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A RARE CASE OF A GIANT VULVAL MASS - CASE REPORT OF VULVAL CELLULAR ANGIOFIBROMA

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ABSTRACT

Background- Cellular Angiofibroma is rare benign tumor of mesenchymal origin. They have a predilection to the genitourinary region and are present in both sexes (Male - scrotum, Female - vulva). **Case presentation-** A 45-year-old female presented with complaints of painless swelling in the Right vulva which has progressively increased in size over the past 4 years. On examination, a large solid pedunculated tumor of 20 x 15 cm was noted arising from the right labia. Ultrasonography showed a well-demarcated, superficial, solid tumor located on the right vulva. The lesion was excised under all aseptic precautions with a rim of normal tissue. The resected mass was 20 x 15 cm in size and weighed 2750 grams. Histological examination shows fibroadipose tissue with the presence of angiomatoid vascular proliferation with areas of myxoma type. No signs of malignancy. The patient is on regular follow-up since the surgery and there have been no signs of recurrence for 6 years. **Conclusion-** Vulval cellular angiofibroma are very rare and knowledge about these is very important for a gynecologist. It should be considered as a differential diagnosis for a painless swelling in the vulva. Every person should have an individualized approach and a multidisciplinary approach is required for these complex cases.

KEYWORDS : Vulval cellular angiofibroma, Vulval mass, excision

INTRODUCTION

Vulval angiofibroma is a rare benign tumor characterized by the proliferation of fibrous and vascular elements within the vulvar region. While typically presenting as a solitary lesion, it can occasionally manifest as multiple lesions or involve other anatomical sites. Here, we present a case of vulval angiofibroma in a 45-year-old woman, discussing the clinical presentation, diagnostic challenges, and management strategies employed to achieve successful outcomes.

Case Report:

A 45-year-old otherwise healthy woman presented with a gradually enlarging painless mass in the vulvar region which has progressively increased in size over the past 4 years. The physical examination revealed a well-defined, pedunculated, firm, non-tender subcutaneous mass measuring approximately 20 x 15 cm in diameter on the right labia majora. No significant abnormalities were observed in the rest of the genital examination. The patient reported no urinary or sexual dysfunction. A detailed medical and family history did not reveal any predisposing factors or significant past medical conditions. Ultrasonography has shown a solid soft tissue tumor that was well-demarcated. The lesion was excised under all aseptic precautions with a rim of normal tissue. The resected mass was 20 x 15 cm in size and weighed 2750 grams. Histological examination shows fibroadipose tissue with the presence of angiomatoid vascular proliferation with areas of myxoma type. No signs of malignancy. The patient is on regular follow-up since the surgery and there have been no signs of recurrence for 4 years.

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DISCUSSION

Vulval angiofibroma is a distinctive and rare benign neoplasm primarily affecting the vulvar region. It is characterized by the proliferation of fibrous and vascular elements within the dermis and subcutaneous tissue. Cellular Angiofibroma is a rare benign tumor of mesenchymal origin. Vulval angiofibroma was initially thought to be an uncommon condition, but in recent years it has become more widely known, thanks in large part to improvements in histological analysis and a greater emphasis on a thorough inspection of vulvar lesions. Although only one lesion is normally present, reports have also been made of several lesions or the involvement of other anatomical areas. They have a predilection to the genitourinary region and are present in both sexes (Male - scrotum, Female - vulva). To achieve an accurate diagnosis and the proper course of therapy, it is essential to comprehend the clinical presentation, diagnostic problems, and management techniques.

Genetics

The majority of vulval angiofibroma cases are sporadic and manifest as solitary lesions. The RB1 gene mutations have not been identified as a significant factor in the development of this specific illness, despite the fact that genetic changes have been examined in a number of tumors, including vulval angiofibroma. In instances linked to the Tuberous sclerosis complex (TSC), the genetic component of vulval angiofibroma is more well-established. We still don't fully understand the precise molecular processes that lead to vulval angiofibroma formation. The mammalian target of the rapamycin (mTOR) pathway and the vascular endothelial growth factor (VEGF) pathway are two signaling pathways linked to cell proliferation and angiogenesis that may be dysregulated. However, the precise genetic modifications or mutations linked to vulval angiofibroma are not fully understood.

Additional study is required to properly understand the genetic variables causing sporadic cases. Modern genetic and molecular research holds promise for more accurate diagnosis, tailored treatments, and better management of this uncommon vulvar tumor.

Clinical Presentation

Although cases have been observed in individuals of various age groups, vulval angiofibroma most frequently affects women during their reproductive years. It often presents clinically as a distinct, single, firm, or rubbery, subcutaneous nodule. The lesion is typically slow-growing and painless, although different diameters have been noted in the literature. The labia majora and surrounding vulvar area are the most often implicated sites. However, extragenital areas such as the abdominal wall, groin, and thigh may also be impacted. Even while vulval angiofibroma is typically sporadic, there

have been a few recorded cases where it has been linked to Tuberous sclerosis complex (TSC).

Diagnostic Challenges

The diagnosis of vulval angiofibroma might be difficult because of its rarity and the fact that it has clinical and histological characteristics with other vulvar lesions. Clinically, it may resemble a number of illnesses, including aggressive angiomyxoma, fibroma, and angiomy ofibroblastoma.

To assess the size and features of the vulval angiofibroma, imaging modalities such as ultrasound, magnetic resonance imaging (MRI), and computed tomography (CT) scans may be used. In order to help with surgical planning and rule out the presence of any invasive or metastatic lesions, these imaging modalities provide extensive information about the size, depth, and involvement of nearby tissues.

Histopathological analysis of a biopsy specimen continues to be the gold standard for vulval angiofibroma diagnosis confirmation. The diagnosis of vulval angiofibroma is confirmed by the presence of fibrous and vascular components in the tissue sample. To distinguish vulval angiofibroma from other mimicking lesions, the immuno histochemical investigation may also be used.

Genetic Testing:

In cases where there is a suspicion of an underlying genetic syndrome, such as tuberous sclerosis complex (TSC), genetic testing may be conducted.

Management Strategies

A multidisciplinary strategy involving gynecologists, dermatologists, and pathologists is the most effective way to treat vulval angiofibroma. Complete surgical excision is the main course of treatment due to the condition's benign nature. However, the extent of the operation may change based on the size, location, and patient preferences of the lesion. There have been successful uses of a number of surgical methods, including simple excision, local excision, and extensive local excision. To keep an eye out for recurrence or the emergence of numerous lesions, especially in individuals with tuberous sclerosis complex, long-term follow-up is crucial.

CONCLUSIONS

The benign vulval tumor known as vulval angiofibroma is rare but gaining in popularity. For proper therapy and to minimize unnecessary procedures, an early and precise diagnosis is essential. To distinguish it from other vulvar lesions, medical professionals must be aware of its clinical presentation and histological characteristics. In order to improve patient outcomes and quality of life, the above research seeks to increase the body of information on vulval angiofibroma and aid physicians in making knowledgeable decisions about diagnosis and care.

Figure 1 - Right vulval mass

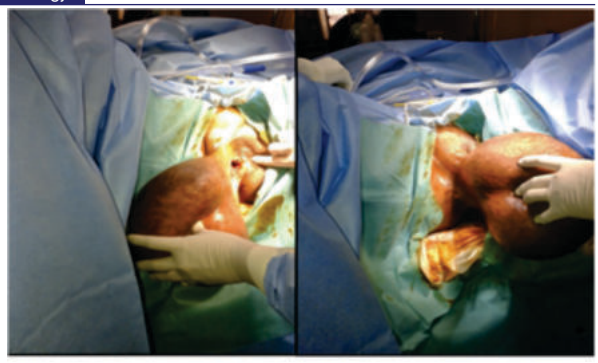

Figure 2 - Medial and Lateral view of the vulval mass

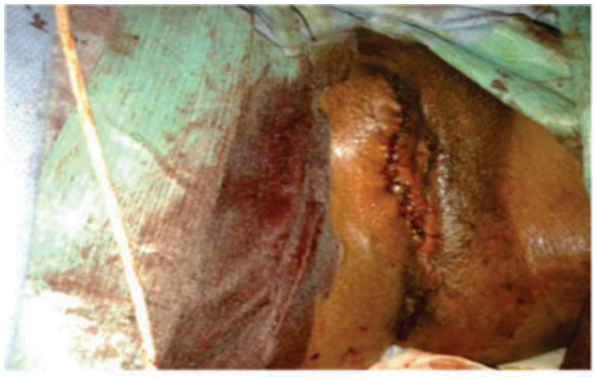

Figure 3 - Vulva after excision

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