

## Case report

## Surgical management of a retroperitoneal pelvic desmoid tumour involving the sacrifice of external iliac vein and internal iliac vessels

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## 1. Introduction

Retroperitoneal pelvic desmoid tumours are rare with limited reports in the literature. They are locally aggressive neoplasms arising from deep muscle fascia, aponeurosis and tendons and do not have metastatic potential. They are associated with significant morbidity and mortality.

Desmoid tumours have an incidence of 0.2–0.4/100,000 and present at any age, most commonly between 25 and 35 years (Eastley et al., 2015). They represent less than 2% of all soft tissue sarcomas (Litchman, 2008). Pelvic desmoid tumours account for less than 20% of patients in most large retrospective series and are associated with female sex, hormonal factors, surgery or trauma, familial polyposis coli, Gardner's syndrome and high local recurrence (Fakhar et al., 2010; Marianai et al., 2000).

Management is commonly wide local excision (WLE), frequently challenging and associated with high morbidity, mortality and recurrence. We present a case of a retroperitoneal pelvic desmoid tumour, requiring the sacrifice of the left external iliac vein and left internal iliac vessels, the first to our knowledge reported. Our patient has had minimal morbidity and no evidence of recurrence.

## 2. Case report

A 65 year-old woman was referred with a left complex adnexal mass. Her history included lower abdominal pain of one year's duration and 3–4 weeks of unilateral oedema to her left leg. Medical background included medicated hypertension, appendectomy and a history of smoking. At presentation she was 10 years post menopausal and had a history of one normal vaginal delivery and one caesarean section. Gynaecological history was unremarkable. Family history included ovarian cancer in the patient's mother (died aged 67).

On examination the patient had a relatively fixed mass in the left iliac fossa. Ca-125 was normal (16) and magnetic resonance imaging (MRI) showed a fibroid uterus displaced by a large cystic complex mass in the left pelvis measuring 9 × 7 cm (Fig. 1). At radiological review the nature of the mass was unclear and was thought to be ovarian. Chest X-ray and computed tomography (CT) scan confirmed the above (Figs. 2–3).

A diagnostic laparoscopy was performed revealing the tumour to be retroperitoneal, emerging from the left obturator fossa. A midline laparotomy was proceeded to and following pelvic side-wall dissection it was realised the external iliac artery and vein were stretched over the medial aspect of the tumour. True cut biopsies were taken and frozen section showed a myxoid spindle cell tumour possibly reactive but unable to exclude a leiomyosarcoma. A second Gynaecological Oncologist and Vascular Surgeon attended. In order to excise the tumour the left

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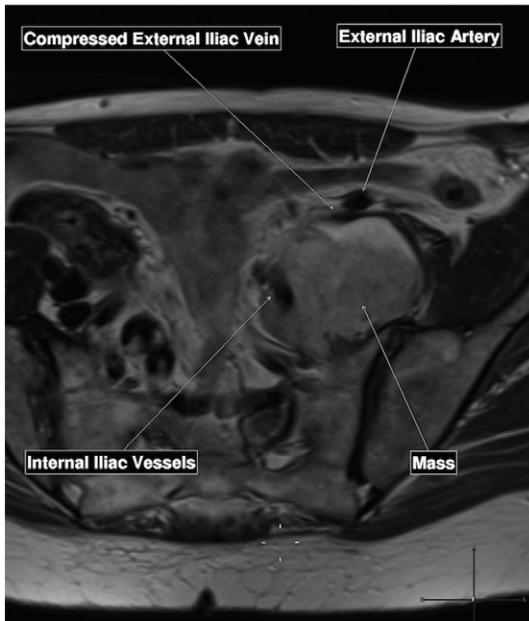


Fig. 1. Axial MRI.

external iliac vein and internal iliac vessels were sacrificed. Placing an interposition graft was considered however obtaining a clean anastomosis would have required a long graft. Given the diagnosis was uncertain and good collateral circulation present this was decided against. The obturator nerve was identified and kept intact.

Surgery took 8 h and estimated blood loss was 1.5 L. The patient received 3 units of packed red blood cells and required a short period of vasopressor support. Initially post-operatively she had gross oedema of her left leg but recovery was otherwise uncomplicated. Final histopathology revealed aggressive fibromatosis. Surgical margins were clear except for the left lateral resection margin where the tumour abutted the sacrum.

The patient was reviewed by physiotherapists and fitted for a full-length compression garment. She was given a 3-month course of prophylactic low molecular weight heparin and referred for Familial Adenomatous Polyposis (FAP) screening. Currently, 18 months post-operatively she is well, without recurrence and has minimal morbidity from leg oedema attributable to good collateral circulation (Fig. 4).

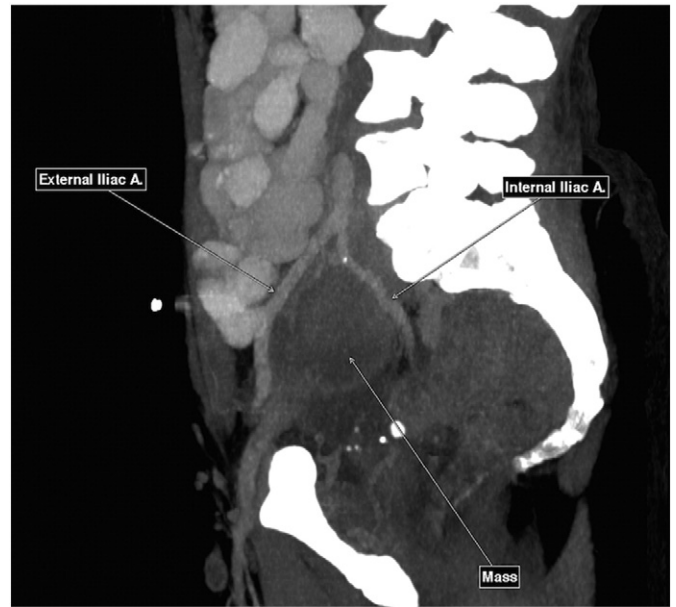


Fig. 3. Sagittal T2 CT demonstrating the displacement of the iliac vessels by the tumour.

There are no consensus guidelines regarding medical imaging in routine follow up however as our patient had positive margins her risk of recurrence is high and we have chosen to do so with regular MRI's.



Fig. 4. The patient's lower limbs post surgery, demonstrating excellent collateral circulation.

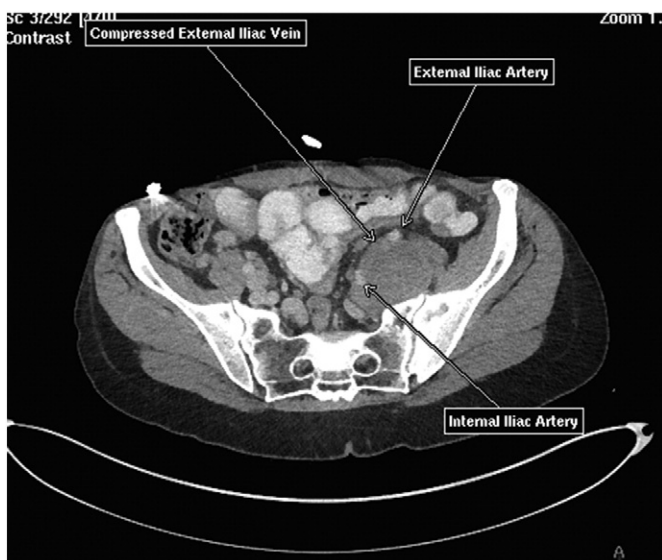


Fig. 2. Axial T2 CT demonstrating the vessels traversing the tumour.

### 3. Discussion

Desmoid tumours are the result of deregulation of the wnt signalling pathway secondary to the accumulation in B-catenin d, a protein involved in cellular proliferation and differentiation (Eastley et al., 2015). This occurs in two ways, most commonly (85%) due to sporadic somatic mutations in the gene encoding for B-catenin d (CTNNB1), and less commonly due to mutations in the gene that encodes for adenomatous polyposis coli (APC), which under normal circumstances binds to B-catenin d signalling for its breakdown (Eastley et al., 2015). Immuno-histochemical analysis of the tumours is positive for b-catenin and vimentin, in addition to COX-2 and the tyrosine kinase receptor PDGFR $\beta$ ; they are also frequently positive for the androgen, oestrogen beta and progesterone receptors (Al-Jazrawe et al., 2015).

Regardless of their anatomical location desmoid fibromatoses share a common macroscopic and microscopic appearance (Litchman, 2008). They are frequently large tumours (less than 5 cm to greater than 20 cm in dimension) with infiltrative borders and gross surfaces that are white or tan in colour and fibrous with trabeculations (Litchman, 2008). Histologically desmoid tumours are comprised of dense spindle-like fibroblastic cells within a collagenous matrix lacking a pseudocapsule, hence their tendency to infiltrate (Al-Jazrawe et al., 2015).

Presentation is characterised by where tumours arise, with locations divided into: intra-abdominal, abdominal wall and extra-abdominal (Litchman, 2008). Regarding intra-abdominal pelvic desmoid tumours patients remain asymptomatic until the growth of the tumour leads to intestinal, neuronal, vascular and ureteric symptoms (Fakhar et al., 2010). The most common presenting complaint is pain (pelvic, abdominal, leg or vulval) (Marianai et al., 2000). The natural history of desmoid tumours is not fully understood, with some spontaneously regressing and others progressing. There is a paucity of data regarding prognostic factors that differentiate between the two (Al-Jazrawe et al., 2015).

Risk factors for pelvic desmoid tumours include female sex, hormonal factors, surgery or trauma, FAP and Gardner's syndrome (Fakhar et al., 2010; Marianai et al., 2000; Weiss and Goldblum, 2008). In support of their hormonal influences, pelvic desmoid tumours are commonly described in young women during or after pregnancy and are reported to regress after menopause (Fakhar et al., 2010). However this is controversial with a recent review disputing the female hormonal environment to have a major aetiological role; with recurrence after surgery not being associated with gender, and pregnancy not being associated with an increased risk of tumour progression or recurrence after surgery (Al-Jazrawe et al., 2015). The review commented on Schiessling et al.'s work (Schiessling et al., 2013), which found that within FAP patient's males have more frequent and larger desmoid tumours compared with females. The authors then cited Fiore et al. (2014) who suggest the higher frequency of tumours in females is likely to reflect higher imaging rates, especially surrounding pregnancy (Al-Jazrawe et al., 2015).

Management options include radiotherapy, chemotherapy, non steroidal anti-inflammatory drugs, Tamoxifen, watchful waiting and WLE (Firoozmand and Prager, 2001; Al-Jazrawe et al., 2015). WLE is common however watchful waiting is advocated for asymptomatic stable tumours to avoid high morbidity and mortality rates associated with surgery (Al-Jazrawe et al., 2015). Surgery is frequently challenging due to the location of tumours and positive resection margins not uncommon, which when coupled with trauma being a known stimulant for desmoid

growth likely contributes to high local recurrence rates of up to 65–88% (Firoozmand and Prager, 2001).

Recurrences frequently occur within 2 years. There is increased risk with extra-abdominal tumours, positive surgical margins and patients with FAP syndrome (Litchman, 2008 & Al-Jazrawe et al., 2015). Presently, there are no reliable genomic or clinical markers that predict tumour behaviour in regards to recurrence or regression and there is a growing field of research investigating this (Eastley et al., 2015). Regarding adjuvant therapy to reduce recurrence radiotherapy is possibly useful and targeted therapies focussing on the PDGFR $\beta$  pathway have been explored but response rates are low (Al-Jazrawe et al., 2015).

As this case illustrates pelvic desmoid tumours are a challenge for the Gynaecological Oncologist. They are rarely encountered and not normally considered during the differential diagnosis of a pelvic mass. Had the team realised the possibility of a desmoid tumour preoperatively CT guided biopsy (as per sarcoma protocol) may have been contemplated. In retrospect, the vessels can be seen traversing the medial surface of the tumour, a finding that was overlooked preoperatively (Fig. 2). Subsequently intra-operative decision making and excision was challenging. The patients history of unilateral leg oedema and the paucity of blood in her compressed external iliac vein overlying the tumour encouraged the surgeons to believe that sacrificing the vessels may not be too morbid as significant collateral circulation had likely developed. Fortunately this proved to be true, as the patient now has minimal peripheral oedema attributable to the insidious onset of these tumours (Fig. 4).

Regarding the vascular involvement of this tumour, one series by Marianai et al. (2000) describes two cases in which the external iliac artery was encountered during tumour resection however to our knowledge there are no reported cases where the sacrifice of the left external iliac vein and left internal iliac vessels has been required, making this case unique. The paucity of data regarding the behaviour of desmoid tumours highlights the importance of publishing rare cases to help guide clinical practice and research.

*Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. All authors report no conflicts of interest in relation to this manuscript.*

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