Angiomyofibrosarcoma: a rare ischiorectal fossa swelling

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INTRODUCTION

Tumours of the ischiorectal fossa are rare. These are often mistaken for inflammatory pathology and the diagnosis is often delayed. This article reports a case of a soft tissue sarcoma mimicking an abscess of the ischiorectal fossa. To date, only one case of sarcoma involving the ischiorectal fossa has been reported in the world literature (Long Pretz et al, 1998). This is a rare lesion in an unusual location, which has limited treatment options.

DISCUSSION

The ischiorectal fossa is the largest of the anorectal spaces. This anatomical location and the rich lymphovascular network makes this a common site for infection. The true size of any lesion in this area is commonly underestimated. Its relatively weak medial wall and communication with the opposite ischiorectal fossa through the post-anal space provides sufficient space for lesions to expand without being symptomatic. Tumours involving the space can put pressure on the pudendal nerve which runs on the lower lateral wall of the space to cause referred pain.

The possible differential diagnoses for an ischiorectal swelling include an abscess, tail gut cyst, a tumour (sarcoma) or spread from rectal neoplasm.

An ischiorectal abscess is the most common pathology encountered in this region. Fluctuation is a late sign in the abscess presenting in this region. The radiological features in this patient were consistent with an abscess, however, the long duration of her symptoms made this less likely. Nevertheless, this could have been a delayed infection in a haematoma following vaginal delivery.

Tail gut cysts occur mainly in the retrorectal space, however, on rare occasions these can extend laterally from the presacral space to involve the ischiorectal fossa. Other uncommon swellings in this location include lipomas, liposarcomas or even an epidermoid cyst (Fujimoto et al, 1993; Llauger et al, 1998). All of these contain fat and would have appeared at the same signal intensity as that of the subcutaneous fat in all signal sequences on magnetic resonance imaging. Hence these were excluded.

The differential diagnosis of a primary tumour in the ischiorectal fossa which is consistent with such radiological findings could include angiomyxoma or a peripheral nerve sheath tumour arising from branches of pudendal nerve. A Trucut biopsy would help in identifying the subtype and grade of tumour in 80% of cases (Kissin et al, 1986).

Direct spread from rectal carcinoma is a well-known occurrence, which can present as ischiorectal sepsis such as an abscess (Avill, 1984) or fistula (Shinohara et al, 2001).

All the swellings mentioned above are very uncommon which explains why, despite a long-standing history, this

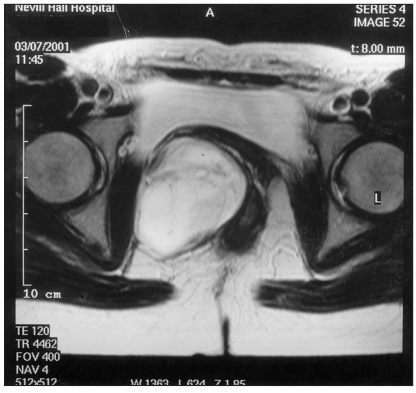
CASE REPORT

A 33-year-old woman presented with a painful lump in the right gluteal region, 7 months after normal vaginal delivery of her child. There was no history of altered bowel habits, bleeding per rectum, dyspareunia or difficult labour. On examination she was afebrile. Local examination revealed asymmetry with a tender lump in the right gluteal region and some redness of the overlying skin. Rectal examination revealed a tender right lateral rectal wall. Magnetic resonance imaging (*Figure 1*) revealed a 12x8x8 cm complex 'cyst' in the right ischiorectal fossa displacing the levator ani upward and the rectum medially, with an apparent pseudo capsule. In the T1-weighted sequences, it had higher signal intensity than the bladder. On T2-weighted sequences, the lesion was of very high intensity. Also in the surrounding fat were multiple linear foci of high intensity, representing oedema or increased vascularity. In view of the location, it was suggested that this could be an abscess.

Examination under anaesthesia revealed a boggy swelling in the ischiorectal fossa. An incision to drain this abscess revealed no pus, however, a soft fatty lump protruded from the incision. The incision was extended and a soft tissue mass 12 cm in diameter was enucleated. The cavity was packed and skin incision closed after 3 days. Histopathology revealed an angiomyxoid tumour, extending to the resection margins. Subsequently she was sent to a specialist centre. Computed tomography of the chest was normal. After discussion with the patient about the possibility of incontinence and stoma, re-excision was performed.

Histology suggested low grade, sarcomatous changes in a pre-existing myofibroblastic lesion, either angiomyxoma or angiomyofibroblastoma. Immunostaining was positive for desmin and weakly positive for actin and calponin. Staining for caldesmin and myogenin were negative. CD34 was present in few scattered spindle cells. No adjuvant therapy was given. She had one admission since then with a wound collection, which was drained.

Figure 1. Magnetic resonance imaging scan showing the tumour involving the right ischiorectal fossa.



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patient underwent surgery under the probable diagnosis of chronic abscess. Even when surgery revealed presence of a soft tissue mass, only a conservative excision was done. For a soft tissue sarcoma, a wide excision with clear margin is essential and the first chance is the best chance. The absence of tissue diagnosis, the inability to achieve wide resection margins because of the close proximity to the sphincter mechanisms and the nature of consent limited the procedure in this case.

Angiomyxoma is a rare, slow-growing tumour, predominantly occurring in women in the anogenital region. This tumour is focally infiltrative and it may recur many years after initial resection, but it has no metastatic potential. Angiomyofibroblastoma is a distinctive benign tumour that most commonly arises in the vulva and vagina (Avill, 1984).

The location, morphological and immunohistochemical studies of these two tumours suggest a common origin from a myofibroblastic progenitor cell (Nielsen et al, 1996; Shinohara et al, 2001). They are differentiated from each other on the basis of histomorphology. Malignant change in angiomyxomas has not been described. In angiomyofibroblastoma it is extremely rare: only one case has been reported which was a recurrence of a vulval tumour with no metastasis (Granter et al, 1997).

The efficacy of radiotherapy in low-grade tumours with relatively complete excision is doubtful. There have been varying reports about the hormonal receptor status of such tumours, namely oestrogens and progesterone (Nielsen et al, 1997; Bigotti et al, 1999). There have been anecdotal reports about a complete response to hormonal therapy in patients with angiomyxomas (Fetsch et al, 1996). However, because of their rare nature, there is still no well-defined protocol for adjuvant therapy in such tumours.

CONCLUSION

The clinical presentation of sarcomas can be very deceptive and can mimic a benign lesion. This case is a pertinent reminder that all swellings encountered in the ischiorectal fossa are not abscesses. HM

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