**SUPERFICIAL ANGIOMYXOMA WITH EPITHELIAL COMPONENT – A RARE CUTANEOUS MYXOID TUMOR**

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**ABSTRACT:**
Superficial angiomyxoma is a benign soft tissue tumor that has a predilection for the head and neck, trunk and lower extremities. We report a case of 30 year old female who presented with a history of nodular swelling on the medial side of the right toe. Pathological analysis confirmed the lesion to be superficial angiomyxoma. We use this case to highlight the features of this rare entity, which is still a poorly recognized cutaneous tumor, despite its description since 1988.

**KEY WORDS:**
Superficial angiomyxoma, cutaneous myxoma.

**1. INTRODUCTION:**
Superficial angiomyxoma, also known as cutaneous myxoma is a rare myxoid tumor of the skin and subcutis, which was first described by Carney and colleagues in 1986.[1] This benign tumor usually presents in middle aged adults as single nodule or polypoid lesion that may be clinically confused with skin tag or neurofibroma.[2] When this lesion occurs in the setting of Carney’s complex (Cutaneous and cardiac myxomas, spotty pigmentation and endocrine overactivity), they are often multiple and frequently occur in the eyelid or external ear.[3] Histologically this tumor has a lobular low power appearance with a circumscribed border that involves the dermis and sometimes cutis. Distinctive histologic features included extensive myxoid stroma with stellate or bipolar fibroblastic cells, small blood vessels, and mixed inflammatory cell infiltrate with notable neutrophils.[4] 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.

**2. CASE REPORT:**
A 30 year old female presented with a painless soft tissue mass on the medial aspect of the right toe, which had been slowly growing for 2 years. She denied any trauma at that site. Physical examination revealed a firm immobile, ovoid nodule attached on the medial aspect of the right toe. The overlying skin was unremarkable. No further lesions were seen. The clinical diagnosis of neurofibroma was made.

Local resection was performed and at surgery the tumor was centred in subcutaneous tissue. Macroscopically, the resected lesional tissue received was a single firm nodule which was skin covered measuring 2.5 X 2.5 X 1.5cms. Cut surface of the lesion was gray to white with glistening surface (Figure-1).

Microscopic examination revealed a circumscribed myxoid lesion within the dermis. Lesion has a lobular appearance at low magnification. The lobules are separated by fibrous septa (Figure-2). The lesional cells are small, cytologically bland and spindled to stellate in shape embedded in the vascularized myxoid stroma (Figure-3). In some foci, the lesion was seen extending into the epidermis (Figure-4). Entrapped adnexal glands (Figure-5) and prominent neutrophilic infiltrate were also seen. Immunohistochemistry showed focal positivity with CD34 (Figure-3). The histological and immunohistochemical features were consistent with superficial angiomyxoma.
3. DISCUSSION:
Superficial angiomyxoma (Cutaneous myxoma) is benign soft tissue tumor which was first described by Carney and later more fully characterized by Allen et al in 1988 [2] and Calonje and colleagues in 1999 [4].

Cutaneous myxoma arises more commonly in males, predominantly middle aged adults with a peak incidence between 20 and 40 years of age. [5] Rare congenital examples have been described. [6] 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). [7]

Cutaneous myxomas associated with Carney’s syndrome are often multiple and involves the eyelids and ear. [8]

The clinical history is typically that of a long standing painless nodule or mass. Most of them are solitary lesions varying in size from 1-5cms. Grossly, cutaneous myxomas are usually well circumscribed tumors. They have a gray white, glistening, gelatinous cut surface. Thin fibrous septa traverse the neoplasm, resulting in a vaguely multinodular tumor. Cysts that are sometimes filled with keratinous debris may be identified grossly. [9] Histologically, this lesion has a lobular or multinodular appearance at low magnification. Most are histologically poorly circumscribed with extension into the underlying subcutaneous tissue and rarely skeletal muscle. A sparse proliferation of spindled to stellate shaped cells are deposited in an extensive myxoid stroma, sometimes forming cysts or irregular clefts, that is sensitive to hyaluronidase digestion. The cells have indistinct cell borders and nuclei with inconspicuous nucleoli. Mitotic figures are rare. Binucleated and multinucleated cells may be seen. There are often small vascular channels seen in myxoid stroma. A mixed inflammatory infiltrate is common, particularly stromal neutrophils, a feature unique to this tumor when compared to other cutaneous myxoid lesions. Upto one quarter of these tumors have epithelial structures consisting of basoloid buds, epithelial strands or epidermoid cyst, possibly as a result of entrapment of adnexal structures by the neoplasm.

Immunohistochemically, the tumor 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. [10]

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

Aggressive angiomyxoma usually tends to be larger, involves deeper structures, usually in the female pelvic region and has a vascular pattern that differs from that of cutaneous myxoma. Focal cutaneous mucinosis lacks lobular architecture, stromal neutrophils, and epithelial structures found in cutaneous myxoma. Superficial acral fibromyxoma arises almost exclusively on the fingers and toes of middle aged adults and lacks neutrophilic infiltrate. [11] Myxoid neurothekeoma has a more pronounced lobular growth pattern and is characterized by more plumper cells that are usually positive for S-100 protein. Myxoid neurofibroma is composed of cells with wavy or buckled nuclei that are also S-100 positive.
**Figure 1** - Skin covered nodular lesion with cut section showing gray-white glistening surface.

**Figure 2** - Section showing lobular arrangement of tumor cells separated by fibrous septa (H&E, X50).
Figure 3- Section showing small, cytologically bland and spindled to stellate shaped cells embedded in the vascularized myxoid stroma (H&E, X100). Left upper: spindle shaped and stellate cells (H&E,X400). Left lower: Immunohistochemistry showing focal positivity with CD34 (CD34,X400)

Figure 4- Section showing the lesion extending into the epidermis (H&E,X50)
Myxoid liposarcomas are usually more deeply located and larger than cutaneous myxomas and is characterized by a “chicken-wire” plexiform vasculature with scattered lipoblasts. Myxofibrosarcoma has a greater degree of nuclear atypia and hyperchromasia as well as curvilinear vessels often lined by hyperchromatic tumor cells.

4. CONCLUSION:
Superficial angiomyxoma is a rare 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 is recommended.

5. REFERENCES:

