Case Report - Angiomyofibroblastoma

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ABSTRACT

Angiomyofibroblastoma is a rare benign mesenchymal tumor occurs in the vulvo-vaginal region of women. Rarely it occurs in scrotum and inguinal region in males. We report a case of 47 year old woman at our hospital with complaint of painless swelling over vulva which gradually enlarged over last 13 years. There is history of trauma 13 years back. No other complaints. Hysterectomised 27 years back for abnormal uterine bleeding. No other comorbidities. Local examination of vulva showed a well circumscribed swelling of size 4 cms x 3 cms with a peduncle over lower 1/3rd of left labia majora. Soft and elastic in consistency, no tenderness, no change in skin color or temperature over the swelling. No palpable inguinal lymph nodes. Surgical excision was done and specimen was sent for histopathological examination. HPE report showed spindle shaped cells arranged in alternating hyper and hypocellular areas with collagen fibrils admixed with adipocytes and thin walled blood vessels. No evidence of malignancy. Confirmed it as Angiomyofibroblastoma.

Keywords : Vulval swelling, Mesenchymal tumour, Pedunculated, Surgical Excision, Spindle shaped cells, Angiomyofibroblastoma.

INTRODUCTION

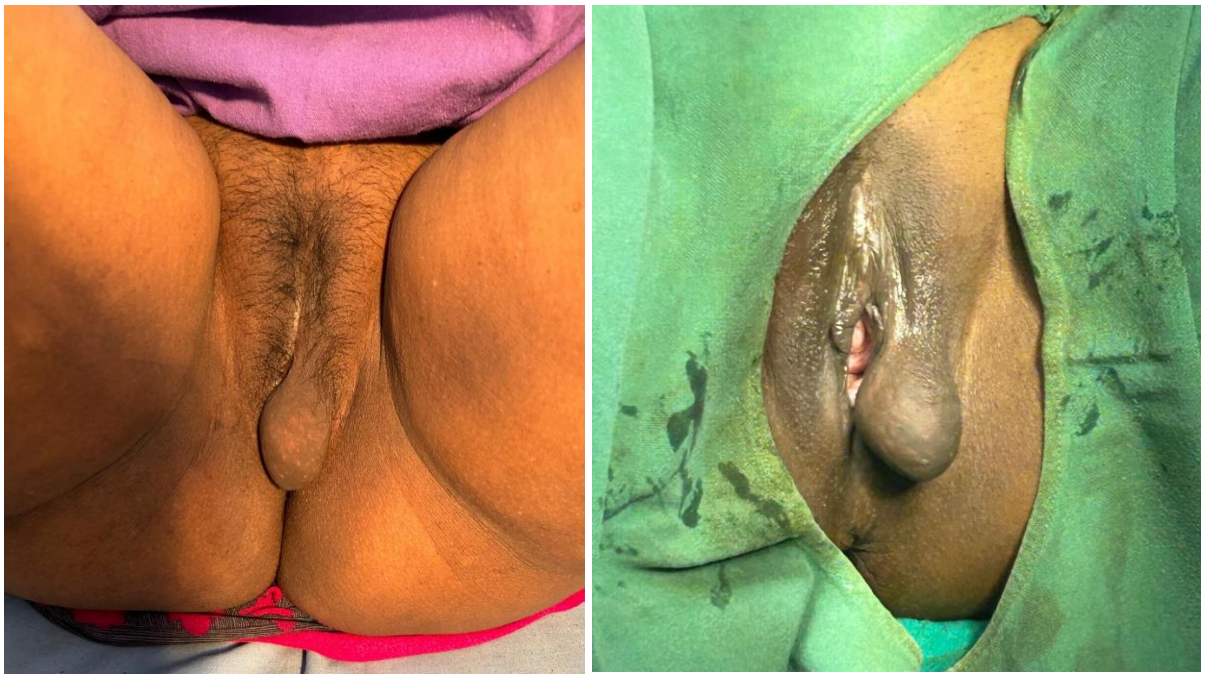
Angiomyofibroblastoma(AMFB) is a rare benign mesenchymal tumor mostly occurs in women of reproductive age in vulva, vagina, perineum and retroperitoneum.[1] Rarely occurs in males in the inguinoscrotal region, scrotum and spermatic cord.[2]

It is a well circumscribed lesion due to which it resembles Angiomyxoma.[3] It has slow growth, painless, pedunculated in some cases, less tendency for recurrence and metastasis.[4] It is often misdiagnosed as Bartholin cyst due to its slow growing nature.[5] Hence it is often difficult to diagnose preoperatively. The two major components are prominent blood vessels and stromal cells.[6] Histopathological findings show abundant thin walled blood vessels with hypo and hypercellular areas[7] which had immunoreactivity for desmin, vimentin, positive for estrogen and progesterone receptor. Negative for S – 100 and CD – 34.[8]

CASE REPORT

A 47 year old, parous woman came to our Gynaecology outpatient department with complaint of swelling in the vulval region which is painless and gradually increased. It was first observed by the patient 13 years ago following trauma over vulval region due to slippage. No other complaints. She was Hysterectomised 27 years back for abnormal uterine bleeding. No significant past history. Family and Medical history was not significant.

On local examination of vulva, showed a well circumscribed swelling of size 4 cms x 3 cms with a peduncle over lower 1/3rd of left labia majora. Soft and elastic in consistency, no tenderness, no change in skin color or temperature over the swelling. On pelvic examination, no palpable pelvic masses and vault is intact. There are no palpable inguinal lymph nodes. Upon examination her vitals and systemic examination was normal.

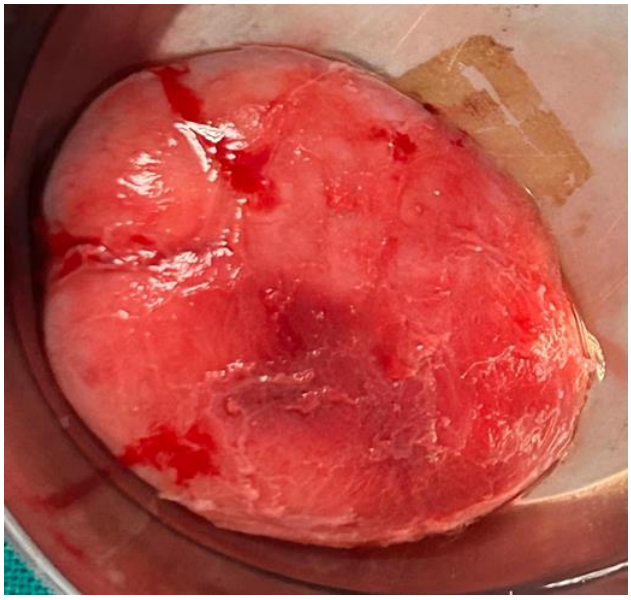


Pre operative images of the tumor

Transperineal Ultrasound was done in which mass appeared solid, well demarcated, vascularized lesion, hyperechoic areas with irregular and small hypoechoic cystic spaces scattered within homogenous echogenic stroma.

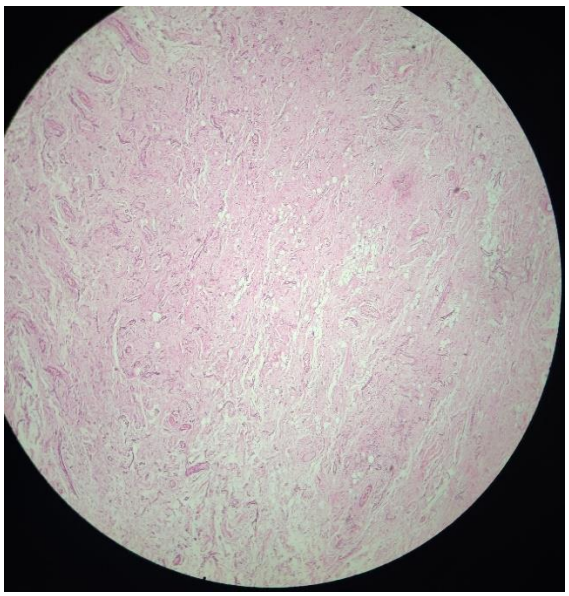
We planned out to excise the mass and send for Histopathology. Patient was admitted and all routine investigations were done which are within normal limits.

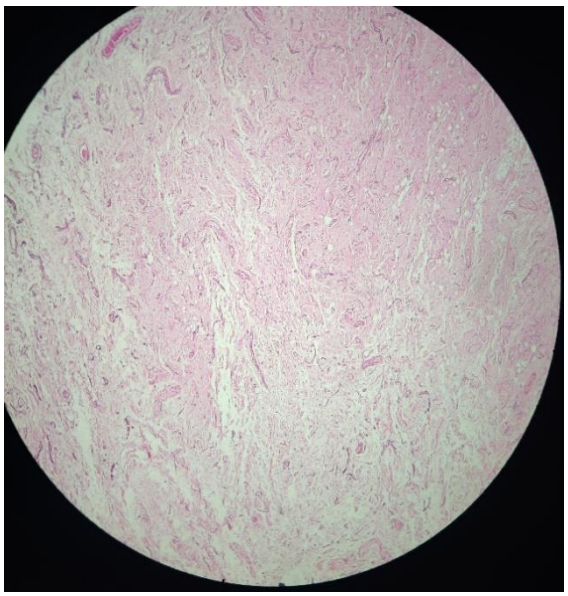
Surgery was performed under Spinal Anaesthesia. Surgical margins of the lesion were clear. The resected specimen was a well circumscribed and non-encapsulated mass, soft and elastic in consistency measuring 4 cms X 4 cms X 3 cms. No areas of haemorrhage or necrosis noted. Specimen was sent for Histopathological examination.



Gross Specimen of the tumor

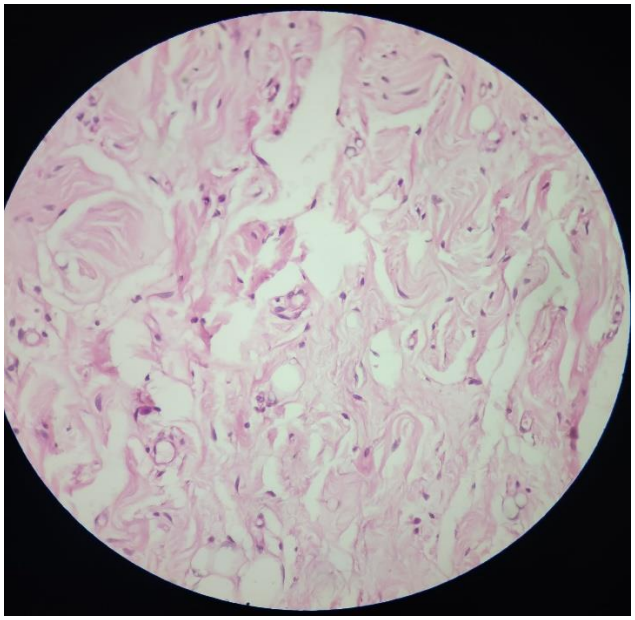
Histopathology sections showed circumscribed tumour composed of spindle shaped cells arranged in alternating hyper and hypocellular areas with collagen fibrils admixed with adipocytes and thin walled blood vessels. No evidence of any malignant changes noted. Thus diagnosis of Angiomyofibroblastoma was confirmed.





a) Shaply circumscribed unencapsulated

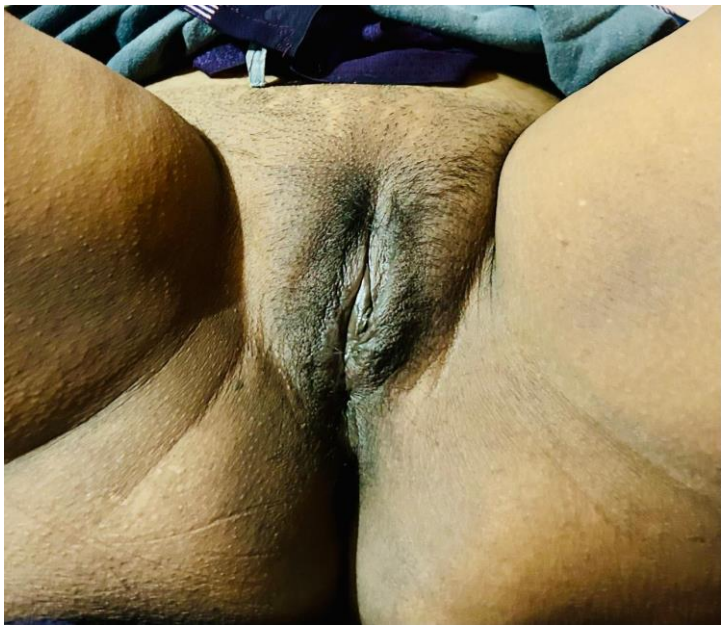
b) Hyper and hypocellular areas, abundant irregularly distributed capillary blood vessels or small veins in myxoedematous to fibrous collagenous stroma.



c) Bland spindle to epithelioid to plasmacytoid tumor cells with eosinophilic cytoplasm arranged in a nest, a cord-like pattern or singly and concentrated around blood vessels and between fat cells.

POST OPERATIVE FOLLOW UP

The patient came for follow up to OPD 1 week after surgery. Her vital signs were normal and there was no evidence of bleeding or infection at the surgical site. She was followed up after 3 months, patient was well and asymptomatic.



Follow up after 3 months

DISCUSSION

Angiomyofibroblastoma (AMFB) is a rare, benign soft tissue tumor that was first brought to attention by Fletcher et al. in 1992 through the report of ten cases localized to the vulva.[9] Though predominantly affecting the vulvo-vaginal region of middle-aged women, there have been rare instances of AMFB occurring in the scrotum and inguinal region of males.[10] The biologic behavior of AMFB is generally benign and the clinical outcome for most patients is favorable. Sarcomatous transformation is exceedingly rare, with only one case documented, and recurrence has not been a common feature.[11]

The average age of onset in females is reported as 45 years and AMFB tumors are characteristically slow-growing, often developing over weeks to several years without associated pain. Due to the subtle and nonspecific nature of these tumors, they are frequently misdiagnosed as more common conditions such as Bartholin gland cysts, inguinal hernias or benign mesenchymal tumors like lipomas or leiomyomas. Grossly, AMFB typically presents as well-circumscribed, rubbery masses with sizes ranging from 0.5 to 12 cm and a characteristic pink coloration.

Histologically, AMFB tumors demonstrate well-demarcated lesions with alternating hypercellular and hypocellular areas. The presence of abundant blood vessels and clusters of tumor cells around these vessels is a notable feature. Adipocytes may occasionally be observed within the mass, leading to the classification of certain cases as the "lipomatous" variant. A key histologic distinction between AMFB and aggressive angiomyxoma is the circumscribed borders of AMFB and the characteristic stromal cell clusters around blood vessels.

Immunohistochemical analysis of AMFB frequently reveals positive staining for **VIMENTIN** and **DESMIN**. Additionally, variable expression of muscle actin, as well as positivity for estrogen and/or progesterone receptors, suggests that the tumor may originate from hormonally responsive mesenchymal cells.

The preferred treatment for AMFB is simple surgical excision with clear margins, which typically leads to a favorable prognosis. In the case we present, the patient underwent successful excision of the tumor.

This case highlights the importance of recognizing AMFB as a distinct, rare clinical entity, particularly in the differential diagnosis of vulvar masses. Surgical excision remains the treatment of choice, and recurrence is not common when clear margins are achieved.

CONCLUSION

In conclusion, Angiomyofibroblastoma which is a rare tumour often misdiagnosed preoperatively. The presented case is a classical presentation of Angiomyofibroblastoma. Diagnosis is confirmed by histology. Managed by complete surgical excision.

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CONSENT FORM

Patient's / Participant's consent to publication of information about them (including image / photograph)

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Article : ANGIOMYOFIBROBLASTOMA

Journal name : JOURNAL OF CARDIOVASCULAR DISEASE RESEARCH

Title of article : CASE REPORT: ANGIOMYOFIBROBLASTOMA

Corresponding author : DR. K. VASUDHA BHARGAVI

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(CH. Venkata Lakshmi)

Signed :

Date : 22/10/2024

సమ్మతి ఫారమ్

వారి గురించిన సమాచారాన్ని (చిత్రం / ఫోటోతో సహా) ప్రచురించడానికి రోగి / పాల్గొనేవారి సమ్మతి

వ్యక్తి పేరు : చింత వెంకటలక్ష్మి

వ్యాసం : ఆంజియోమయోఫైబ్రోబ్లాస్టోమా

పత్రిక పేరు : జర్నల్ ఆఫ్ కార్డియోవాస్కులార్ డిసీజ్ రీసర్చ్

వ్యాసం శీర్షిక : కేస్ రిపోర్ట్ – ఆంజియోమయోఫైబ్రోబ్లాస్టోమా

సంబంధిత రచయిత : డా. కె. వసుధ భార్గవి

నేను CH.వెంకట లక్ష్మి పైన పేర్కొన్న విషయానికి సంబంధించిన సమాచారం మరియు/లేదా చిత్రాలు / ఛాయాచిత్రాలు/ నా/ నా బిడ్డ/ నా బంధువు గుర్తించబడిన జర్నల్ మరియు అనుబంధిత ప్రచురణలలో కనిపించడానికి నా సమ్మతిని తెలియజేస్తున్నాను. జర్నల్‌కి సమర్పించాల్సిన మెటీరియల్‌ని నేను చూశాను.

నేను ఈ క్రింది వాటిని అర్థం చేసుకున్నాను:

* సమాచారం నా పేరు లేకుండా ప్రచురించబడుతుంది. పూర్తి అనామకత్వం హామీ ఇవ్వబడదని మరియు ఎవరైనా నన్ను గుర్తించగలరని నేను అర్థం చేసుకున్నాను.

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* ప్రపంచవ్యాప్తంగా పంపిణీ చేయబడిన జర్నల్‌లో సమాచారం ప్రచురించబడవచ్చు. పాఠకులలో వైద్యులు మరియు పాత్రికేయులతో సహా ఇతర ప్రజా సభ్యులు కూడా ఉన్నారు.

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(CH. వెంకట లక్ష్మి)

సంతకం :

తేదీ : 22/10/2024