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A Rare Presentation of Angiomyofibroblastoma in the Mons Pubis

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Abstract **Case Report**

Angiomyofibroblastoma (AMF) is rare mesenchymal tumor. Most commonly reported in middle aged female, with predilection for the vulvovaginal region. To the best of our knowledge, no case of angiomyofibroblastoma involving the mons pubis has been reported in the literature yet. We present a rare case of angiomyofibroblastoma occurring in the mons pubis of 51year old premenopausal women. The case emphasizes the importance of considering angiomyofibroblastoma in the differential diagnosis of soft tissue masses in unusual locations and highlights the role of clinical assessment, imaging and histopathological evaluation in establishing the diagnosis and guiding appropriate management.

Keywords: Angiomyofibroblastoma, mons pubis, mesenchyaml tumor, stromal tumor.

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INTRODUCTION

Angiomyofibroblastoma (AMF) is a rare benign soft tissue tumor that most frequently affects the lower genital tract of young to middle-aged women.[1] It commonly arises in the vulva but can also occur in perineum, vagina, retroperitoneum and inguinoscrotal region, scrotum, and occasionally spermatic cord in men.[2] We encountered with rare mesenchymal tumor-angiomyofibroblastoma, at mons pubis, in 51year old premenopausal female.

CASE REPORT

A 51-year-old female patient presented with asymptomatic mass on the mons pubis on left side, which she noticed a year back and was gradually increasing in size since then, without any complication. On examination, the mass was measuring 2.5cm × 2.5 cm, soft in consistency. No inguinal lymph nodes were palpable. No other family or medical history was present. Ultrasonography of local area was suggestive of fairly well-defined oval, heterogenous hypochondriac lesion of 3*2.1cm noted in subcutaneous plane in the mons pubis. There was minimal vascularity noted. No obvious evidence of calcification or surrounding fat stranding noted. Simple excision was performed from the site. Gross appearance of 2*2cm specimen was, externally congested with cut surface was whitish solid homogenous. [Fig:I] Microscopy section studied shows fascicles of spindle cells interspersed by thin-walled blood vessel and hypo cellular edematous areas. The hyper cellular areas shows crowding of spindle cells around blood vessels. Mild cytological atypia seen. No areas of hemorrhage or necrosis or abnormal mitotic figures. Features suggestive of benign spindle cell tumor likely to be angiomyofibroblastoma. [Fig: II and III] Immunohistochemical examination suggestive of PR (progesterone receptor) positive and SMA negative. [Fig IV and V]

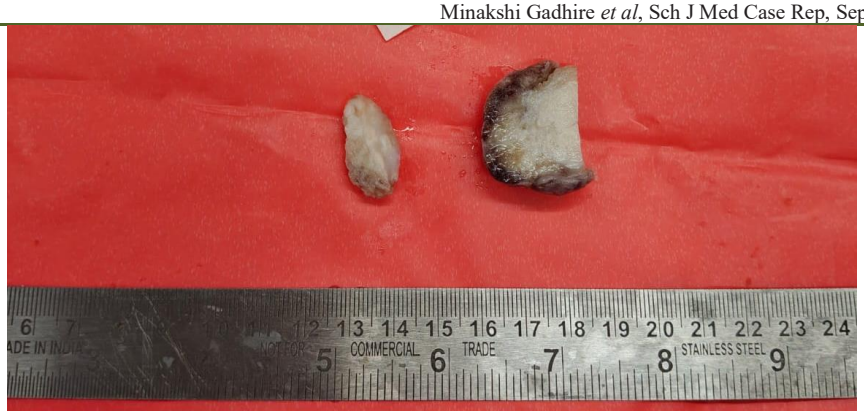

Fig: I – Gross appearance of specimen

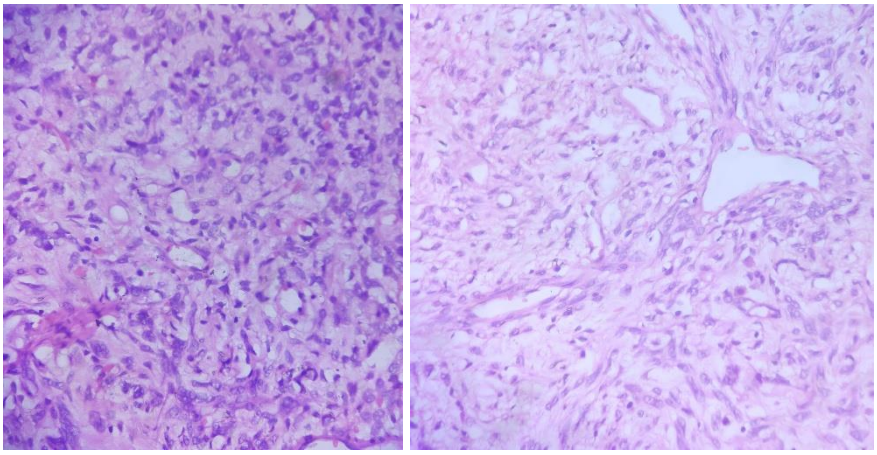

Fig: II and III – Microscopic picture of specimen

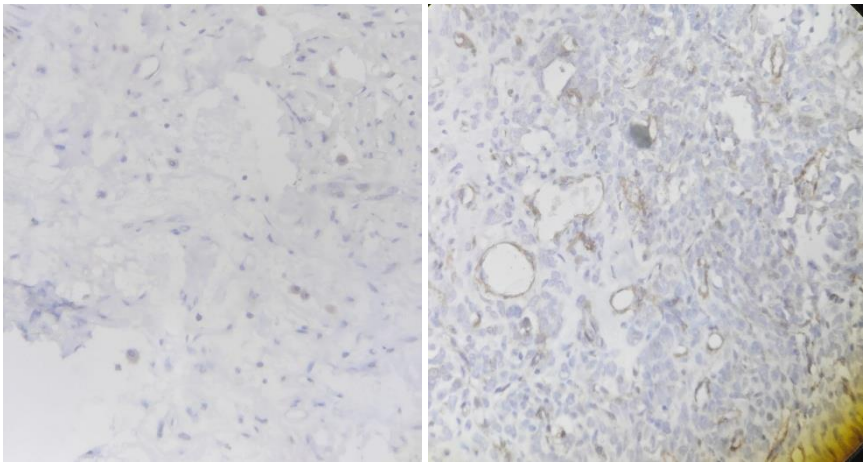

Fig: IV - PR – positive **Fig: V - SMA - Negative**

DISCUSSSION

Angiomyofibroblastoma is a well-circumscribed, nonencapsulated soft tumor, and microscopically, it consists of two components: stromal cells and numerous thin-walled blood vessels.[3] Angiomyofibroblastoma is composed of polygonal cells with myoid appearance in a myxoid matrix.[4], which differentiate the angiomyofibroblastoma from other mesenchyaml tumor. Due to the overlapping of histopathological picture, diagnostic perplexity often arises between AMF and aggressive angiomyxoma (AAM).[1] which require further investigation like immunohistochemical marker investigation. Because AMF being benign in nature is treated by local excision.[1] SMA+, ER+, AE1/AE3-, CD34-, S100- is the immunohistochemical staining for angiomyofibroblastoma.[4] Negative expression of epithelial immunostains, such as keratin ruled out a possibility of a carcinoma in that tumour, which was another differential diagnosis.[4] The cell origin of AMF remains unclear. Some studies consider AMF to be from myofibroblastic origin, where strong immunoreactivity for vimentin and variable expression for desmin and alpha-smooth muscle actin support this hypothesis.[5] It

has also been proposed that AMF might originate from perivascular stem cells with a capacity for myofibroblastic and fatty differentiation which is supported by capillary-like pattern demonstrated by stromal cells and obvious CD34 positivity around the vessels.[6] Since this tumor shows positivity for ER/PR, it provides the possibility of role of hormonal manipulation in its management.[7] angiomyofibroblastoma is managed primarily via local excision. (Nielsen *et al.,* 1996).[4] where as aggressive angiomyxoma requires extensive surgical approaches with postoperative radiotherapy or hormonal therapy due to greater risk of local recurrence upto 72%.[8]

CONCLUSION

Mons pubis stromal tumours are rare but distinct mesenchymal tumours, primarily affecting middle-aged women. They might misdiagnosed as other mesenchymal tumors such as lipoma, liposarcoma, aggressive angiomyxoma or superficial myofibroblastoma. Accurate diagnosis requires detailed histopathological evaluation along with immunohistochemical investigation. It is essential to differentiate angiomyofibroblastoma from aggressive angiomyxoma, as this significantly alters the subsequent management plan.

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