

Case Report

Combined surgical and medical management for recurrent cellular angiofibroma of vulva in a young female - a case report

Sasmita Das^{1*}, Manisha Sahu¹, Amit k. Adhya², Ranjan K. Sahoo³

¹Department of Obstetrics and Gynaecology, IMS and SUM Hospital, Siksha 'O' Anusandhan University, Bhubaneswar-751003, Odisha, India

²Department of Pathology, Kalinga institute of medical Sciences, KIIT University, Bhubaneswar, Odisha, India

³Department of radiology, IMS and SUM Hospital, Siksha 'O' Anusandhan University, Bhubaneswar-751003, Odisha, India

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*Correspondence:

Dr. Sasmita Das

E-mail: sasmitadas74@yahoo.co.in

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ABSTRACT

Cellular angiofibroma of the vulva is a benign tumor of mesenchymal origin in female and male genital tract composed of stromal spindle cells and mixture of adipose tissue, prominently vascular in nature. Here we present a case of recurrent cellular angiofibroma in a 27 years old young woman which was the 5th time recurrence when she presented to us. Diagnosis was confirmed by MRI, histopathology study and immunohistochemistry. With wide excision of tumor followed by GnRH analogue therapy post operatively, she is now tumour free even after 20 months of surgery.

Keywords: Vulva, Angiofibroma, GnRH analogue, Histopathology

INTRODUCTION

Cellular angiofibroma of the vulva is a rare benign growth of mesenchymal origin which was initially described by Nucci in 1997. This chiefly involves vulvar region. Original report described only 4 cases of this distinctive lesion, all in middle aged women i.e. 39-50 years.¹ These tumours occur in premenopausal women. A case reported in a 77 years old female, who had received long term estrogen treatment. Case of recurrence reported even after excision with a rim of uninvolved tissue. Long term follow up of cases is not available currently. Very few cases reported so far. We are describing a case of vulvar cellular angiofibroma that occurred in a young woman of 27 years which was a recurrent lesion, a 5th time recurrence in the same site.

CASE REPORT

A 27 year old married woman attended to our gynaecology OPD on 21st April 2013 with complain of swelling over left aspect of vulval region (Figure 1) for last one year. She described that, she had been operated 4 times for a similar lesion at the same site in last 3 years elsewhere. Checking her previous documents, we found that she was operated in 2007, 2008, 2010 and 2011 at different private hospitals. Previous documents mentioned, the mass removed was around 4x4 cm and histopathology study report of 2011 revealed it was cellular angiofibroma. Considering her history of previous surgeries and HP study report, the case of recurrent aggressive angiofibroma was suspected.

Her menstrual cycle was regular; she had one baby of 3 years of age, which was by normal delivery. Her general examination findings were normal except moderate

pallor. Systemic examination findings were within normal limit. Her clinical examination of local area revealed a mass of 10x7 cm over left labia majora extending to left ischio-rectal fossa laterally, up to middle half of the vagina medially and up to sub urethral space anteriorly. The mass was variegated in consistency, non-tender. Rectal mucosa was free. Ultrasound of perineum revealed a soft mass in left labia majora with extension to vaginal wall and ischio-rectal fossa. In MRI, a 102X46X48 mm size T1 isointense, T2/STIR hyperintense lobulated soft tissue mass noted in left side perineum, left labia majora, left side of mons pubis, left lateral wall of vagina up to lateral fornix. The mass was infra levator in location, extended to left ischio-rectal fossa and pushed the urethra to right side. Urinary bladder, urethra, uterus & cervix were not involved. There was no regional lymphadenopathy (Figure 2).



Figure 1: Pre-operative photograph showing mass over left aspect of vulvar region.

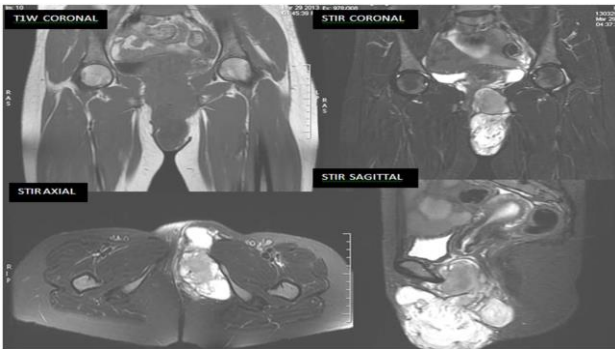


Figure 2: MRI picture of pelvis and perineum of patient.

All her general investigations like complete blood count, renal function tests, serology were normal except anemia (haemoglobin 5.7 gm%). Preoperatively 4 units of blood were transfused and planned for wide excision of mass. Incision of around 7 cm given over maximum projection of the tumour to the lateral aspect of left labia majora. Intra-operative finding revealed a mass of 10X8 cm with lobulation, variegated consistency extending upwards and displacing urethra towards right. Its left upper margin towards symphysis pubis and left ischiopubic rami, medial part was extending up to lower 2/3 of vagina and laterally up to ischio-rectal fossa. Wide excision of mass

could be done keeping tumor free margin. Skin wound closed keeping a drain. Postoperative period was uneventful (Figure 3 and 4).



Figure 3: Intraoperative photograph after taking out of the tumour.

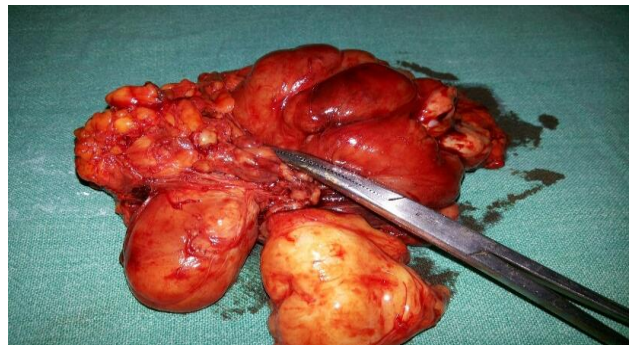


Figure 4: Gross picture of the tumour.

Histopathology of biopsy specimen was composed of bundle of plump spindle cells in background of collagen. Cells showed mild to moderate nuclear pleomorphism and prominent nucleoli without mitosis (Figure 5). Histological diagnosis was cellular angiofibroma/angiofibroma. Immunohistochemistry revealed tissue negative for smooth muscle actin, S-100P, pan cytokeratin, CD34. Tissue was focally positive for desmin and KI 67. Immunohistochemistry for tumor cells shown moderate positivity for estrogen and progesterone receptors. So diagnosis of cellular angiofibroma was confirmed.

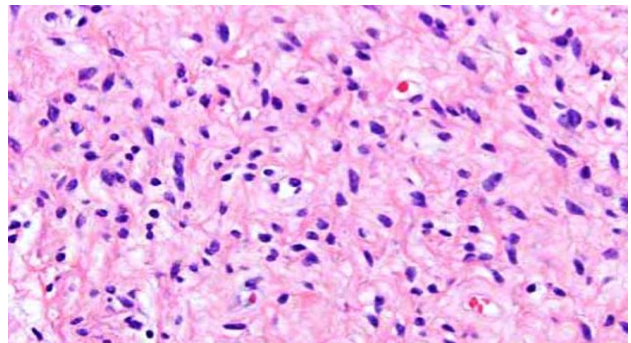


Figure 5: Histopathological picture of tumour tissue.

Considering recurrent nature of tumour and hormone receptor positivity, postoperative GnRh analogue therapy planned. Three doses of GnRh analogue (3.75 mg) instituted at monthly interval. The case was followed up at 3rd, 6th, 12th, 18th and 20th months post-surgery without any recurrence (Figure 6).



Figure 6: Post-operative photograph at 6 month follow-up.

DISCUSSION

Cellular angiofibroma is a benign mesenchymal lesion that was first described in 1997, and which chiefly involves the vulvar region.¹ The original report described four cases of this distinctive lesion, all occurring in middle aged women, and the authors considered this to represent a benign neoplasm with little or no potential for local recurrence if excised with a rim of uninvolved normal tissue. Since then, an identical lesion has been described in a woman involving the subcutaneous tissue of the chest wall.²

Mesenchymal tumors of vulva include leiomyoma, haemangioma, lipoma, Spindle cell tumors include neurofibroma, schwannoma, smooth muscle sarcoma. Unusually encountered masses are angiofibroma, angiofibroma, angiofibroma, cellular angiofibromas. Morphology, histopathology and immunohistochemical studies confirm the diagnosis. Exclusion of other Vulvovaginal soft tissue tumors particularly sarcomas and aggressive angiofibromas are mandatory to frame line of management. Simple excision with tumor free margin is adequate in all other cases.^{1,2}

Wide excision of cellular angiofibromas with clear margins is usually adequate treatment of these lesions. Data on long-term follow-up of cases is not currently available.^{3,4} Recently, one report of local vulvar recurrence after six months of surgery reflects continued growth of residual disease, rather than local aggression.^{3,5} Complete surgical removal is of paramount importance in angiofibroma as this tumour is almost always encapsulated and vascular.³ Surgical removal with its complete capsule ensures complete removal of tumour with added advantage of minimizing blood loss.

Identifying the capsule is most important early step in surgical removal.

Although there is some morphologic overlapping of cellular angiofibroma with the above mentioned differentials but bland spindle cells and prominent hyalinized blood vessels along with wispy collagen are distinctive for cellular angiofibroma.^{1,2,5} Moreover, immunohistochemistry plays an important role in differentiating this tumour from other vulvo-vaginal tumours. Cellular angiofibromas are consistently positive for vimentin with variable expression of CD34 and desmin, and negative for smooth muscle actin (SMA) and S-100.^{6,7}

This hormonally responsive tumour is believed to arise from specialized mesenchymal cells of the pelvic – perineal region or from the multipotent perivascular progenitor cells, which often display variable myofibroblastic and fibroblastic features.

Immunohistochemically, most AA express different combinations of estrogen and progesterone receptors, vimentin, desmin, smooth-muscle actin CD34 and CD44 but all are invariably negative for S-100, CEA and keratin.⁸⁻¹⁰ It has been postulated that the expression of estrogen and progesterone receptors suggest a role in pathogenesis of the tumor.⁵

Differential diagnoses of cellular angiofibroma are spindle cell lipoma, angiofibroma, spindle cell lipoma. They have both morphological and immunohistochemical similarities. They have common genetic origin as documented by Hameed M et al.¹² Probably, any tumour can be combination of different cell lines. In our case, this may be more towards angiofibroma because of its aggressiveness and recurrences.

In our case, the tumor is negative for CD34, smooth muscle actin, S-100P and pan cytokeratin and it is focally positive for desmin and KI-67, and moderately positive for estrogen and progesterone receptors. Danesh et al in 2007 has reported dramatic response of hormone positive tumors to GnRH analogue.¹¹ Considering recurrent nature of tumor in our case and hormone receptor positivity, GnRH analogue given post operatively, 20 month post-operative she is now tumor free.

CONCLUSION

Cellular angiofibromas are benign tumors. They can affect male and females of reproductive age group. Wide excision of tumor with clear margins is treatment of choice. Cosmetic and sexual function preservation should be kept in mind during surgery. For incomplete excisions and recurrent tumors, medical management with GnRH analogue is helpful.

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REFERENCES

1. Nucci MR, Granter SR, Fletcher CDM. Cellular angiofibroma: a benign neoplasm distinct from angiofibroma and spindle cell lipoma. *Am J Surg Pathol.* 1997;21:636-44.
2. Lane E, Walker AN, Mullis EN, Etheridge JG. Cellular Angiofibroma of the Vulva. *Gynecologic Oncology.* 2001; 81(2): 326-9.
3. Dikmen Y, Yucebilgin MS, Kazandi M, Zekioglu O, Akalin T, Ozdemir N. Cellular angiofibroma of the vulva: report of a case. *Eur J Gynaecol Oncol.* 2004;25(2):242-4.
4. Dufau JP, Soulard R, Gros P. Cellular angiofibroma, angiofibroma and aggressive angiofibroma: members of a spectrum of genital stromal tumours? *Ann Pathol.* 2002;22(3):241-3.
5. Dargent JL, de Saint Aubain N, Galdon MG, Valaey V, Cornut P, Noel JC. Cellular angiofibroma of the vulva: a clinicopathological study of two cases with documentation of some unusual features and review of the literature. *J Cutan Pathol.* 2003;30(6):405-11.
6. Flucke U, van Krieken JH, Mentzel T. Cellular angiofibroma: analysis of 25 cases emphasizing its relationship to spindle cell lipoma and mammary-type myofibroblastoma. *Mod Pathol.* 2011;24(1):82-9.
7. McCluggage WG, Perenyi M, Irwin ST. Recurrent cellular angiofibroma of the vulva. *J Clin Pathol.* 2002;55(6):477-9.
8. Dierickx I, Deraedt K, Poppe W, Verguts J. Aggressive angiofibroma of the vulva: a case report and review of literature. *Archives of Gynecol and Obstet.* 2008;277(6):483-7.
9. Dahiya K, Jain S, Duhan N, Nanda S, Kundu P. Aggressive angiofibroma of vulva and vagina: a series of three cases and review of literature. *Archives of Gynecol and Obstet.* 2011;283(5):1145-8.
10. Sun NX, Li W. Aggressive angiofibroma of the vulva: case report and literature review,” *Journal of International Medical Research.* 2010;38(4):1547-52.
11. Danesh A, Sanei MH. Aggressive angiofibroma of the vulva: dramatic response to gonadotropin-releasing hormone agonist therapy. *Journal of Research in Medical Sciences.* 2007;12(4):217-21.
12. Hameed M, Clarke K, Amera HZ, Mahmetc K, Aisner S. Cellular angiofibroma is genetically similar to spindle cell lipoma: a case report. *Cancer genetics and Cytogenetics.* 2007;177(2):131-4.

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