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## **Pre-sacral schwannoma**

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## Main Text

Tumours of the pre-sacral space are rare, with an incidence of 1/40,000-1/60,000.<sup>1</sup> Despite this, a general surgeon can expect to encounter at least one case throughout a typical career.<sup>2</sup> We describe a case of pre-sacral schwannoma, an uncommon tumour in this location, which illustrates diagnostic and operative challenges of pre-sacral tumours.

### Case Report

A 52-year-old female was referred to the surgical department with an incidental pre-sacral mass on MRI, requested for investigation of heavy menstrual bleeding. Alongside uterine fibroids, MRI revealed a retroperitoneal, solid, well-circumscribed, homogenous 21x21x20mm mass at S1-2 (Fig. 1). It appeared to be displacing the pre-sacral vessels (Fig. 2). The case was discussed at a Multidisciplinary Meeting, with an impression of a benign lesion and a recommendation for excision during a planned hysterectomy for uterine fibroids. The patient denied abdominal, neurological or bowel/bladder symptoms. On examination there was tenderness in the lower abdomen. Digital rectal examination (DRE) was normal. Flexible sigmoidoscopy and pre-operative CT Chest/Abdo/Pelvis were unremarkable.

She underwent laparotomy, subtotal hysterectomy, bilateral salpingectomy and excision of pelvic mass. Left oophorectomy was performed as the left ovary was asymmetrical. The mass was more adhered to the pelvic side wall than anticipated, sitting in close proximity to the right internal iliac vein, presacral veins and superior rectal vessels. It was dissected meticulously

(Fig.3). Blood supply was encountered at the inferior border and cauterised. After complete excision the mass measured 30x20x16mm. Histology was consistent with a schwannoma (Doc. S1). Histology of the ovary was benign. The patient had no peripheral stigmata of Neurofibromatosis I or II. MRI brain excluded acoustic neuroma. The patient reported no symptoms post-operatively.

## Discussion

Pre-sacral masses are rare. The pre-sacral space is a potential space bordered anteriorly by the mesorectal fascia, posteriorly by the presacral fascia, laterally by ureters, iliac vessels and sacral nerve roots, superiorly by the peritoneal reflection and inferiorly by Waldeyer's fascia.<sup>2,3</sup> It contains embryologic remnants derived from a variety of tissues; masses which arise here reflect that.<sup>1</sup> They are classified as congenital (most common), inflammatory, neurogenic, osseous or miscellaneous, and can be benign or malignant.<sup>2</sup> Our case, a schwannoma, is a benign tumour which arises from Schwann cells.<sup>4,5</sup> Alongside other neurogenic entities they account for 10% of pre-sacral lesions.<sup>2</sup> The majority are benign; 5-18% are associated with Neurofibromatosis I, which may undergo malignant changes.<sup>6</sup>

This case demonstrates the challenges of pre-sacral tumours, from diagnosis to operative management. Symptoms, if present, tend to be non-specific and insidious, most common being pelvic or back pain (38%).<sup>2,7</sup> Many patients are asymptomatic and the mass is found incidentally.<sup>6</sup> Detection by DRE is highly variable (35-97%).<sup>2,3,7</sup> MRI is the preferred imaging modality, however is unreliable in distinguishing between benign and malignant tumours.<sup>1,4</sup> Surgery is therefore usually required for definitive histological diagnosis. Surgical approaches

include posterior, for small lesions below S3-S4; and anterior or combined, for lesions above S3-S4, depending on the size of the tumour and sacral involvement.<sup>2,3</sup> Laparoscopic cases have been described.<sup>3</sup> Excision can be challenging, and minimal bleeding can obscure the operative field within this narrow space. Life-threatening bleeding has been described,<sup>3</sup> and should be pre-empted, particularly if the surgery is prolonged.<sup>10</sup> Preoperative embolisation of larger tumours can be considered.<sup>8</sup> Neurological injury is higher with larger tumours,<sup>10</sup> and complex surgery such as sacrectomy.<sup>2</sup> Recurrence rates vary depending on tumour type, with around 11% for benign<sup>7</sup>, and as high as 84% for malignant lesions<sup>3</sup>. A recent meta-analysis showed no difference in progression-free survival for en bloc or gross-total resection for Ancient Schwannomas (Schwannomas involving  $\geq 2$  contiguous vertebrae or with a diameter  $\geq 5$ cm). Gross-total resection therefore can be performed in challenging Schwannoma cases. Multidisciplinary approach is crucial, with consideration of Vascular, Orthopaedic and Colorectal expertise.

Pre-operative biopsy is controversial. Historically, biopsy was contraindicated due to risks of faecal fistula, meningitis, abscess formation and malignant seeding.<sup>2,3</sup> Recent literature however suggests no evidence of increased risk of seeding,<sup>1,7</sup> and biopsy may be appropriate in tumours where neoadjuvant therapy has a role, or tumours which could be treated with chemotherapy alone (GIST, lymphoma).<sup>1,7,9</sup>

With liberal use of imaging, the authors estimate that pre-sacral tumours will be encountered more frequently in the future. A high index of suspicion is required in patients presenting with non-specific symptoms or abnormal DRE. The role of pre-operative biopsy remains controversial; authors suggest consideration of biopsy only when it is likely to change management. Surgery carries significant risks, including bleeding and neurological injury,

however is generally recommended to exclude malignancy. Authors emphasize thorough pre-operative planning, Multidisciplinary Team involvement and meticulous dissection to avoid complications.

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## Figure Legends

Fig. 1. – Magnetic Resonance Imaging Pelvis. Pre-sacral mass, hypo-intense on T-1 weighted imaging (short arrow). Uterus with numerous bulky fibroids (long arrow).

Fig. 2. – Magnetic Resonance Imaging Pelvis. Right pre-sacral mass (short arrow), compressing a dilated pelvic vein (long arrows). The mass is hyper-intense on T2 weighted imaging.

Fig. 3. – Pre-sacral mass following meticulous dissection with essentially no blood loss during the operation. Mesorectal fascia is displayed (arrow).

## Supporting Information

Doc. S1. – Histology.