CASE REPORT

Presacral tumours: a rare case of a dermoid cyst in a paediatric patient

Matthew Jones,¹ Japinder Khosa²

¹Department of General Surgery, Fremantle Hospital, Perth, Western Australia, Australia

²Department of Paediatric Surgery, Princess Margaret Hospital, Perth, Western Australia, Australia

Correspondence to

Dr Matthew Jones, matthew.lloyd.wynne.jones@googlemail.com

SUMMARY

Presacral tumours are considered very rare tumours with their incidence being around 1:40 000. Presacral dermoid tumours, part of the family of presacral tumours, usually present in the female adult population. They are benign tumours arising from all the three germ cell layers. Presentation in a paediatric population, defined as an individual under the age of 18 years, is extremely rare. A 15-year-old girl presented with abdominal pain present for 5 years diagnosed as a somatisation disorder. Upon further investigation with abdominal CT and MRI a diagnosis of a presacral mass was made. A laparoscopic-assisted transabdominal excision of the mass was performed and histopathology confirmed the radiological suspicion of a presacral dermoid cyst. As there are no reports of a presacral dermoid cysts being excised and reported in the paediatric surgical literature, we hope to highlight this pathology as a potential cause of abdominal pain in the paediatric population.

BACKGROUND

Presacral tumours are considered very rare tumours with their incidence being around 1:40 000.¹ Presacral dermoid tumours, part of the family of presacral tumours, usually present in the female adult population. They are benign tumours arising from all the three germ cell layers.² Presentation in a paediatric population, defined as an individual under the age of 18 years, is extremely rare.

CASE PRESENTATION

A 15-year-old Caucasian girl presented with a 24 h history of acute lower abdominal pain with nausea and vomiting. She had a background history of longstanding lower pelvic pain which was thought to be psychosomatic in nature.

Upon examination she was tender in the right iliac fossa with guarding. Her blood tests were unremarkable except slightly raised C reactive protein and urinalysis was clear. A clinical diagnosis of appendicitis was made for which she underwent diagnostic laparoscopy and appendicectomy.

Intraoperatively she had a normal looking appendix but free fluid in the pelvis and evidence of a ruptured ovarian cyst.

Postoperatively she continued to have pain and on day 3 she complained of dysuria for which a urinalysis was performed, which showed microscopic haematuria.

A CT KUB was organised to rule out renal calculus. It showed an incidental finding of a 75×55 mm

well-circumscribed left pararectal/adnexal lesion which intraoperatively was not seen in the pelvis.

An MRI showed a presacral cystic mass with foci of calcification and fat within, suggestive of a dermoid cyst.

The tumour markers in particular α -fetoprotein (AFP) and β -human chorionic gonadotropin (β -HCG) were normal.

A laparoscopic-assisted transabdominal approach was used to remove the presacral tumour in toto.

Histopathologically the lesion was reported to be a presacral dermoid cyst with no evidence of malignancy.

INVESTIGATIONS

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An MRI showed a presacral cystic mass with foci of calcification and fat within, suggestive of a dermoid cyst.

The tumour markers in particular AFP and β -HCG were normal.

DIFFERENTIAL DIAGNOSIS

- Presacral teratoma
- Ovarian cyst

TREATMENT

A laparoscopic-assisted transabdominal approach was used to remove the presacral tumour in toto.

Histopathologically the lesion was reported to be a presacral dermoid cyst with no evidence of malignancy.

OUTCOME AND FOLLOW-UP

The patient's abdominal pain has now resolved and she had no postoperative complications.

DISCUSSION

The presacral space, defined as the area bounded by sacrum posteriorly, rectum anteriorly, levator muscles inferiorly and superiorly by the peritoneal reflection, can herald different types of cystic lesions.

The differential diagnosis of a presacral cystic lesion is large including both benign and malignant pathologies. Benign lesions such as a presacral dermoid, epidermoid cysts or duplication cysts can occur in this region. Malignant teratomatous lesions are the most common presacral lesions in the paediatric population. Dermoid cysts can be differentiated from epidermoid cysts because they

To cite: Jones M, Khosa J. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008783

are not only composed of stratified squamous epithelium, but also have skin appendages like sweat glands, hair follicles or sebaceous cysts.³ Dermoid cysts can be unilocular or multilocular with the contents varying from clear fluid to blood and mucus. Caudal embryonic vestiges are postulated to result in dermoid cysts; however, teratomatous lesions are thought to be derived from primordial germ cells which have deviated from a common pathway of development.⁴

Most presacral dermoid cysts present asymptotically and are found incidentally. Symptomatic patients with presacral dermoid tumours can complain of vague symptoms such as perineal or back pain and some patients present with more specific symptoms such as constipation or urinary retention. Presacral dermoid cysts can become infected in up to 30% of cases.⁵ These patients can be mistaken for infected pilonidal disease, perirectal abscesses or fistula-in-ano when presenting with perineal discharge. Diagnosing presacral dermoids by ultrasound can be very difficult as proximity to the ovary can lead to a misdiagnosis of an ovarian cyst. Diagnostic laparoscopy can result in false negatives as lesions are frequently missed in the retrorectal space. Therefore other imaging modalities like CT and MRI are advocated if there is suspicion of a presacral mass.⁶ The CT features are a benign, hypodense lesion with well-demarcated edges with no adjacent visceral invasion (figure 1). The cyst signal on MRI depends on its content (figure 2). Malignant changes are suspected in a case of focal parietal thickening or with invasion of the adjacent organs.⁷

Surgery is the mainstay of treatment in presacral dermoid lesions. The ultimate goal is to remove the lesion from the presacral space in its entirety without any morbidity to the patient. Simple marsupialisation of the cyst is not adequate and has high recurrence rates.

The approach can be transabdominal, perineal or combined. A transabdominal approach is best used for lesions above the level of S4 as was the case in this patient. In paediatric patients

Figure 1 The sagittal CT slice showing a large presacral, benign, hypodense lesion with well-demarcated edges with no adjacent visceral invasion.

Figure 2 The T2-weighted sagittal MRI slice showing a large cystic presacral mass with high signal.

a transabdominal surgical approach is taken either through an open incision or a laparoscopic incision. A laparoscopic approach is less invasive and offers excellent visualisation of the presacral space for dissection. However, this is a more challenging operation technically and results in high conversion rates to an open transabdominal procedure.⁵ Particular attention and emphasis on careful dissection away from the rectum is required as postoperative fistulas can occur in up to 3% of cases.⁴ Recurrence rates are higher in malignant teratomas with recurrence rates being very low in dermoid lesions once all the tissues are removed.

In conclusion, presacral dermoid tumours are very rare in a paediatric population and either present incidentally or with vague symptoms of perineal pain. Diagnosis can sometimes be difficult, however CT and MRI help assess morphology and location. Surgery is the mainstay of treatment with low recurrence rates.

Learning points

- ▶ Presacral masses found on imaging can be secondary to dermoid cyst formation; however, more common differentials such as presacral teratomas and ovarian cysts are more widely reported.
- ▶ Continued, vague, abdominal pain should be investigated thoroughly before diagnosing somatisation disorder.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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