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MASQUERADING MAMMARY HAMARTOMAS

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ABSTRACT: INTRODUCTION: Hamartoma of the breast is a rare entity, frequently underdiagnosed by pathologists. It is a benign tumour-like nodule with overgrowth of mature cells that normally occur in the affected part. Patients present at an average of 45 yrs. with a unilateral, well defined, painless mass or enlargement; incidence being 0.1% to 0.7%. The aim of our study was to investigate clinical and histopathological features of breast hamartomas. **MATERIALS AND METHODS:** Eighteen cases of breast hamartomas operated our hospital, between Jan. 2005 to Dec. 2012 were included in this study. The clinicopathological features and follow up data were analysed in all the cases. **RESULTS:** All cases in our study were female patients with age group ranged between 23-40 yrs. All patients presented with unilateral breast masses. Clinically majority of patients were suspected to have fibroadenoma with only 2 cases suspected to be cystosarcoma phyllodes. Lumpectomy was performed in all the cases. Grossly all tumors were well circumscribed, grey white, lenticular masses with some showing cystic changes, myxoid areas and nodular areas. The maximum dimension of the tumors ranged from 4 to 16 cms. Microscopically, all hamartomas demonstrated good demarcation with fibrous tissue condensation and hyalinisation. Epitheliosis, apocrine metaplasia, pseudo angiomatous stromal hyperplasia, atrophy and cystically dilated ducts were among the major microscopic features observed. No recurrence or malignant components were noted in our case series. **CONCLUSION:** The diagnosis of hamartoma is difficult as no specific diagnostic histological features are present. The presence of fibrous tissue within the lobules, or fibrous tissue and fat in the stroma, with or without pseudoangiomatous changes, should alert the pathologist to the possibility of a hamartoma. The role of FNAC and needle core biopsy is limited. Coincidental malignancy can occur in hamartomas and hence the importance of its diagnosis.

KEYWORDS: Mammary hamartoma, Lenticular, Pseudoangiomatous change.

INTRODUCTION: Breast hamartoma is a rare, uncommon, frequently underdiagnosed breast tumor. It approximately accounts for about 4.8% of all benign breast lesions.^[1,2] The word hamartoma was first coined and described by Arrigoni et al. in 1971.^[1,3] Patients usually present with a unilateral, well defined, painless mass or enlargement at an average age of 45yrs. The reported incidence of this entity ranges between 0.1% to 0.7%.^[1,3,4] Histologically it is a well encapsulated lesion composed of an overgrowth of mature cells that normally occur in the affected part. The pathologic features of hamartomas are not well known and hence it is often underdiagnosed.^[1,5] This retrospective study was designed to present the clinicopathologic findings of eighteen patients with breast hamartoma.

MATERIAL AND METHODS: Eighteen cases of breast hamartomas operated at our hospital, between Jan. 2005 to Dec. 2012 were included in this study. The clinicopathological features and follow up data were analysed in all the cases.

ORIGINAL ARTICLE

RESULTS: All cases reviewed in the present study were female patients. The age ranged between 23-40 yrs. Majority of the patients presented with unilateral painless breast masses. One patient had bilateral masses. The lesion was associated with pain only in 2 cases. Ten hamartomas were on the left and eight were on the right side. Clinically majority of patients were suspected to have fibroadenoma with only 2 cases suspected to be cystosarcoma phyllodes. Fine needle aspiration cytology was performed in two cases, which did not yield significant results. Lumpectomy was performed in all the cases.

Grossly all tumors were well circumscribed, grey white, lenticular masses with some showing cystic changes, myxoid areas and nodular areas. [Fig. 1] The maximum dimension of the tumors ranged between 4 to 16cms. Microscopically, all hamartomas demonstrated good demarcation with fibrous tissue condensation [Interlobular fibrosis] and hyalinisation. Epitheliosis, apocrine metaplasia, pseudo angiomatous stromal hyperplasia, atrophy and cystically dilated ducts were among the major microscopic features observed. [Fig. 2, 3, 4, 5, 6] One case showed xanthogranulomatous change due to cyst rupture. [Table 1] One patient who had bilateral masses was diagnosed to have fibroadenoma in the contralateral breast. This was also confirmed on histopathological evaluation. None of the cases showed any adjoining malignant component.

Gender	All female patients
Age	Mean: 29 [23-40]
Clinical diagnosis	Fibroadenoma [16/18] 88.8% Cystosarcoma phyllodes [2/18] 11.1%
Surgical procedure	Lumpectomy in all cases
Gross features	All cases the tumor was well circumscribed and lenticular Few cases showed myxoid degeneration, cystic areas, nodular areas and slit like spaces
Microscopy	
- Interlobular fibrotic stroma and hyalinisation	18 [100%]
- Adipose tissue	18 [100%]
- Pseudoangiomatous stromal hyperplasia	10 [55.5%]
- Cystic change	11 [61.1%]
- Apocrine metaplasia	11 [61.1%]
- Atrophy	2 [11.1%]
- Epitheliosis	13 [72.2%]
- Adenosis	7 [38.8%]
- Blunt duct adenosis	3 [16.6%]
- Xanthogranulomatous change,	1 [5.5%]
- Chondromyxoid change,	1 [5.5%]
- Myoepithelial cell hyperplasia	1 [5.5%]
Total No. of cases	18

Table 1: Clinicopathological Features of 18 Patients with Breast Hamartomas

ORIGINAL ARTICLE

DISCUSSION: Hamartomas are well-circumscribed, rare lesions composed of benign mammary tissue often underdiagnosed by pathologists. Prym et al initially defined them as mastomas. They are also known adenolipomas, fibroadenolipomas or lipofibroadenomas. In 1973, Davies and Riddell used the term myoid hamartoma to a rare form of hamartoma with significant smooth muscle component.^[1,5,6,7]

In the case series of breast hamartomas by Charpin et al and Sevim et al, the age ranged between 19-56 yrs. and 33.5-66.5 yrs respectively. In our study the age ranged between 23-40 yrs. few authors have noted them to occur typically in women older than 35 years of age. In the present series 6 patients were above 35 years of age.^[1,5,8,9] 16 cases in the present study were suspected to be fibroadenomas and 2 cases were cystosarcoma phyllodes on clinical examination.

Ultrasound examination was performed only in 10 cases which showed features suggestive of fibroadenoma/hamartoma. On ultrasonography, mammary hamartomas show sharp definition and displacement of surrounding structures. They contain sonolucent fat and echogenic fibrous components with a heterogeneous internal echo pattern. The mammographic appearance is typically described as 'slice of containing fat, varying radiodense fibrous & 'salami' or 'piece of cut sausage' or 'breast in breast' appearance.^[9,10,11] Mammography was not done in any of the cases in the present study.

No specific histopathologic features of hamartomas are known. However, on macroscopic examination, hamartomas are typically well-circumscribed, lenticular, round or oval masses with heterogeneous features on cut section, depending on the particular prominence of histological components, mainly white and/or pinkish, with yellow islands.^[11,12,13,14] These features were also noted in the present study.

Hamartomas of the breast are thought to arise from dysgenesis and sex steroids have also known to be implicated in their development. On microscopy, the presence of fibrotic and hyaline stroma which surrounds and extends between individual lobules leading to obliteration of the usual interlobular specialized loose stroma, is observed by many authors to be the most characteristic feature.^[1,5,7] This is referred to as interlobular fibrosis. However this feature is also seen in other benign proliferative conditions like sclerosing adenosis. This feature along with the presence of adipose tissue is helpful in distinguishing hamartomas from fibroadenomas microscopically.^[5,8]

Adipose tissue within the stroma is also commonly reported in hamartomas. In most series,^[2,3,5,6] adipose tissue is present in more than 90% of the cases. In the present study we found adipose tissue to be present in all the cases similar to the study of Tse, Law, Ma, et al.^[7]

McGuire et al. and Jones et al have classified hamartomas of the breast based on fibrous and adipocytic components. However these criteria are not used by pathologists and are not known to have an impact on the clinical management or prognosis of these patients.^[8,10]

Pseudoangiomatous stromal hyperplasia is another notable feature of breast hamartomas. This feature has also shown to vary among different studies. Daya et al and Tse, Law, Ma, et al and Fischer et al have observed pseudoangiomatous stromal hyperplasia in 20%, 32% and 71% of hamartomas respectively in their case series. This feature was noted in 55.5% of cases in the present study which was consistent with the rates reported in literature.^[7,9,11]

Cystic changes and apocrine metaplasia were noted in more than 50% of cases in our study. However this feature was observed to a lesser extent by few other authors.^[5,7,8] Other rare features that have been described in the literature include microcalcification, myxoid differentiation, stromal oedema and stromal giant cells.

ORIGINAL ARTICLE

Some special features observed in our study were xanthogranulomatous response to cyst rupture, chondromyxoid change and myoepithelial hyperplasia. No malignant lesions or recurrence were noted in our study. However invasive ductal carcinoma and myofibroblastoma are some tumors known to arise from mammary hamartomas.^[15,16,17]

CONCLUSION: Due to nonspecific diagnostic histological features, the diagnosis of hamartoma is challenging. Macroscopic examination of lumpectomy specimens should be performed with caution. Lenticular mass on gross examination and the presence of fibrous tissue within the lobules, or fibrous tissue and fat in the stroma, with or without pseudoangiomatous changes, should alert the pathologist to the possibility of a hamartoma. The role of FNAC and needle core biopsy is limited. Recurrence and coincidental malignancy can occur in hamartomas and hence the importance of its diagnosis.

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ORIGINAL ARTICLE

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Fig. 1: Characteristic Lens-Like Morphology of Breast Hamartoma with Abundant Fibrous Tissue.

Fig. 2: Photomicrograph Shows Interlobular Fibrotic Stroma along with Adipose Tissue [H & E x40].

Fig. 1

Fig. 2

Fig. 3: Stroma Showing Pseudoangiomatous Hyperplasia [H & E x400].

Fig. 4: TDLU's with Mild Epitheliosis with Surrounding Stroma Showing Pseudoangiomatous Hyperplasia [H&E x200].

Fig. 3

Fig. 4

ORIGINAL ARTICLE

Fig. 5: Photomicrograph Showing Focal Apocrine Snouting [H & Ex400].

Fig. 6: Photomicrograph Showing TDLU's with Adenosis [H & E x200].

Fig. 5

Fig. 6

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