CASE REPORT

Cotyledonoid dissecting leiomyoma as a possible cause of chronic lower back pain

David O Onu,¹ Lisa M Fiorentino,² Michael W Bunting¹¹Department of Gynaecological Oncology, Royal Hobart Hospital, Hobart, Tasmania, Australia²Department of Anatomical Pathology, Royal Hobart Hospital, Hobart, Tasmania, Australia

Correspondence to

Dr David O Onu,
dr_donu@yahoo.com

SUMMARY

Uterine fibroids having the distinct pathological and immunohistochemical features of cotyledonoid dissecting leiomyoma have been reported infrequently. We describe a postmenopausal woman with an incidental finding of an abdominopelvic mass arising from the uterine fundus on routine radiological imaging of the lumbar spine. The imaging was performed for the investigation of chronic radicular lower back pain refractory to usual pain management. However, the woman did not manifest any gynaecological symptoms. Intraoperatively, the pelvic mass appeared malignant and a frozen section suggested uterine sarcoma. As such, the mass was radically resected, resulting in significant resolution of the back pain. To the authors' knowledge, this is the first report of cotyledonoid dissecting leiomyoma presenting solely as chronic lower back pain, and also the first report of this fibroid variant in Australasia. We discuss the diagnostic and operative challenges, emphasising the role of radiological imaging and immunohistopathology in such cases and review current literature.

BACKGROUND

Uterine leiomyomas are benign neoplasms of smooth muscle cells arising from the myometrium. They most commonly occur in women of reproductive age and tend to regress following the menopause. They are remarkably common pathological specimens, occurring in over 75% of hysterectomy specimens and are typically well-circumscribed, solid white tumours.¹ One of the most striking features of this benign tumour is the sharp line of demarcation between the tumour and the surrounding myometrium, forming a plane of cleavage which enables the leiomyoma to be easily shelled out at myomectomy.²

However, there is a range of histological variants of leiomyoma, some having very unusual growth patterns, among which is the cotyledonoid dissecting leiomyoma.³

In this report, we describe cotyledonoid dissecting leiomyoma presenting as an incidental finding of an abdominopelvic mass and as an unusual cause of chronic radicular lower back pain in a postmenopausal woman without gynaecological symptoms. We discuss the challenging surgical treatment, emphasising the role of radiological imaging and immunohistopathology in diagnosis of such cases and review the current literature.

CASE PRESENTATION

Our patient was a postmenopausal woman in her 50s who was referred for gynaecologic oncological evaluation after a large complex pelvic mass was incidentally found on a CT scan of the lumbar spine. The scan had been performed by the patient's local doctor for the investigation of her chronic lower back pain that radiated to the lower limbs and was refractory to a cocktail of pain medications. On a critical retrospective review of lumbar spine X-rays and MRI performed 3 years previously, also for the lower back pain, the pelvic mass was apparent. The mass did not appear to have any significant interval change in size over the 3-year period. Importantly, our patient did not manifest any gynaecological symptoms and did not have any significant medical history. She was on hormonal replacement therapy over the past 3 years for treatment of menopausal symptoms. She had a positive family history of colon and lung cancers but previous colonoscopy that was performed for the investigation of anaemia was normal. Recent Papanicolaou test and mammography were normal but Pipelle endometrial sampling was undiagnostic.

On examination, a large soft mobile abdominal mass was palpable. The pelvic examination was normal.

INVESTIGATIONS

CT showed a large soft tissue mass with few interspersed hypodense regions occupying the anterior abdominal cavity extending from the inferior aspect of the right lobe of liver down into the pelvis. It was well circumscribed and blended into the uterus and there was no evidence of ascites, peritoneal thickening or significant lymphadenopathy (figure 1).

Ultrasound confirmed the same findings, outlining a plane of cleavage between the single contiguous mass and the uterus with echogenicity similar to the myometrium (figure 1). The mass had a moderate blood supply arising from the pelvis but was thought to be independent of the uterine blood supply. There was a 4 cm solid fibroid on the anterior wall of the uterus.

MRI confirmed the same findings above and did not show any evidence of spinal canal stenosis or nerve root impingement (figure 2).

DIFFERENTIAL DIAGNOSIS

A differential diagnosis of gastrointestinal stromal tumour was considered before surgery and a colorectal opinion sort.

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Figure 1 Preoperative abdominopelvic CT (A and B) and ultrasound scan (C). (A) Coronal view of CT showing the complex pelvic mass filling the entire abdominal cavity (red arrows) and displacing the bowels upwards B, axial view of CT also showing the mass located more on the right and centre (red arrows). (C) Ultrasound scan again showing the same mass in the pelvis (red arrows).

TREATMENT

The patient underwent diagnostic laparoscopy at which time a large, complex, mucoid, soft tissue mass that appeared malignant and seen to be arising from the left uterine fundus was evident. There was also small volume nodular/adhesive disease on the peritoneum of the vesicouterine and Douglas pouches. The ovaries and bowel loops appeared normal. The frozen section of the lesion suggested a low-grade sarcoma arising from the uterus. Subsequently, a laparotomy was performed at which the multinodular, mucoid mass with irregular margins firmly attached to the left cornu of the uterus was completely resected weighing 1351 g (figure 3). Additionally, a class 2 radical hysterectomy with excision of the contiguous pelvic peritoneum, bilateral salpingo-oophorectomy, omentectomy and appendicectomy were also performed. The uterus weighed 210 g and harboured a well-circumscribed, whitish, submucous fibroid measuring

40×30×25 mm in dimension. No residual disease was found at the end of the radical surgery.

Postoperative management included analgesics, graded oral intake, physiotherapy as well as antibiotic and deep vein thrombosis prophylaxis.

Histopathological findings

Multiple cross-sections showed multinodular proliferation of smooth muscle tissue with an unusual cotyledonous architecture (Figure 4). Within these perinodular areas, a background of hypocellular and hydropic connective tissue was apparent. There was proliferation of epithelioid to spindle-shaped smooth muscle cells with regular round to ovoid nuclei and complete absence of mitotic figures. Extensive stromal hyalinisation was seen in some areas but there was no evidence of tumour necrosis or haemorrhage (figure 5).

Figure 2 Preoperative axial views of T2-weighted MRI of the lumbar spine at L2 level (A) and L4 level (B). The lesion can be seen occupying most of the abdominopelvic cavity and displacing the bowel loops posteriorly.

Figure 3 (A) Intraoperative image showing the red spongy multinodular lesion filling up the pelvis. (B) The lesion can be seen arising from the left cornu of the uterus displacing the left fallopian tube (black arrow).

Immunohistochemical staining showed strong positivity to oestrogen receptors (moderate nuclear staining in more than 50% of tumour cells), positivity to smooth muscle actin and desmin, but very low Ki67 proliferation index and negativity to CD10 (figure 6).

These histological features were in keeping with a diagnosis of cotyledonoid dissecting leiomyoma of the uterus without any morphological features of malignancy.

OUTCOME AND FOLLOW-UP

At 2-month follow-up clinic visit, the patient reported feeling much better than she had previously expected. Interestingly, she had near-complete resolution of the debilitating radicular chronic lower back pain without the need for her usual cocktail of analgesics. She had just recently resumed hormonal replacement therapy and continues to do well.

DISCUSSION

Cotyledonoid dissecting leiomyoma is an unusual variant of uterine leiomyoma which was first described by Roth *et al*³ in 1996 and named after the late Dr William H Sternberg who had painstakingly collected cases of what he called 'red seaweed lesion' from 1940s to 1970s. Only 45 cases of this rare condition had been reported in the literature.^{4–11}

This variant merits description because it is a rare benign tumour with bizarre gross and radiological appearances which may raise the possibility of malignancy.

Cotyledonoid dissecting leiomyomas are exophytic multinodular tumours, which generally exceed 10 cm in size.⁷ As was

seen in our case, the surface of the tumour is often red, congested and spongy, giving it a resemblance to placental tissue hence, described as cotyledonoid. These leiomyomas tend to arise in the subserosal myometrium, from the lateral aspect of the uterus around the uterine cornu and extend into the extra-uterine adnexal structures and pelvic cavity. Such lesions are aptly described as dissecting. Some case reports describe the exophytic leiomyomatous component as being continuous with an intramural dissecting leiomyoma, however, this was not seen in our case. In 20% of reported cases, an intravascular growth pattern was also identified.⁷

Histologically, this variant of leiomyoma, tends to show a dissecting growth pattern. As previously described, the tumour in our case was composed of epithelioid to spindle smooth muscle cells growing in sweeping and disorganised fascicles. Immunohistochemically, the cells expressed smooth muscle actin and desmin. There was no expression of CD31 or CD10. There was no nuclear atypia and no increase in mitotic activity. The proliferation index was less than 1% and there was no coagulative tumour necrosis. Extensive hyalinisation of the tumour stroma was evident and multifocal areas of hydropic degeneration were seen. These stromal findings are consistent with those reported in other cases, including perinodular hydropic degeneration and distinctive connective tissue alterations such as a highly vascularised stroma.

The epithelioid cytomorphology raises a relevant discussion point, since the inexperienced eyes might mistake this neoplasm for a true epithelial neoplasm, particularly during intraoperative frozen section examination. However, the macroscopic appearance and microscopic architecture and stromal features, are characteristic and diagnostic.

Clinically, cotyledonoid dissecting leiomyoma present commonly in the reproductive or postmenopausal period with abnormal uterine bleeding and abdominal or pelvic masses. The tumour was incidentally detected in only three previous reports either during radiological investigations, as in our case, or during surgery for other conditions.^{4–6} This fibroid variant being the sole cause of chronic lower back pain has never been reported. The authors strongly believe that it is feasible that a 1.35kg abdominopelvic mass clearly arising from the uterus may cause chronic lower back pain given the well described nervous innervation of the pelvis.

Surgical excision, in the form of hysterectomy, remains the mainstay of treatment for uterine sarcomas. In our case, the lesion macroscopically looked malignant and the frozen section diagnosis was of a uterine sarcoma. While we recognise the limitation of

Figure 4 Cross-section of the exophytic tumour showing multinodular proliferation of smooth muscle tissue with an unusual cotyledonous architecture.

Figure 5 Micropictographs. (A) Multinodular architecture of the cotyledonoid dissecting leiomyoma (H&E, $\times 100$). (B) The smooth muscle cells within the leiomyoma are epithelioid to spindled, with regular ovoid nuclei. Extensive stromal hyalinisation is present in the background (H&E, $\times 400$).

Figure 6 Immunohistochemical staining. (A) The cells express smooth muscle actin, strongly and uniformly ($\times 400$). (B) The cells do not express CD10 ($\times 400$).

frozen section in differentiating between benign and malignant smooth muscle tumours of the uterus, in our case, radical surgical resection was required to obtain macroscopic clearance of the disease. In other cases of this disease, a total abdominal hysterectomy and resection of the extrauterine tumour should be performed to avert the low risk of tumour recurrence which has been reported in a single case initially treated more conservatively by myomectomy and extrauterine tumour excision.¹¹ There are however no reported cases of metastases also supporting the benign nature of this variant of leiomyoma.

Learning points

- ▶ Cotyledonoid dissecting leiomyoma is an unusual variant of uterine fibroid, which is rarely reported in the literature.
- ▶ This benign tumour may demonstrate bizarre malignant appearance and may be difficult to differentiate from malignant tumours on frozen section at the time of surgery.
- ▶ Surgical treatment is justified given the malignant macroscopic appearance and very low risk of recurrence if more conservative approach is taken.
- ▶ Radiological imaging and immunohistochemical staining are very useful in diagnosis.
- ▶ Cotyledonoid dissecting leiomyoma can be an unusual cause of chronic lower back pain.

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Contributors DOO is the major contributor who analysed the patient's data and prepared the manuscript. LMF is the pathologist who prepared the histopathology

report and contributed to literature review. MWB is the gynaecologist who performed the procedure, provided patient's data and responded to the reviewers' comments. All authors read and approved the manuscript.

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REFERENCES

- 1 Cramer SF, Patel A. The frequency of uterine leiomyomas. *Am J Clin Pathol* 1990;94:435–8.
- 2 Anderson MC, Robboy SJ, Russell P. *Pathology of the female reproductive tract*. 1st edn. China: Churchill Livingstone, 2002, Chapter 15, Uterine smooth muscle tumours; 389–414.
- 3 Roth LM, Reed RJ, Sternberg WH. Cotyledonoid dissecting leiomyoma of the uterus. The Sternberg tumor. *Am J Surg Pathol* 1996;20:1455–61.
- 4 Menolascino-Bratta F, García de Barriola V, Naranjo de Gomez M, et al. Cotyledonoid dissecting leiomyoma (Sternberg tumor): an unusual form of leiomyoma. *Pathol Res Pract* 1999;195:435–8.
- 5 Kim MJ, Park YK, Cho JH. Cotyledonoid dissecting leiomyoma of the uterus: a case report and review of the literature. *J Korean Med Sci* 2002;17:840–4.
- 6 Gurbuz A, Karateke A, Kabaca C, et al. A case of cotyledonoid leiomyoma and review of the literature. *Int J Gynecol Cancer* 2005;15:1218–21.
- 7 Smith CC, Gold MA, Wile G, et al. Cotyledonoid dissecting leiomyoma of the uterus: a review of clinical, pathological, and radiological features. *Int J Surg Pathol* 2012;20:330–41.
- 8 Chavez Martinez S, Arias Gonzalez ML, Silva Lopez RM, et al. [Cotyledonoid dissecting leiomyoma of the uterus. A malignant-looking benign tumor]. *Ginecol Obstet Mex* 2012;80:528–33. Leiomioma cotiledonoide disecante del útero. Un tumor benigno de apariencia maligna.
- 9 Makharoblidze E, Goishvili N, McHedlishvili M, et al. Unusual types of smooth muscle tumors of uterine corpus: case reports and literature review. *Georgian Med News* 2013;7–11.
- 10 Tanaka H, Toriyabe K, Senda T, et al. Cotyledonoid dissecting leiomyoma treated by laparoscopic surgery: a case report. *Asian J Endosc Surg* 2013;6:122–5.
- 11 Roth LM, Kirker JA, Insull M, et al. Recurrent cotyledonoid dissecting leiomyoma of the uterus. *Int J Gynecol Pathol* 2013;32:215–20.

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