Angiomyofibroblastoma of vulva- a case report and review

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Received August 03, 2014; Revised September 20, 2014; Accepted September 20, 2014; Published Online September 21, 2014

Case Report

Abstract

Angiomyofibroblastoma is an uncommon benign mesenchymal tumor that has a tendency to occur mostly but not solely in the vulvar region. It is common in vulvar region of premenopausal women and rarely also in inguinioscrotal region of man. Mostly it is asymptomatic and resembles with Bartholin’s gland cyst. Due to overlapping of histopathological picture, diagnostic perplexity often arises between angiomyofibroblastoma (AMF) and aggressive angiomyxoma (AAM). Aggressive angiomyxoma is a more infiltrative lesion that has a higher tendency for local recurrence whereas AMF is benign in nature.

Keywords: Angiomyofibroblastoma; Aggressive Angiomyxoma; Vulva

Introduction

Angiomyofibroblastoma is a rare tumour of the vulva, the more commonly encountered being fibroma, lipoma, haemangioma and leiomyoma.¹ ² Angiomyofibroblastoma was first described by Fletcher et al.³ This is a distinctive mesenchymal tumour almost exclusively occurring in the vulva.⁴ A few cases have been reported to occur in vagina, fallopian tubes, cervix, inguinioscrotal region and occasionally of spermatic cord and scrotum.⁵ ⁶ ⁷ ⁸ ⁹ ¹⁰ Angiomyofibroblastoma are well circumscribed, amenable to local excision, compared to the aggressive angiomyxoma which are more often infiltrative requiring wide excision with a tendency to recur in more than 70% of cases within 2 years.³ Between 1999 and 2013, only 57 cases of AMF have been reported in English literature.⁴ We report herein a case of angiomyofibroblastoma of vulva in a 34-year-old woman. Though histologically tumour showed characteristic features, it also expressed S-100 protein in addition to Vimentin and Desmin.

Case Presentation

A 34-year-old woman presented with a painful swelling of the left labia majora for the last 3 months. The lesion has slowly increased in size. On clinical examination, a 2 × 2 cm, well circumscribed, mobile swelling was felt on the left labia majora. The preoperative diagnosis was a Bartholin cyst. Local excision was done with primary closure. The tumour was very vascular. Postoperative period was unventful. Microscopically, the tumour was not well encapsulated but well demarcated. The tumour was composed of short spindle shaped cells arranged loosely in edematous, collagenous rich loose stroma with distinct perivascular pattern. The tumour was highly vascular with large number of focally anastomosing channels around which the spindle shaped cells were arranged in incomplete concentric pattern. Spindle cells embedded in collagen rich stroma form slot like spaces and channels. There were a few fat cells embedded in the tumour mass. Three to five mast cells were seen per high power field all throughout the stoma. There was no appreciable mitosis, there were no nuclear atypia, pleomorphism or an evidence of tumour necrosis (Figure 1). Immunohistochemically, the tumour cells expressed Vimentin, Desmin and S-protein. The tumour cells were negative for smooth muscle actin [SMA].

Discussion

Solid benign tumors of the vulva are rare. Two contrasting mesenchymal lesions of the female genital tract featuring loosely textured collagenous stroma and a prominent vascular component have been reported in the literature. In 1983, Steepar and Rosai ¹¹ described a locally infiltrative but nonmetastasizing tumour of the pelvic soft tissues with a high rate of recurrence and designated as aggressive angiomyxoma (AAM). In 1992, Fletcher et al. ³ described ten cases of a clinicopathologic entity distinct from aggressive angiomyxoma and designated as Angiomyofibroblastoma (AMF). The tumour has distinct histopathology by which it can easily be identified and easily distinguished from aggressive angiomyxoma and other tumors occurring in this region. Between 1999 and 2013, only 57 cases of AMF have been reported in the literature.⁴

The characteristic clinical features of these neoplasm is that it occurs in middle aged woman and often misdiagnosed as Bartholins gland or labial cyst. It is a slow growing, asymptomatic pink color tumor with hard and rubbery consistency. The size of the lesion varies from 0.5 to 12 cm in diame-

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Cite this article as: Goyal S, Singla S, Singla M, Vyas D. Angiomyofibroblastoma of vulva- a case report and review. J Case Rep Onc Ther 2015; 1(1):112

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ter and is often of <5 cms 4,12, whereas most of the aggressive angiomyxomas measure more than 5 cms (one reached a size of 60 cm). When the size of the tumor is big enough, it can be also puzzled with perineal hernia. Rarely AMF can present as pedunculated mass and can also occur in inguinoscrotal region of man. 13 The tumor is usually asymptomatic and the patients often complain pelvic discomfort or cosmetic problem. It also has low tendency for local recurrence. 6 The vulval mass like cellular angiofibroma, fibroepithelial stromal polyp, or superficial angiomyxoma can resemble clinically with AMF and should be differentiated. 14

Mostly angiomyofibroblastoma is located in the superficial soft tissues of the vulva and well circumscribed but it is not uncommon for aggressive angiomyxoma to affect the deep pelvic soft tissues, i.e. infiltrative in nature such as para-vaginal and pararectal region, particularly at recurrence. 11 so, it requires wide excision. Angiomyofibroblastoma is an important entity to be recognized, because it requires only simple excision and there are currently no published reports of local recurrence or metastatic disease when excised completely. Although Weiss et al. 12 mentioned a few cases with recurrence where the tumour was not completely excised. There are few cases on record where Angiomyofibroblastoma showed sarcomatous differentiation in parts. 14 When tumor is of large size then transperineal ultrasonography or magnetic resonance imaging (MRI) is useful and precisely tell about extent of tumor. A Computed Tomography (CT) can reveal the involvement of neighboring pelvic organs and can differentiate it from a perineal hernia. Due to accurate radiologic- pathologic correlation, MRI or ultrasonography is superior to CT in differential diagnosis of various vulvar tumors. CT findings of AAM and AMF are similar but Gadolinium- enhanced T1 image can differentiate between the two. 15

The methods other than surgical excision such as needle biopsy or aspiration cytology are not sufficient to preoperative diagnosis. Histopathology of excised tumor is important because AMF has to be differentiated from AAM. The histologic findings of AMF are alike but different from AAM as there is false capsule around AMF. In contrast, AAM infiltrates to surrounding tissue and the border is not clear. Microscopically the tumors are characterized by high cellularity, numerous blood vessels, and plump stromal cells. 15

FIG. 1: Slide showing histopathology of tumor (low power and high power microscopic field).
Immunohistochemically, the tumour expresses Vimentin with focal positivity for Desmin, Smooth Muscle Actin [SMA]. The tumour cells are known to be negative for S-100 protein, cytokeratin and myoglobin. The present case expressed S-100 protein differently in addition to Vimentin and Desmin while all the cases of Angiomyofibroblastoma recorded in literature till to date are known to be S-100 negative. We have no explanation for the strange observation. AMF also has expression of estrogen and/or progesterone receptors and due to this it might arise from hormonally responsible mesenchymal cells.

AMF and AAM may have common origin as there is morphological overlap. On the contrary, another study revealed that AMF and AAM are biologically different tumors as HMGAA2 gene arrangement is established in 1/3 of AAM but not in AMF.

Surgery is mainstay of treatment. Diagnosis of AMF should be confirmed as it requires local excision whereas AAM is a locally invasive tumor which requires a wide excision. Prognosis is good if proper excision is done.

Conclusion

We conclude that Angiomyofibroblastoma is a rare mesenchymal tumour of the vulvar region. It is important to distinguish it from the Aggressive angiomyxoma, as Angiomyofibroblastoma requires only simple excision with less or no tendency to recur. Pelvic sonography, CT or MRI imaging modality is useful in differential diagnosis.

Conflict of interest

The authors declare that they have no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References