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Case Report

Angiomyofibroblastoma of scalp: a rare soft tissue tumour at an unusual site

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ABSTRACT

Angimyofibroblastoma is a very rare, benign mesenchymal soft tissue tumour which involves the vulvovaginal area of females of reproductive age group. These tumours are usually slow growing and painless which has a very low tendency for local recurrence. In women it is common in female genital tract, vulva, and posterior perivesical space. In men it occurs in spermatic cord. Here a report of 47 year old male patient from VMKV Medical College and Hospital, Salem was presented with a swelling in the posterior aspect of scalp from 6 months with no any associated symptoms. A preoperative diagnosis of dermoid scalp was made and patient was subjected for excision.

Keywords: Angiomyofibroblastoma, Angiomyofibrosarcoma, Vulva, Scalp

INTRODUCTION

Angimyofibroblastoma is a very rare, benign mesenchymal soft tissue tumour which involves the vulvovaginal area of females of reproductive age group. It was first described by Fletcher et al in 1992.[1] It is a benign tumour, but there has been reported case of recurrence and sarcomatous transformation.[2,3]

These tumours are usually slow growing and painless which has a very low tendency for local recurrence. In women it is common in female genital tract, vulva and posterior perivesical space.[4-6] Angiomyofibroblastoma consists of two components blood vessels and stromal cells, with prominent vascular component.

Angiomyofibroblastoma typically involves the valvular soft tissue of females of reproductive age group. These tumour present as a painless, small, slowly progressive and well circumscribed mass, usually less than 5 cm, white to yellowish brown and sponge like or myxoid on cross section. Histologically, the tumour cells are oval to spindle shaped with areas of high and low cell density and stromal proliferation of small vessels.

Generally, these tumours are reported to be benign, with no local recurrence or metastasis. Surgical excision is sufficient.

CASE REPORT

A 47 year old male presented with a swelling in the posterior aspect of scalp (occipital region) since 6 months. Patient was having difficulty in sleeping due to the location of swelling. There was no associated history of pain, fever or any dischare from the swelling. The swelling was slowly increasing in size for past 6 months. On local examination the swelling was soft to firm in consistency, around 8 cm × 6 cm in size, immobile with overlying skin stretched. There was no associated warmth or tenderness. Patients general physical examination and other systemic examination was normal. USG report showed a soft tissue lesion in subgaleal region of scalp tissue in occipital region with scalloping of underlying bone, possibly dermoid or lipoma. Screening CT was in favour of benign subgaleal lesion in right occipital region, possibly dermoid cyst. On the basis of clinical and radiological examination, a diagnosis of dermoid cyst

was made. Haematological and other routine investigations were normal.

After adequate preparation, the swelling was carefully dissected and excised under local anaesthesia. The resected tumour was well circumscribed, soft and measured 5*4 cm, and was sent for histopathological examination.

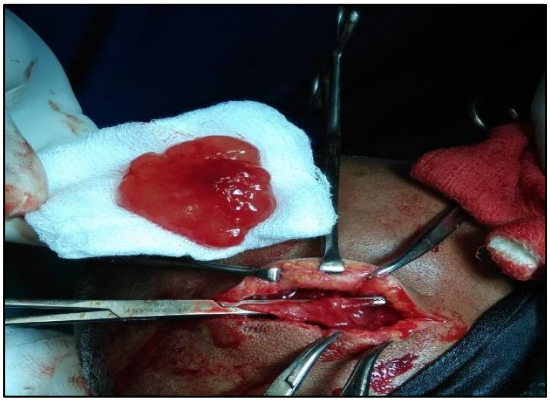

Figure 1: Intra-op photo showing tumour after excision from scalp.

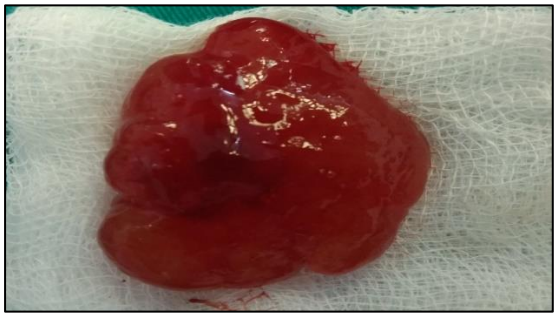

Figure 2: Tumour excised from scalp.

Post operatively patient had an uneventful recovery. Suture removal was done on postoperative day 10. Patient was followed in OPD for 6 months after surgery where the patient was asymptomatic and there was no sign of recurrence. HPE report showed a well circumscribed lesion with alternating hypocellular and hypercellular areas .Individual cells have oval to spindle shaped nuclei with moderate amount of eosinophilic cytoplasm with no atypia. Thin and ectatic blood vessels were seen. Mast cells were seen. Features were suggestive of angiomyofibroblastoma of scalp.

DISCUSSION

Angiomyofibroblastoma is a very rare mesenchymal soft tissue tumour which arises as a superficial soft tissue tumour in female genital tract of women of reproductive age group and in early menopause. The average age of onset is in the late 30's. The tumour are restricted to external genitalia and perineal soft tissue, and very rarely seen in males. The lesions are painless, slow growing, well circumscribed with a thin pseudocapsule and smooth surface, usually less than 5 cm in diameter. The cut surface appears white to yellowish brown and sponge like or myxoid. Histologically, they are characterised by alternating areas of hypercellularity and hypocellularity and randomly distributed thin walled vessels .The lesions are occasionally sclerotic, but no necrosis. The lesion is made up of spindle to epitheloid stromal cells, many stromal cells are plasmacytoid. Plenty of mast cells are seen scattered in the plasma. Eosinopjilic cytoplasm, which are occasionally hyaline. Multinucleated giant cells have also been described. An adipocyte component is seen in 10% of cases (lipomatous variant).[7,8] Immunohistochemically, the tumour cells are usually positive for desmin, vimentin, oestrogen receptor and progesterone receptor.

In present case, the lesion was present in the subgaleal region of scalp tissue in right occipital region, which is a very rare area as till now no case is reported of this tumour in scalp. The tumour was 5×4 cm in dimensions, was grey-brown in colour and soft. Our differential diagnosis was dermoid and lipoma. Characteristic histological feature of angiomyofibroblastoma was found. well circumscribed lesion with alternating hypocellular and hypercellular areas. Individual cells were having oval to spindle shaped nuclei with moderate amount of eosinophilic cytoplasm with no atypia. Thin and ectatic blood vessels were seen Mast cells were present. Absence of features like necrosis and mitosis were also in favour of angiomyofibroblastoma.

CONCLUSION

In present case, the tumour location is a very rare location, as most of the angiomyofibroblastoma till now has been reported in the female genital tract, and is seen mostly in women of reproductive age group. But in present case, the tumour was in subgaleal region of scalp in a male patient, which is an unusual site for angiomyofibroblastoma.

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