Presacral myelolipoma: A gynaecological disorder mimicry

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SUMMARY

Presacral myelolipomas are benign and do not warrant surgical resection unless symptomatic. If the mass exhibits suspicious imaging features or if there are any lingering suspicion, percutaneous biopsy can be performed to aid in the diagnosis. Our patient had classic CT and MRI imaging features of presacral myelolipoma. The percutaneous biopsy was performed due to the clinical suspicion of liposarcoma. The pathology was hence extremely helpful in determining the subsequent management of the patient, who was otherwise considered for preoperative radiotherapy for suspicion of pelvic liposarcoma.

INTRODUCTION

Myelolipoma is a benign tumour consisting of mature fat tissue and normal haemopoietic elements that is commonly found in the adrenal glands. It also can occur outside the adrenal glands—extra-adrenal myelolipoma (EAM). EAM is commonly localised in retroperitoneum, particularly in the presacral region. Myelolipoma is usually a slow-growing and asymptomatic tumour but they can become symptomatic when the size exceeds 10 cm.[1] Herein, we present a case of a 7 cm presacral myelolipoma in a 69-year-old female who presented with painful post-menopausal per vaginal bleeding.

CASE PRESENTATION

A 69-year-old female presented with 3 months history of post-menopausal per vaginal bleeding and suprapubic abdominal pain with pain score of 0 at rest, 3 during strenuous activity. She had no fever, constitutional symptoms, lower urinary tract symptoms, bowel symptoms and bone pain. Her past medical and surgical history were unremarkable. She is a mother to three sons with a single partner. On physical examination, she was hemodynamically stable, abdomen was soft and no mass was palpable. Per vaginal examination was normal. Serum full blood count, renal profile, coagulation profile, erythrocyte sedimentation rate and C-reactive protein were normal. Serum tumour markers such as carcinoembryonic antigen, CA19-9, CA125, beta-human chorionic gonadotrophin (bhCG) and alpha-fetoprotein were normal as well.

An initial ultrasound of the pelvis was performed and demonstrated uterine fibroids as well as a solid mass posterior to the uterus, which appeared to be arising from the left adnexa and was thought to be ovarian in origin (Figure 1A). A computed tomography (CT) study showed that the lesion was presacral in location and measured approximately 3.8 × 4.1 × 2.9 cm. The mass was predominantly of fat attenuation and some soft tissue components (Figure 1B). The magnetic resonance imaging (MRI) study showed a 4.8 × 3.4 × 7.3 cm well-circumscribed, lobulated, predominantly fatty mass in the presacral space extending from the level of the mid S1 to the mid S5 vertebral bodies. The sacral nerve roots were not involved, and no bony extension was appreciated. Internal soft tissue component which does not suppress fat saturated sequences was seen with questionable mild enhancement. The impression was that the imaging features were most consistent with a myelolipoma, although a liposarcoma could not be excluded (Figure 1C).

A CT-guided core needle biopsy of the presacral mass was performed, and the tissue samples were submitted for histology (Figure 2A). Microscopy sections show a fatty tumour containing areas of erythropoiesis featuring scattered megakaryocytes, erythroblasts and differentiating myeloid cells. The trilineage haematopoiesis present is most consistent with a myelolipoma (Figure 2B and 2C). Additional stains showed some of the larger mononuclear cells present within the infiltrate are positive for haematopoietic precursors. These stains confirmed the impression of a myelolipoma (Figure 2D). Fluorescence *in situ* hybridisation (FISH) analysis for Mouse Double Minute-2 Homolog (MDM2) was negative for amplification and thus supported the impression of myelolipoma.

Her per vaginal bleeding was treated with 1 week of oral route Tranexamic Acid 500mg TDS and Celecoxib 200mg BD. Pap smear test was negative. She was offered surveillance versus surgical resection since it was a biopsy-proven benign disease and the symptoms had already resolved. She opted for surveillance. She had undergone her 5th year of surveillance as of this writing. She was asymptomatic, and serial (yearly) ultrasound study showed no interval changes.

DISCUSSION

Myelolipomas are benign lesions which contain mature adipose cells and trilineage haematopoietic cells (red blood cells, white blood cells and platelets).[2] The adrenal glands are the most common site of occurrence, and the pre-sacral region is the most frequent extra-adrenal location. Other extra-adrenal locations include the pelvic retroperitoneum, mediastinum, musculofascial tissue, liver, kidney and stomach.[3] Presacral myelolipomas classically occur in older patients with a female predominance of approximately 2:1.[4] The incidence at autopsy ranges from 0.08% to 0.4%.[5] These lesions are usually asymptomatic and incidentally

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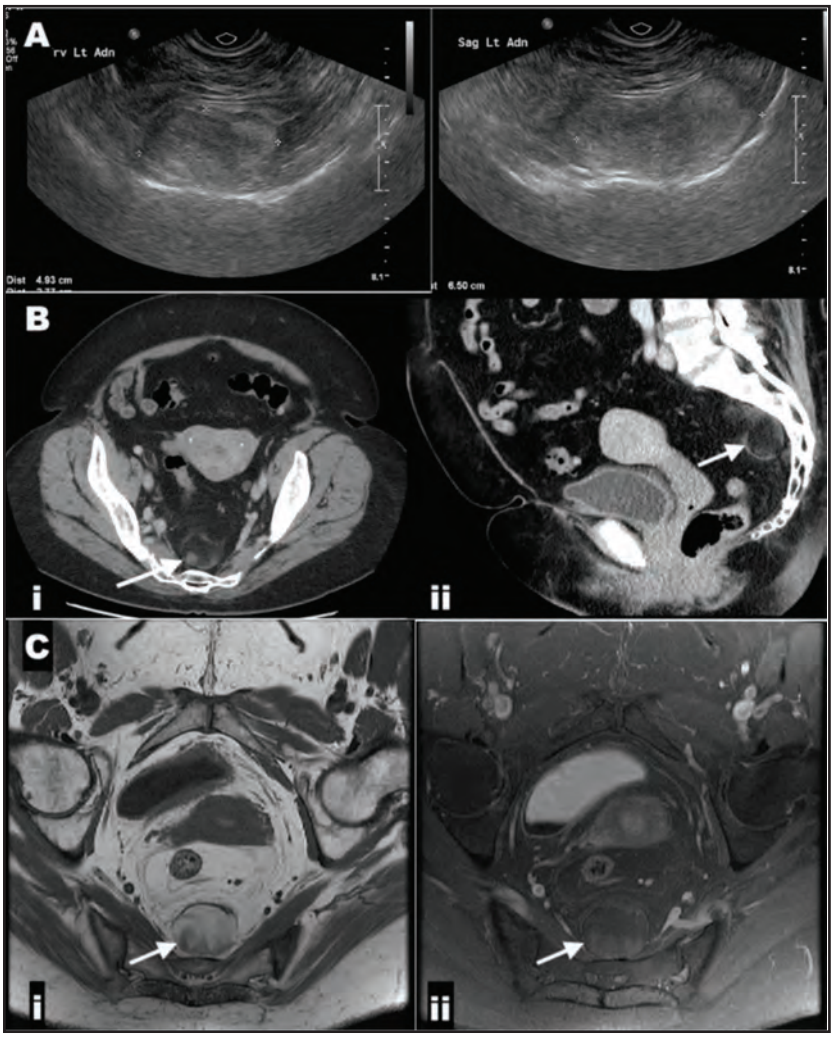

Fig. 1: Multimodality diagnostic imaging to assess the presacral tumour. (A) Ultrasound of the pelvis demonstrated a solid hyperechoic lesion posterior to the uterus. (B) Contrast-enhanced CT IN (i) axial and (ii) sagittal views demonstrated pre-sacral fat-containing lesion. The arrow illustrates the soft tissue component. (C) Axial T1-weighted MR image of the presacral tumour. (i) The arrow shows high-signal-intensity presacral lesion with some regions of intermediate signal intensity within. (ii) Axial T1-weighted fat-saturated gadolinium-enhanced MR image, arrow shows mild enhancement of tissue within the presacral lesion

discovered, as in our case. However, the lesion could exert a mass effect on adjacent structures, including the bladder, ureters, sacral nerve plexus and rectum. These lesions have an association with Cushing syndrome, Addison disease, adrenal hyperplasia and chronic exogenous steroid usage.[4]

The imaging appearances of a presacral myelolipoma is that of a well-encapsulated round or ovoid mass containing varying amounts of fat and soft tissue within. The soft tissue component may enhance post-contrast enhancement and small areas of haemorrhage within the lesion may give rise to calcifications on imaging. The lesion typically does not invade the surrounding bony architecture. As such, the lesion typically appears hyperechoic on ultrasound due to the varying fat content. On CT imaging, the fat-containing component of the myelolipoma will be of low attenuation (≤ –20HU). On MRI, the lesion will appear hyperintense on T1W sequences and hypointense on fat-suppressed T1W sequences.[6]

The main differentials for a fat-containing lesion in the pre-sacral region would be that of a liposarcoma, teratoma, extramedullary haematopoiesis or neurogenic tumour.[3,7] A combination of imaging findings and the clinical history aids in the diagnosis. Liposarcomas are the most common fat-containing retroperitoneal tumour. A liposarcoma typically has ill-defined margins with no surrounding capsule.[7] Neurogenic tumours are the second most common type of presacral tumours; they contain neural elements which are not seen in myelolipomas. On imaging, macroscopic fat is also not typically seen for neurogenic tumours.[8] A teratoma may contain areas of fat, soft tissue and calcium. However, they contain mesenchymal tissue elements on microscopy, which helps differentiate this from a myelolipoma.[6] Extramedullary haematopoiesis may resemble myelolipoma microscopically; however, imaging characteristics are typically ill-defined, multifocal lesion with a lack of macroscopic fat.[9] Clinically, these also occur with a male preponderance, in association with myeloproliferative disorders and chronic haemolytic anaemias.

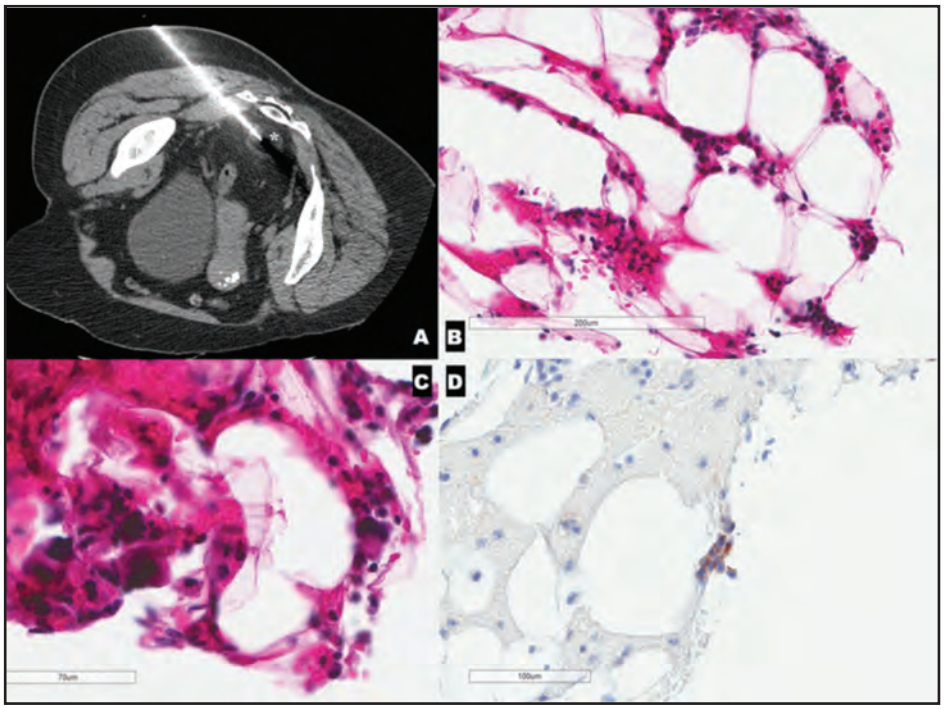

Fig. 2: Histopathological examination of the biopsied presacral tumour specimen. (A) CT-guided biopsy of the presacral lesion (asterisk). (B) Photomicrograph from the obtained biopsy specimen using hematoxylin and eosin (H&E) staining (200× magnification) showing haematopoietic cells in the background of fat. (C) Photomicrograph from the obtained biopsy specimen using H&E staining (200 × magnification) showing megakaryocytes. (D) Photomicrograph from the obtained biopsy specimen using immunostain for E-Cadherin (400× magnification) labelling a cluster of erythroblasts

With respect to management, The AACE/AAES Guideline (2009) recommends that myelolipomas that are observed (not receiving surgical excision) should undergo radiological evaluation at 3 and 6 months, continued by an annual interval for 1–2 years. Accepted indications for the surgical excision of myelolipomas are symptomatic tumour, size > 4 cm, metabolically active tumour, and a suspicion of malignancy on an imaging study. Malignant degeneration has not been documented.[10]

CONCLUSION

Presacral myelolipomas are benign and should be considered in the differential diagnosis during the evaluation of presacral fatty masses. The definitive diagnosis of presacral myelolipoma relies on histopathologic evaluation. Biopsy is necessary to exclude other malignant pathology that requires more aggressive management.

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