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Original Research Paper

General Surgery

HILLOCK ON THE BACK- A RARE CASE OF ANGIOMYXOMA OF THE BACK

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ABSTRACT Angiomyxoma is a locally invasive rare neoplasm having mesenchymal origin. The reported cases mostly occur in females originate in the pelvis. We present a case of a 50 year old female, from Udupi, who presented with c/o swelling over the upper back, in the interscapular area since 8 years. The swelling gradually progressed in size & it was not associated with pain. Intraoperatively swelling was deep to the deep fascia, and trapezius muscle and intramuscular (rhomboideus major). On microscopy there were features of Angiomyxoma. Due to the rare site of presentation we have decided to report this case

KEYWORDS :

BACKGROUND

In 1983 Steeper and Rosai first coined the term 'aggressive angiomyxoma' which was later changed to 'deep angiomyxoma' by WHO.[1] A locally aggressive neoplasm of mesenchymal origin occurring mainly in elderly adult, Angiomyxoma commonly involves the vulva in women; spermatic cord and inguinal region in men.[2] Only a small number of cases have been reported in literature. We encountered a case of a large deep angiomyxoma of the back which was diagnosed with histopathology.

CASE PRESENTATION

A 50 year old female, from Udupi, a homemaker, Presented with complaints of swelling over the upper back since 8 years, swelling gradually progressed in size. It was not associated with pain and there was no history of trauma to the back

INVESTIGATIONS

On inspection, a 10x 6 cm swelling was noted in the upper back in the interscapular region, it was ovoid in shape. The skin over the swelling was normal with no visible scars, sinuses or diated veins (fig 1). On palpation, there was no local rise in temperature or tenderness. 10x 6 Cm swelling with smooth surface, firm consistency and no significant mobility was palpated in the upper back over the interscapular region. On putting the Trapezius muscle into contraction, swelling became less prominent indicating that it was deep to the Trapezius(fig 2).A clinical diagnosis of intramuscular lipoma was made.

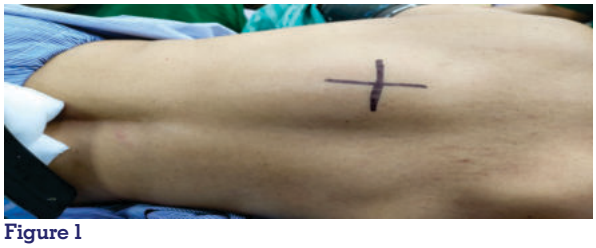

Figure 1

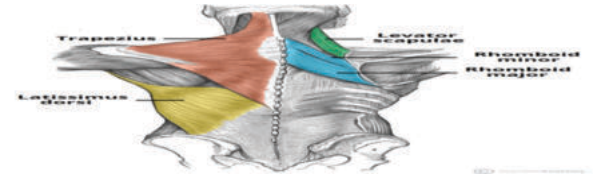

Figure 2

On MRI of upper back, evidence of a well encapsulated T1 hypointense, T2/STIR hyperintense cystic lesion measuring 11x7x3.6cm was seen in the left posterior paraspinal muscle within the erector spina and deep to the subcutaneous plane showing multiple internal septations, likely epidermoid / ? sebaceous cyst. (non neoplastic) (fig 3). Histopathology correlation was suggested.Ultrasound guided FNAC was suggestive of lipomatous lesion and a wide local excision was planned

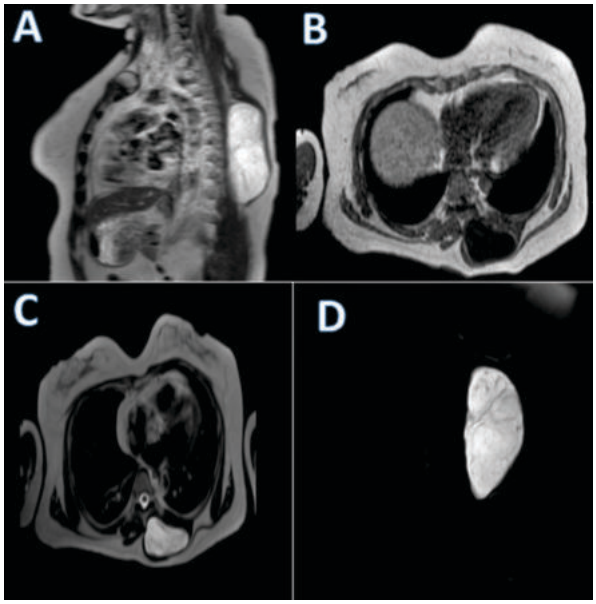

Figure 3: Showing T1 hypointense(A),T2 hyperintense(B) and (C) ; internal spetations on T2 W SPAIR, lesion over the left posterior paraspinal muscle with mild contrast enhancement

TREATMENT

Excision of the lesion was done under general anaesthesia. Intraoperatively a 10 x 6 x 3 cm cystic lesion, reiniform shaped, nodular swelling deep to the deep fascia, trapezius muscle and intramuscular (rhomboides major) plane was excised and specimen was sent for histopathological evaluation.Post operative period was uneventful.A drain placed in situ was removed on 3rd day and sutures were removed on day

12.Patient was discharged and came for follow up after one week.Recovery period was uneventful

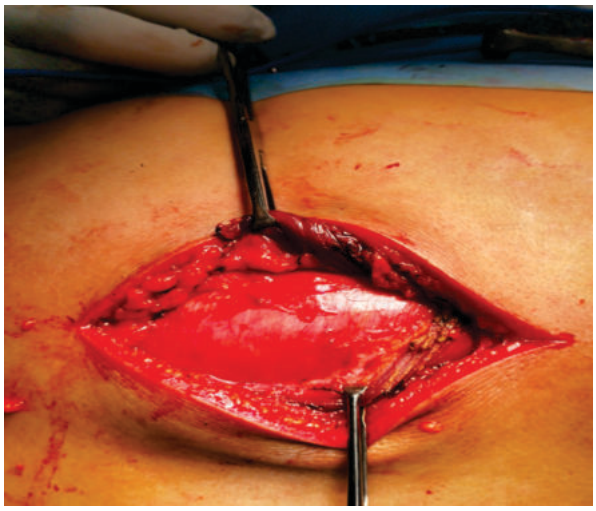

Figure 4

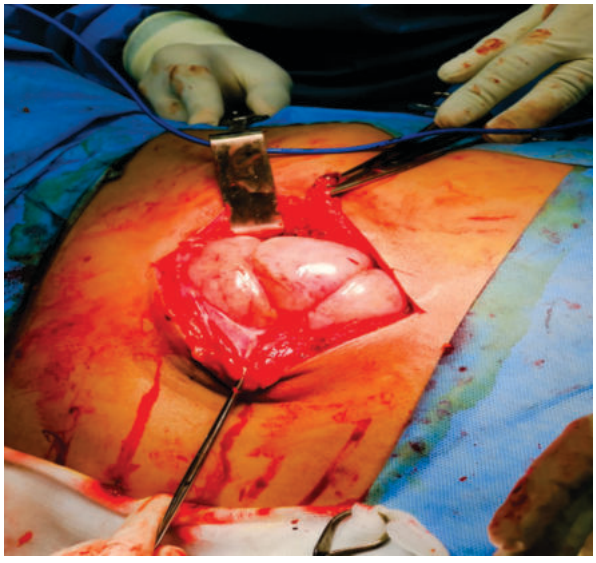

Figure 5

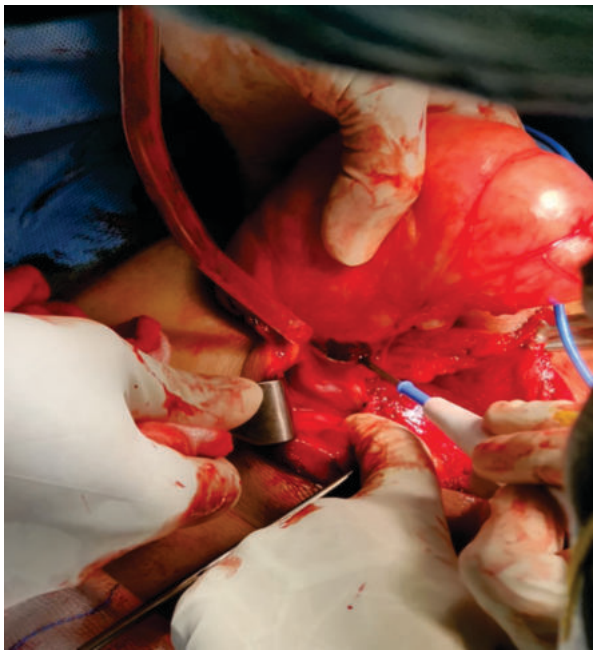

Figure 6

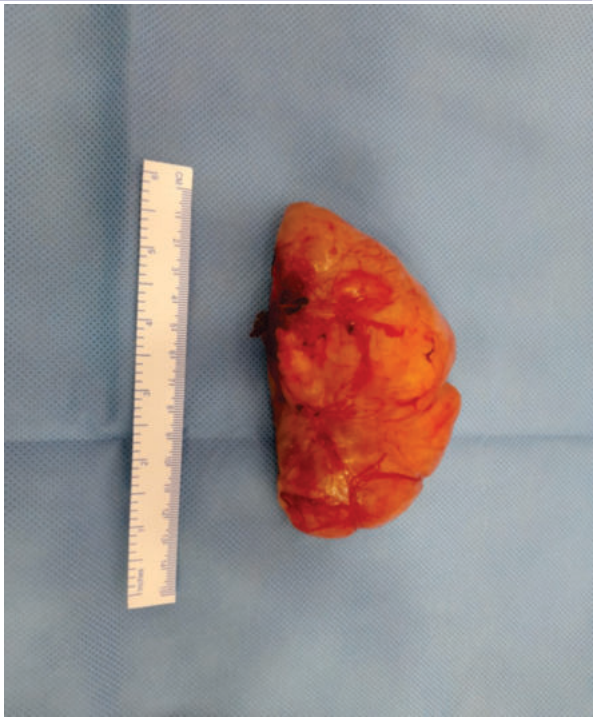

Figure 7

HISTOPATHOLOGY

Grossly: Sections studied showed a poorly circumscribed tumor extending into surrounding fat. Microscopically: the Tumor was composed of proliferating spindle cells deposited in a myxoid stroma. The cells had indistinct cell border, oval nuclei and inconspicuous nucleoli. Numerous blood vessels were seen(fig 8) .A diagnosis of Angiomyxoma was made

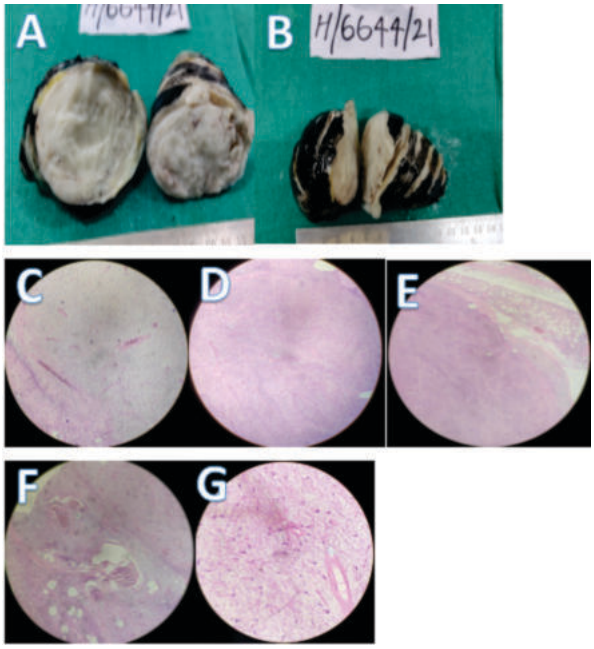

Figure 8: (A) Shows gross appearance of well circumscribed mass of 11x6.5x3.5 cms with smooth surface.Cut surface (B) wash homogenous,glistening and smooth. Scanner View (C) revealed hypocellular lesion with myxoid background (D). Interspersed in these cells were variable-sized thick-walled and thin-walled vessels (E) and stroma entrapping surrounding fat(F) with collagen fibres(G)

DISCUSSION

Angiomyxoma is a rare soft tissue tumor that has a female

predominance (6:1) ; most commonly in the third to fifth decade of life.[2] The tumour occurs mostly in the perineal region. Relative incidence in scrotum, spermatic cord, perineum and pelvis are 38%, 33%, 13% and 8%, respectively.[3] About 200 cases of angiomyxoma reported in the literature till date.They usually grow quite slowly and they don't usually spread to other parts of the body.[4] They can be superficial or deep (aggressive). Superficial type is generally seen on the skin or just below the surface of the skin. It can affect anywhere on the trunk or lower part of the body, the genitals , and head and neck.

The name 'aggressive angiomyxoma' is due to the high local recurrence and the histological findings of myxoid stroma, rich in collagen fibre with stellate-shaped cells. There are numerous thick-walled blood vessels with perivascular hypercellularity with focal areas showing lymphoid aggregates.

Immunohistochemically tumour cells are positive for CD34, desmin, estrogen, progesterone and SMA . A few have been reported to have t(12:21) and HMGA2 rearrangement.[4] Estrogen and progesterone positivity points towards hormonal influence on the tumour.[5] However, it occurs in both sexes and in all age groups

Wide local excision of these tumours is mainstay of treatment.There is a chance of recurrence in 30%–70% of cases and can occur as early as within 1 month.[5] This is usually due to its local infiltration and the difficulty in excising the tumour completely.Hormonal therapy has advocated due to hormonal sensitivity in cases of recurrences.Occasional case of metastasis has been reported.[6]

Deep angiomyxoma is a rare benign neoplasm that can mimic many clinical and histological entities. It's aggressive but indolent course leads to late diagnosis. Surgery is the only treatment available but with a high risk of recurrence, thus requiring close follow-up.

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