CASE REPORT

Large perineal and gluteal angiomyxoma: the role of diagnostic imaging and literature review

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SUMMARY

Aggressive angiomyxoma is a benign soft tissue tumour usually affecting the pelvis and perineum predominantly in women. Because of its variable presentation, this tumour is often clinically misdiagnosed as liposarcoma. We describe a case of a 38-year-old woman who presented with a large perineal and gluteal mass which increased in size in one year. Ultrasound showed hypoechoic mixed solid and cystic mass. Contrast CT and MRI examinations showed typical appearance of swirling and layering pattern. She had undergone TAHBSO as the mass was difficult to dissect intra-operatively. The post-operative specimen confirmed to be an aggressive angiomyxoma.

BACKGROUND

We describe a very rare and an aggressive angiomyxoma presented with a huge perineal and gluteal mass. We also highlight the importance of imaging features, including the characteristic findings of 'swirling' appearance in post-contrast MRI images which aid in pre-operative diagnosis and planning. The pre-operative core biopsy was not representative as the mass was huge.

CASE PRESENTATION

A 38-year-old lady presented with painless perineal and gluteal mass which progressively increased in size for the past 1 year. She denied any symptom related to gynaecological problems like dysmenorrhoea, menorrhagia, and irregular menses; or urological symptoms like dysuria and incontinence; or bowel related symptoms like constipation and obstructive bowel symptom.

Physical examination revealed a large soft to firm mass at the left perineal region extending to the left gluteal region. The left labia majora was bulging with fullness of the ischiorectal fossa.

INVESTIGATIONS

Ultrasound showed a large hypoechoic and heterogeneous pelvic mass which consisted of cystic and solid component (figure 1). Contrast enhanced CT showed a huge heterogeneously enhanced well-defined mass occupying the perineum, involving the vulva, the left ischiorectal fossa and also the subcutaneous space of the left gluteal region (figure 2). The mass measured 17 cm×24 cm×21 cm (width×anterior–posterior×craniocaudal). There was no evidence of lung or bone involvement.

MRI was carried out and the mass demonstrated homogeneously a low signal on T1-weighted image (T1WI), heterogeneously high signal on T2WI and

enhanced intensely following contrast administration. There were also 'swirling' and layering strands appearances within the mass in the post-contrast images (Figure 3). The mass showed a clear fat plane and displacement of the adjacent structures. The cervix and uterus were displaced supero-posteriorly. The anal canal and distal rectum also were compressed without causing significant bowel obstruction. Postero-inferiorly, the mass insinuates through the left ischiorectal fossa and medial aspect of the left gluteus maximus and extends posterolaterally into the subcutaneous tissue of the left gluteal region. Antero-inferiorly the mass extends to the level of mid thigh.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses which were considered in this patient were liposarcoma and malignant fibrous histiocytoma.

TREATMENT

She underwent laparotomy which required long operation hours by a conjoint venture of a gynaecologist and a surgical team. Intra-operatively there was a huge left labial mass extending to the left gluteal region. It was lobulated, had a well-defined margin, and consisted of cystic and solid component with a smooth surface. This mass was extending to the left lateral vaginal wall and bladder base up to the level of the pelvic cavity. Total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAHBSO) was later performed as the mass was difficult to dissect. There was no invasion into the vital structures. The rectum was not involved. The mass weighed 2.5 kg.

OUTCOME AND FOLLOW-UP

Histopathologically, sections from the nodular tissue and cystic spaces showed a tumour composed

Figure 1 Ultrasound of the pelvis showed a huge mixed solid and cystic component within the perineal lesion. Uterus was normal.

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Figure 2 Contrast enhanced CT of the abdomen (axial and sagittal section) showed a heterogeneously enhancing lesion arising from the perineal region displacing the uterus and rectum. There was clear fat plane between these structures.

of hypocellular, bland looking stellate and spindle-shaped mesenchymal cells embedded mainly in collagen and focally myxoid stroma. There was no nuclear atypia seen. The degenerative cystic spaces were present. Mitosis and necrosis were absent. Invasion into the skeletal muscle was present and the margins were involved. Overall features were consistent with aggressive angiomyxoma.

The postoperative period was uneventful. There was no post-operative complication. The patient was discharged 8 days post-surgery. Two months follow-up showed no evidence of recurrence.

DISCUSSION

The majority of aggressive angiomyxoma occurs among premenopausal women in the pelvis and perineum which is frequently misdiagnosed pre-operatively because of its rarity and no specific and typical clinical symptom. Typically, the lesion is slow growing hence the patient may present with a large mass once clinically symptomatic.¹ The smaller lesions at presentation usually are of incidental findings. It may also locally infiltrate the adjacent structures like the bladder, bowel and the pelvic bone.^{2,3}

The CT and MRI beforehand would give a detail of tumour extension, involvement of the pelvic floor and adjacent organs.

Figure 3 (A) MRI of the axial T1-weighted image (T1WI) showed a huge homogeneously low signal intensity mass (red arrow); (B) MRI of the axial T2WI showed a heterogeneous mass at the perineal region which shows predominantly high signal on T2WI (red arrow); (C) the axial and (D) the coronal T1WI post-gadolinium images showed an avidly enhancing perineal mass with 'swirling' and 'layering' pattern (yellow arrow) which are the typical characteristics in angiomyxoma.

This information will also aid in deciding the surgical approach. The pre-operative tissue diagnosis was a challenge as the biopsy sample was not representative particularly in this patient with a huge mass. The definitive tissue diagnosis was obtained post-operatively.

Radiologically, in this case and other reported cases, they typically have a well-defined margin for displacing rather than infiltrating the adjacent organs.⁴ Ultrasound imaging showed a complex heterogeneously hypoechoic mass which even appears cystic.^{4,5} The mass had attenuation lower than that of a muscle on CT as reported previously.⁵ The characteristics on MRI are isointense relative to the muscles on T1WI, high on T2WI and intensely enhanced following contrast with the internal 'layering' or the 'swirling' pattern. These swirling and layering strands pattern in our patient on the contrast-enhanced images is a characteristic and also were demonstrated in approximately 83% of the reported cases.³⁻⁵ The MRI fat saturation sequence was also important to differentiate the tumour from lipoma or liposarcoma.

The high water content and the loose myxoid matrix attributed to the appearance of hypoechoic on ultrasound, hypodensity on CT and hyperintensity on T2-weighted MRI. These previous reported findings are consistent with those of this patient.³⁻⁷ Despite the rarity, the potential of local infiltration and distant metastases should be considered.^{5,6} Conventional vascular angiogram also can be performed and usually would reveal a hypervascular mass as reported previously. This was not performed in our centre, because intravascular embolisation was not considered as part of the treatment.⁷

The tissue histopathological and immunohistochemical examinations are the gold-standard for the final diagnosis. Histopathologically, it is usually composed of fibroblasts within a strong myxoid background, prominent proliferation of vascular tissue and almost an absence of mitotic activity. Desmin in the myxoid bundles and/or stromal cells are positive in majority of the cases, but variable of positivity for the actins and CD34.⁸

Surgical resection is the most acceptable mode of treatment. This patient was initially planned for a complete surgical resection. However, owing to the difficulty of resection intra-operatively TAHBSO was performed. The likelihoods of the disease-free for wide local excision were not statistically different from those who had complete resection (50% vs 40%). The disease relapse is usually asymptomatic resembling the initial presentation of the primary disease thus necessitating clinical and diagnostic imaging follow-up.^{8,9}

Learning points

- ▶ Aggressive angiomyxoma is a rare benign pelvic tumour and radiologically it has a typical character of displacing rather than infiltrating the nearby organ.
- ▶ Internal 'layering' and 'swirling' appearance on a contrasted study is characteristic but not specific. Histopathological examination remains the gold-standard for the diagnosis.
- ▶ Angiomyxoma should be considered as a differential in a female patient presented with a massive perineal and gluteal mass.
- ▶ Proper diagnostic imaging is essential for diagnosis and disease extension which determines the surgical approach.

Competing interests None.

Patient consent Obtained.

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