

### **Primary Chondrosarcoma of the Breast: A Rare Case Report**

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#### **Abstract**

Pure sarcomas are very uncommon tumours of the breast. They represent a diverse and heterogeneous group of malignant neoplasms that arise from the mammary stroma. A diagnosis of primary mammary sarcoma can be established only after excluding metaplastic carcinoma and malignant cystosarcoma phyllodes where malignant cartilaginous areas may be present, by extensive sampling for evidence of ductal carcinoma in situ or invasive mammary carcinoma. This case report describes a very rare primary chondrosarcoma of the breast in a 50 year old female diagnosed precisely on fine-needle aspiration cytology and confirmed by histopathological examination after total mastectomy.

**Keywords :** Breast, Chondrosarcoma

#### **Introduction**

Primary pure sarcomas of the breast are extremely rare ranging from 0.5% to less than 1% of all breast malignancies. Of these, pure chondrosarcomas without any other area of epithelial or mesenchymal differentiation are even rarer.<sup>[1]</sup> They are non-epithelial

malignancies and contain chondrosarcomatous areas throughout the tumour, arising from mesenchymal mammary stroma.<sup>[2]</sup> Differential diagnosis involves cystosarcoma phyllodes and metaplastic carcinoma of breast with chondroid differentiation.<sup>[3]</sup> Although their clinical features mimic breast carcinomas in some ways, therapy and prognosis differ dramatically.<sup>[4]</sup>

#### **Case report**

A 50 years old female patient presented at a rural hospital with a lump in the right breast since 10 months. There was no history of lump or pain anywhere else in the body. There was neither family history of breast cancer nor history of any exposure to radiation. On examination, a lump of size 7X8 cm involving whole of the right breast was palpable. It was hard, non-tender and fixed to the underlying structures. The overlying skin and nipple-areola complex appeared normal. There was no axillary lymphadenopathy. The contralateral breast was normal. Rest of the systemic and general examination was unremarkable. Her routine laboratory investigations were normal. Chest radiograph was also normal with normal ribs. A provisional clinical diagnosis of carcinoma of

breast was made and patient was advised fine needle aspiration cytology (FNAC). The FNAC smear showed few clusters and scattered pleomorphic tumour cells against a chondroid and chondromyxoid background (Figure 1). High power examination of FNAC smear showed tumour cells with hyperchromatic nucleus enclosed in a lacunae around them. This led to a diagnosis of chondroid tumour probably chondrosarcoma of the right breast. A diligent search for any chondrosarcomatous lesion in bone was made and none found. Patient underwent a right mastectomy. The right mastectomy specimen was received in formalin for histopathological examination. Grossly, the mastectomy specimen measured 10x7x6 cm with no skin covering. On cutting, gritty sensation was felt and a tumour of size 9x6x5 cm, was noted. Cut surface was translucent white, lobulated with areas of haemorrhage (Figure 2). The tumour was seen extending close to the surgical margins. Extensive sampling of the tumour was done for histopathological examination. No lymph nodes were identified in the attached fibrofatty tissue. Microscopic examination revealed tumour mass composed entirely of lobules of chondrocytes separated by fibrous septae. Chondrocytes showed large hyperchromatic nuclei with anisonucleosis and pleomorphism (Figure 3). Areas of calcification, surrounding fibroadipose tissue and skeletal muscle fibres were also seen. The tumour extended upto the posterior margin of surgical excision. Careful examination didn't show any malignant epithelial component. A final diagnosis of primary chondrosarcoma of the breast was made. Patient did not develop any post-operative complications. She was advised follow up.

### Discussion

The term 'stromal sarcoma' was introduced in 1962, the first case of breast angiosarcoma was reported by

Kennedy and Biggart in 1967. As a primary breast tumour, chondrosarcoma may occur in three different forms: as a pure neoplasm (pure chondrosarcoma), as a stromal component of malignant phyllodes tumour or as chondrosarcomatous differentiation in a metaplastic carcinoma.<sup>[5]</sup> Breast sarcomas are a heterogeneous group of neoplasms. Majority of these are malignant fibrous histiocytoma, fibrosarcoma, liposarcoma, angiosarcoma and rhabdosarcoma etc.<sup>[1,2]</sup> They can be primary or secondary. The prevalence, risk factors and clinical course of breast sarcomas and other primary non-epithelial malignancies are less well characterised than are the breast tumours arising from epithelial tissue. A causative factor is not identified in majority of cases. In contrast secondary breast sarcomas are associated with primary radiation therapy or conditions causing chronic lymphedema.<sup>[1]</sup>

Primary and pure chondrosarcoma of the breast is an extremely rare entity. It arise from the breast itself and contains chondrosarcomatoid areas throughout the tumour.<sup>[3]</sup> Very few cases of pure and primary chondrosarcomas of the breast have been reported so far. These tumours are usually large sized, occur in woman more than 40 year old and do not invade the overlying skin. Regional lymphadenopathy is expected in 14-29% of the cases, most of which are reactive hyperplasias.<sup>[7]</sup>

A preoperative clinical and cytological diagnosis of such cases is usually not reached, both due to marked similarity in clinical behaviour to identify ductal carcinoma and low index of suspicion.<sup>[8]</sup> In our case, the presence of pleomorphic tumour cells with hyperchromatic nucleus enclosed in a lacunae around them against abundant chondroid background raised a strong suspicion thereby confirming the diagnosis of primary chondrosarcoma on cytological basis. The malignant chondroid component

can be mistaken for ductal carcinoma, however one should pay attention to the background matrix material and typical chondroblastic morphology. Only occasional stromal fragments can be seen which is nonfavourable for phyllodes tumour, although it is very difficult to distinguish only by cytology.

Even on histopathology, it is sometimes very difficult to distinguish a primary chondrosarcoma from metaplastic breast carcinoma and malignant phyllodes tumour if the chondrosarcomatous component is predominant. In such cases wide sampling is important in order to rule out any foci of in situ or invasive epithelial malignancy. Any such foci would lead to a diagnosis of metaplastic breast carcinoma.<sup>[9]</sup> Another important differential diagnosis in such cases is primary sarcoma of the chest wall metastasising to the breast. The only method to rule out this possibility is thorough search for lesions in chest wall preoperatively through imaging and during surgery.<sup>[8]</sup> They are generally hormone receptors (ER, PR and HER2) negative and show positivity for S-100 and vimentin.<sup>[3]</sup>

The origin of primary sarcoma of the breast is obscure and speculations abound. Misplaced mesenchymal rests, chondroid differentiation of tumours of mesenchymal origin and pluripotent cells from periductal and acinar mesenchymal cells undergoing chondroblastic differentiation have been proposed. The study of microenvironment of sarcomas reveal an interesting situation where cancer cells of mesenchymal origin are surrounded by stromal cells of same origin. At present, the number of documented cases of breast chondrosarcomas is too small to permit a critical discussion of their origin.<sup>[10]</sup>

Surgery remains the treatment of choice for most sarcomas. The role of chemotherapy and radiotherapy is

not yet established due to limited number of cases reported so far. They generally have good prognosis once completely excised as compared to ductal carcinoma.<sup>[8]</sup>

### Conclusion

Preoperative cytological diagnosis is challenging as primary chondrosarcoma of breast usually will not be considered in the differential diagnosis. We offered the precise diagnosis in our case. Histologically extensive sectioning of the tumour is very essential to rule out metaplastic carcinoma and malignant phyllodes tumour. Careful search for ductal epithelial component is mandatory to confirm primary sarcoma of breast. This is important because chondrosarcoma are primarily managed surgically and have good prognosis.

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Figure 2: Gross mastectomy specimen, cut surface of tumour showing lobules of translucent white tissue along with areas of haemorrhage.

#### Figures



Figure 1 : FNAC smear showing cell rich aspirate containing pleomorphic tumour cells against chondromyxoid background (H & E, 10X)

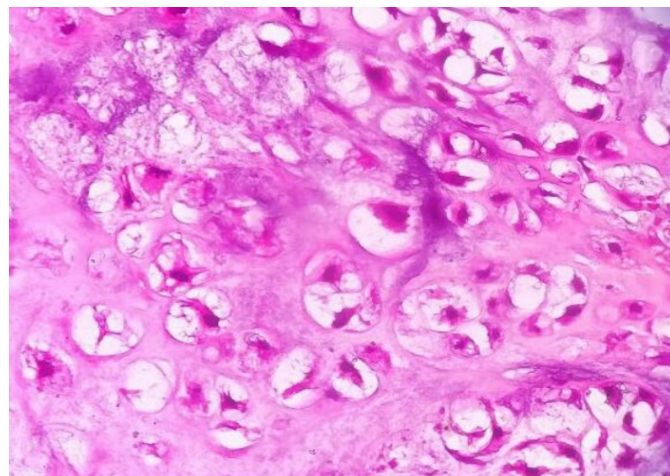


Figure3 : High power examination showing pleomorphic tumour cells with large hyperchromatic nuclei enclosed within lacunae suggestive of chondroblasts.(H & E, 40X)