

Clinical, Cytopathological and Histopathological correlation of Angiofibroma in Head and Neck, a rare Case Report.

¹Dr Richa Gupta, ²Dr Shaweta, ³Dr Pratima Thakur

1) Senior Resident at Department of Pathology, Shri Lal Bahadur Shastri Government Medical College And Hospital, Mandi, Himachal Pradesh, India.

2) Senior Resident at Department of ENT, Shri Lal Bahadur Shastri Government Medical College And Hospital, Mandi, Himachal Pradesh, India.

3) Senior Resident at Department of Pediatrics, Government Medical College and Hospital, sector-32, Chandigarh.

Corresponding Author- **Dr Richa Gupta**, Senior Resident at Department of Pathology, Shri Lal Bahadur Shastri Government College And Hospital, Mandi, Himachal Pradesh, India. Phone no- 7018536414

E-mail id: - **thakur.drricha@gmail.com**

Abstract

Angiofibroma is a benign mesenchymal neoplasm¹ of fibrous tissue and blood vessels. It usually occurs in adolescent males however it can be seen in older males at 6th -7th decade, as in our case. It is described as a rare neoplasm with frequency < 1% at Head & Neck region although prevalence seems to have geographic differences⁴. It is locally aggressive with mortality < 3%. Etio-pathogenesis is yet unexplained. Fine-needle aspiration cytology (FNAC) has a supplementary role for confirmation of diagnosis^{2,3}.

Immunohistochemistry confirms and characterizes the exact nature of this tumor. Angiofibroma is rarely seen in head and neck region, therefore it is important to know differential diagnosis and IHC when dealing with soft tissue mass at nape of neck.

Keywords: Angiofibroma Neck, benign vascular tumor, Soft tissue mass of head and neck, fibrovascular neoplasm.

1) Introduction

Angiofibroma is an uncommon benign fibrovascular neoplasm characterized by specific cytogenetic aberrations. The tumor has a unique feature comprising of prominent spindle cell component and prominent stromal vasculature. Men are affected more than women however examples in females have been described as well. It shows strong positivity for CD₃₄, a marker for endothelial cells of blood vessels, desmin and vimentin for mesenchymal tissue including muscle and collagen and smooth muscle actin (SMA) positivity for smooth muscles and fibroblasts⁴.

2) Case Report

Our report refers to 62 years old male patient who presented in our hospital at ENT OPD with the history of painless slow growing mass at posterior triangle of neck over the period of 5-6 months. He was asymptomatic and family history of any past illness was absent.

On clinical examination, a well defined mass measuring 1*1 cms was seen at posterior triangle of neck which was mobile, non tender and firm in consistency. Overlying skin was normal in color and texture. Clinically it was suspected to be **Lipoma**.

3) Method

FNAC was performed & blood mixed smears were prepared and examined. Later formalin fixed biopsy specimen of the same patient from above mentioned lesion was received at the histology section and slides were prepared after hematoxyline and eosin staining & reviewed. We describe here clinical, cytological and histopathological co-relation of tumor along with its differential diagnosis.

4) Result

Microscopic examination of FNAC showed cellular smears (Fig.1) comprising of small clusters, aggregates of isolated ovoid to spindle cells having bland chromatin, inconspicuous nucleoli embedded in myxoid stroma. Multiple prominent blood capillaries were seen surrounding these cells in a hemorrhagic background (Fig.2). No atypia / mitosis noticed. Cyto-morphological features were suggestive of **Benign Spindle Cell Lesion**.

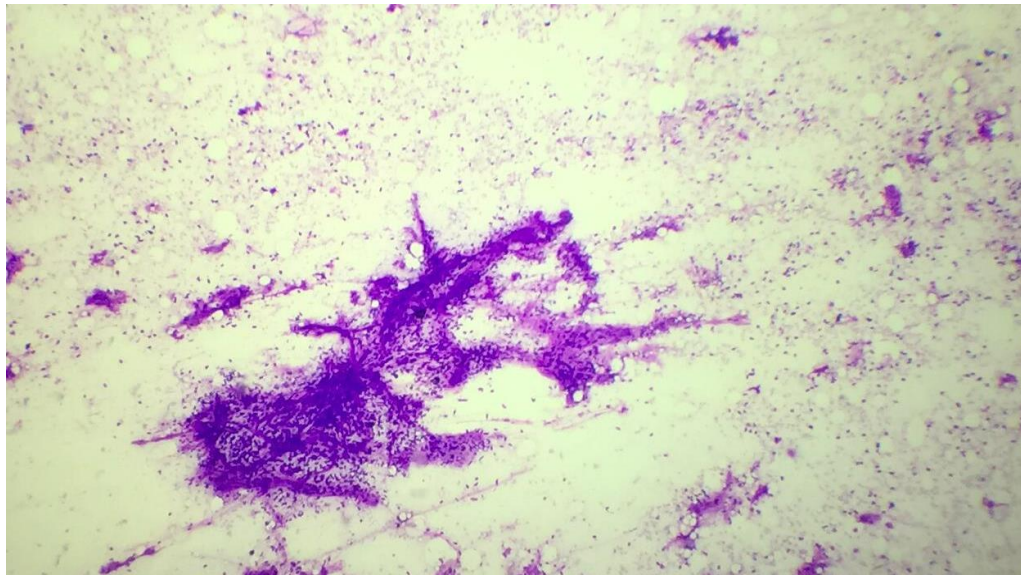


Figure:1 scanner view of benign spindle cell tumor

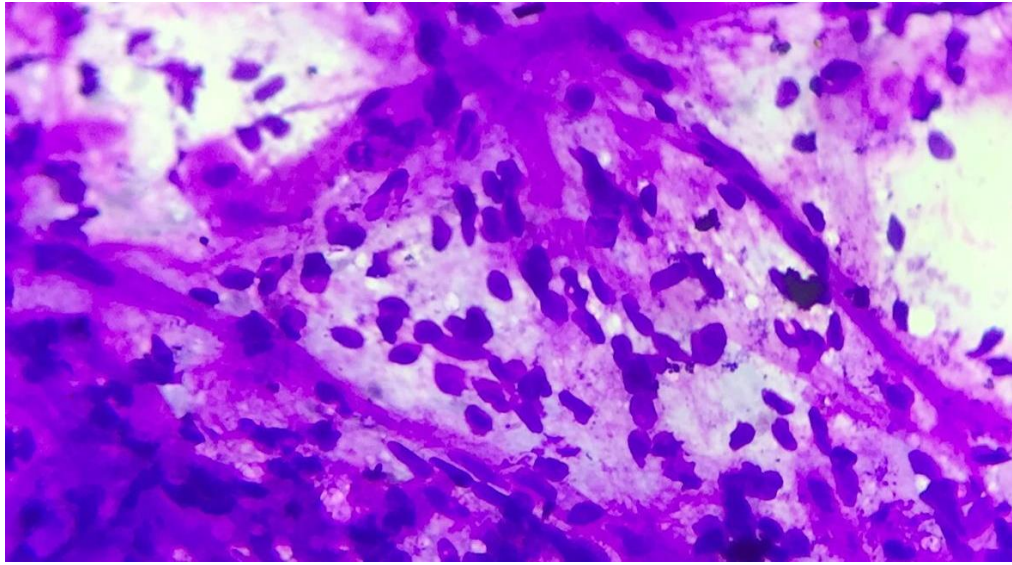


Fig: 2 High power view of benign spindle cell tumor showing spindle to ovoid cells and myxoid stroma

On gross examination- a single, well circumscribed, grey white to tan brown to yellowish, polypoidal soft tissue mass measuring 2*1.5* 0.9 cm³ was received. Cut surface showed well circumscribed, grey brown – tan brown areas measuring 9 mm in diameter having rubbery surface.

2 blocks with 2 pieces were prepared.

All tissue was used up.

Microscopic examination of H&E stained sections showed proliferation of thin walled blood vessels lined by endothelial cells surrounded by uniform bland spindle shape cells with oval nuclei and scant cytoplasm in the myxoid and collagenous stroma (Fig. 3,4 & 5). Focal areas showing adipose tissue were identified.

Histopathological features were suggestive of Angiofibroma, neck.

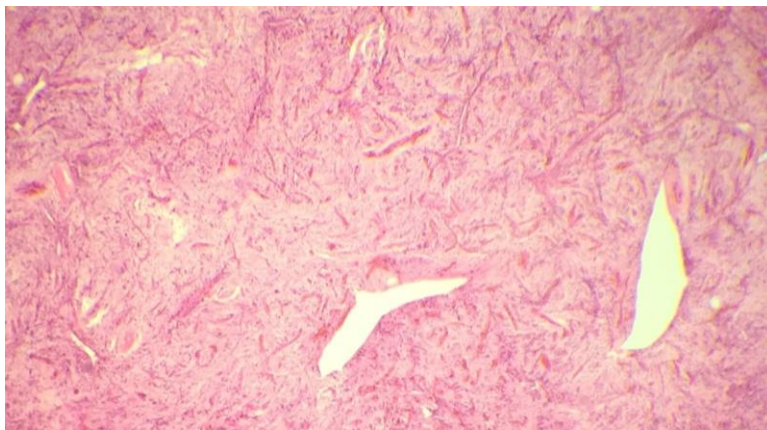


Fig: 3 scanner view of proliferation of thinned walled vessels along with myxoid stroma

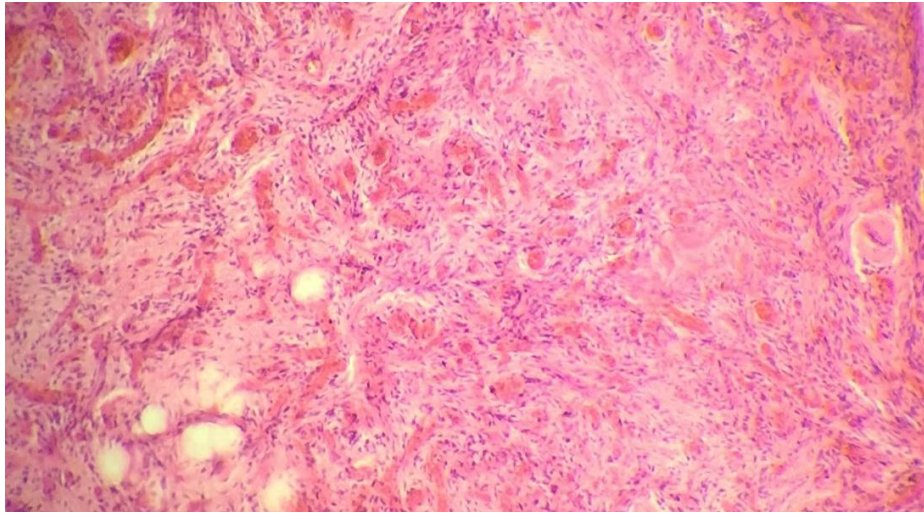


Fig: 4 low power view of numerous proliferating blood vessels having spindle shape endothelial cells.

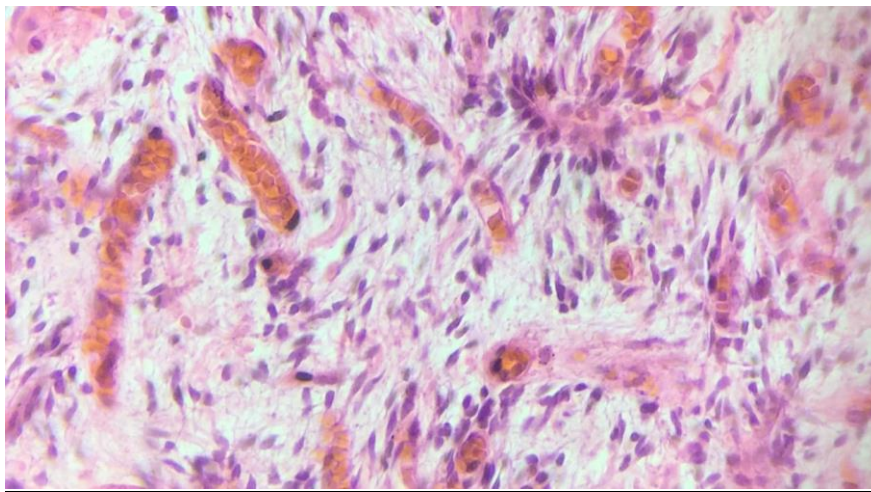


Fig: 5 high power view showing numerous proliferating blood vessels and stromal cells.

5) Discussion

Angiofibroma is a benign neoplasm of mesenchymal origin. Its presentation as soft tissue mass on head and neck region is a rare finding. Site of origin – extremities (esp. lower limbs -62% > upper limb – 16% > other sites including abdomen, back and pelvis). Head, orbit and neck involvement is ~ 0.05 -0.5%. AF is a locally aggressive benign soft tissue tumor with rare recurrence.

Lipomatous lesion are usually seen at the nape of neck in males having similar clinical presentation of mobile, non tender, soft tissue mass at posterior neck with slow growing history which was why it was suspected to be Lipoma.

Angiofibroma is a vascular tumor comprising of increased number of blood filled capillaries that leads to occurrence of profuse bleeding while performing routine FNAC procedure in this case. Cytologically these capillaries are lined by endothelial cell which are spindle to ovoid shape having bland chromatin giving it a suspicion of benign spindle cell tumor.

Histologic examination of the soft tissue mass revealed proliferation of thin walled blood vessels lined by endothelial cells surrounded by uniform bland spindled cells with oval nuclei and scant cytoplasm in the myxoid and collagenous stroma suggesting it to be Angiofibroma. Positive IHC marker -CD34 confirmed our case to be Angiofibroma.

6) Differential diagnosis based on histopathological findings

A) Adipose tissue tumors (lipoma) – is characterized by benign fragment of mature adipose tissue.

- **Spindle cell Lipoma**- is a benign fatty tumor
Location: regions of the shoulder and posterior neck of adults.
C/F: painless slow growing mass
Gross: yellow, soft lobulated
Histologically It is composed of an admixture of mature lipocytes and uniform spindle cells set in a mucinous and fibrous background
- **Angiolipoma**^{5,6}- These well-circumscribed small tumors occur shortly after puberty.
C/F: painful and characteristically multiple.
Location: sub cutis, most commonly on the trunk or extremities.
Gross: encapsulated, yellow red nodule
M/E: mature adipose tissue, branching capillaries and thick walled blood vessels with pericytes at periphery. Hyaline/fibrin thrombi constitute important diagnostic feature.
- **Myxolipoma**^{5,6}- This tumor is characterized by an admixture in variable proportions of mature adipose tissue and bundles of well-differentiated smooth muscle having myxoid change in the background

B) Neural tumors –

- **Neurofibroma**- is a benign tumor of neural origin.
Location: orbit, neck, back, and inguinal region
C/F: painless slow growing solitary, skin colored nodule which invaginates at pressure.
Gross: firm to rubbery soft mobile mass.
C/S: grey white homogenous
M/E: cellular element having markedly elongated nuclei, with a wavy, serpentine configuration and pointed ends.
Special stains and IHC for confirmation: NSE, (neurofilaments)& S-100
- **Schwannoma (neurilemoma)** is truly encapsulated neoplasms and is almost always solitary.
Location: extremities, neck, mediastinum, retro peritoneum, posterior spinal roots, and cerebellopontine angle.
C/F: pain and neurologic symptom are uncommon unless large

Grossly: solid and cystic areas.

Microscopically it shows two different patterns - *Antoni A and B*. The type A areas, are quite cellular, composed of spindle cells often arranged in a palisading fashion k/a (*Verocay bodies*). In type B, areas the tumor cells are separated by abundant edematous fluid that may form cystic spaces.

C) Vascular tumors -

- *Capillary Hemangioma* - Benign tumor of vascular origin.

Site: anywhere in the body especially face, scalp, chest and back. It is usually not associated with pain.

Gross: polypoid reddish brown mass which bleeds easily. Cut surface shows rubbery tan brown areas with blood filled dilated spaces.

Microscopically it contains closely packed spindle cells with spaces containing little blood. Organized or thrombosed vascular channels lined by plump endothelial cells and scant fibrous stroma.

- *Lymphangioma*- Benign lymphatic tumor

Sites of predilection are the neck, axilla, breasts, chest, buttocks, and thighs, usually not associated with pain.

Grossly tumor is soft in consistency. Cut surface shows grey white solid areas with multiple cystic spaces.

Microscopically tumor contains large and small communicating cysts or sponge like areas composed of small cavernous spaces. These lymphatic channels are lined by attenuated endothelium. Cystic spaces may contain lymphoid aggregates.

- *Pyogenic Granuloma* –

C/F: presents as rapidly growing polypoid red mass, surrounded by a collarets of thickened epidermis, which has traumatic or hormonal etiology.

Location: anywhere over skin and mucosa

Gross: it is a red elevated and ulcerated lesion.

Microscopically: it is a pedunculated lesion composed of granulation tissue separated by band of connective tissue. There is *vascular proliferation, edema, and inflammation*. The epidermis is thinned and sometimes ulcerated at the top, and there is acanthosis and hyperkeratosis at the sides.

D) Fibroblastic/ Myofibroblastic tumors-

- *Solitary Fibrous Tumor*⁷- Deep tissue tumor of uncertain histogenesis.

Sex: adults affecting both sexes equally.

Location: deep tissue of thigh, neck, trunk, pelvis, retroperitoneum & serosal surface.

C/F: painless swelling with compression symptoms.

Gross: large, solid, well circumscribed & unencapsulated.

C/S: Grey white to tan brown, with hemorrhagic or cystic areas.

M/E: hypo and hyper cellular areas separated by hyalinised blood vessels showing stag horn appearance, spindle cells dispersed in thick collagen with elongated nucleus, indistinct nucleoli & minimal cytoplasm.

- ***Epidermoid Cyst***- Common keratin filled epithelial lined cyst.

Location: anywhere in the dermis or sub cutis.

Sex: both sexes are equally affected.

C/F: painless slow growing mass.

Gross: round, pearly white well define mobile mass, soft to firm.

C/S: pultatious material

M/E: fibrous wall lined by keratinized squamous epithelium, contains anucleated squams but no adnexa or hair seen.

7) Conclusion

Based on our findings we conclude that Angiofibroma in head and Neck region is seldom but they represent an important differential diagnosis to adipose tissue tumors and especially to lipoma⁵. Therefore one must keep in mind the important differential diagnosis since misdiagnosis of these neoplasms as more aggressive tumors can lead to unnecessary treatment⁶.

8) Conflict of interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

9) Ethical Approval/ Funding

Ethical Approval – not required & no funding received for this article to be published.

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