ejpmr, 2017,4(8), 217-219

SJIF Impact Factor 4.161

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Review Article

ISSN 2394-3211

EJPMR

ANGIOMYOFIBROBLASTOMA OF VULVA: A STUDY OF 4 CASES WITH REVIEW OF LITERATURE.

***[1]Dr. Sonia Singh, [2]Dr. Meenu Gill, [3]Dr. Abha Chandra and [4]Dr. Suman Tomar**

[1]Adesh Medical College.
[2,3,4]PGIMS, Rohtak.

***Corresponding Author: Dr. Sonia Singh**
Adesh Medical College.

Article Received on 05/06/2017 **Article Revised on 25/06/2017** **Article Accepted on 15/07/2017**

ABSTRACT

Angiomyofibroblastoma is a rare benign mesenchymal tumor that occur mostly in the vulvovaginal region in premenopausal females and also rarely involves inguinoscrotal region in males. It resembles Bartholin's gland cyst due to its location. It usually occur in middle age group females. We hereby report 4 cases of angiomyofibroblastoma who presented with vulval mass. One of them was a postmenopausal female while another one was a 20 year old young woman, an uncommon finding. Rest two were middle aged females. Due to overlapping histological features, angiomyofibroblastoma(AMFB) should be distinguished from aggressive angiomyxoma(AAM) which is a locally aggressive tumor and has tendency to recur whereas AMFB is benign in nature and does not recur.

KEYWORDS:- Angiomyofibroblastoma, postmenopausal, young female, aggressive angiomyxoma, vulva.

INTRODUCTION

Angiomyofibroblastoma is an extremely uncommon benign mesenchymal neoplasm. It was first described by Fletcher and his colleagues in 1992.[1] It occurs primarily in vulval region in females although unusual sites are vagina, cervix, fallopian tubes in females and rarely in inguinoscrotal area, spermatic cord and scrotum in males.[2] It occur mainly in middle aged women and is less common in young females as well as in postmenopausal age group.[3] Due to its slow growing nature it is confused clinically with Bartholin's gland cyst. Another important differential diagnosis to be excluded is aggressive angiomyxoma, an infiltrative lesion with high propensity of local recurrence.[1]

CASE HISTORIES

Case 1

A 34 year old female presented with complaints of painful swelling of right labia majora since 4 months. The swelling was gradually increasing in size. On examination, 2.5x 2 cm well circumscribed soft to firm, mobile, tender mass was felt on right labia majora. Her general and systemic examination were within normal limits. The mass was locally excised and the specimen was sent for histopathological examination. Microscopically the tumor was well demarcated and consisted of large number of anastomosing blood vessels around which spindle shaped cells were arranged in perivascular pattern embedded in loose edematous collagenous stroma. Few mast cells were also seen. Nuclear pleomorphism, atypia, mitosis were absent. Immunohistochemical staining show diffuse positivity for vimentin, desmin, CD34 and negative for S 100 and SMA. Thereby a final diagnosis of angiomyofibroblastoma was made. Patient was followed up for 26 months with no evidence of recurrence.

Case 2

A 52 year old female came with history of swelling in left vulval region since 7 months. The swelling was painless and was gradually increasing in size. Her past medical, menstrual and family history were not significant. On examination, 4.5x 3 cm soft, non tender, mobile mass was present in anterior aspect of labia majora. Inguinal lymph nodes were non palpable. Her general and systemic examination were within normal limits. Local excision of mass was done and the specimen was sent for histopathological examination. Microscopic and immunohistochemical features confirmed to the diagnosis of angiomyofibroblastoma. Patient was kept on regular follow up for 2 years with no evidence of recurrence.

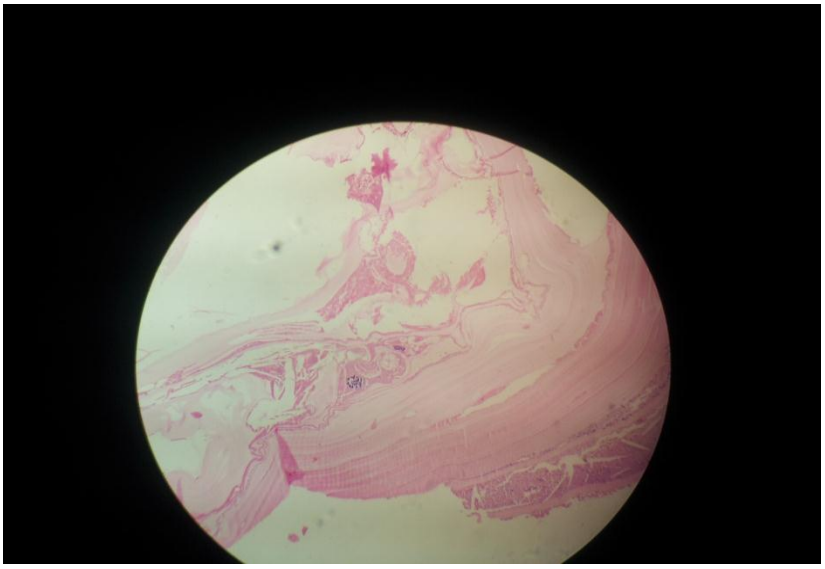
Case 3

A 32 year old woman, gravida 2 para 2 presented with a painless swelling of labia majora since 14 months. The swelling was pea sized when started and gradually progressed to the present size. Her family history and medical history were unremarkable. Ultrasonography showed normal uterus and bilateral adnexae and inguinal lymph nodes were not enlarged. A 5.5x3.5 cm hyperechoiec mass with few hypoechoiec areas was

present in labia majora. Clinical examination showed 5x4 cm grey pink, soft, nontender mass on lateral margin of labia majora. Histological examination of excised mass revealed irregularly distributed edematous hypocellular areas admixed with hypercellular areas with spindle shaped cells. Few small clusters of adipose tissue was also present. Tumor showed positive staining for vimentin, desmin, Factor VIII, ER, PR. MIB index was less than 1%. Tumor was negative for S-100, SMA, CD 34. The diagnosis of angiomyofibroblastoma was derived. Patient was kept under follow up with no evidence of recurrence for 1 year.

Case 4

A 20 year old female reported to the department of obstetrics and gynaecology complaining of swelling in vulvar area since 2 years. It was increasing in size since 6 months. The swelling was painless but was causing discomfort to the patient. Her personal history, menstrual history and family history was unremarkable. Clinical examination showed a 6 x 3 cm reddish, mobile, non tender mass, soft in consistency. Her vaginal ultrasound showed no abnormality and inguinal lymph nodes were also not enlarged. Immunohistochemical and morphological features were consistent with angiomyofibroblastoma. Her postoperative period was uneventful. Patient was followed up for 15 months with no evidence of recurrence.



DISCUSSION

Angiomyofibroblastoma is an extremely rare tumor of the vulva; more common tumors being fibroma, lipoma and leiomyoma.[4,5] It occurs primarily in middle aged women. In our study one of the patient is of 52 years old and another one was very young of 20 years of age which is a less common finding. Most of the cases measure less than 5 cm in greatest dimension with previous reported cases ranging between 0.5cm to 13 cm. It occurs as sharply circumscribed mass in the vulval subcutaneous tissue, few of the reported cases also showed a pedunculated mass.[6] Most of the times tumor is slow growing and painless. In our case also, 3 out of 4 patients presented with slow growing painless mass. Patient normally complains of pelvic discomfort and swelling. Clinically angiomyofibroblastoma can be misdiagnosed as Bartholin's gland cyst, inguinal hernia, labial cyst and tumors like lipoma, leiomyoma and liposarcoma.[7] The cell of origin of angiomyofibroblastoma is unclear but some consider it to be myofibroblastic in origin.[6] Grossly angiomyofibroblastoma is well circumscribed, pink in colour with soft to rubbery consistency. Microscopic examination of the tumor show a well demarcated lesion with hypocellular and hypercellular areas alternating with each other. Stroma is edematous, loose and contain abundant blood vessels around which tumor cells are arranged. Tumor cells are spindle to ovoid with eosinophilic cytoplasm. Nuclear atypia, pleomorphism and mitotic figures are rare to absent. Adipocytes can be sparsely scattered within the tumor but if fat cells predominates in such instances, these tumors are classified as lipomatous variant of AMFB.[8] Mast cells can also be seen scattered within stroma. Majority of the cases show immunoreactivity for desmin, vimentin, estrogen and progesterone receptors, occasionally positive for CD34 and negative immunostaining for S 100, cytokeratin, smooth muscle actin.[9] The positive staining for estrogen and progesterone receptors shows hormone dependent nature of these tumors.

Angiomyofibroblastoma should be distinguished from leiomyoma, aggressive angiomyxoma and cellular angiofibroma. Leiomyoma was ruled out due to absence of arrangement of cells in whorls and fascicles and tumor cells were negative for smooth muscle actin. Another differential diagnosis to be excluded is aggressive angiomyxoma. There is false capsule in AMFB while

AAM infiltrates into the surrounding tissues and the border of aggressive angiomyxoma is not clear.[2] AMFB demonstrate alternating hypo and hypercellular areas with prominent perivascular accentuation whereas in AAM there is no hypercellular areas and the stromal cells are uniformly distributed with no prominent perivascular distribution of cells. Though immunohistochemical findings are overlapping in both but absence of stromal mucin favors AMFB over AAM.[6] It is very important to distinguish between AAM nd AMFB as their biological behaviour differs markedly. AAM occurs in deep soft tissue of pelvic region and have a strong propensity for local recurrence if excised incompletely therefore a wide excision is advised in AAM. The infiltrative nature of AAM in surrounding tissue results in its incomplete surgical excision. In contrast, AMFB is located in superficial pelvic soft tissues and is a well circumscribed lesion. Hence a local surgical excision with clear margins suffice with less chances of recurrence. Another differential diagnosis to be considered is cellular angiofibroma. Although there is no such clinical significance to differentiate them. The round cells arranged around vessels with absence of numerous thick walled hyalinised blood vessels help in distinguishing AMFB from cellular angiofibroma. Also AMFB show variable positivity for desmin, CD34 whereas cells in cellular angiofibroma are frequently immunoreactive for CD34 than desmin.[11] Cellular angiofibroma is mitotically active while AMFB has low mitotic figures. If tumor is of large size then magnetic resonance imaging (MRI) or transperineal ultrasonography is useful in describing the extent of tumor. Computed tomography (CT) scan help in differentiating it from perineal hernia and involvement of neighbouring pelvic organs. Though these investigations may be of some help but histopathological examination of an excised tumor is important along with immunohistochemical analysis of the tumor to help in establishing the diagnosis of angiomyofibroblastoma.[2] Malignant transformation is extremely rare, only one case of AMFB has reported to have undergone sarcomatous transformation.[12]

Surgical excision of the tumor is mainstay of treatment. But diagnosis should be confirmed as local excision suffice in case of angiomyofibroblastoma but aggressive angiomyxoma requires wide surgical excision in order to prevent recurrence. All of our patients were followed up and none of them show recurrence.

CONCLUSION

Angiomyofibroblastoma is an extremely rare benign vulval mesenchymal tumor with rare chances of recurrence. Radiological, histopathological and immunohistochemical findings are necessary to establish the diagnosis and also to exclude other differential diagnosis especially AAM which have chances of recurrence.

REFERENCES

1. Fletcher CD, Tsang WY, Fisher C, et al. Angiomyofibroblastoma of the vulva. A benign neoplasm distinct from aggressive angiomyxoma. *Am J Surg Pathol*, 1992; 16: 373-82.
2. 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.
3. Hisaoka M, Kouhu H, Aoki T, Daimaru Y, Hashimoto H. Angiomyofibroblastoma of the vulva. A clinicopathologic study of seven cases. Pathol Int, 1995; 45: 487-92.
4. Fu Y, Reagan JW. Pathology of the uterine cervix, vagina and vulva. Major problems in pathology. Philadelphia: WB Saunders, 1989; 21: 336-79.
5. Newman PL, Fletcher CD. Smooth muscle tumours of the external genitalia:clinicopathological analysis of a series. *Histopathology*, 1991; 18: 523-9.
6. Omori M, Toyoda H, Hirai T, Ogino T, Okada S. Angiomyofibroblastoma of the vulva: A large pedunculated mass formation. Acta Med Okayama, 2006; 60(4): 237-42.
7. Tochika N, Takeshita A, Sonobe H, Matsumoto M, Kobayashi M, Araki K. Angiomyofibroblastoma of the vulva: report of a case. Surg Today, 2001; 31: 557-9.
8. Seo JW, Lee KA, Yoon NR, Lee JW, Kim BG, Bae DS.A ngiomyofibroblastoma of the vulva. Obstet Gynecol Sci., 2013; 56(5): 349-51.
9. Barat S, Tirgar-Tabari S, Shafaee S. Angiomyofibroblastoma of the vulva. Arch Iran Med, 2008; 11: 224-6.
10. Sims SM, Stinson K, McLean FW, Davis JD, Wilkinson EJ. Angiomyofibroblastoma of the vulva: A case report of a pedunculated variant and review of the literature. J Low Genit Tract Dis., 2012; 16: 14954.
11. Iwasa Y and Fletcher CDM: Cellular angiofibroma: Clinicopathologic and immunohistochemical analysis of 51 cases. Am J Surg Pathol, 2004; 28: 1426-35.
12. Nielsen GP, Rosenberg AE, Young RH, Dickersin GR, Clement PB, Scully RE. Angiomyofibroblastoma of the vulva and vagina. Mod Pathol, 1996; 9: 28491.