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Pathology

PERIDUCTAL STROMAL SARCOMA (LOW GRADE) WITH PROGRESSION TO BORDERLINE PHYLLODES TUMOR- A RARE CASE REPORT.

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ABSTRACT

INTRODUCTION: Periductal stromal sarcoma is a rare fibroepithelial tumor of the breast that exhibits low grade malignancy and intermediate behaviour. It is characterized by proliferation of atypical spindle cells surrounding benign ductal elements and infiltrating adjacent adipose tissue. Periductal stromal sarcoma is distinguished from phyllodes tumor by its lack of leaf like architechture.

CASE REPORT: This case describes a 40 years old lady who presented with a lump in the right breast since 5 months and had undergone lumpectomy. Microscopic features and immunohistochemical findings are that of periductal stromal sarcoma (low grade) with progression to borderline phyllodes tumor.

DISCUSSION: The number of available studies on PDSS is currently limited and therapeutic strategy yet to be determined. Histological diagnosis of PDSS is based on the criteria established by the Armed Forces Institute of Pathology (AFIP) and surgery with significant free margin is the cornerstone of treatment.

KEYWORDS : Fibroepithelial, phyllodes, PDSS.

INTRODUCTION

Periductal stromal sarcoma is an extremely rare malignant fibroepithelial tumor of the breast which is characterized by its biphasic histology with benign ductal elements and a surrounding sarcomatous stroma made of spindle cells and lacking phyllodes architecture. The clinical presentation of a breast mass is non-specific and does not aid in the differentiation of this tumor from benign or malignant lesions. Similar to phyllodes tumors, PSS has a tendency for local recurrence when incompletely excised, and a potential to develop specific soft tissue sarcomas, as well as metastasis in cases harboring more aggressive sarcomatous patterns.

CASE REPORT

This case describes a 40 years old lady who presented with a lump in the right breast since 5 months. The patient had undergone lumpectomy 7 years back in the same breast and a diagnosis of fibroadenoma was histopathologically confirmed. On presentation of recurrence of lump in the right breast 5 months back, she has again undergone lumpectomy. Received multiple grayish pieces of tissue aggregate measuring $11\times6\times3cm^3$,largest one measures $5\times3\times2cm^3$.On cut open whitish homogenous area noted.

Microscopically, multiple random section from tumor show multiple nodular structure composed of central open tubules or glands with periductal proliferation of spindle cells with mild cellular atypia (3 to 4 mitotic count/10HPF) separated by adipose tissue and fibrous stroma. Focal area show leaf like stromal proliferation with cleft like spaces also noted. Areas of moderate stromal cellular atypia with (mitotic figure 7 to 8/10HPF) also seen. Microscopic invasion of tumor border noted. No vascular and neural invasion seen No areas of haemorrhage and necrosis noted. Immunohistochemistry for CD34 is focally positive in stromal component and CK is positive in epithelial and myoepithelial cells but negative in stromal component.

The above histopathological features and IHC findings are that of periductal stromal sarcoma (low grade) with progression to borderline phyllodes tumor.

DISCUSSION

PSS was previously considered to be a variant of cystosarcoma with adipose metaplasia ; however, currently, PSS is recognized as a separate entity and was classified by the World Health Organization in 2002. PSS occurs in perimenopausal and postmenopausal women (median age, 55.3 years) who are a decade older compared with those presenting with phyllodes tumor (median age, 45 years), however it has also been described in a 14 year old boy. The common symptoms of PSS are similar to other benign and malignant breast tumors and have no radiological specificity. This tumor is characterized by a hypercellular proliferation of spindle cells forming cuffs around well-preserved ductal units with infiltration of the fat and surrounding tissue. Adjacent cuffs may coalesce to form large nodules and extend into lobules surrounding open tubules and ducts. This is in contrast to mammary stromal sarcomas, which displace normal mammary tissue, entrapping ducts and lobules peripherally.

The histological characteristics of PSS were defined by the Armed Forces Institute of Pathology (AFIP) as follows: i) A predominant spindle cell proliferation of variable cellularity and atypia around open tubules and ducts devoid of a phyllodes tumor pattern; ii) One or multiple nodules separated by adipose tissue; iii) Stromal mitotic activity of ≥3/10 high-power fields; and iv) Stromal infiltration into the surrounding breast tissue.

The histological grading depends on atypia and mitotic activity and ranges from low- to high-grade PSS. Immunohistochemically, PSS is CD34-positive and lacks S-100, ER and PR expression. The tendency of PSS to recur and progress into phyllodes tumor or soft tissue sarcomas, as well as the occasional appearance of intraepithelial changes ranging from ordinary hyperplasia to intraductal carcinoma , indicate that close follow-up is required. PSS is a tumor of intermediate behavior, resection with significant margins is generally considered sufficient and axillary lymphadenectomy is not required. With regard to adjuvant therapy, the currently available literature does not suggest any benefit of radiotherapy or chemotherapy.

CONCLUSION

The number of available studies on PSS is currently limited and a therapeutic strategy for PSS has yet to be determined. Histological diagnosis of PSS is based on the criteria established by the AFIP, and surgery with significant margins is the cornerstone of treatment. The prognosis of patients with PSS remains unclear; thus, increased experience of such cases and a longer follow-up period are required to investigate the optimal management and clinical behavior of this neoplasm.

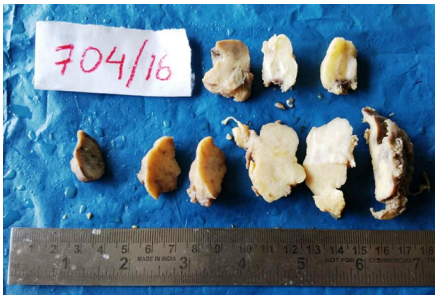

Fig:1 Picture showing cut section of the specimen.

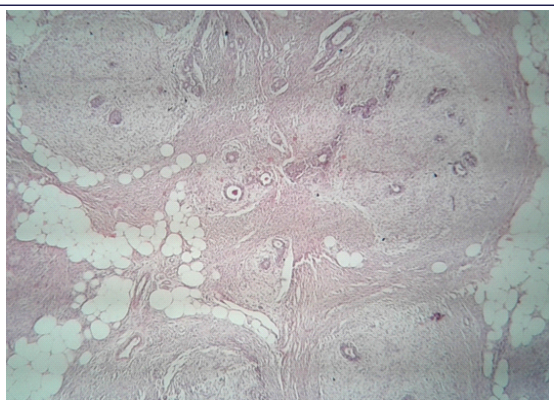

Fig 2; Photomicrograph showing multiple nodular structure composed of central open tubules or glands with periductal proliferation of spindle cells separated by adipose tissue and fibrous stroma. (H&E,4X)

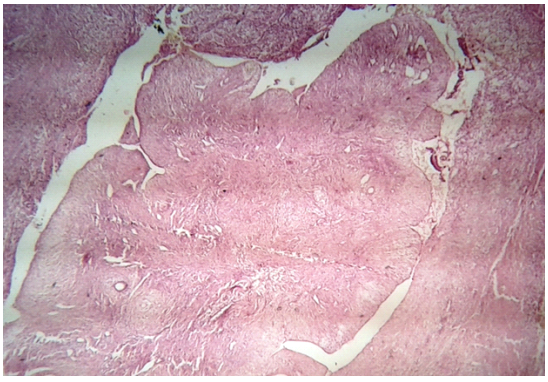

Fig 3; Photomicrograph showing area of leaf like stromal proliferation with cleft like spaces. (H&E,4x)

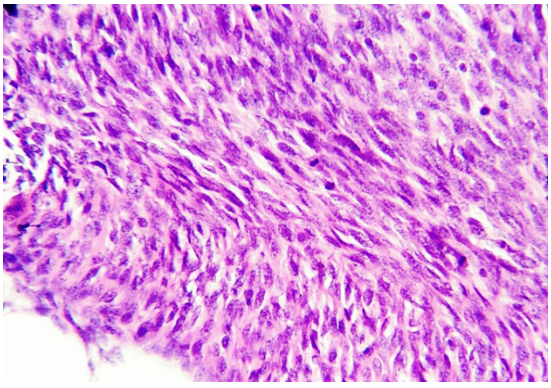

Fig 4; Photomicrograph showing areas of moderate stromal cellular atypia with mitosis. (H&E,40x)

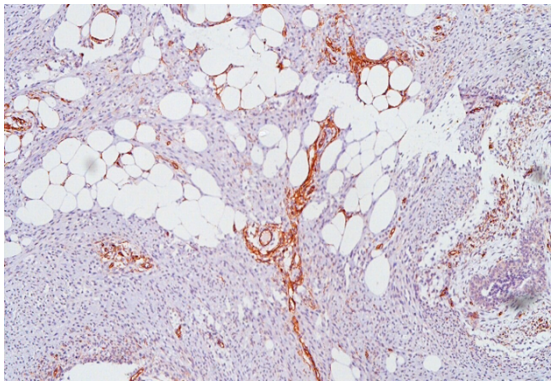

Fig 5; Photomicrograph showing focal positivity of CD34 in stromal cells.(IHC,10x)

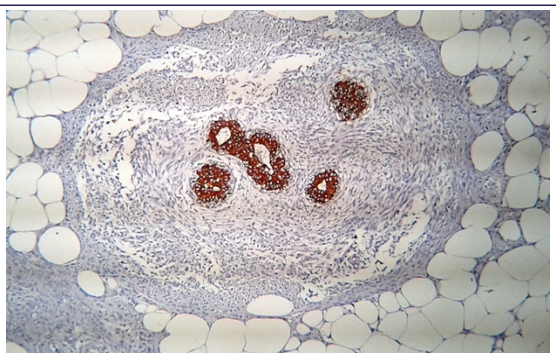

Fig:6 Photomicrograph showing CK negative (stromal component) and positive in epithelial & myoepithelial component (IHC,10X).

REFERENCES:

1. Periductal stromal tumor: a rare lesion with low-grade sarcomatous behavior AM Burga, FA Tavassoli - The American journal of surgical …, 2003 - journals.lww.com.
2. Low‐grade periductal stromal sarcoma of the breast with myxoid features: Immunohistochemistry D Tomas, D Janković, Marušić- Pathology …, 2009 - Wiley Online Library.
3. Cystosarcoma phyllodes: a clinicopathologic study of twenty-six hypercellular periductal stromal tumors of the breast WR Hart, RC Bauer… - American journal of …, 1978 - ajcp.oxfordjournals. org
4. Periductal stromal sarcoma in a child: a case Report . Ouafae Masbah, Issam Lalya, Nawfel Mellas et al.
5. Periductal stromal sarcoma of the breast: A case report and review of the literature YANDAN LAN*, JIA ZHU*, JIANLUN LIU, HUAWEI YANG, YI JIANG and WEI WEI Department of Breast Surgery, The Affiliated Cancer Hospital of Guangxi Medical University.