INTRODUCTION

About 157 cases of aggressive angiomyxoma, a rare soft tissue tumour, have been reported. This was first described in 1983 by Steeper et al. Aggressive angiomyxoma is a locally invasive mesenchymal tumour, occurring predominantly in the pelvic-perineal region of adults and carries a high risk for local relapse and hence needs to be differentiated from other mesenchymal tumours occurring in this region. Except for positive surgical margins, there are no clinical or histological means for predicting the tumour recurrence. A diligent long-term follow-up is mandatory though rarely possible in our setting. This tumour needs to be considered in the differential diagnosis of vulval polyps. This case report describes this tumour involving the vulva of an adult lady.

CASE REPORT

A 44 years old lady presented with a slowly growing mass in the right labia majora over a duration of one year. The patient was apparently well one year back when she noticed a small pea size painless swelling on the right labium major. This swelling then progressively increased in size over one year. The mass measured about 24 cm x 15 cm in diameter on presentation and was attached to right labia by a thick stalk (Figure 1).

Ultrasound (USG) showed a large right perineal mass which was predominantly echogenic. Low resistance type of arterial blood flow was seen at places. It was not possible to characterize the lesion on USG. MRI was suggested for function characterization and elucidation. It showed a 24 x 14 x 8 cm sized soft tissue intensity mass originating from right ischiorectal fossa extending down into right labial region through a narrow neck and hanging in between thighs (Figure 2). A few necrotic areas were also seen. No lymph nodes were seen. Cervix, both adnexa, visualized bones, urinary bladder and rectum appeared normal.

Tumour was completely resected, including 2 cm of surrounding normal skin margins. Dissection was done upto the right ischeorectal fossa to remove the base of the pedicle. Patient's recovery was smooth and uneventful.

Gross examination of the lesion showed a well circumscribed pedunculated skin covered mass measuring 25 cm in diameter with a thick, wide stalk measuring 6 x 5 cm and having a length of 5 cm. The overlying skin was normal looking and there was no ulcer formation. Cross-section of mass showed solid whitish homogeneous appearance.

Microscopically, the tumour composed of numerous small and medium sized blood vessels. These were separated by abundant myxoid stroma with stellate cells. There was no atypia. No mitosis was seen. Immunohistochemical markers showed actin positivity in blood vessel walls. The stroma was positive for vimentin and negative for cytokeratin.

The patient followed-up for the last 3 years has not shown any evidence of recurrence.

DISCUSSION

Aggressive angiomyxoma was described by Steeper and Rosai in 1983. It mostly occurs in women in...
reproductive age. The peak incidence is during the third decade of life suggesting that estrogen may stimulate its growth. It generally involves the genital, perineal and pelvic region, with vulvar region being the most common site of involvement. Tumours occurring during pregnancy show a rapid growth as there is a state of high estrogen and progesterone production during this period. This patient also belonged to the fertile age group, who presented with a pedunculated mass in the right labium major.

Aggressive angiomyxomas (AAs) are very rare tumours and frequently clinically mistaken for cysts of Bartholin duct or for abscesses, vaginal cysts, lipomas, vulvar masses or abscesses, vaginal cysts or hernias. Differential diagnosis is important because of its tendency for recurrence. The size of the tumour is often significantly underestimated by physical examinations. On CT scan, these tumours have a well defined margin with attenuation less than that of muscle. On MRI, these tumours show high signal intensity. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high water content of angiomyxoma.

Grossly, these tumours are soft, partly circumscribed, polypoidal lesions with gelatinous appearance on cut section. Microscopically these lesions are composed of many thick walled vessels of varying sizes in loose myxoid and collagenous stroma with spindle and stellate shaped neoplastic cells. The tumour cells express vimentin, desmin and SMA (Smooth Muscle Actin) and are negative for S-100 immuno-histochemically. These tumours express estrogen and progesterone receptors. Suggesting that they may be hormone dependent.

These tumours have to be differentiated from angiofibroma fibroblastoma. These are small well circumscribed tumours composed of plump epithelial cells arranged in perivascular distribution and are locally invasive.

The treatment of choice is wide local excision. The excision of these tumours is difficult as they have the same consistency as that of normal connective tissue and, therefore, have propensity for local recurrence (36 – 70%). Recurrence could occur after 10 – 15 years of primary excision. GnRH agonist therapy may help complete resection of large tumours because of shrinkage and provide avoidance to radical surgery when administered pre-operatively. It can be an alternative, if patient is reluctant to undergo radical surgery or when the surgery is not feasible. Postoperative administration may be useful for the treatment of residual tumours or recurrences. On the other hand, GnRH agonists also have some disadvantages such as menopausal symptoms and bone loss especially associated with long-term therapy. Resistance to GnRH agonist therapy remains unclear and further concern is that withdrawal of medication may also result in re-growth of the tumour.

In this case, the lesion being pedunculated, the excision was complete and postoperative period was uneventful. There has been no evidence of any local recurrence. Hence, long-term follow-up is necessary. Magnetic resonance imaging is the preferred method for detecting recurrences.

REFERENCES