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AN UNUSUAL CASE OF *SUPERFICIAL (CUTANEOUS) ANGIOMYXOMAS*

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Abstract
Cutaneous myxomas also called angiomyxomas are rare benign connective tissue tumours, composed of stellate cells, set in a loose mucoid stroma. These lesions have been recognized as part of Carney complex.
We report a 12 year old boy affected by multiple Superficial Angiomyxomas without any other components of Carney complex.

Key words: Superficial Angiomyxomas; Carney complex; Myxoid stroma

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Introduction

In 1988, Allen et al, proposed the disease entity 'superficial angiomyxoma which is a dermal or subcutaneous tumour composed of a mixture of small blood vessels and sparse spindle-shaped cells in a prominent myxoid stroma. Cutaneous myxomas have been well recognized in recent years, especially as part of the autosomal dominant complex of endocrine hyperactivity now known as Carney's complex.
We herein report a case of Cutaneous Angiomyxomas limited to skin and subcutaneous tissue for its rarity.

Case Report

A 12 year old boy presented with asymptomatic multiple swellings of 15 days duration (Fig. 1a - c). The swellings appeared over both the hands and spread to forearms, forehead, chin, nose, and lower back. They were painless, non tender, non reducible, non pulsatile and soft to firm in consistency measuring about 1x1 to 2x2 cm in size. Skin over the swellings appeared normal.
A differential diagnosis of Cysticercosis cellulose cutis, Cutaneous myxomas, and Lipomas was considered.
Physician and Cardiologist were consulted.
Routine blood, urine and stool examination was within normal limits, except raised triglycerides. Serological test for rheumatoid factor, ASLO titer was negative. ECG and ECHO were normal.
Radiological examination of hands, skull and chest was normal. On USG, nodules were found in the subcutaneous plane, hypoechoic with no extension in to the muscular plane. No evidence of Cysticercosis cellulose cutis found.
Fine Needle Aspiration Cytology (FNAC) - aspirates from the lesion showed predominantly many lobules of mature adipocytes, thin capillary channels seen traversing among them. A few ovoid to spindle cells seen encircling and traversing in between adipocytes. Background showed myxoid substance, collagen fibrils and lipid droplets.
Excision biopsy and histopathological examination with H&E, special stains Alcian blue and PAS revealed myxoid matrix in which were noted ovoid to stellate cells with numerous thin walled blood vessels, few of which were arborising in nature, seen punctuating the lesion. Scattered inflammatory cells were noted (Fig. 2a, b).
A diagnosis of Cutaneous Angiomyxomas was made based on clinical and histological picture.

Discussion

In 1957, Herbert Z. Lund classified myxomas of the skin into cutaneous myxoid cysts of the fingers, mucoceles of the lip, and myxomatous reactions associated with epithelial elements [1].
Superficial angiomyxoma was first described as a cutaneous myxoma complex by Carney et al in 1986. 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 over activity such as Cushing's syndrome and Acromegaly [2].

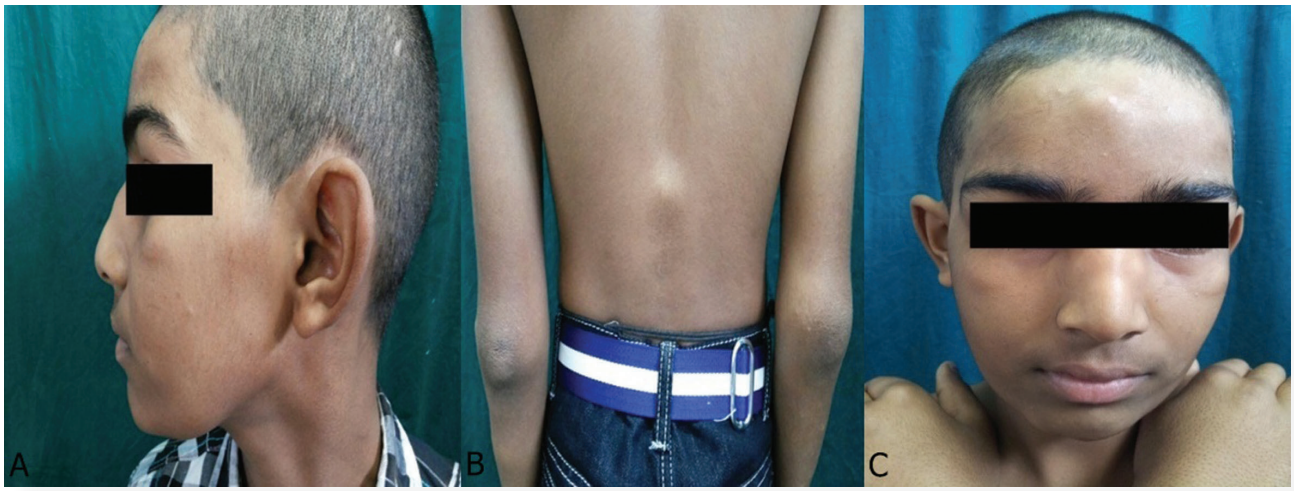

Figure 1A - C. Multiple swellings present over face, back, and upper limbs.

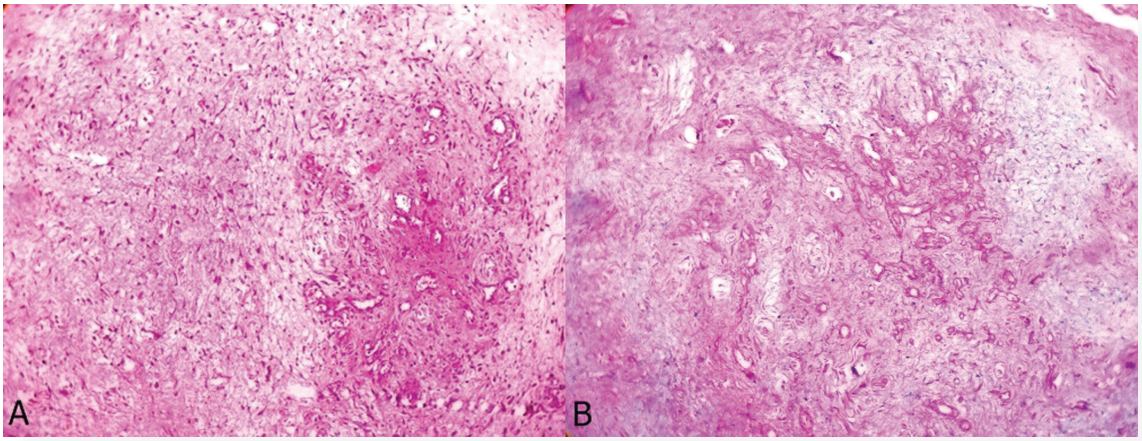

Figure 1A, B. Myxoid matrix with numerous thin walled blood vessels, stellate cells and scattered inflammatory cells.

Carney complex, previously reported as NAME (Nevi, Atrial myxoma, Myxoid neurofibromas, and Ephelides) and LAMB (Lentigines, Atrial myxoma, Mucocutaneous myxoma, and Blue nevi) syndromes, is inherited as either an AD or X-linked disorder as a result of mutations in the PRKAR1A gene [3].

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.

The skin condition was diagnosed as superficial angiomyxoma based on clinical and histologic findings of the tumor.

In 1988, Allen et al, proposed the disease entity „superficial angiomyxoma", which was a benign myxomatous neoplasm characterized by moderately to sparsely cellular angiomyxoid nodules with scattered small vessels [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].

In our case age of onset was 12yrs and lesions were measuring about 1-3 cm with predilection for face, forearms, and hands.

Histologically superficial angiomyxoma is a dermal based lesion with frequent extension to the subcutis. Tumours are multilobulated, with copious myxoid

stroma, numerous delicate small blood vessels and spindle-shaped or stellated bland cells, probably representing fibroblasts. Aggregates of inflammatory cells, mainly neutrophils, are frequent [1,5].

Individual nodules are moderately to sparsely cellular with copious basophilic interstitial material that is PAS negative, hyaluronidase sensitive and alcian

blue positive at pH 1-4. Spindle- and stellate-shaped cells are scattered in the myxoid stroma [1,5].

In our case histopathological feature was consistent with the literature except occasional mitotic figure and adipose tissue seen entrapped within the lesion at places.

Conclusion

Contrary to the typical description, the patient here is an adolescent boy. The clinical features, histopathology are consistent with description of superficial angiomyxomas except for the sites of predilection.

Patient is on follow up and there is no new lesion.

Cutaneous Angiomyxomas limited to skin and subcutaneous tissue is presented for its rarity.

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REFERENCES

1. Allen PW, Dymock RB, MacCormac RB. Superficial Angiomyxomas with and without Epit helial Components. Am J of Surg Pathol. 1988;12:519-30.

2. Yamamoto K, Kondo A, Iwashita K, Umezawa Y, Ohta Y, Matsuyama T, et al. A case of Superficial Angiomyxoma. Tokai J Exp Clin Med. 2006;31:43-5.

3. O'Neill JL, Narahari S, Sane DC, Yosipovitch G. Cardiac manifestations of cutaneous disorders. J Am Acad Dermatol. 2013;68:156-66.

4. Nakayama H, Hirol M, Kiyoku H, Naruse K, Enzan H. Superficial Angiomyxoma of the Right Inguinal Region: Report of a case. Jpn J Clin Oncol 1997;27:200-3.

5. Calonje E. Soft Tissue tumours and tumour like conditions In: Rooks Burns T, Breathnach S, Cox N, Griffiths C, editors. Rooks textbook of Dermatology. 8th edition, vol 3 Oxford: Blackwell; 2010. p56.59.