal image identifies a thickened cyst wall with hypointense signal, representative of myometrium hypertrophy and demonstrate two normal uterine horns, distinct from the cyst.

DISCUSSION

The cystic myometrial lesion, in this case, is most likely a cystic adenomyoma. The cyclic nature of symptoms, ultrasound appearance, similarity of the lesion and endometrium in MRI signal intensity and response to hormone suppression are all consistent with this suspected diagnosis.¹⁰

Severe dysmenorrhea and pelvic pain may be due to intracystic bleeding with progressive increase of the cyst size.

The differential diagnosis can be done based on MRI. In the case of haemorrhagic leiomyoma, methemoglobin accumulates in obstructed veins at the periphery of the mass, producing a rim that is T1-hyperintense and T2-hypointense. This is distinct from the T1-hypointense and T2-hypointense rim of haemosiderin in cystic adenomyosis. Fat and blood can be distinguished by the use of fat-suppression images, excluding the hypothesis of fibroid with fatty degeneration.^{3 11} Unless both uterine horns are well visualised in MRI, it is difficult to exclude isolated congenital anomaly with haematometra in a non-communicating horn.¹² In these cases, hysterosalpingography can be useful. Congenital uterine cysts and intramyometrial hydrosalpinx can also appear as cystic lesions within the myometrium; however, these entities contain simple fluid and lack of haemosiderin rim.

Figure 4 A T2-weighted axial image after 2 years of follow-up demonstrates the lesion with smaller size, less than 1.5 cm.

Treatment depends on the age of the patient, severity of her symptoms and size and localisation of the cyst. Medical treatment may be suppressive therapy with gonadotropin-releasing hormone agonists or continuous oral contraceptive pills, but mostly with temporary effect. Non-steroid anti-inflammatory drugs and analgesics are used for relief of symptoms. In young patients, if the medical treatment is not effective, conservative surgery for excision of the lesion with the minimally invasive procedure should be preferred, always taking into account fertility preservation in young patients.^{8 13}

Cystic adenomyoma, although a rare lesion in young girls, may be considered when severe dysmenorrhoea is associated with uterine cyst diagnosed by ultrasound and MRI. The use of radiological imaging modalities enables a non-invasive and a conservative approach to this disease management, which is particularly important in paediatric population.

Learning points

- ▶ Cystic adenomyoma, although a rare lesion in young girls, may be considered when severe dysmenorrhoea is associated with uterine cyst.
- ▶ Juvenile cystic adenomyoma can mimic uterine malformation. The principal differential diagnosis includes congenital anomaly with haematometra in a non-communicating horn.
- ▶ The use of radiological imaging modalities enables a non-invasive and a conservative approach to this disease management.

Competing interests None.

Patient consent Obtained.

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