CASE REPORT

AGGRESSIVE ANGIOMYXOMA AS A PEDUNCULATED GROWTH OF VULVA: A RARE CASE REPORT

Anitha N[1], Prathibha S. D[2], Rashmi Sinha[3], Pavana H. H[4]

HOW TO CITE THIS ARTICLE:
Anitha N, Prathibha S. D, Rashmi Sinha, Pavana H. H. "Aggressive Angiomyxoma as a Pedunculated Growth of Vulva: A Rare Case Report". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 6, February 9, 2015; Page: 742-745.

ABSTRACT: Aggressive Angiomyxoma is a slow growing, uncommon myxoid neoplasm occuring chiefly in the genital, perineal and pelvic regions of adult women.(1,2) It is considered as a non-metastasizing tumour with locally aggressive nature and propensity for local recurrence.(1) Hence it is important to differentiate from other mesenchymal tumours in this region and follow up is equally essential. We report a case of 37 years old lady with a pedunculated growth in the left labia majora. Our case highlights the variable presentation of these tumours.
KEYWORDS: Aggressive Angiomyxoma, mesenchymal vulval tumours, local recurrence.

INTRODUCTION: First described by Steeper and Rosai in 1983, Aggressive Angiomyxomas are rare mesenchymal tumours.(3) It often presents as a vulval mass and clinically simulates Bartholin's cyst mostly presenting in second or third decade of life.(4) The female: male ratio is more than 6: 1.(1) It is a locally infiltrative, non-metastasizing neoplasm. Tumours are of varying size often relatively larger than 10 centimeter's.(5) About one fourth of these tumours is pedunculated.(6) Imaging (CT and MRI) helps to identify the extent of pelvic involvement as clinical examination may underestimate the size of tumour. Wide local excision is now the treatment of choice. Local recurrence rate is about 30% of cases.(1,6) Recurrence occurs within few months to even decades after initial excision. More recently some patients have been treated for initial and recurrent disease with Gonadotrophic releasing hormone agonists.(1)

CASE REPORT: A 37 year old lady, P4L4, sterilized, presented with history of growth in the labia majora for past 10 years. Initially it was about pea size and has increased to present size of 5 X 6 centimeters over 10 years. There was no history of pain, bleeding or discharge from the swelling. She mainly presented due to mechanical discomfort of the swelling during walking. No history of any other similar swellings. On local examination there was a well circumscribed polypoidal swelling of 5 X 6 centimeters attached to the mid portion of labia majora with a pedicle measuring 1.5 centimeter thick and 4 centimeters long. (Figure 1). The swelling was non-tender, rubbery in consistency. The inguinal lymph nodes were not enlarged. Her pre-operative investigations were normal. She was taken up for surgery and a wide margin local excision of the pedicle done (Figure 2). There was abnormally increased vascularity at the base and the patient withstood procedure well.

On histology the tumour composed of widely scattered spindle and shaped cells in myxoid stroma with abundant blood vessels. Thickened hyalinised medium to large vessels noted with foci of hemorrhage. The entire specimen was covered by normal skin. (Figure 3 and 4) This was suggestive of Aggressive Angiomyxoma. Her post-operative period was uneventful. She has not presented with recurrence during our 4 month follow up.

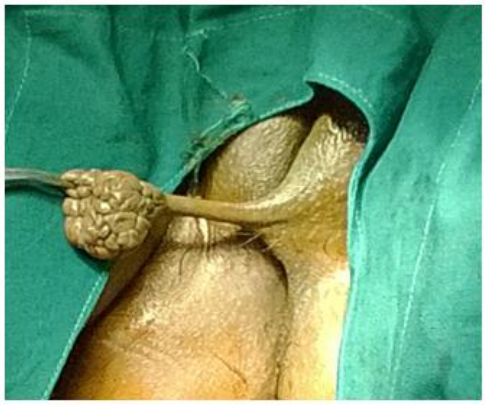

Figure 1

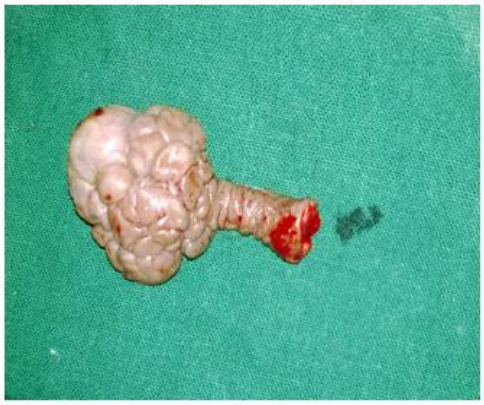

Figure 2

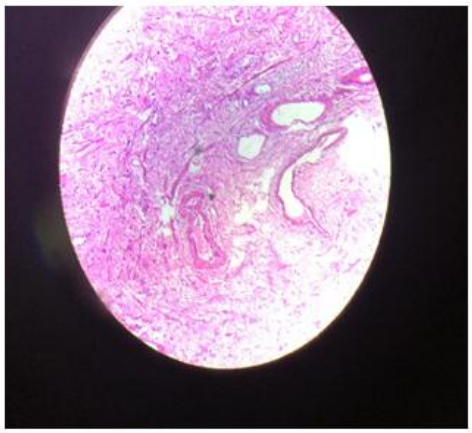

Figure 3

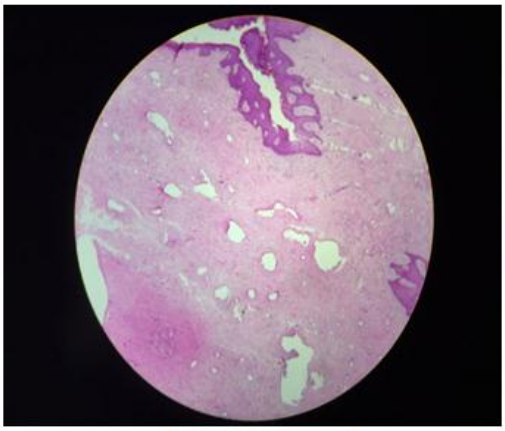

Figure 4

DISCUSSION: The term Aggressive Angiomyxoma was coined by Steeper and Rosai in 1983.[3] About 150 cases have been reported till 2012. It is slow growing, locally infiltrating tumor typically arising in the pelvis and perineal regions of women, may also arise in the inguino scrotal region of men. Described as non-metastasizing neoplasm but few cases of metastasis to the inferior venacava, right atrium[7] and mediastinum and lungs[8] have been reported. The tumor can usually grow to unusual sizes of more than 10 centimeters pushing the normal structures of the pelvis without invading them.

Clinically the differential diagnosis for Aggressive Angiomyxoma may be Bartholin's cyst, Lipoma, Labial cyst, Gartner duct cyst. Our case was a pedunculated growth at the vulva. Histologically Aggressive Angiomyxoma must be differentiated from other benign myxoid neoplasms like Angiomyofibroblastoma given its propensity for local recurrence. Aggressive Angiomyxoma is poorly circumscribed, infiltrative > 10 centimetres whereas Angiomyofibroblastoma is circumscribed < 3 centimetres. The cell shape is spindled with variable

thick walled vessels in Aggressive Angiomyxoma and epitheloid with numerous thin walled vessels in Angiomyofibroblastoma.[1]

Wide local excision with negative margins has been the treatment of choice.These tumours tend to recur locally after excision as early as 2 months to even decades. Recurrence also depends on the initial surgery. Sometimes because of infiltrative borders complete excision is often difficult and likely accounts for high rate of local recurrence.[1] Recent studies have shown a recurrence rate of less than 10 % following aggressive surgical excision and most patients had only one recurrence.[1]

Hormonal therapy with Gonadotropin Releasing hormone agonists has been used to reduce the tumour size to facilitate complete surgical excision and also for treatment of recurrences.[6] Till date there is no general consensus for the prevention or management of the recurrence because of rarity of the tumour. Gonadotropin Releasing hormone agonists have also been tried for prevention of recurrence in few cases based on fact that these tumours are Estrogen and Progestrone receptor positive.[2] Efficacy of Chemotherapy and Radiotherapy is not yet defined. In our case the patient is under meticulous follow up for last four months without any intervention as there is no defined protocol for prevention of recurrence.

CONCLUSION: Aggressive Angiomyxoma is a slow growing benign, rare mesenchymal tumour with local infiltrative properties with tendency for local recurrence. The clinical presentation may be varied. Histological diagnosis is the most important aspect as these patients need meticulous follow up for recurrence.

REFERENCES:

1. Sharon W Weiss M D. Aggressive Angiomyxoma. Enzinger and Weiss Soft Tissue Tomours 5 th Edition. Elsevier; 2008: 1081 – 88.
2. R. Elkanttah, O. Sarkodie, H. Otteno and A. Fletcher. Aggressive Angiomyxoma of the Vulva: A Precis for Primary Care Providers. Case Reports in Obstetrics and Gynecology. Vol 2013, 4 pages.
3. Steeper TA, Rosai J. Aggressive Angiomyxoma of the female pelvis and perineum. Report of nine cases of a distinctive type of gynecological soft tissue neoplasm. Am J Surg Pathol. 1983; 7: 463-475.
4. Juan Rosai M D. Aggressive Angiomyxoma. Rosai and Ackerman's Textbook of Surgical Pathology 9 th Edition. Mosby; 2009: 1495 – 96
5. Christopher D. M. Fletcher. Tumours of Female Genital Tract. Diagnostic Histopathology of Tumours 4 th Edition. Elsevier; 2013: 863 – 864
6. Mahendra M. Kura, Saurabh R. Jindal and Usha N. Khemani. Aggressive Angiomyxoma of the Vulva: An uncommon entity. Indian Dermatol Online J. 2012 May – June; 3(2): 128 – 130.
7. Junzu Geng M D, Bofeng Cao M D and Liping Wang M D. Aggressive Angiomyxoma: An unsual presentation. Korean J Radiol. 2012 Jan – Feb; 13(1) 90 – 93.
8. Ramin Michael Siassi M D, Thomas Papadopoulos M D Ph D , Klaus E Matzel M D Ph D. Metastasizing Aggressive Angiomyxoma. N Engl J Med 1999; 341: 1772.

AUTHORS:

1. Anitha N.
2. Prathibha S. D.
3. Rashmi Sinha
4. Pavana H. H.

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Obstetrics & Gynaecology, Dr. B. R. Ambedkar Medical College & Hospital.
2. Professor, Department of Obstetrics & Gynaecology, Dr. B. R. Ambedkar Medical College & Hospital.
3. Assistant Professor, Department of Obstetrics & Gynaecology, Dr. B. R. Ambedkar Medical College & Hospital.
4. Junior Resident, Department of Obstetrics & Gynaecology, Dr. B. R. Ambedkar Medical College & Hospital.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Anitha N,
1085, 12th Main,
2nd Stage, WOCR,
Mahalakshmipuram,
Bangalore-560086.
E-mail: dranitha15@gmail.com

Date of Submission: 15/01/2015.
Date of Peer Review: 17/01/2015.
Date of Acceptance: 28/01/2015.
Date of Publishing: 06/02/2015.