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 colleagues in 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 subcutis. 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 histologic features included extensive myxoid stroma with stellate or bipolar fibroblastic cells, small blood vessels, and mixed inflammatory cell infiltrate with notable neutrophils. Cutaneous myxoma should be distinguished from the other cutaneous myxoid lesions with which it may be confused because it has a propensity 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 cardiovascular 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 of lympho-vascular swelling was made.

Local radiograph was unremarkable. Ultrasonography revealed, well defined, heterogeneously echohogenic, 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 cutaneous myxoma is benign soft tissue tumour which was first described by Carney and later more fully characterized by Allen et al in 1988 as well as Calonje and colleagues in 1999.

Cutaneous myxoma arises more commonly in males, predominantly middle aged adults with peak incidence between 20 and 40 years of age. Rare congenital examples have been described. These lesions can arise essentially anywhere in the superficial tissue but there is a predilection for the trunk, lower extremities, 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 often are solitary lesions varying in size from 1-5cms. Grossly, cutaneous myxomas are usually well circumscribed tumours. They have a grey white, glistening, gelatinous cut surface. Thin fibrous septa traverse the neoplasm, resulting in a
vaguely multinodular tumour. Cysts that are sometimes filled with keratinous debris may be identified grossly. Histological, this lesion has a lobular or multinodular appearance at low magnification. Most are histological

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

<table>
<thead>
<tr>
<th>Spindled to stellate shaped cells</th>
<th>Inflammatory infiltrate, particularly stromal neutrophils</th>
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<tbody>
<tr>
<td>Extensive myxoid stroma</td>
<td>Cysts/irregular clefts sensitive to hyaluronidase digestion</td>
</tr>
<tr>
<td>Mitotic figures rare</td>
<td>Indistinct cell borders</td>
</tr>
<tr>
<td>Binucleated and multinucleated cells</td>
<td>Nuclei with inconspicuous nucleoli</td>
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<tr>
<td>Small vascular channels in myxoid stroma</td>
<td>Epithelial structures due to adnexal entrapment in stroma</td>
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Table 2: Differential diagnosis of superficial angiomyxoma

<table>
<thead>
<tr>
<th>Aggressive angiomyxoma</th>
<th>Larger, deeper structures involved, genital, perinea, pelvic region, larger caliber vessels, local recurrence</th>
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<tbody>
<tr>
<td>Angiomyofibroblastoma</td>
<td>Small, non-infiltrative, highly cellular, lack large caliber vessels, RBC extravasation</td>
</tr>
<tr>
<td>Myxofibrosarcoma</td>
<td>Multinucleated atypical cells, hyperchromasia, curvilinear vessels</td>
</tr>
<tr>
<td>Myxoid liposarcoma</td>
<td>Deeper, larger tumors, Flexiform (chicken wire) vasculature</td>
</tr>
<tr>
<td>Myxoid neurofibroma</td>
<td>Cellular, S100 protein +</td>
</tr>
<tr>
<td>Cellular angiofibroma</td>
<td>Small, &lt;3cm, hyalinised blood vessels</td>
</tr>
<tr>
<td>Focal cutaneous mucinosis</td>
<td>Lacks lobular architecture, epithelial elements, neutrophil Infiltrate</td>
</tr>
<tr>
<td>Superficial acral fibromyxoma</td>
<td>Hands and toes</td>
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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 acralfibromyxoma and 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 patient are recommended.

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