CASE REPORT

Angiomyxolipoma of the Upper Lip-A Rare Tumor at an Uncommon Site with Review of Literature

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Abstract:

Angiomyxolipoma is a very rare benign mesenchymal tumor characterized by admixture of adipose tissue and myxoid stroma along with multiple vascular channels. It should be differentiated with other benign and malignant variants of lipoma. We are reporting a case of 70 years old male with a history of a painless swelling in upper lip for 5 years.

Keywords: Angiomyxolipoma, Benign, Lip, Myxoid

Introduction

Lipoma is a common benign soft tissue neoplasm consisting of mature adipocytes usually found in the subcutaneous tissue of the trunk, proximal limbs, thigh and neck. In oral cavity, lipomas are the most common mesenchymal neoplasm. Variant of lipoma are much less common and differ from ordinary lipoma by a characteristic microscopic picture and specific clinical setting. These include angio-, fibro-, myo-, angiomyo-, myelo-, chondroid, spindle cell/ pleomorphic, hibernoma and lipoblastoma [1]. Angiomyxolipoma or vascular myxolipoma was first described by Mai et al in 1996 as a rare variant of lipoma [2]. On reviewing the literature, only 17 cases of angiomyxolipoma were reported in the past out of which only 2 were intraoral [2-18].

Case Report:

A 70 year male presented with history of a painless swelling on the undersurface of upper lip for 5 years. There was no history of trauma and discharge. On physical examination, a solitary, well defined, firm, non-tender, submucosal mass was identified on upper lip. Overlying mucosa is normal in color and texture. Clinical diagnosis of mucocele was made.

Total excision of the swelling with wide margins was done under local anaesthesia. Grossly, the specimen was spherical, firm, capsulated mass measuring 3.0x2.4x1.0 cm with external surface smooth and pink (Fig.1a). Cut surface shows grayish white coloured solid mass with mucoid shiny surface (Fig.1b). Histopathological examination revealed admixture of lipomatous areas and myxoid stroma containing abundant small to medium sized vessels. Lipomatous component consists of mature adipocytes distributed randomly throughout the tumor. Myxoid areas are hypocellular and contain

bland spindle cells. Scattered mast cells and few histiocytes were also identified in myxoid areas (Fig. 2). Myxoid areas stained strongly with alcian blue (pH 2.5) while spindle cells of myxoid areas and endothelial cells are CD34 positive (Fig. 3a and Fig.3b). There was no evidence of necrosis, haemorrhage, atypia, mitotic figures, chondroid metaplasia or lipoblast. Diagnosis of angiomyxolipoma of upper lip was made. Patient was followed up for 8 months and no recurrence was noted.

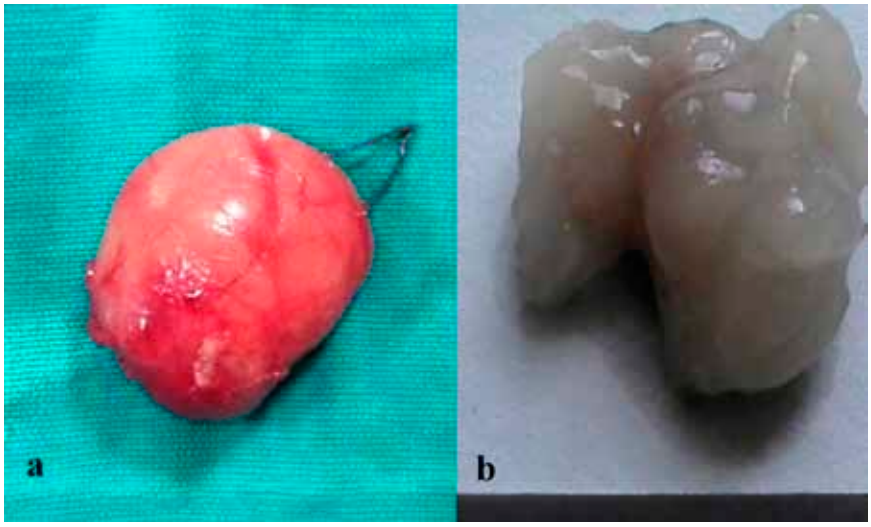

Fig.1: Photograph Showing (a) A 3x2.4x1cm Spherical Mass with External Surface Smooth and Pink (b) Cut Surface is Grayish-White in Colour, Shiny and Mucoid

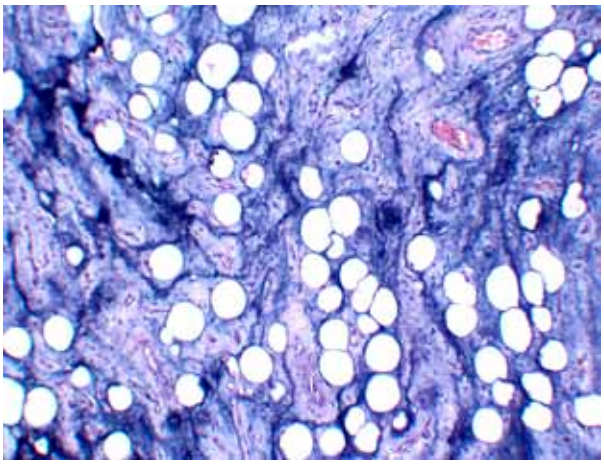

Fig. 2: Microphotograph Showing Admixture of Mature Adipocytes, Myxoid Stroma and Small to Medium Sized Vessels (H&E X 100)

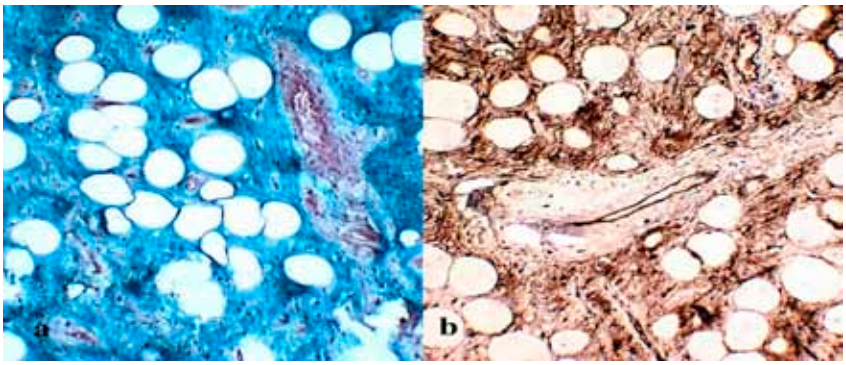

Fig. 3: (a) Microphotograph Showing Myxoid Stroma Staining Positive for Alcian Blue (AB X200). (b) Immunohistochemical Reactivity for CD34 in Spindle Cells of the Myxoid Stroma and Endothelial Cells (X200)

Discussion:

Angiomyxolipoma is a rare variant of lipoma. It usually presents as a solitary, slow growing painless mass with male predominance [2, 8, 11]. The most common location has been the subcutaneous tissue in the scalp, arm, wrist, thigh and gluteal region with one case each at spermatic cord and subungual area [3, 6, 9, 11]. The tumor does not invade locally or reveal any malignant transformation [18]. Until now, only two cases of angiomyxolipoma in oral cavity were reported, one in buccal mucosa and another in floor of mouth [13, 18]. Our patient presented with a history of painless lip swelling for 5 years. The histopathologic features of angiomyxolipoma include admixture of mature adipose tissue without lipoblast, myxoid stroma with few spindle cells, and multiple thick and thin walled blood vessels [2, 8, 11, 13]. Although the histologic features are characteristic, the lesion needs to be differentiated from other variants of lipomas. With the help of immunohistochemical staining, we can not only identify the various elements in the tumor but also differentiate it from other tumors. The spindle cells of the myxoid areas usually express CD 34 and Vimentin and not Desmin, S100 or SMA. The vascular smooth muscle cells express SMA and Vimentin whereas the mature adipocytes are S100 positive [11, 18]. In our case, spindle cells show positivity for CD34 and adipocytes are positive for S100.

The differential diagnosis includes myxolipoma, angiolipoma, angiomyolipoma, superficial angiomyxoma, myxoid spindle cell lipomas, myxoid liposarcoma and low grade myxoliposarcoma [11]. The histopathological characteristics of AML is the presence of vascular, myxoid and lipomatous elements, one of which is usually absent in angio, myo or myxolipoma. The tumors with smooth muscle like myolipoma and angiomyolipoma can be differentiated with HMB 45 expression. Myxoid spindle cell lipoma can be differentiated with the characteristic collagen

bundles, sparse vascular features with mild positivity to CD 34, whereas absence of myxoid areas and vascular structures help in differentiating vascular and pseudoangiomatous spindle cell lipomas respectively [18]. Absence of lipoblast and a diffuse capillary plexiform pattern rules out the diagnosis of myxoid liposarcomas [14].

Radiological investigations like CT & MRI are useful in defining the extent of lesion and thus help in planning the surgery [18]. Treatment is wide local excision. Recurrence was not seen in any of the reported cases in the past [2-18]. On reviewing the literature, only 17 cases of angiomyxolipoma were found to be reported in past (Table 1). The age of patients ranged from 9 to 81 years with most cases occur during 6th and 7th decade. Tumour shows male sex predilection with M: F ratio being 4.7:1. The common sites involved are scalp and upper extremities. Only 2 cases of oral cavity were reported in the past [13, 18]. Angiomyxolipoma is a rare benign lipomatous tumor with characteristic histopathological and immunohistochemical features. It should be differentiated from other lipomatous and myxoid lesions so that it can be managed surgically with complete excision.

Table 1: A Review of Reported Cases of Angiomyxoma from the Published Literature

Sr. No.	Author	Age	Sex	Site	Year
1	Mai KT *et al*	32	M	Spermatic cord	1996
2	Zamecnik M *et al*	57	M	Scalp	1999
3	Okafor O *et al*	50	M	Upper back	2000
4	Scio R *et al*	60	F	Thigh	2001
5	Tardio JC *et al*	66	M	Scalp	2004
6	Lee HW *et al*	44	M	Arm	2005
7	Sanchez Sambucety P *et al*	57	M	Wrist	2007
8	Kang YS *et al*	43	M	Subungual area, gluteal area	2008
9	Usta U *et al*	36	M	Neck	2009
10	Song M *et al*	69	M	Hip	2009
11	Kim HJ *et al*	09	M	Knee	2010
12	MartAnez –Mata G *et al*	12	M	Oral cavity	2011
13	Anand M *et al*	81	M	Elbow	2012
14	Pukar M *et al*	15	F	Colon	2012
15	Hantous- Zannad S *et al*	49	F	Posterior mediatinal	2012
16	Faten hammed *et al*	50	M	Thigh	2013
17	Nair SA *et al*	70	M	Floor of mouth	2014

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