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Case Report

A rare tumor at an unusual site-angiomyolipoma vulva

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ABSTRACT

An angiomyolipoma is one of the very rare benign tumors. All three elements, that is, blood vessels, adipose tissue and smooth muscles, should be present to qualify it to be labelled as an 'angiomyolipoma'. Although it has been reported at other sites also like mediastinum, heart, spermatic cord, fallopian tube, oral cavity, penis, salivary glands, and skin.

Keywords: Angiomyolipoma, Vulva, Extra-renal

INTRODUCTION

Angiomyolipoma is rare benign mesenchymal tumor. It was previously considered as hamartoma. It most commonly occurs in kidney but extra-renal tumors location like liver, mediastinum, fallopian tubes, oral cavity and skin have also been reported.[1] Very few cases of angiomyolipoma with vulval involvement have been reported.

CASE REPORT

The 26 years old female presented with growth in the vulva for 2-3 years, gradually increasing in size. It is associated with pain in the vulval region at the time of menstrual cycle. Obstetric history P1 A0 L1. Patient has menorrhagia for 4-5 years (cycle of 28 days, duration 15-20 days). Local examination shows swelling approximately 10×5 cm on the right side of groin extending from lower 1/3rd of labia majora anterior to anal canal. Ultrasonography revealed 9.4×4.8 cm heterogenous lesion with focal cystic areas interspersed within solid areas with focal minimal vascularity d/d- 1) Lipoma with secondary degeneration 2) complicated Bartholin cyst/ abscess. FNAC done from the lesion was suggestive of benign cystic lesion.

Grossly the tumor was pedunculated, bosselated and firm measuring 9×9×4.5 cm. the stalk measured 3.5 cm. cut surface was greyish to yellow in color with thick fibrous septae traversing through it and multiple cysts with largest measuring 1 cm in diameter. No necrosis or hemorrhage noted.

Microscopic examination showed encapsulated tumor showing predominantly mature adipose tissue fragments, numerous thick and thin-walled congested vessels of variable caliber and fascicles of smooth muscle cells radiating away from the blood vessels. Also, seen are cystic spaces lined by flattened epithelium with areas of hemorrhage, infarct, chronic inflammatory cell infiltrate also noted. Periphery of the lesion shows normal skene gland with focal squamous metaplastic change. No mitosis, necrosis or hemorrhage noted.

Impression: Morphology suggestive of angiomyolipoma-vulva.

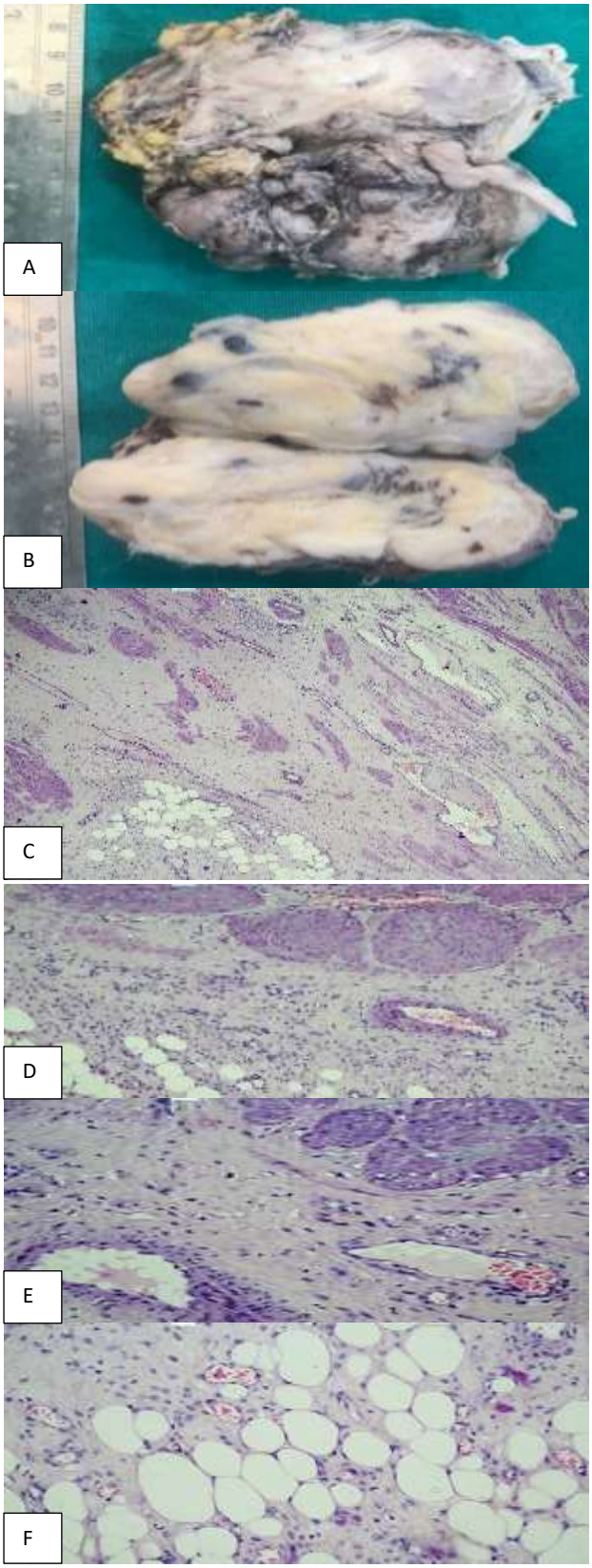

Figure 1 (A-F): Pedunculated mass, cut surface was greyish to yellow in color with areas of congestion and haemorrhage, fascicles of smooth muscle bundles, variably sized thick-walled blood vessels and sheets of mature adipose tissue, thick-walled blood vessels and high power of mature adipocytes.

DISCUSSION

Angiomyolipoma is an uncommon and benign mesenchymal tumor. It was first described by Martignoni in kidney 1951.[2] Kidney is the most common site but rarely angiomyolipoma can also be found in liver, mediastinum, fallopian tubes, oral cavity, and skin. Renal Angiomyolipoma is common in middle age females with median age of 46 years.[3] The tumor is mostly sporadic in origin however, 20% cases are associated with Tuberous sclerosis.[4-6] Sporadic cases are unilateral, single, and large while the tumor associated with tuberous sclerosis are bilateral, multiple, and small. Grossly the tumor is unencapsulated, circumscribed, yellow to grayish white in color depending upon the major component of the tumor with extensive areas of hemorrhage. Microscopically, angiomyolipoma is unencapsulated, circumscribed and is composed of smooth muscle, fat, and thick-walled blood vessels in varying proportion. Features such as >70% atypical epithelioid cells, ≥2 mitosis per 10 HPF or atypical mitosis and necrosis are associated with malignant behavior.[2,7-9] On immunohistochemistry, the smooth muscle component in the tumor shows positivity with Cathespin K smooth muscle marker (vimentin, actin and desmin) and melanoma associated markers (HMB-45, Melan-A and microphthalmia transcription factor).[5,11] Extra renal angiomyolipoma differs from renal angiomyolipoma as it occurs in older age group and is HMB negative, however those that are associated with tuberous sclerosis are positive for HMB-45.[3,12] Treatment includes resection, embolization, ablative therapy or mTOR inhibitors with close follow up especially in renal angiomyolipoma.

CONCLUSION

Angiomyolipoma at vulva location is very rare even among the extra-renal angiomyolipoma and should be kept as differential when dealing with vulval lesion.

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