

Superficial angiomyxoma of the external ear not associated with Carney's complex: A case report

Khadilkar UN¹, Khadilkar NP², Rao PS³, Chakravorty S⁴, Goel G⁵

¹Professor, ³Assistant Professor, ⁵Postgraduate Student, Department of Pathology, Kasturba Medical College, Mangalore ²Associate Professor, Sint Eustatius School of Medicine, Sint Eustatius

Abstract

Superficial angiomyxomas are rare tumours and multiple tumours occurring in the external ear are invariably associated with Carney's complex. In the present study, a solitary superficial angiomyxoma was found in a 20year old lady; and there was no evidence of any of the components of Carney's complex at the time of presentation or at the end of 2 years of follow-up, after the surgical excision of the tumour.

Key words: Solitary, Superficial angiomyxoma

Superficial angiomyxomas are rare cutaneous tumours with a male preponderance and a tendency for local recurrence. Despite being first described in 1988, it is still a poorly recognized cutaneous tumour. Superficial angiomyxomas can present either as solitary or multiple lesions. Multiple myxomas of the external ear are clinically significant as they are almost always associated with Carney's complex.¹ In the present study, the patient presented with a solitary angiomyxoma in the external ear without evidence of Carney's complex and without any recurrence for one year after the surgical removal of the tumour.

Case Report

A 20year old lady presented with a right external ear mass of 3years duration. It was small to begin with and was slowly progressive, associated with decreased hearing, tinnitus and off and on bleeding. On clinical examination, a lobulated, polypoid mass measuring 5cms in its greatest dimension was noticed. The surface was ulcerated and there was evidence of bleeding on touch (Fig. 1). The post auricular lymph nodes were palpable. There was no evidence of facial weakness or nystagmus. A CT scan revealed a large lobulated and pedunculated irregular soft tissue mass in the anterosuperior aspect of the right external ear, blocking the external auditory canal. FNAC of the mass yielded only hemorrhagic and acellular material. During surgical excision, there was profuse bleeding from the base of the pedicle which was cauterized by bipolar diathermy (Fig. 2). The patient was investigated for endocrine overactivity, pigmentation, myxomas and

schwannomas at other locations. Since all these manifestations were not found in the present case, Carney's complex was ruled out.

Pathological findings

Grossly, the lesion was irregular, nodular, pedunculated and grey-white measuring 4.5x3.5x3 cms. Cut surface showed soft grey white areas with cystic and myxoid changes. Multiple sections from the centre and periphery of the mass showed an unencapsulated tumour, composed of many ectatic blood vessels, separated by fibromyxoid stroma. No mitoses were seen in the stroma. Neither nuclear hyperchromasia nor pleomorphism was apparent. Stellate cells, muciphages, mucin, neutrophils and lymphocytes were seen in the stroma (Fig. 3). There was polypoid elevation of the epidermis which was hyperplastic and focally ulcerated. No epithelial or adnexal structures were detected in the lesion. The defect in the external auditory canal completely healed after six months and on follow up for two years, there was no recurrence or evidence of tumour on CT scan.

Correspondence

Dr. Urmila N. Khadilkar
Professor of Pathology
Kasturba Medical College
Mangalore-575001 Karnataka State, India
Email: urmilank@yahoo.co.in



Fig 1: Gross photograph of the tumour protruding from the ear

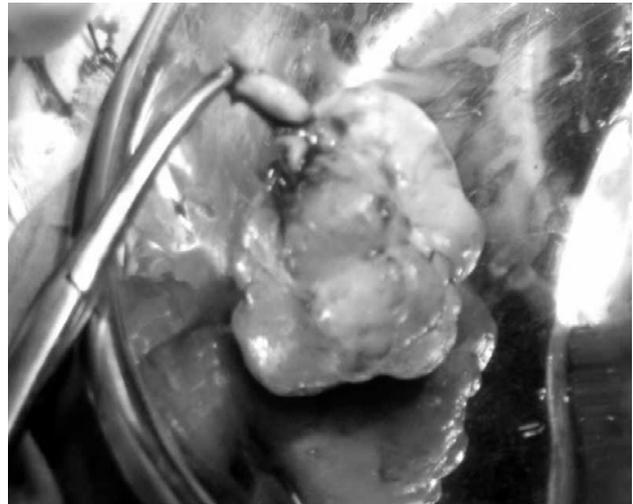


Fig 2: Excision of tumour mass with pedicle

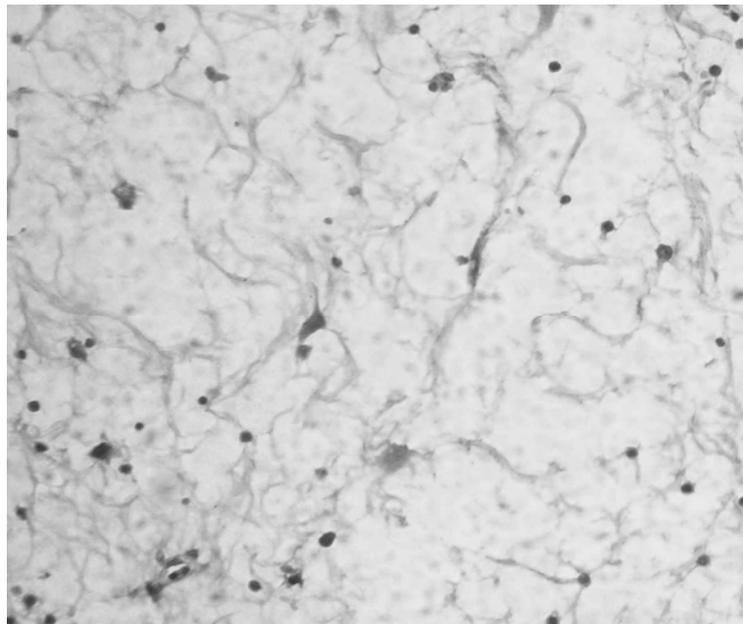


Fig 3 High power view of the tumour showing stellate cells and inflammatory cell infiltration (H and E stain 45x)

Discussion

Superficial cutaneous angiomyxoma is a distinctive clinicopathologic entity, first described in 1988.² It is a rare benign tumour in adolescents and adults, with a predominant manifestation in the region of the maxillary and mandibular bones.³ It can arise in the genital region, trunk, lower and upper extremities. In the head and neck region the tumour can be seen on the head, lower eyelid, preauricular location, pinna and the external auditory canal. Multiple myxomas occurring in the external ear are almost pathognomonic of Carney's complex or syndrome.⁴ Carney's complex is a familial disorder transmitted as an autosomal dominant trait; it includes myxomas, spotty pigmentation, endocrine overactivity and schwannomas. In addition to Carney's the other closely related syndromes are NAME and LAMB syndromes.⁵ In the present study the tumour was a solitary angiomyxoma located in the external ear and a clinical evaluation did not reveal the syndromic components associated with Carney's complex.

Clinically superficial angiomyxoma is a polypoid or papulonodular cutaneous lesion which may be confused with a cyst, skin tag, or neurofibroma.² The symptoms of decreased hearing or deafness due to occlusion of the canal relieved by removal of the tumour, bleeding after minor trauma, pain and ear canal discharge may be present as seen in the present case.⁶

Histologically superficial angiomyxoma is a dermal based lesion with frequent extension to the subcutis. It is multilobulated with poorly defined margin. 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. Mitoses are sparse, numbering less than one per 10 HPF. The occasional findings in the superficial angiomyxoma are perivascular hyalinization, perivascular lymphocytes, fibrin thrombi, interstitial haemorrhage with blood lakes, and hemosiderophages. One unique feature in contrast to other cutaneous myxoid tumours is the presence of neutrophils, eosinophils, lymphocytes and mast cells in the stroma as seen in the present case. In approximately 20% of cases, the primary lesion or its recurrence contained epithelial structures including epidermoid cyst, thin strands of squamous epithelium and small buds of basaloid cells. The epithelial component may represent entrapped adnexal structures or adjacent squamous epithelium.^{2, 5} Stromal cells are immunoreactive for vimentin and focally reactive for SMA representing focal myofibroblastic

differentiation, and negative for desmin, KP1, MAC387, factor XIIIa, CD34, Leu-7, pankeratin and S-100 protein.^{2, 7}

The other cutaneous myxoid tumours in the differential diagnosis include, cutaneous focal mucinosis, cutaneous myxoid cyst, trichodiscoma, trichofolliculoma, dermal nerve sheath myxoma (neurothekeoma), myxoid neurofibroma, myxoid liposarcoma, and myxofibrosarcoma^{4, 5}. Focal cutaneous mucinosis lacks the lobular architecture, stromal neutrophils, and epithelial structures found in cutaneous myxoma. Cutaneous myxoid cyst is easily distinguished given its exclusive location on the fingers. Neurothekeoma has a more pronounced lobular growth pattern and is characterized by plumper cells that are invariably positive for S-100 protein. Myxoid neurofibroma is composed of cells with wavy or buckled nuclei that are also S-100 protein positive. Myxoid liposarcoma is more deeply located and larger than cutaneous myxoma, and it is characterized by a plexiform vasculature with scattered lipoblasts. Myxofibrosarcoma has a greater degree of nuclear atypia and hyperchromasia as well as curvilinear vessels often lined by hyperchromatic tumour cells.^{5, 8}

Aggressive angiomyxomas differ from the superficial angiomyxomas by the deeper locations, occurring in the genital, pelvic and perineal regions as single and larger nodules. Histologically aggressive lesions show variably sized vessels ranging from small thin walled capillaries to large vessels with secondary changes including perivascular hyalinization, medial hypertrophy and a prominence of mast cells. Focal areas of hypercellularity and associated nuclear hyperchromasia and pleomorphism are also seen.⁹ Cutaneous myxoma has a propensity for local recurrence if incompletely excised. In the series by Allen et al., 8 of 20 (40%) tumours recurred, including five of eight tumours with epithelial components.⁵ Calonje et al, reported a recurrence rate of 30%, in incompletely excised tumours in a median interval of 12 months following initial excision.² No metastasis has been reported with cutaneous myxomas.⁸

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