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Intrinsic Features of Vulval Angioleiomyoma: Case Report and Mini Review.

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Abstract

Vulvar angiomyolipoma is a rare, benign mixed mesenchymal pathology, composed of smooth muscle, vascular and mature adipose tissue in varying proportions. It may stand for an incidental diagnosis; and the treatment can be conservative or surgical by enucleation. The definitive diagnosis is made by anatomopathological and immunohistochemical examination. The authors report a case of angiomyolipoma located in the labrum vulvar major and conduct a mini review.

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Keywords: Angiomyolipoma; Mesenchymal neoplasm; Gynecological Neoplasm.

Introduction:

The diseases classified as vaginal tumors are divided into epithelial, mesenchymal, melanocytic, germ cell, lymphoid and tumors with secondary origin.

Vulvar angiomyolipomas are rare benign mesenchymal lesions, composed of smooth muscle tissue, vascular and adipocytes in varying proportions. They are usually insidious, asymptomatic, but can cause soreness and cosmetic changes depending on size1. Their size varies from 0.4 cm to 20 cm, and bigger lesions may cause vaginal bleeding, dyspareunia, abdominal or back pain and urinary symptoms. The mean age of presentation is around 40 years old2.

Leading risk factors associated with the development of angiomyolipomas include tuberous sclerosis, which is a genetic disease that is correlated with the growth of other tumors, such as astrocytoma, rhabdomyomas, oncocytomas, and angiofibroma. In addition, they may be associated with Von Recklinghausen disease (neurofibromatosis type I), von Hippel-Lindau disease, Sturge-Weber syndrome, and autosomal dominant polycystic kidney disease. Some reports suggested that events which cause a hormonal imbalance in the body, such as puberty or pregnancy in women, may be a risk factor 1,2,3.

The most common site of angiomyolipomas is in the kidneys, followed by the liver. Also, can rarely affect the fallopian tubes, ovaries, spermatic cord, and colon. A spontaneous, isolated angiomyolipoma accounts for 80% of cases, on the other hand the associated with tuberous sclerosis handles 20% of cases1,3.

For the correct diagnosis of vulvar angiomyolipoma it is fundamental to consider, and exclude premalignant and malignant mesenchymal pathologies, which will help to define the biological behavior, treatment, and prognosis.
Considering the above we present a clinical case of vulvar angiomyolipoma.

Clinical Case:

A 41-year-old patient reports the presence of a nodule in the left labia majora, for past two years with complaints of mild local discomfort, without other symptoms. She presented irregular menstrual cycles and denied urinary changes. During the examination of the vulva and perineum, a nodule measuring approximately 3.0 cm x 2.0 cm was seen in the left labia majora, characterized as mobile, painless, with fibroelastic consistency.

Magnetic resonance imaging described the presence of a solid nodule with hypo signal on T1 and T2 weights, measuring 2.5 cm, with important enhancement by paramagnetic contrast medium, found in the vaginal introitus, next to the left lip, indeterminate.

Surgical excision of the nodule was performed by the assistant team.
The macroscopic examination showed a nodular fragment of light brown tissue, elastic, partially encapsulated, measuring 2.8 cm x 2.2 cm x 1.7 cm. On cuts, it was grayish and shiny.

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Histological analysis and diagnosis: Proliferation of smooth muscle cells, fusiform, vascular and adypocits without atypia (figure1).

An immunohistochemical study was suggested for differential diagnosis and confirmation of a benign lesion.

Immunohistochemical study showed: Desmin (D33) positive; SMA (1A4) positive; Caldesmon (h-CD) positive; Ki67 (30-9) positive 9%; S100 (P) positive in adipocytes; CD34 (QBEnd-10) positive in blood vessels; ERG (EPR3864) positive in blood vessels and HMB45: negative. (Figures 2, 3 and 4).

The histological picture complemented by the immunohistochemical study are compatible with vulvar angiomyolipoma.

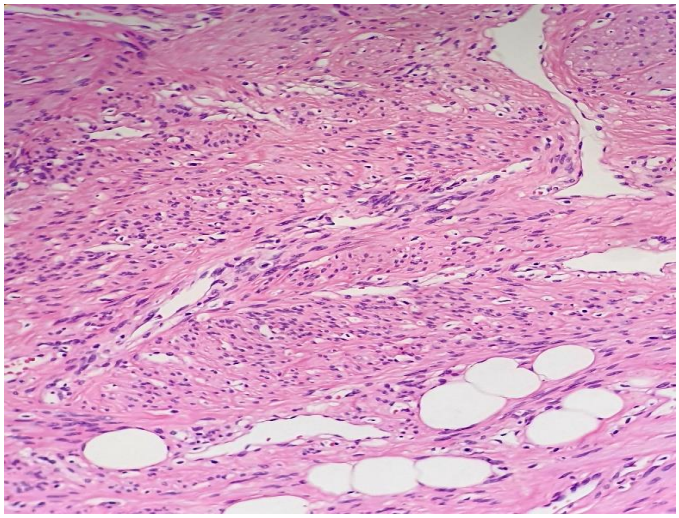

Figure 1: H&E. Proliferation of muscle cells, vessels, and adipocytes

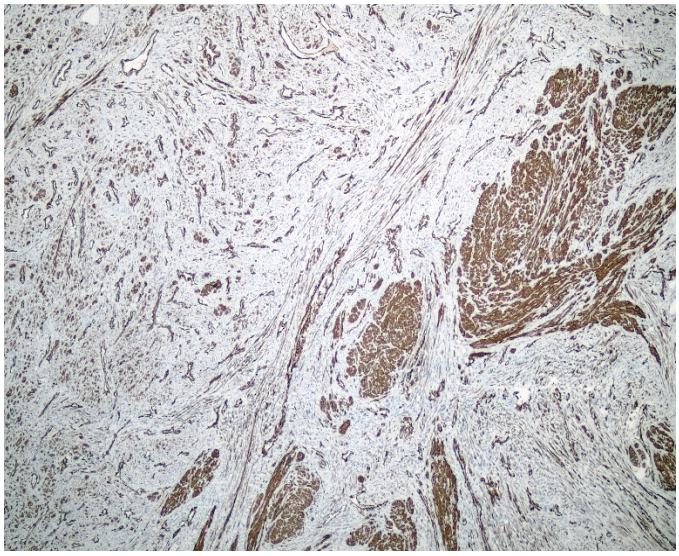

Figure 2: Smooth muscle actin SMA.Positive for smooth Muscle.

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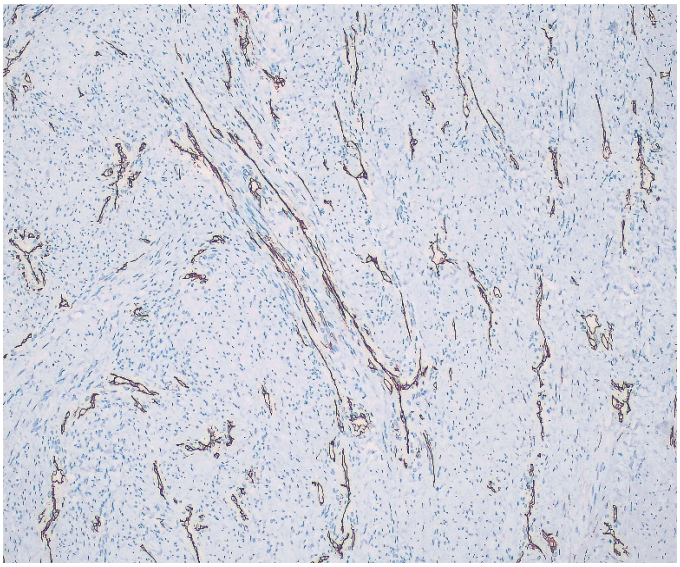

Figure 3: CD34. Positive for blood vessel

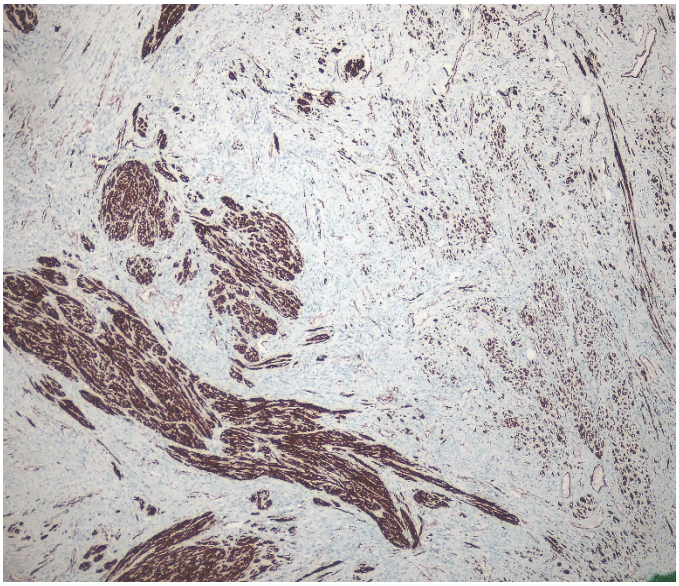

Figure5: Desmin. Positive for smooth muscle.

Discussion:

Angiomyolipoma is a benign mesenchymal neoplasm of preferential renal location. In the vulva and/or vagina, it is an extremely rare entity. Research set forth that these tumors arise from cells called perivascular epithelioid cells (PEC), which surround blood vessels. Thus, angiomyolipoma is a kind of tumor known as PEComa4. The exact etiology and mechanism of development of vulvar/vaginal angiomyolipoma, in most cases, are unknown4.

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Macroscopically, they manifest as an incidental tumor found on physical examination or incidentally during imaging tests. They are usually asymptomatic, but patients may experience discomfort and local pain. If they are large, with a vaginal location, they can cause dyspareunia, vaginal bleeding, local pain, pelvic and lumbar pain 2,5.

Histologically, they are triphasic, consisting of smooth muscle, adipocytes, and blood vessels. The smooth muscle part can be fusiform, intermediate, epithelioid, or pleomorphic. Nuclear pleomorphism, cell atypia, and mitosis can occur. The vascular part includes 4 types of neoplastic blood vessels (cellular, hemangiopericytic, glomeruloid, aneurysmal) as well as non-neoplastic sinusoidal type and collagenous blood vessels; large, dilated vessels may be present at the tumor/non-tumor interface 5.

The medical diagnosis uses radiological exams, such as ultrasound, computed tomography, or magnetic resonance, which takes part in the preoperative characterization, confirming the location and excluding the presence of lymphadenopathy and ascites. For a definitive diagnosis, the histological analysis of the lesion is necessary6.

The treatment of vulvar angiomyolipoma depends on the size and symptoms of the patient. It can be conservative or making use of surgical procedures such as enucleation in cases that the absence of clinical features does not suggest a malignant behavior. The complications of the surgery involve the non-desirable damage to muscles, nerves and blood vessels and post-surgical infection at the wound site6.

Former studies have not conclusively proved that angiomyolipoma can turn into malignant process, but some cases of sarcoma are known to be the originally an angiomyolipoma. Tumors with characteristics of hemorrhage, necrosis, local invasion, and high mitotic activity are likely to have a poor outcome7,8. Some cases of local recurrence have also been described, especially when in association with larger first lesions9.

Extrarenal angiomyolipomas are generally positive for smooth muscle actin and negative for HmB45 and have no association with tuberous sclerosis10. In our clinical case, SMA was positive and HmB was negative. Extrarenal angiomyolipomas are generally positive for smooth muscle actin and negative for HmB45 and have no association with tuberous sclerosis10. In our clinical case, SMA was positive and HmB was negative. The dimensions of lesion, absence of necrosis low rate of cell proliferation confirmed benign behavior.

Conclusion:

Despite the rarity of vulvar and vaginal angiomyolipomas, it is necessary to consider this entity when analyzing lesions in the topography of the labia majora and around the Bartholin's gland without inflammatory signs. The recognition of this pattern and the knowledge of the differential diagnosis allow the pathologist to suggest it precociously as a diagnostic hypothesis and helps to guide the treatment.

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The definitive diagnosis is multidisciplinary, the clinical, imaging, anatomopathological and immunohistochemical must be combined in benefit of the patient.

Interest Conflicts: The authors declare that they have no conflict of interest.

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