Aggressive angiomyxoma of the vulva

S Baruah, P Latthe, NR Bhatti, SS Ghataura

INTRODUCTION

This article describes an unusual myxoid and vascular-appearing vulval neoplasm termed aggressive angiomyxoma of the vulva in a 33-year-old woman. The histopathology and treatments are discussed.

DISCUSSION

There has been a small number of case series of aggressive angiomyxoma involving the vulva. All presented as painless slow-growing polypoid cyst-like masses in women between the ages of 21 and 38 years. Aggressive angiomyxoma is a locally infiltrative tumour which has a tendency to recur (30–50%). Recurrence is secondary to incomplete excision. There have been no reports of any metastasis.

It has a predilection for the pelvis and perineal region in females as well as males. It is more common on the right side of the vulva. The treatment is wide excision. It is quite difficult to assess surgically the extent of lesion because of imperceptible infiltration of adjacent soft tissue. Resection of tumour with wide tumour-free margins is the best means of preventing recurrence (Hilgers et al, 1986). Multiple organs are rarely involved and computed tomography (CT) or magnetic resonance imaging (MRI) is advised to help identify invasion (Smith et al, 1991). As this tumour has not been shown to cause death, treatment such as pelvic exenteration is not indicated. Long-term follow up is indicated because of the known risk of delayed recurrence. The role of hormonal manipulation remains to be determined (Fetsch et al, 1996).

This tumour has been seen to recur from between 9 months to many years. There is a report in the literature of recurrence after 144 months.

Clinically the differential diagnosis includes vulval abscess, Bartholin abscess, Gartner's duct, vaginal cyst and vaginal polyp (Hilgers et al, 1986).

These neoplasms form large gelatinous masses which infiltrate the soft tissues of the female pelvis and perineum. They have a deceptively bland histological appearance, with many small blood vessels set in an abundant myxoid stroma which contains scattered stellate and spindle-shaped cells (*Figure 1*). Delicate elongated bundles of smooth muscle cells are often present in the stroma, especially around vessels. Immunohistochemically, the spindle cells may stain for actin and desmin but are negative for S-100 protein, factor VIII-related antigen, carcinoembryonic antigen and keratin (Begin et al, 1985). Removal of these neoplasms is sometimes difficult because of their infiltrative nature, and they tend to recur, sometimes repeatedly.

The tumour should be distinguished from angiomyofibroblastoma (AMF), a benign, predominantly superficial tumour of the vulva, canal of Nuck or perineum. The latter has circumscribed borders, frequent plump stromal cells of epithelioid and spindle-celled types, preferentially arranged around the numerous, non-hyalinized blood vessels. There is minimal stromal mucin, and rarely extravasation of erythrocytes (Fletcher et al, 1992). Intralesional fat may be present – the lipomatous variant of AMF (Laskin et al, 1997). Tumour cells express vimentin, oestrogen

CASE REPORT

A 33-year-old nulliparous women presented with a 4-week history of a painful vaginal swelling. There had been no relevant medical or gynaecological history of note and she was using the combined oral contraceptive pill (Cilest, Janssen-Cilag, High Wycombe, Bucks) for contraception. On pelvic examination, she had a tender right labial swelling 3 x 3 x 2 cm in size. The surrounding area showed no evidence of inflammation.

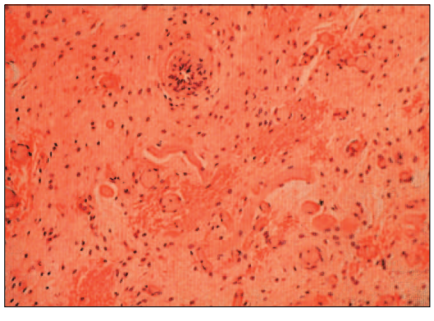
Systemic examination was unremarkable. The clinical diagnosis made was of a right Bartholin cyst. At examination under anaesthesia and excisional biopsy, the lesion appeared to be a cystic, fleshy-looking lesion located on the right labia majora, but was also seen to be extending towards the right ischiorectal fossa. The area was excised and multiple grey fragments (the largest being 110 x 45 x 20 mm) were submitted for pathological examination.

The lesion was well sampled and consisted of a mass of rather featureless spindled and stellate cells set in a myxoid and fibrillary background. It was highly vascular with considerable areas of red cell spillage into the stroma and some of the larger vessels showed medial hypertrophy. There was evidence of infiltration into adjacent muscle.

The other main differential diagnosis considered was angiomyofibroblastoma, but the perivascular condensation and zonality was not seen, nor was the plasma cell or epithelioid component. The tumour was positive for desmin (marker for intermediate cytoskeletal filaments), but negative for smooth muscle actin, S100 and epithelial membrane antigen. The appearance was in keeping with an aggressive angiomyxoma.

On confirmation of histology, the patient was sent to the gynaecological oncology team for further follow up, and magnetic resonance imaging was planned for identification of any invasion by tumour in case of recurrence.

Figure 1. Pauci-cellular myxoid stroma including scattered spindle cells infiltrating among striated muscle fibres: characteristics of an aggressive angiomyxoma.



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receptor protein, progesterone receptor protein, desmin, and occasionally smooth muscle actin and CD34. Transformation to angiomyofibrosarcoma has been described (Nielson, 1997). Aggressive angiomyxoma and AMF are probably related neoplasms in a spectrum of tumours derived from myofibroblasts or perivascular stem cells. The demonstration of immunoreactivity for desmin in aggressive angiomyxoma means that this antibody is not helpful in discriminating between these two tumours, and the principal means of distinction remains histomorphological analysis. HM

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IMAGES IN MEDICINE

Cystic duct stump stone

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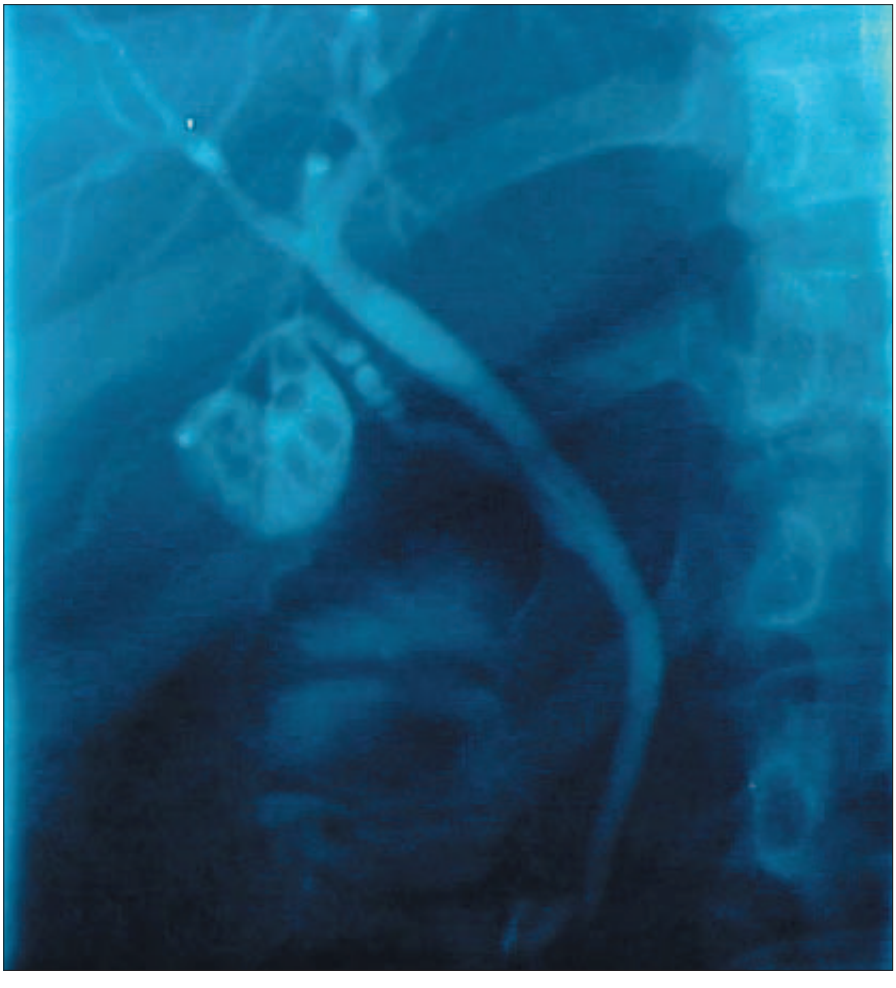

Figure 1. Endoscopic retrograde cholangiography showing a long and dilated cystic duct stump with multiple stones within it. The common bile duct is normal.

A 47-year-old woman was admitted to hospital with fever, jaundice, and upper abdominal pain. Laboratory tests revealed biochemical evidence of cholestasis together with leukocytosis suggesting acute cholangitis. She had had open cholecystectomy for acute cholecystitis 14 years ago, and had had no subsequent complaint until this presentation. Right upper quadrant abdominal sonography was unremarkable.

On endoscopic retrograde cholangiography the common bile duct was normal, but there was a long and dilated cystic duct stump with multiple stones within it (*Figure 1*). The patient underwent surgical resection of the stump, and then remained free of symptoms at 1 month follow up. This rare complication should be considered in patients presented with cholestasis or biliary colic following cholecystectomy. HM

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