International Journal Dental and Medical Sciences Research
Volume 5, Issue 2, Mar - Apr 2023 pp 906-908 www.ijdmsrjournal.com ISSN: 2582-6018

Anangiomyxoma of the Vulva- A Bizarre Perineal Lesion A Case Report

Dr Swarna Lakshimi, Dr Usha Rani
Department of obstetrics and gynaecology
Sri Ramachandra medical college andHospital ,Chennai .

Date of Submission: 20-04-2023 Date of Acceptance: 30-04-2023

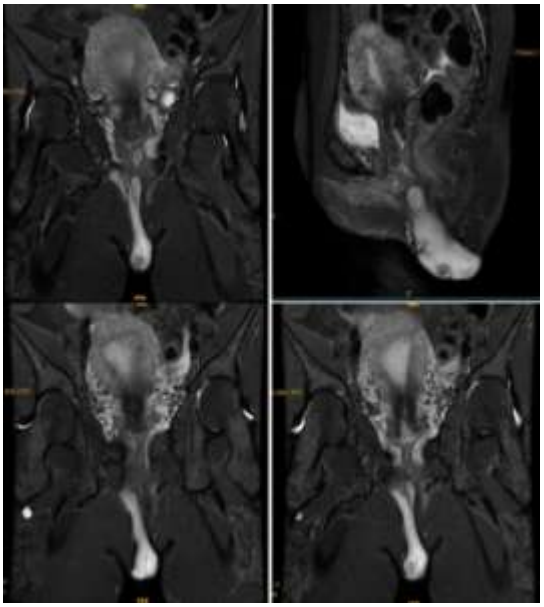
ABSTRACT
Angiomyxoma is a rare,slowly growing and benign tumour of mesenchymal origin, which affects women of reproductive age group and it is associated with a high risk of local recurrence.Here we present a 38 years old Para one live one female with a 10-12 cm swelling over the right labia majora extending upto the perianal region.Histopathologically,the lesion was composed of spindleandstellate shaped cells embedded in a myxoid matrix.Another specific feature is the presence of variable sized thin walled capillaries and thick walled vascular channels.The patient underwent wide local excision of the tumour with clear margins .Angiomyxoma of the vulva needs to be distinguished from benign myxoid tumours with a low risk of local recurrence as well as from malignant myxoid neoplasms. Wide local excision with tumour free margins and occasionally hormonal manipulation is the treatment of choice.

I. INTRODUCTION

Angiomyxoma is a rare mesenchymal neoplasm.Primarily it arises from the soft tissues of the pelvis and perineum of adults.It is a benign looking myxoid and vascular which is infiltrative with a propensity for local recurrence.It affects female 6.6 times more when compared to males and affects a majority of premenopausal group in their mid thirties to forties.It has a moderate to high risk for local relapse.It has a recurrence rate of 30 %.Very few cases have been reported in men involving the scrotum.The origin of the tumour is most likely fibroblastic or myofibroblastic origin.

II. CASE PRESENTATION

A 38 years old female, para one live one, previous normal vaginal delivery, last child birth five and half years ago ,known case of hypothyroid on medications was admitted to the hospital with complaints of swelling in the vulval region for the past two years.The swelling was insidious in onset,initially peanut size and gradually progressed to lemon size.The swelling was not associated with pain or discharge.Therewere no history of excessive white discharge per vaginum or dyspareunia.She had regular menstrual cycles bleeding for four to five days once in 30 days.There were no bowel or bladder disturbances.On clinical examination a diffuse swelling of size was noted over the right side of the perineum extending from the right labia majora to the right side perianal region.No scars, sinuses or ulcers were noted over the surface of the swelling. The swelling was mobile and nontender , soft and spongy in consistency with no associated lymphadenopathy. On gynaecological examination of the cervix , vagina and uterus were unremarkable. Her laboratory investigations were within normal limits and ultrasonography of the abdomen was normal.Moreover , MRI abdomen and pelvis showed loculated fluid collection measuring 18x15x10 mm seen in the right labia with a fluid filled tract of 65mm length,thickness of 20x12 mm seen to extend inferiorly from the labial collection to the right gluteal surface.The collection was 30x30 mm seen just deep to the skin surface.



MRI ABDOMEN AND PELVIS -SHOWS LOCULATED FLUID COLLECTION IN THE RIGHT LABIA

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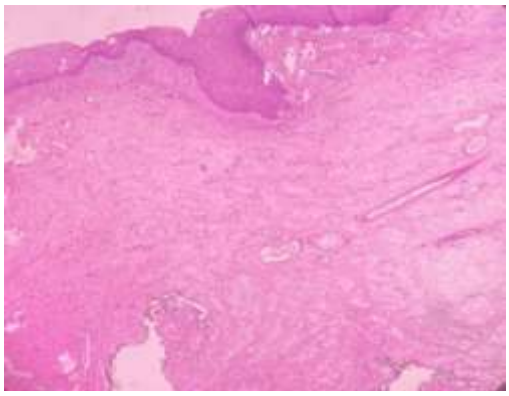
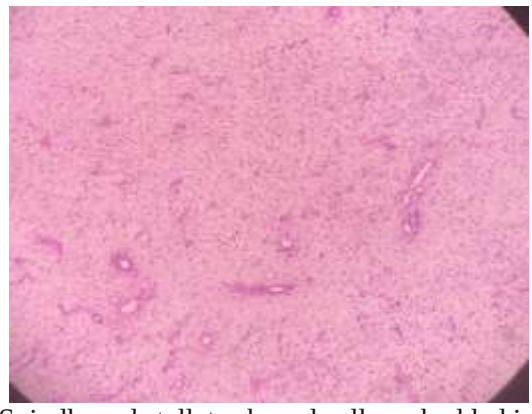
After obtaining anesthesia fitness and consent,the patient was taken up for wide local excision of the tumour and the specimen was sent for frozen section.On cut section, multiple cystic and solid loculations were seen within the swelling.A solid component was cut and pus was drained.Frozen section reported as angiomyxoma of vulva.Perfect hemostasis was secured.

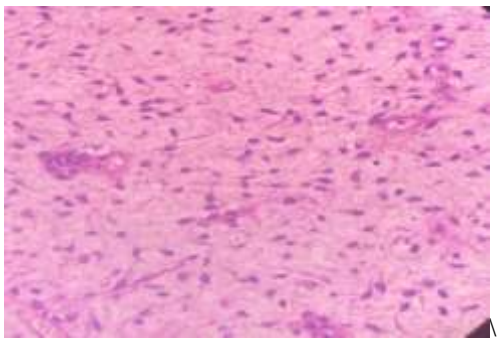
Histopathological examination revealed an encapsulated neoplasm composed of stellate and spindle cells, embedded in a loose matrix with wavy collagen.The lesion contained thin walled capillaries and thick walled vascular channels.Mitosis was 1/10 high power field.No evidence of atypia or necrosis were noted.Immunohistochemistry , the cells were positive for vimentin, actin,desmin and progesterone but negative for S100 protein, CEA,CKAE1/AE3,factor VIII.The background stroma stained positive with alcian blue. Combining the above features , the diagnosis of angiomyxoma was made out.



Spindle and stellate shaped cells embedded in the myxoid stroma



Spindle and stellate shaped cells embedded in the myxoid stroma.Presence of variable sized thin walled capillaries and thick walled vascular channels.



Presence of mitotic figures 1/10 HPF and no evidence of atypia or necrosis

Postoperative period was uneventful.Patient is on a regular follow up every 6 months without any treatment for further,prevention of recurrence and showing no signs of relapse as far and in good health till date.

III. DISCUSSION

In general angiomyxomas are classified as superficial (also known as cutaneous myxoma) or aggressive angiomyxoma.Superficial angiomyxoma occurs in the setting of Carney complex.The tumour usually presents as an asymptomatic mass in the genital area among the women of reproductive age group.Clinically these tumours can be misdiagnosed as BartholinsLipoma ,Gartner Cyst,angiofibroblastoma,cellular angiofibroma and other smooth muscle tumours need to be ruled out.The tumour is hormonally responsive and arises from specialized mesenchymal cells of the pelvis, perineal region or from the multipotent perivascular progenitor cells,which often display variable myofibroblast and fibroblastic features.(Alameda.F et al.).

Recent studies suggest the involvement of chromosome 12 at 12q13-15 which is called high – mobility group protein isoform 1-C (HMG1-C) that encodes the protein involved in the transcriptional regulation.On subjection to immunohistochemistry,most express different combinations of estrogen and progesterone receptors,vimentin,desmin, smooth muscle actin CD34 and CD44,but are invariably negative for S-100, carcinoembryonic antigen and keratin.

Radiologiocallythese tumour have a well defined margin with attenuations less than that of the muscle on CT scan and high signal intensity on MRI that are attributed to the presence of loose myxoid matrix and high water content.

Han-Geurtiset alproposed the following guidelines for treating angiomyoma as follows;

- ✓ Complete excision of the lesion when possible, avoiding multilating surgery.

- ✓ Adjunct therapy using arterial embolization and/or hormonal treatment needed in case of partial resection of the tumour.
- ✓ Radiotherapy is reserved for cases that are resistant to embolization and/or hormonal therapy and still symptomatic.

Hormone therapy with tamoxifene, raloxifene and gonadotrophin releasing hormones has been tried and results in downsizing of the tumour in the neoadjuvant settings.Long term follow up includes MRI for detecting recurrences as the most effective imaging modality.

IV. CONCLUSION

The case report demonstrated the challenges that could arise when dealing with a vulvar swelling.High level of suspicion is needed to make a clinical diagnosis.These tumours must be treated via surgical resection avoiding mutilating surgeries granting a lower recurrence rate.They require close and long term follow up.

There are no conflict of interest.

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