CASE REPORT

Vulvovaginal Angiomyofibroblastoma: An Uncommon Benign Mesenchymal Tumour

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ABSTRACT

The female genital tract angiomyofibroblastoma (AMFB) is a rare benign mesenchymal tumour that mostly affects premenopausal women. Although it is most commonly seen in the vulvovaginal region, it has also been documented in the inguinosacral region, pelvis, and retroperitoneum (1,2). Patients usually present with indolent, painless swelling in the vulvovaginal region, or a pedunculated lesion on rare occasions. Those that appeared in the vulva were frequently misdiagnosed as Bartholin cysts. Leiomyoma is a differential in the vaginal area. AMFB must be recognised from aggressive angiomyxoma (AA) due to the latter's potential for recurrence with typical infiltrative boundaries. We discuss a case of a 46-year-old woman who had a slow-growing paravaginal swelling incidentally discovered during a total hysterectomy for leiomyoma five years prior.
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INTRODUCTION

Angiomyofibroblastoma is a benign soft tissue tumour that affects reproductive-aged women's lower genital tract, mainly involving the vulva and less frequently the vagina. Other than these sites, rare incidences in fallopian tube, uterine cervix, urinary bladder, and inguinoscrotal in men have been documented in the literature (1-4). Although its histogenesis is unknown, it is thought to be derived from the subepithelial stroma or perivascular stem cells (3,5). The tumour is normally small, less than 5cm in diameter, however past reports have described tumours as large as 23cm (1,4). On clinical examination, it could be mistaken for a cyst. It often manifested clinically as an indolent, painless swelling that sometimes resulted in dyspareunia or vaginal discharge (1,4,5). It can also present as a pedunculated mass that may prolapse (3,4).

The tumour is well-circumscribed macroscopically, with a soft to firm grey-white to tan cut surface (1,2). Due to the limited clinical and radiological characteristics of AMFB, a precise preoperative diagnosis of it is frequently difficult. Aggressive angiomyxoma (AA), cellular angiofibroma (CAF), myofibroblastoma (MFB), prepubertal vulval fibroma (VF), fibroepithelial stromal polyp (FP) and leiomyma are among the possibilities in the differential diagnosis (1-4). It is vital to differentiate between AMFB and AA because both tumours are mesenchymal tumours occurring at the same site with similar immunophenotype but behave quite differently. With infiltrative margins, AA tends to recur (1-5).

CASE REPORT

A 46-year-old para 3 female came in with a 5-year history of painless paravaginal swelling. She denied any associated weight reduction, appetite loss, or changes in bowel movements or bladder function. The swelling was identified during a total abdominal hysterectomy for leiomyoma 5 years prior. Subsequently, the patient was monitored on an annual basis without any active intervention. Over the course of five years, the swelling gradually increased in size. There was no associated vaginal discharge or skin changes. She did not complain of dyspareunia. A fluctuant cystic-like lesion with restricted movement was discovered at the pouch of Douglas during a pelvic examination. The cervix was unremarkable. A large complex cystic tumour measuring 13x10x9cm was noted in the pelvic cavity by magnetic resonance imaging (MRI). There was an area suspicious of an aggressive focus seen as papillary lobulated enhancing solid component. Although it was completely encapsulated, the lateral borders appeared to be adherent to the pelvic and iliopsoas muscles. There

was no evidence of lower paraortic lymphadenopathy.

Vaginal mass excision, bilateral salpingoophorectomy, and omentectomy were performed on the patient. Dense adhesion with endometriotic spots were seen intraoperatively, and the mass was entrenched by the bladder anteriorly, sigmoid colon, and rectum posterolaterally. The mass was totally removed with intact capsule. The resected specimen measured 110x50x50mm on gross inspection. A cut section revealed two well-circumscribed masses with smooth greyish pink surfaces, each measuring 50x50x32mm and 90x42x30mm. There was no evidence of haemorrhage or necrosis.

The tumour was clearly separated from the surrounding soft tissue microscopically. The histology of both masses was comparable, with alternating hypocellular and hypercellular foci and conspicuous randomly dispersed vascular channels of variable diameter. Scattered clusters of spindle to ovoid cells were detected aggregating around thin walled blood vessels in the hypercellular zones (Fig. 1A, 1B). These cells display fine chromatin, inconspicuous nucleoli, and eosinophilic cytoplasm. No nuclear atypia, mitosis, or necrosis were observed.

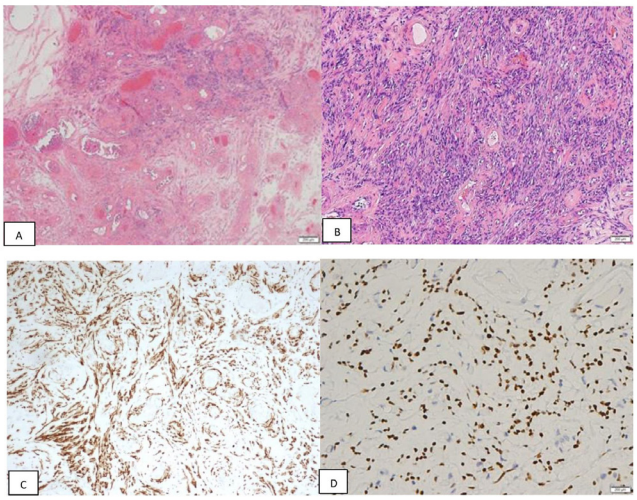

Figure 1: (A) Mesenchymal lesion with alternating hypocellular and hypercellular areas (40x magnification, H&E). (B) Bland spindle to ovoid cells clustered around small blood vessels (100x magnification: H&E stain). The spindle cells are immunoreactive for Desmin (C) and Oestrogen receptor (D).

A few mast cells were seen scattered within the mass. Dilated and thrombosed vascular channels were also identified in occasional foci. The hypocellular areas were largely oedematous, with collagenous stroma lacking myxoid alterations. Strong and diffuse immunostaining for smooth muscle actin (SMA), desmin, progesterone receptor, and oestrogen receptor were identified in the spindle and ovoid cells (Fig. 1C, 1D), however CD34 immunohistochemistry was negative.

Thus, a diagnosis of benign mesenchymal neoplasm in favour of angiomyofibroblastoma was made. Patient was followed up post-surgery with no evidence of tumour recurrence.

DISCUSSION

The vulvovaginal area is home to various benign mesenchymal tumours. There have been a few benign mesenchymal tumours documented that arise or differentiate from the specialised stroma of the female vaginal tract. AMFB, CAF, AA, and MFB are among the lesions that fall under this category. These lesions share morphological and immunophenotypic characteristics to some extent (1-5). In addition, because its common clinical appearance was that of a cystic lesion, clinical impression would include Bartholin cyst, Gartner duct cyst, epidermal inclusion cyst, lipoma, and hemangioma. Due to its predisposition for the vulvovaginal region, AMFB is a diagnosis to consider in reproductive-age individuals who presents with vulvovaginal mass.

AMFB is a benign soft tissue tumour with myofibroblastic differentiation accompanied by blood vessels. It is defined by conspicuous thin-walled blood vessels surrounded by plump, ovoid to spindle stromal cells placed within a variable oedematous to collagenous matrix with alternating zones of cellularity. The stromal cells usually expressed oestrogen receptor (ER), progesterone receptor (PR), and desmin. Even though they are usually negative for actin, their response to this marker can be varied (1,3-5). AMFB infrequently has CD34 positivity (4). This tumour did not belong to the myofibroblastic family of angiofibroma (AF) and breast MFB, which are characterised by monoallelic loss of genetic material at FOXO1 loci (13q14) (2,4). A single report of sarcomatous transformation in angiomyofibroblastoma has been reported despite its indolent behaviour (2-3). Characteristically, the tumour shows prominent nuclear atypia and abnormal mitosis, or histologically resembled leiomyosarcoma or an undifferentiated sarcoma (4).

The most important differential diagnosis is AA, which has considerable risk for local invasion and a reported rate of recurrence of more than 33% (1,2). It is most commonly found in deep soft tissue and has infiltrative border (1,3,5). There is no alternating zones of cellularity in AA or tendency for perivascular spindle cells concentration. The stroma is usually myxoid, with collagen fibres aplenty. In contrast to capillary-sized capillaries seen in angiomyofibroblastoma, it contains larger, thicker-walled vessels. It was also reported that entrapped mucosal glands and nerve bundles were observed in AA (1). Both tumours may have a similar immunophenotypes, therefore the difference is mostly based on morphology (1,3).

Both AMFB and CAF are characterised by circumscribed superficial nodules composed of spindle cells and blood vessels. The less visible vascular component in CAF, which is usually made up of medium-sized vessels with

hyalinized walls, is its distinguishing feature. MFB of the mammary type is a benign spindle cell tumour that can occur anywhere in the genital tract. When CAF and MFB were compared to AMFB, the spindle cells are more often CD34 positive in the former (2,4). The former affects both men and women in the inguinoscrotal region with a comparable frequency. The strong vascular component and perivascular distribution of epithelioid cells found in AMFB are usually absent in MFB.

When situated in the vagina, the main differential diagnoses are likely to be leiomyoma with prominent vascularity or an angiomyoma. They however, share the same immunophenotypic characteristics with AFMB, hence morphology is the best distinguishing feature. Fibroepithelial stromal polyp (FEP) is also included in the differential since it is a stromal tumour with a wide range of morphological appearance (1). It is polypoidal macroscopically, however lack a visible interface with the mucosa or overlying skin. In contrast to AMFB, the absence of a well-defined interface in FEP is a valuable signal to its diagnosis. Multinucleated stromal cells are also seen in FEP, which are situated along the epithelial stromal interface (1,2). It is much more similar to superficial angiomyxoma in that both are hypocellular and have a myxoid stroma with dispersed vessels of various sizes.

CONCLUSION

Angiomyofibroblastoma is an uncommon benign non-recurring mesenchymal tumour with predilection to occur in the lower female genital tract. An accurate preoperative diagnosis is challenging as clinical and radiological features are not distinctive. Histopathological examination is essential in differentiating from other mesenchymal tumours with high recurrence potential.

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