



SOLITARY CUTANEOUS MYXOMA (SUPERFICIAL ANGIOMYXOMA) - A PRECURSOR TO CARNEY'S COMPLEX.

Pathology

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KEYWORDS

Cutaneous myxomas also called as superficial angiomyxomas are rare benign connective tissue tumours, composed of stellate cells to spindle cells, set in a loose mucoid stroma. Cutaneous myxomas are the earliest presentations in Carney's complex and may herald potentially fatal cardiac myxomas which is responsible for serious embolic complications. Cutaneous Angiomyxomas limited to skin and subcutaneous tissue is discussed here for its rarity.

35 year old male presented with slowly progressive swelling on scalp since 3 yrs. It was pedunculated, soft in consistency, measuring 3cm diameter in long axis and was nontender [Fig 1]. Patient was then send for FNAC to ur department. Mild blood tinged gelatinous material was aspirated from the swelling [Fig 2]. Cytosmear showed ample amount of pale and deeply stained myxoid material in which scattered population of short spindle cells with bland looking nuclei was seen. Final cytodagnosis was suggestive of solitary cutaneous myxoma Fig 3 (a)(b)(c). Patient was advised for biopsy. Routine hematological investigations of the patient were within normal limits. There was no hyperpigmented spotty lesions in the entire body. Chest X ray and 2D Echo cardiogram were also within normal limits thus ruling out possibility of Carneys complex.

Excisional biopsy was performed and sample was sent to our department for histopathological study. Grossly it was a skin lined globular mass measuring 3cm in diameter, cut section was gelatinous [fig 4]. Histopathology showed normal looking epidermis with hypocellular dermis containing spindle to stellate cells set in a mucinous matrix background. Few delicate small blood vessels and aggregates of inflammatory cells were seen frequently [Fig 5 (a)(b)(c)]. Based on clinical, cytological and histopathological features a diagnosis of solitary Cutaneous Myxoma was rendered.

In 1957, Herbert Z. Lund classified myxomas of the skin into cutaneous myxoid cysts of the fingers, mucocoeles of the lip, and myxomatous reactions associated with epithelial elements [1]. Superficial angiomyxoma was first described as a cutaneous myxoma of Carney's complex by Carney et al. in 1986 [2]. Carney's complex is an autosomal dominant syndrome characterized by myxomas of the heart, skin, and breast; spotty pigmentation of the mucous membrane; and endocrine overactivity such as Cushing's syndrome and acromegaly [2]. The syndrome is transmitted as an autosomal dominant trait and is genetically heterogeneous, linkage to two loci at chromosomes 2p16 and 17 q22 was reported[3]. In addition to the cutaneous and mammary locations, myxomas in the Carney complex have also occurred in the abdomen and pelvis[4]

Most cases occur in adults as an asymptomatic solitary papule or nodule with equal sex incidence. Lesions are usually less than 3 cm and have a wide anatomical distribution with a predilection for the trunk, head and neck and genital skin [5]. Cutaneous myxomas are noted in 81% of cases prior to the diagnosis of cardiac myxoma. To the best of our knowledge, only three cases of coexistent cutaneous myxoma and cardiac myxoma are reported in the Indian literature[6,7].

Microscopically, cutaneous myxomas in the Carney complex have sharp circumscription, hypocellularity, abundant myxoid stroma, prominent capillaries which was similar to our findings that helped us in arriving at correct diagnosis. Our patient did not have Carney's complex as his cutaneous tumor was not associated with non-cutaneous myxomas, pigmentation of the skin or mucous membrane, or endocrine disorders. It is important to correctly identify these cutaneous myxomas because the tumor may be a harbinger of cardiac myxoma, which is observed in one-fifth of patients with the Carney complex and has also been responsible for serious embolic complications in equal number[8,9]. So a vigilant eye and adequate follow up is essential to overcome the complications associated with this entity. Our patient is on follow up since past 6 months and is apparently asymptomatic till date.



Fig 1- Soft pedunculated lesion on scalp.

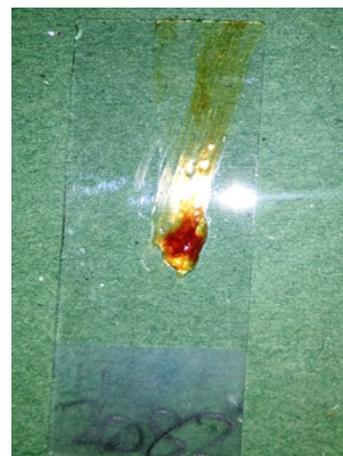


Fig 2- Blood tinged gelatinous aspirate.

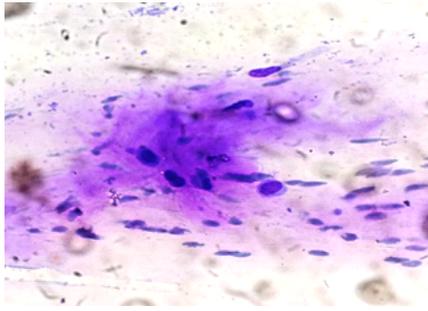


Fig 3 (a) Cytosmears showing bland looking spindle cells in myxoid background.

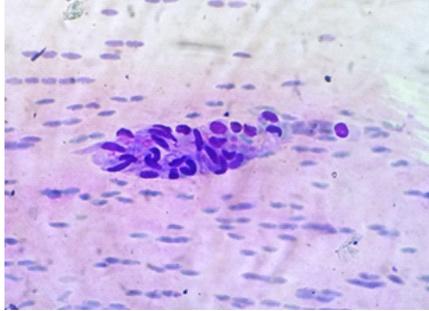


Fig 3 (b) Cytosmears showing bland looking spindle cells in myxoid background.

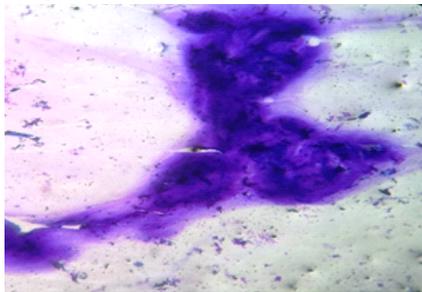


Fig 3 © Cytosmears showing bland looking spindle cells in myxoid background.

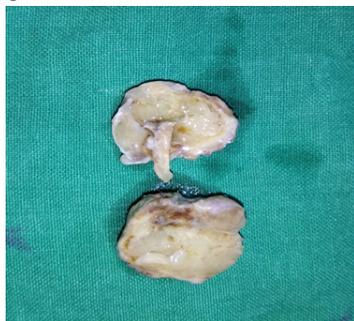


Fig 4 - Gross pic-Showing gelatinous material on cut section.

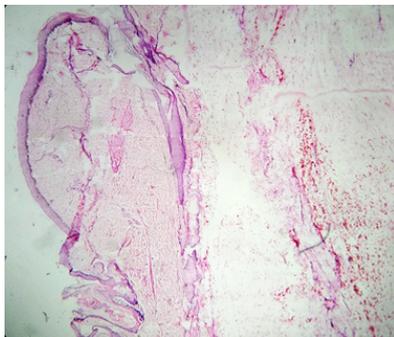


Fig 5(a)-40x showing Normal epidermis with underlying hypocellular dermis.

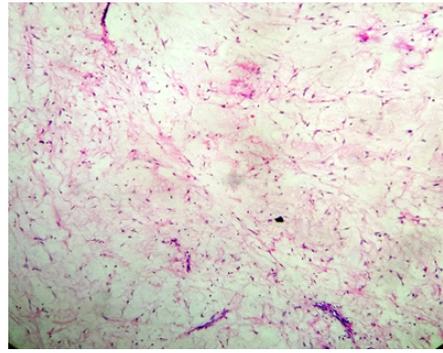


Fig 5 (b)-100x- Hypocellular dermis showing spindle cells.

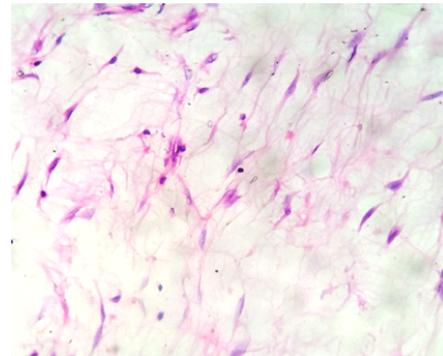


Fig 5 (c)-400X showing spindle to stellate cells in myxoid background.

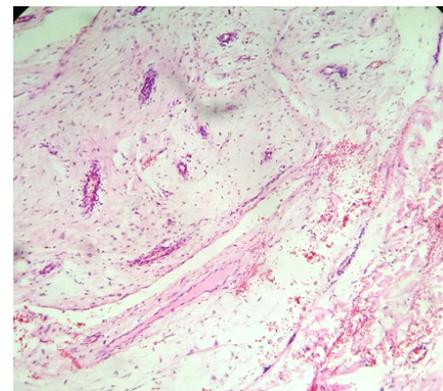


Fig 5 (d)- Thin walled delicate vasculature seen admixed with spindle cells in the dermis.

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