

An Uncommon Case of Vulvar Epithelioid Leiomyoma - Case Report

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Abstract: Genital leiomyomas are rare tumours that can often be misdiagnosed as Bartholin cyst. Epithelioid Leiomyoma of vulva is distinctly uncommon when compared with uterine leiomyoma. We report a case of 18 year old girl who had a nodulocystic swelling over left vulva suggestive of Bartholin Cyst. A local excision procedure was performed. Microscopy and IHC findings favoured a diagnosis of Epithelioid Leiomyoma of Vulva. The macroscopic features of cystic lesion of vulva made it difficult to differentiate between leiomyoma and Bartholin cyst. Therefore, histopathologic examination is often recommended for such cases.

Keywords: Leiomyoma, Bartholin cyst

1. Introduction

Smooth muscle tumors of the vulva are very uncommon entities and this rarity could lead to misdiagnose these tumors as other benign condition including Bartholin cyst or abscess. Considered to originate from a variety of tissues including smooth muscle in erectile tissues, blood vessel walls, the round ligament, the dartos muscle, or the erectorpili muscle.

Histologically, smooth muscle tumors of vulva show various morphological features of spindled, epithelioid and myxohyaline patterns.

Due to rarity and morphological complexity, it is difficult to attain accurate diagnosis and predict the prognosis.

2. Case Details

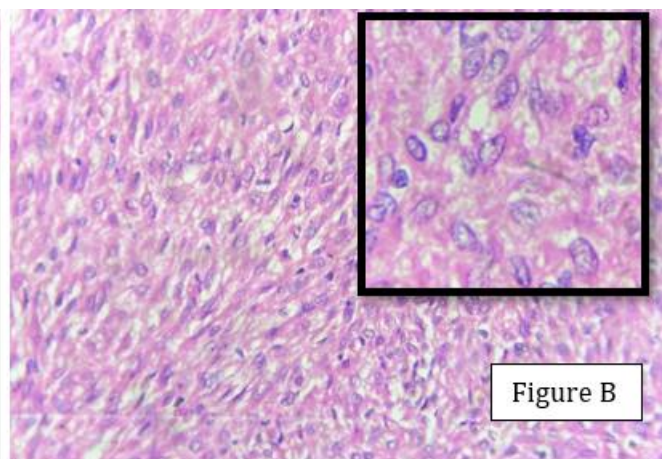
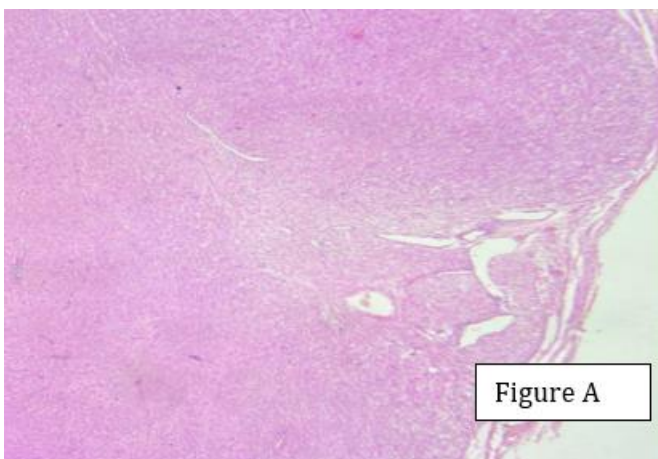
18 year old girl presented with history of painless swelling over left vulva for about 4 months duration. Swelling gradually increased in size. Clinical diagnosis was made as Bartholin cyst. Hence initially an attempt for aspiartion was done. Followed by which she underwent local excision

procedure and the sample was sent for histopathological analysis.

We received multiple gray-white tissue bits aggregate measuring 4.5 x4x1.5cm. Cut section was homogenous gray-white glistening. Microscopy showed a neoplasm composed of cells arranged in sheets and bundles. Individual cells are plump spindly to epithelioid with moderate eosinophilic cytoplasm and the nucleus is plump, ovoid and bland looking. . Mitotic activity noted was <4 /10hpf. No areas of necrosis noted. No normal tissue was seen beyond the Lesion. Immunohistochemistry studies showed Desmin, H-caldesmon and SMA positivity. ER, S-100, CD68 and CD34 were negative. Ki67 index was <5%. With the histopathological morphology and IHC findings, final diagnosis suggestive of Epithelioid Leiomyoma of Vulva was given.

Advised to closely follow up the patient as the completion of excision cannot be assessed and there was a high chance of recurrence.

Microscopy & Immunohistochemistry:



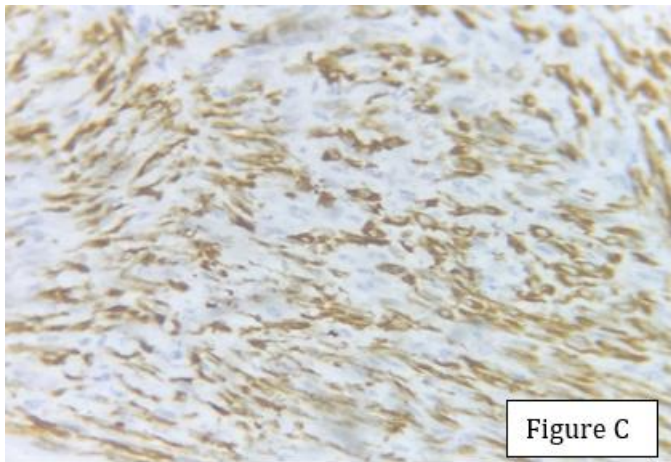


Figure C

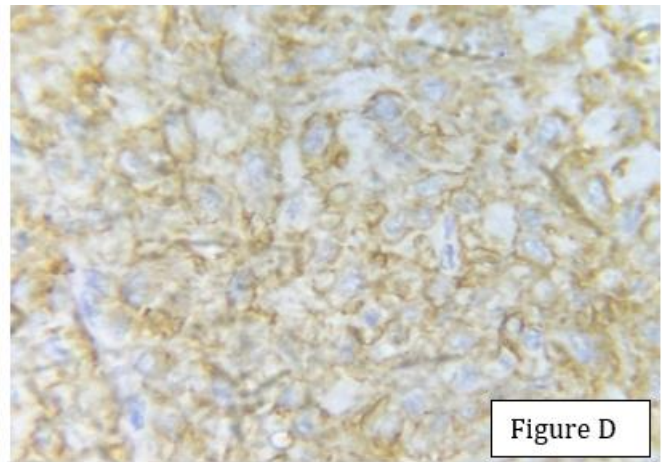


Figure D

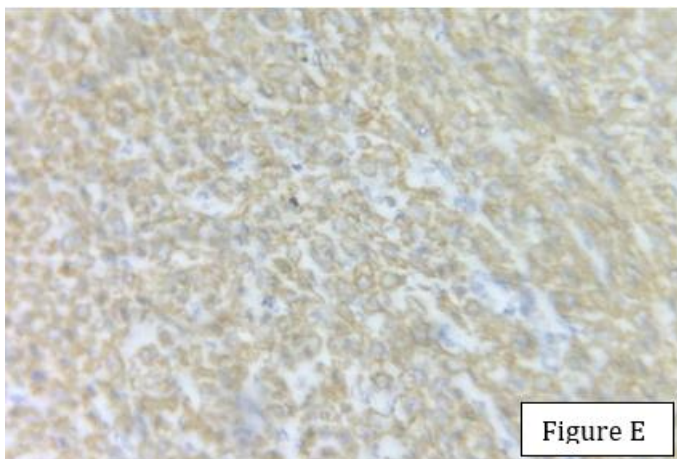


Figure E

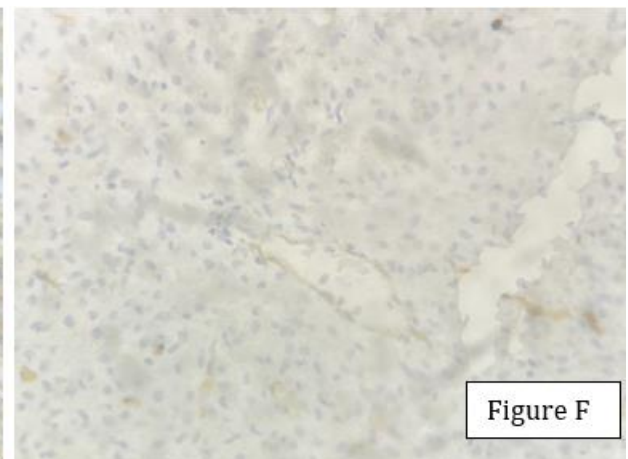


Figure F

Figures: Figures A&B showing H&E pictures of the neoplasm with inset highlighting the epithelioid morphology of the neoplastic cells. Figure C to E shows the IHC positivity for Desmin, H-Caldesmon and SMA respectively. Figure F shows the Ki67 index.

3. Discussion

Vulvar leiomyomas are infrequent tumors accounting for 4.2% of cutaneous leiomyomas and 0.07% of all vulvar tumors.

Among them, vulvar leiomyomas comprising of epithelioid cells as a dominant histologic component are of very rare occurrence.

Vulvar epithelioid leiomyomas were first introduced in 1979 by Tavassoli and Norris.

The age when vulvar epithelioid leiomyomas tend to occur range from 19 to 47 years with a mean age of 31.7 years. The lower genital tract leiomyomas are usually less than 5cm and are slow growing painless mass and clinically suspected as cystic Lesion.

Nielsen et al proposed an expanded and specific criterion paying attention to four characteristics including a large tumor size (≥ 5 cm), an infiltrative margin, a high mitotic activity (≥ 5 mitosis/10HPFs) and moderate to severe nuclear atypia. Researchers then categorized the tumors with 3 or 4 features as leiomyosarcoma, the tumors with 2 features as atypical leiomyoma and the tumors with none or one as leiomyoma.

4. Conclusion

In conclusion, we report a very uncommon case of vulvar smooth muscle tumor with a predominant epithelioid cell differentiation and our case shows the typical histologic features of vulvar epithelioid leiomyoma including absence of atypia and reduced ($< 4/10$ hpf) mitosis

It is necessary to be aware of epithelioid morphology of vulvar smooth muscle tumors for presenting an appropriate diagnostic criterion to predict the prognosis.

References

- [1] Tavassoli FA, Norris HJ. Smooth muscle tumors of the vulva. *Obstet Gynecol* 1979; 53:213–7.
- [2] Nielsen GP, Rosenberg AE, Koerner FC, et al. Smooth-muscle tumors of the vulva. A clinicopathological study of 25 cases and review of the literature. *Am J Surg Pathol* 1996; 20:779–93.
- [3] WHO Classification of Tumors 5th edition-Soft Tissues and Bone Tumors
- [4] WHO Classification of Tumors 5th edition-Female Genital tumors
- [5] Zhou J, Ha BK, Schubeck D, et al. Myxoid epithelioid leiomyoma of the vulva: a case report. *Gynecol Oncol* 2002;103;342-5