

CASE REPORT

Giant labial fibroepithelial stromal polyp

Michelle CHAN Mei Fung *MBBS, FRCPath*, YONG Tze Tien* *MBBS, MRCOG* and Kesavan SITTAMPALAM *MBBS, FRCPA*

*Department of Pathology and *Department of Obstetrics & Gynaecology, Singapore General Hospital*

Abstract

We report an 18-year-old girl with a four-year history of a slow-growing labial mass with a sudden increase in size in the last year. Examination revealed a large fleshy 20 cm perineal mass centering on the left labia majora and attached to it by a 1cm pedicle. It was associated with pain, ulceration and discharge. The lesion was excised via diathermy at the base of the stalk. The excised specimen weighed 1.112kg and measured 20.5 x 17 x 5cm. The lesion showed a solid, soft whitish, cut surface. Histology revealed a hypocellular tumour with focally oedematous fibrous stroma in which were scattered large and small blood vessels, mast cells and other chronic inflammatory cells. True myxoid matrix was not observed. The stromal cells had a spindle to stellate morphology. There was no significant cytological atypia, mitotic activity or necrosis. The tumour cells were negative for SMA, desmin, CD34, S100 protein, EMA and PR. The diagnosis was clinically and histologically challenging because various vulvovaginal soft tissue tumours often have overlapping clinicopathological features. However, based on strict histological criteria and the absence of worrisome cytological features, a diagnosis of fibroepithelial stromal polyp was rendered despite the unusual size. A review of the literature shows that whilst vulvovaginal fibroepithelial stromal polyps are well described, giant variants are rare. Awareness of the extraordinary size that can be attained by such polyps can facilitate swift clinical and histological diagnosis.

Keywords: Vulvovaginal soft tissue tumour, fibroepithelial stromal polyp

INTRODUCTION

A myriad of mesenchymal tumours can occur in the vulvovaginal region, ranging from benign entities to locally aggressive and malignant tumours. Their clinical and histological features can be overlapping and immunohistochemistry is of limited help. These vulvovaginal tumours include fibroepithelial stromal polyps, aggressive (deep)angiomyxoma, superficial angiomyxoma, angiomyofibroma, cellular angiofibroma and (prepubertal) vulval fibroma. Diagnosis often relies on the identification of subtle morphological differences between these different tumours and clinical correlation is paramount. Arriving at the correct diagnosis becomes problematic when these lesions present with unusual clinical features, as illustrated by our case report. The importance of making the correct diagnosis must be emphasized as these various vulvovaginal tumours have very different biological behavior.

CASE REPORT

An 18-year-old female presented with a four-year history of a labial mass that was initially slow-growing until the last year when there was a sudden increase in size. The mass was associated with pain, ulceration and discharge. On examination, there was a large fleshy mass that covered the entire perineum but centering on the left labia majora and attached to it by a 1cm pedicle. Bedside ultrasound showed a homogenous mass with fluid pockets but no bowel segments were found within. Due to the presence of a pedicle, a congenital lesion was in the clinical differential. However, the large size and the extent of the lesion raised the possibility of an aggressive or even malignant lesion. The patient subsequently underwent local excision of the lesion via diathermy at the base of the stalk.

The patient was well and discharged during her follow-up visit three weeks post-operatively.



FIG. 1. Macroscopic photograph showing large size and diathermised stalk of the labial mass.

To date, she has not presented to the hospital again.

Pathology

The excised specimen weighed 1.112kg and measured 20.5 x 17 x 5cm. The skin surface was focally ulcerated whilst the base of the specimen was cauterized and haemorrhagic, containing a 1cm area representing the stalk. Sectioning of the specimen revealed a solid, soft whitish, cut surface with one haemorrhagic cystic cavity up to 4cm. No necrosis was seen. Figure 1 illustrates the diathermised base of the lesion containing the stalk. Figure 2 shows the homogenous whitish

oedematous cut surface of the lesion.

The histological sections showed a hypocellular tumour with a focally oedematous fibrous stroma (Figure 3) in which were scattered large and small blood vessels, mast cells and other chronic inflammatory cells. True myxoid matrix was not observed. The stromal cells had a spindle to stellate morphology (Figure 4). A perivascular pattern was not seen. There was no significant cytological atypia, mitotic activity or necrosis. Epithelioid stromal cells and alternating cellularity was not seen. The tumour cells were negative for the immunostains SMA, desmin, CD34, S100 protein, EMA and PR. There was,

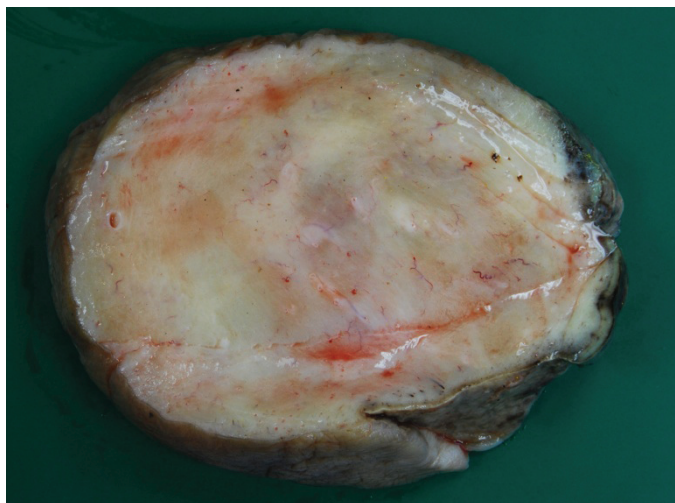


FIG. 2. Labial mass with whitish oedematous cut surface.

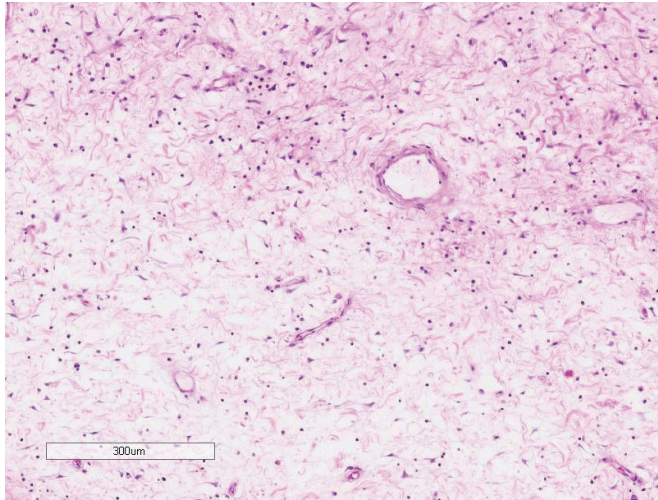


FIG. 3. Photomicrograph showing hypocellular tumour with oedematous fibrous stroma, scattered blood vessels and chronic inflammatory cells. H&E X40.

however, weak immunoreactivity for ER in the stromal cells. The overall morphology together with the pedunculated growth, was most in keeping with a fibroepithelial stromal polyp, despite the striking size.

DISCUSSION

Fibroepithelial stromal polyps are benign proliferations which are usually polypoid or pedunculated, and less than 5cm in size.¹ They are generally single lesions but can be multiple during pregnancy. They typically have a central fibrovascular core and contain stellate and multinucleated stromal cells which are best seen beneath the surface epithelium. True myxoid stroma is absent. Although

vulvovaginal fibroepithelial stromal polyps are well documented, a giant variant such as the one we report here is rather rare. To our knowledge, our case is the largest fibroepithelial stromal polyp compared to others reported in the literature. Other reports which had relatively large fibroepithelial stromal polyps include one case of a 10cm lesion in a young girl with a history of congenital lymphoedema,² and one case of a 15cm lesion occurring in association with vulval psoriasis.³ Another case report documented a 15cm giant fibroepithelial stromal polyp and described the sonographic features as an adjunct to the diagnosis of fibroepithelial stromal polyp in large vulvovaginal lesions.⁴

Other vulvovaginal soft tissue lesions can

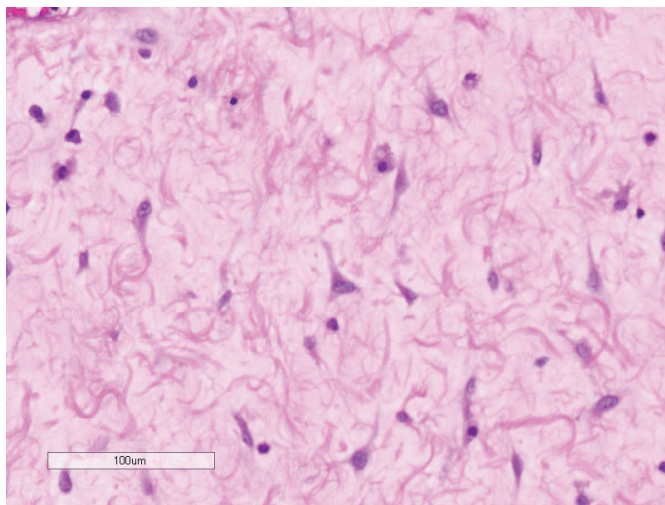


FIG. 4. Spindle to stellate stromal cells. H&E x 200.

share similar features to fibroepithelial stromal polyps. One important differential in our case would be the aggressive (deep) angiomyxoma, due to the very large size at presentation. Histologically, aggressive angiomyxomas have true myxoid stroma and tend to have medium to large thick-walled blood vessels regularly distributed throughout the lesion as opposed to the central fibrovascular core in fibroepithelial stromal polyps. Myoid fibres are often seen surrounding the blood vessels, a feature which is absent in the lesion we described. Challenge arises when fibroepithelial stromal polyps undergo torsion and develop extensive oedema which can mimic the real myxoid stroma in aggressive angiomyxomas, as demonstrated by the fibromyxoid-like stroma in our case of giant fibroepithelial stromal polyp. However, the two entities can still be distinguished because aggressive angiomyxomas have more infiltrative borders and are more deeply located, whereas fibroepithelial stromal polyps are circumscribed and occur in the superficial subepithelial stroma. Aggressive angiomyxomas are prone to local destructive recurrence, requiring adequate excision margins and long-term follow-up, whereas fibroepithelial polyps can be treated by local excision.

Other vulvovaginal lesions that can mimic fibroepithelial stromal polyps because of their well-circumscribed margins are angiomyofibroblastomas, superficial angiomyxomas and cellular angiofibromas. Angiomyofibroblastomas are benign tumours which can be distinguished from fibroepithelial stromal polyps histologically because they tend to have alternating cellularity with clusters of epithelioid stromal cells surrounding small capillaries. They are cured by local excision. Vulvovaginal superficial angiomyxomas are also different histologically because they have a multinodular pattern of growth and abundant true myxoid stroma. Although benign, they have a recurrence rate exceeding 30% therefore requiring clear margins.¹ Cellular angiofibromas are more cellular spindle cell lesions with hyalinised blood vessels. They are benign and are managed by complete local excision. Another consideration in vulvovaginal lesions presenting in young girls is the more recently described prepubertal vulval fibromas. However, these usually have ill-defined margins involving submucosa or subcutis.⁵ In addition, the spindle cells in these tumours are typically positive for CD34.⁶

In summary, the diagnosis of vulvovaginal soft tissue lesions requires careful histological and clinical correlation. Awareness of the giant variants of fibroepithelial stromal polyps can help clinicians and pathologists at reaching the correct diagnosis.

REFERENCES

1. Kurman RJ, Ellenson LH, Ronnett BM, editors. Blaustein's Pathology of the Female Genital Tract. 6th ed. US: Springer; 2011.
2. Orosz Z, Lehoczky O, Szoke J, Pulay T. Recurrent giant fibroepithelial stromal polyp of the vulva associated with congenital lymphedema. *Gynecol Oncol.* 2005; 98(1): 168-71.
3. Dane C, Dane B, Cetin A, Erginbas M, Tatar Z. Association of psoriasis and vulval fibroepithelial polyp: first reported case. *Am J Clin Dermatol.* 2008; 9(5): 333-5.
4. Bozgeyik Z, Kocakoc E, Koc M, Ferda Dagli A. Giant fibroepithelial stromal polyp of the vulva: extended field-of-view ultrasound and computed tomographic findings. *Ultrasound Obstet Gynecol.* 2007; 30(5): 791-2.
5. Iwasa Y, Fletcher CD. Distinctive prepubertal vulval fibroma: a hitherto unrecognized mesenchymal tumor of prepubertal girls: analysis of 11 cases. *Am J Surg Pathol.* 2004; 28(12): 1601-8.
6. Nucci MR. Mesenchymal lesions of the lower genital tract. *Surg Pathol Clin.* 2009; 2(4): 603-23.