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# Myoid Hamartoma: An Exceptionally Rare Breast Lesion

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**Breast hamartomas (BH) account for 3.9-4.8% of all breast tumors. The myoid or the muscular variant, Myoid Breast Hamartoma (MBH) is exceptionally rare with less than 30 reported cases in the English medical literature. This paper reports a case of MBH in 53-year old female who presented with 6-year-old left breast lump which has been gradually increasing in size. Clinically, the mass was suggestive of a fibroadenoma; however, microcalcifications on ultrasonography were an indication for further investigation with Fine Needle Aspiration Cytology (FNAC). Cytologically, the yield was in favor of a fibroadenoma. Lumpectomy was performed without any complication and the mass was finally diagnosed as MBH histopathologically. The histologic diagnosis was based on the findings of predominant stromal smooth muscle bundles (proved by immunohistochemistry) along with scattered intact terminal duct-lobular units, dilated ducts and entrapped islands of mature fibroadipose tissue.**

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The first description of BH has been attributed to Albrecht in 1904. Hamartoma in general refers to a benign localized malformed tumor-like mass that can impinge on almost any part of the body. It is composed of disorganized mature tissue elements normally found at the affected part. Hamartomatous tissue can be either in the form of an abnormal quantity, an abnormal structure, or an abnormal degree of maturation. It usually has one predominant tissue, but with an overall similar growth rate to the surrounding normal tissues. Hamartomas are often asymptomatic especially when small. They can be solitary, multiple and bilateral as well as being part of clinical manifestations of some genetic disorders such as Cowden's disease[1]. In this paper, the author reports a case of MBH, describes the clinico-radiologic presentations with special emphasis on the morpho-immunophenotypic findings.

## The Case

Fifty-three year-old Bahraini woman presented with a slowly growing left breast lump which has been present for the past 6-years. Neither significant past history of breast diseases nor other medical problems were stated. Physical examination revealed a well-circumscribed, 4.5 x 3.5 cm, painless, rubbery mass in the upper central aspect of the left breast. On Ultrasonography, the lump was fairly well-outlined with complex scattered echogenic areas of calcifications. The overall appearance was in

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favor of a Fibroadenoma, however due to the presence of microcalcifications, malignancy could not be excluded and mammography and FNAC were recommended for further assessment. Mammography was not performed. Ultrasound guided FNAC suggested a fibroadenoma. The aspirate was moderately cellular and contains numerous cohesive sheets of benign bimodal ductal epithelial cells, fibroadipose micro-fragments and many bare bipolar myoepithelial nuclei in the background. The lesion was removed by a simple lumpectomy ten days later. The 18 gram mass measured 4.5 x 3.8 x 2.5 cm, and was well circumscribed, rubbery in consistency, resembling a regular fibroadenoma with a yellowish white solid cut surface (Figure 1).

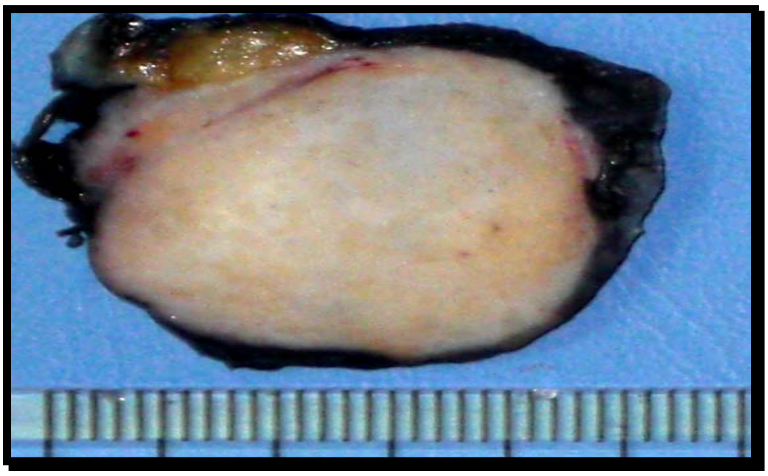

**Figure 1: Myoid Breast Hamartoma: A Well Circumscribed, Non-Encapsulated Lump, Much Resembling a Regular Fibroadenoma with Yellowish White Solid Cut Surface**

Microscopically, the lesion was composed of scattered normal terminal lobular-ductal units within a dominant overgrowth of stromal spindle shaped smooth muscle both in loose fibers and tangled bundles (Figure 2).

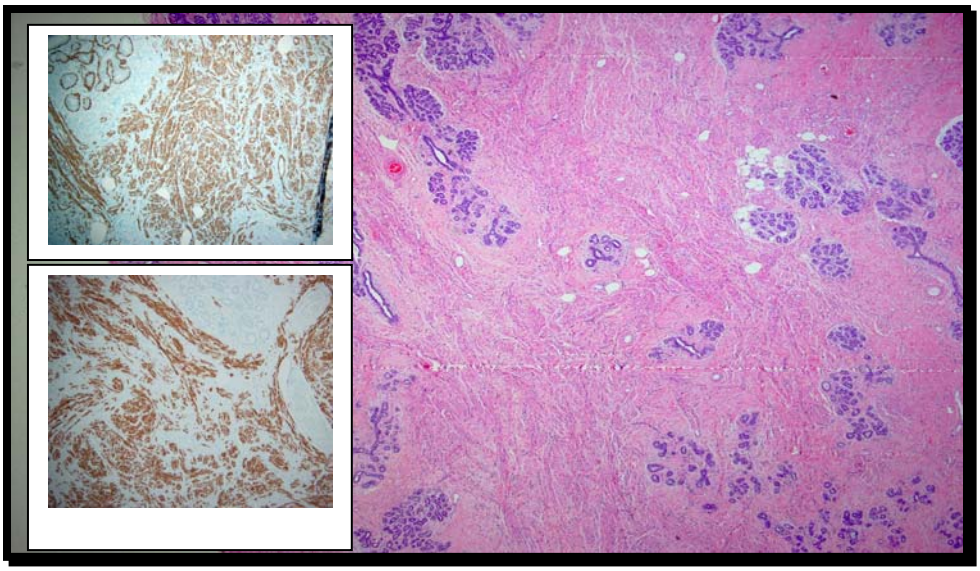

**Figure 2: Microscopically, the lesion was composed of scattered normal terminal lobular-ductal unites within a dominant overgrowth of stromal spindle shaped smooth muscle both in loose fibers and tangled bundles; Insert: The dominating smooth muscle tissue stained diffusely positive for smooth muscle actin (Top) and desmin (bottom), however SMA has also stained the basal myoepithelial cells of the terminal ducts lobular units.**

Occasional islands of mature adipose tissue were present along with benign angiomatous proliferation. No typical pseudo-angiomatous stromal hyperplasia was found. Neither cytoarchitectural atypia nor significant mitosis was seen. The dominating smooth muscle tissue stained diffusely positive for smooth muscle actin (SMA) and desmin; however, smooth muscle actin, has also stained the basal myoepithelial cells of the terminal ducts lobular units. Estrogen receptors and Progestrone receptors were strongly positive. The CD34 marker stained only the specialized periductal and perilobular stroma as well as highlighted the rich vascular intralesional network. Other markers including myoglobin, MyoD1, S-100, CD99 and bcl-2 were all negative. Myoid hamartoma was diagnosed, and as the lesion was completely excised, no further surgical intervention was undertaken and the patient reported no further complaints for the past six months.

## DISCUSSION

Increased patient awareness and the ongoing breast screening programs have resulted in reporting more and more BH in the last two decades. Patients (age range 13-89 years) with BH can present in three patterns, a slow growing well-outlined, palpable, painless mass, macromastia without a palpable localized mass lesion, or with a discrete lesion detected on mammography. BH can be single or multiple[2]. It can be seen in other extra-mammary locations such as the axilla and the inguinal region. BH is similar to the surrounding mammary tissue, which is influenced by hormones and may enlarge during pregnancy and lactation. These lesions are mostly mistaken for other more common mammary lesions such as fibroadenoma, low grade phyllodes tumor or lipoma, depending on the dominating tissue composition[3].

Typical BH has a characteristic appearance on Mammography (MG), Ultrasonography (US) and CT scans (when indicated). On mammography, BH gives the so called 'breast in the breast' appearance; they come out as an encapsulated, round shaped lesions with well-circumscribed borders that usually demonstrate internal fat densities with radiolucent halos. The main familiar sonographic appearance is an oval shaped, well defined lesion with heterogeneous internal echogenicity (either hyperechoic or mixed echogenicity); these changes could be associated with cystic areas, echolucent halos or posterior enhancements (i.e. absent retro tumor acoustic phenomena). In some cases, the mammographic and/or the ultrasonic image can be non-specific and may mimic, to a large extent, a fibroadenoma, juvenile hypertrophy or a phylloides tumor. The change of the mammographic round shaped mass into an elongated oval shaped mass by sonography can largely suggest compressibility and therefore serves as one of the main distinctive features of BH. Internal calcification is not a constant feature but can be seen also[2,4].

For those borderline, atypical or inconclusive lesions with BH-Like appearance on MG and/or US, it is recommended to combine the mammography, ultrasonography and CT studies with the FNAC or Tru-cut biopsy findings to reach a more decisive diagnosis[5].

Grossly, BH looks much like a fibroadenoma, it could be spherical, or disc-shape and range in size from 1.0-14 cm with a mean of 3 cm. The cut surface is solid, well-circumscribed but rarely with indistinct margins, and depending on the tissue component, it could be grayish-white or yellowish-white.

Microscopically, the presence of both intact lobules and ducts seems to be a consistent significant diagnostic feature of BH, unlike fibroadenoma in which intact lobules are often absent. BH consists of varying amounts of normal mammary tissue including fibroadipose tissue, normal lobules and ducts, apocrine cysts, areas of sclerosing adenosis, smooth muscle tissue and probable areas of pseudo-angiomatous stromal hyperplasia[2]. It is important to emphasize that BH lacks the characteristic growth patterns attributed to most described entities arising in the breast like fibroadenoma, myolipoma, fibroadenolipoma, adenolipoma, adenohibernoma, and chondrolipomas; those entities should not qualify as a hematoma. Likewise, MBH can demonstrate a close resemblance to breast myoma or other mammary spindle cell stromal tumors. MBH has a characteristic H&E morphology and beside the usual disorganized indigenous mammary tissue described before, it shows a characteristic stromal dominance of spindle shaped smooth muscle fibers that run in patternless bundles or around blood channels. MBH is positive for alpha-smooth muscle actin (SMA) as a rule; S-100 protein, myoglobin, and keratin are negative, and vimentin is variable[6].

It was postulated that the origin of MBH is derived from the myoepithelium of the vascular blood channels or the muscularis mammillae of the areola[7]. It was also suggested that MBH has a dual cell origin from both myoepithelial and myofibroblastic components in variable combinations. The expression of CD34 antigen in smooth muscle cells suggests that MBH originates from stromal cells via leiomyomatous metaplastic changes[8].

Local recurrence after incomplete excision were reported in some BH, likewise, a coexistent malignant change have also been described; some of those cases were either associated with

Cowden's syndrome or atypical mammographic microcalcifications. Examples of such malignancies are in-situ and invasive ductal carcinoma, invasive lobular carcinoma and lobular neoplasia within a myoid hamartoma. The management of such lesions should follow the standard protocol for managing breast cancers[9,10].

Histological evaluation of any unfamiliar mammary lesion must be attempted in conjunction with the clinical and radiologic findings. Good attention must be paid for the cellular composition of the lesion (biphasic or monophasic), the predominant cellular component (stromal or epithelial), presence of atypia (within the stromal and/or epithelial elements) and the overall disease frequency. No doubt, immunohistochemistry is indispensable.

In the author opinion, the key role of the FNAC is to differentiate between benign (C2) and malignant (C5) breast lesions and to refer equivocal cases (Categories C3 and C4) for further investigations such as an intraoperative frozen section assessment. Presence of certain cyto-morphologic features such as finding intact lobular units and relative paucity of stromal component can favor breast hamartoma over fibroadenoma on FNAC examination. However, the author, like many others, believes it is relatively unfruitful practice to take unnecessary jeopardy of a potential diagnostic pitfall, since it will not affect the immediate clinical management. Nevertheless, it can open the door to undermine the pathologist skills[11]. It is known that BH can be totally overlooked particularly on Tru-cut biopsy. The sampled tissue is either non-evocative 'normal' breast tissue or abundant adipose tissue. In such situations and without considering the clinico-radiologic findings the pathologist may report the findings as inadequate, normal mammary tissue, or even a lipoma. This potential pitfall is inappropriate for a clinically or radiologically overt lesion.

Hamartoma's Tru-cut biopsy could be easily confused with other benign entities such as fibroadenoma, pseudo-angiomatous stromal hyperplasia and fibrocystic change. The Tru-cut biopsy advocators claim that presence of fibroadipose tissue within the breast lobules or fibrous septae, or the presence of fatty islands within the stoma will enable them to confidently diagnose hamartoma, especially when pseudo-angiomatous stromal hyperplasia is encountered within favorable clinico-radiologic findings[12]. The verdict is neither the cytologic features nor histologic appearance in Tru-cut biopsy are enough to distinguish hamartomas from other benign and malignant lesion, in fact many reports have well documented such morphologic overlapping just like in this reported case.

## CONCLUSION

**In this report, the author presented a rare case of myoid breast hamartoma in a 53-year old woman. The clinical, radiologic and FNAC findings were suggestive of a fibroadenoma, however the definitive diagnosis was made on histological examination. Breast hamartomas are rare lesion, but whenever unfamiliar presentation of benign breast lesion is encountered with microcalcifications, this entity should be considered and coexistent malignancy should be ruled out.**

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