Case Report

Concurrent Utero-Vaginal Prolapse with Cervical Angiomyofibroblastoma: A Rare Disease with Distinct Entity

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Abstract

We report the case of a rare, benign mesenchymal tumour arising from the cervix. A 53-year-old post-menopausal woman presented with mass per vagina. Examination revealed stage 2 utero-vaginal prolapse and multiple elongated polyps seen at the cervix. She underwent local excision. Histopathological examination findings and the immunohistochemical studies were consistent with Angiomyofibroblastoma.

Keywords: Angiomyofibroblastoma, cervix, malignant mesenchymal tumour, uterine prolapse

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Introduction

Angiomyofibroblastoma (AMFB) is a unique clinical entity that exhibits myofibroblastic differentiation and stromal cell proliferation (1). Despite the fact that it has been recognized for the past few decades, the cause is still not well defined. It is a rare, benign, mesenchymal tumour and commonly involves female lower genital tract i.e. vulva, vagina and labia majora (2). Cervical AMFB is even rarer. To date, only two cases have been reported (3,4). We report a case of cervical AMFB and illustrate the importance of recognition of this entity with accurate pre-operative diagnosis.

Case Report

A 53-year-old post-menopausal woman, Para 2+1, complained of mass per vaginal which was affecting her daily activity and sexual life. There was no vaginal bleeding, abnormal discharge, urinary or bowel symptoms. She was rather healthy except for well-controlled diabetes mellitus and hypertension. Physical examination demonstrated a stage 2 utero-vaginal prolapse (UVP). There were multiple elongated polyps seen at the cervix (Fig. 1). The largest polyp measured 4x3 cm in diameter. The polyps were excised and sent for histopathological examination. She was counselled that the choices of treatment option for her UVP would depend on the histopathological finding of the cervical polyps.

Histopathological examination (HPE) revealed polypoidal tissues lined by stratified squamous epithelium and were composed of hyper and hypocellular areas admixed with small blood vessel. There were spindle and plump stromal cells aggregated around the vessels (Fig. 2). A focal area of marked neutrophils and lymphocytes infiltrate within the stroma was seen. There was no evidence of malignancy. The immunohistochemical studies showed the tumour cells were positive for Smooth

Muscle Actin (SMA), Desmin and CD34 (Fig. 2) which were consistent with angiomyofibroblastoma.

In view of the benign entity, she was given the option of vaginal ring and surgical intervention. She had an uneventful laparoscopic assisted vaginal hysterectomy and bilateral salphingo-oopherectomy, anterior and posterior colporrhaphy. Histopathological examination of the uterus and both ovaries revealed no evidence of endometrial hyperplasia or malignancy. The cervix showed few Nabothian cysts with no residual polyp. There was no evidence of dysplasia or malignancy. She remained asymptomatic with no recurrence of either angiomyofibroblastoma or pelvic organ prolapse at two-year follow up.

Discussion

Angiomyofibroblastoma was first reported by Fletcher et al. almost two decades ago (5). Following that, several reports had been published where majority was vulvo-vaginal origin (2,6,7). Other rare locations include pouch of Douglas (1) and spermatic cord in male (8). Kitamura et al. (9) reported another interesting case: a young 24-year-old lady presented with abnormal posteriorly directed urinary jet due to urethral AMFB. Angiomyofibroblastoma during pregnancy had been reported as well (10).

Vulvo-vaginal AMFB is easily detectable due to its location. However, AMFB arising from other regions such as pouch of Douglas, may present with huge pelvic mass and pose significant challenge to establish the diagnosis (1). In the present case, the polypoidal growth was easily seen due to the stage 2 UVP. Angiomyofibroblastoma of the cervix is extremely uncommon with only two published reports so far (Table 1) (3,4). Therefore, AMFB of the cervix is rarely being considered pre-operatively. Both reported

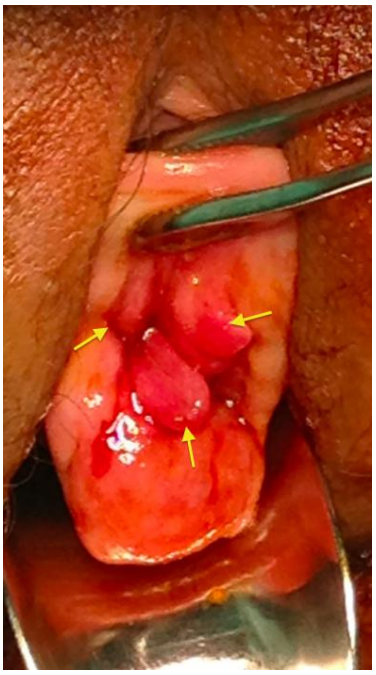

Figure 1: Multiple elongated polyps seen at cervix

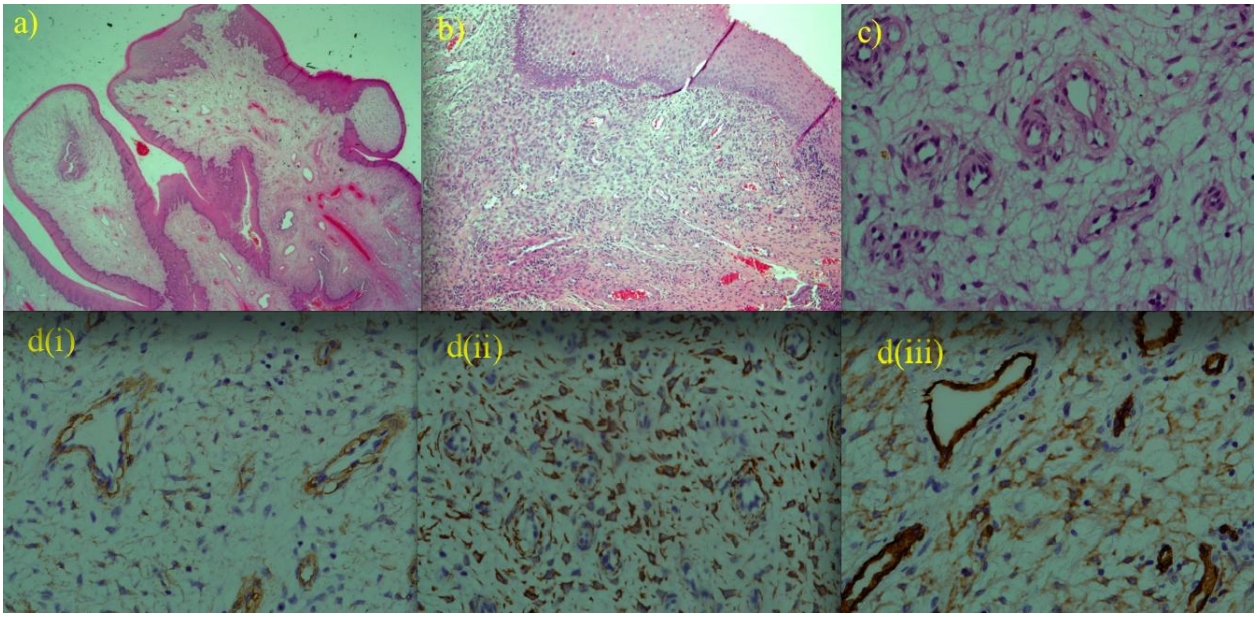

Figure 2: a) Polypoidal tissue lined by squamous epithelium (H&E, 2x) b) Hypocellular and hypercellular area admixed with small blood vessels (H&E, 4x) c) Spindle cells aggregate around blood vessels (H&E, 40x) d) Tumour cells immunopositive for: i) Smooth Muscle Actin (SMA, 40x), ii) Desmin (40x), iii) CD34 (40x)

Table 1: Cervical angiomyofibroblastoma reported in the literature.

Case	Age (years)	Presentation	Size (cm)	Positive IHC	Negative IHC	Treatment	Reference
1	43	asymptomatic cervical myoma	3.0x3.0x2.5	vimentin, desmin	actin, CD34	local excision	Kim et al. 2011 [3]
2	44	polypoidtumour	2.0	vimentin, desmin,CD44	sarkomeric actin, Ki67	N/A	Babala et al 2011 [4]
3	53	vaginal mass	4.0x3.0	SMA, desmin, CD34	N/A	local excision	Present case

IHC: Immunohistochemistry
N/A: Not available

cases presented with asymptomatic mass per vaginal at their fifth decade of life. In fact, one of these cases underwent local excision for suspected cervical myoma, with AMFB diagnosed only after HPE.

Total excision, which is curative, is the treatment for cervical AMFB. There was only one case being reported to have tumour recurrence (11) and the excised recurrent tumour was confirmed to be benign in nature.

Although, almost all reported cases in the literature showed that AMFB exhibited benign course, there was a case of vulva AMFB that underwent sarcomatous transformation (12). In addition, aggressive angiomyxoma (AA), a form of locally invasive malignant mesenchymal tumours, shares many common features with AMFB including presentation and pathological entities. It has high tendency to recur even years after resection (13).

When compared to AMFB, AA tends to be larger (up to 60cm) macroscopically, where as AMFB is smaller and measures average 4cm in size (0.5-12cm) (5,13). Microscopically, AA displays sparse cellularity with stellate cells widely separated by loose myxoid stroma, where as AMFB demonstrates alternating hypercellular and hypocellular zones with irregularly distributed blood vessels which was observed in our case. Immunostaining pattern towards SMA, desmin and CD34 may be helpful, however, there are variable overlapping features in both AA and AMFB. There is emerging potential marker like HMGA2 protein expression that could help in differentiating AA from AMFB. Aberrant nuclear HMGA2 expression was found in 1/3 of cases of AA but not in AMFB (14).

Therefore, a more aggressive approach might be needed if AA is suspected. Hence in the presence case, our pathologist took great effort to exclude possibility of AA, including use of immunohistochemical staining, in order to assist us in formulating the management plan for the stage 2 UVP.

Conclusion

Cervical AMFB is extremely rare. Recognition of this unique clinical entity pre-operatively is important for surgical planning as it shows benign characteristics compared to other malignant tumour like AA. Furthermore, patient's counseling is of paramount important particularly when patient is having concurrent UVP.

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