SUPERFICIAL ANGIOMYXOMA OF AXILLA: A RARE ENTITY

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ABSTRACT

Superficial angiomyxoma, a benign mesenchymal tumour, arises from dermis and subcutaneous tissue. It is characterised by prominent thin walled blood vessels in myxoid matrix. A case of superficial angiomyxoma of axilla is reported to highlight the features of this rare entity, which is still a poorly recognized tumour, despite its first description by Carney and colleaguesin 1985.The clinical presentation, radiological features, intra-operative findings,pathological analysis and literature review are presented.

Key words: Superficial angiomyxoma, cutaneous myxoma

INTRODUCTION:

Superficial angiomyxoma, also known as cutaneous myxoma is a rare myxoid tumour of the skin and sub cutis. This benign tumour usually presents in middle aged adults as single nodule or polypoid lesion that may be clinically confused with skin tag, neurofibroma or other myxoid tumours.

Distinctive histologicfeatures included extensive myxoid stromawith stellate or bipolar fibroblastic cells, smallblood vessels, and mixed inflammatory cellinfiltrate with notable neutrophils.Cutaneous myxoma should be distinguishedfrom the other cutaneous myxoid lesions withwhich it may be confused because it has apropensity for local recurrence.

CASE REPORT

A 21 year old male presented with a painless, slow growing mass in right axillary region since 1 year. He denied any history of trauma, tuberculosis, movement restriction, tingling or numbness in the arm or cardio-vascular symptoms.

Physical examination revealed a soft, cystic, multi-lobulated, mobile, pedunculated mass (figure 1, 2), which was brilliantly transilluminant (figure 3). The overlying skin was unremarkable. No other similar lesions were seen elsewhere. The clinical diagnosis ofLympho-vascular swelling was made.

Local radiograph was unremarkable. Ultrasonography revealed, well defined, heterogeneouslyechogenic, predominantly hyper-echoic lesion with moderate vascularity in subcutaneous plane. Routine pre- operative investigations were in normal limits. Wide local excision was performed and at surgery the tumour was found to be centred in subcutaneous tissue.

Macroscopically, the resected lesional tissue received was a single cystic greyish brown mass which was skin covered measuring 7 X 6 X 3.5cms. (Figure 4, 5)

Cut surface of the lesion was grey to white with glistening surface. Microscopic examination revealed a circumscribed myxoid lesion. Cells are benign looking, stellate in shape embedded in the vascularized myxoid stroma. (Figure 6)

Post-operative recovery and 6 months follow up was uneventful.

DISCUSSION

Superficial angiomyxoma or cutaneousmyxoma is benign soft tissue tumour which was first described by Carney and later morefully characterized by Allen[2] et al in 1988 as well as Calonje[3] and colleagues in 1999.

Cutaneous myxoma arises more commonly inmales, predominantly middle aged adults witha peak incidence between 20 and 40 years ofage. Rare congenital examples have beendescribed. These lesions can ariseessentially anywhere in the superficialtissuebut there is a predilection for the trunk, lowerextremities, and head and neck.

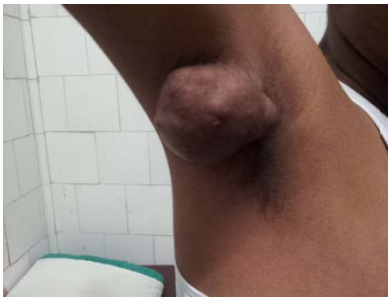
Some arise in the genital region of both males and females (vulva, mons pubis, scrotum, inguinal region).

When this lesion occurs in the setting of Carney's complex (Cutaneous myxomas, cardiac myxomas, spotty pigmentation and endocrine over activity), they are often multiple and frequently occur in the eyelid or external ear.

The clinical history is typically that of a longstanding painless nodule or mass. Most ofthem are solitary lesions varying in size from1-5cms. Grossly, cutaneous myxomas areusually well circumscribed tumours. They havea grey white, glistening, gelatinous cut surface. Thin fibrous septa traverse theneoplasm, resulting in a

vaguely multinodular tumour. Cysts that are sometimes filled withkeratinous debris may be identified grossly.

Histological, this lesion has a lobular or multinodular appearance at low magnification. Most are histological poorly circumscribedwith extension into the underlying subcutaneous tissue and rarely skeletalmuscle.



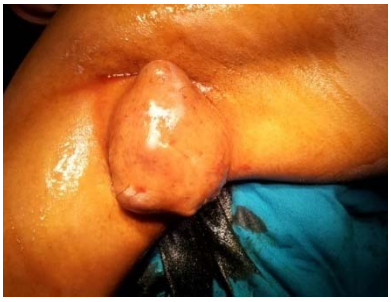


Figure 1 and 2: Clinical appearance of the tumour

Figure 3: Transillumination test

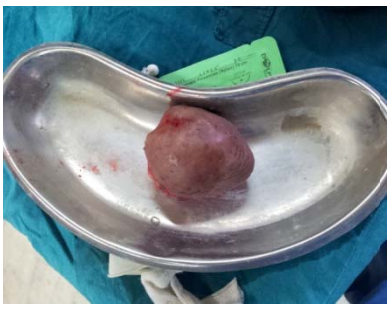


Figure 4: Excised tumour

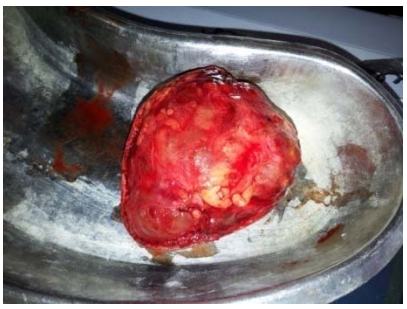


Figure 5: Macroscopic appearance

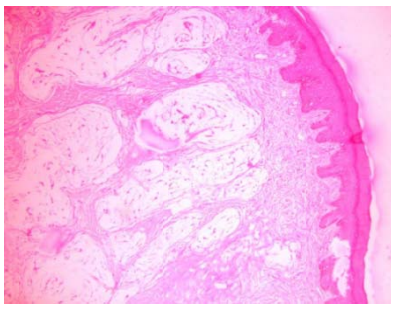


Figure 6: Microscopic appearance

Table 1: Histological features

Spindled to stellate shaped cells	**Inflammatory infiltrate,particularly stromal neutrophils**
Extensive myxoid stroma	Cysts/irregular clefts sensitive to hyaluronidase digestion
Mitotic figures rare	Indistinct cell borders
Binucleated and multinucleated cells	Nuclei with inconspicuous nucleoli
Small vascular channels in myxoid stroma	Epithelial structures due to adnexal entrapment in stroma

Table 2: Differential diagnosis of superficial angiomyxoma

Aggressive angiomyxoma	**Larger, deeper structures involved, genital, perinea, pelvic region, larger caliber vessels, local recurrence**
Angiomyofibroblastoma	Small, non-infiltrative, highly cellular, lack large caliber vessels, RBC extravasation
Myxofibrosarcoma	Multinucleated atypical cells, hyperchromasia, curvilinear vessels
Myxoid liposarcoma	Deeper, larger tumors, Plexiform (chicken wire) vasculature
Myxoid neurofibroma	Cellular, S100 protein +
Cellular angiofibroma	Small, <3cm, hyalinised blood vessels
Focal cutaneous mucinosis	Lacks lobular architecture, epithelial elements, neutrophil Infiltrate
Superficial acral fibromyxoma	Hands and toes

Immuno-histochemically, the tumour cells express vimentin and CD34, but rarely stain for cytokeratin or S-100 protein. Some cells stain for smooth muscle actin, desmin, possibly indicating focal myofibroblastic differentiation.

The differential diagnosis of cutaneous myxomas is extensive and includes benign and low grade malignant myxoid lesions including aggressive angiomyxoma, focalcutaneous mucinosis,myxoid neurothekeomas (dermal nerve sheath myxoma), myxoid neurofibroma, superficial acralfibromyxomaand myxoid liposarcoma.

CONCLUSION

Superficial angiomyxoma is a rare recognized clinicopathologic entity with recurrence potential.

Hence it should be distinguished from other myxoid lesions of the skin. Appropriate diagnosis, complete surgical removal and close follow up of the patientarerecommended.

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