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آموزش مهارت های کاربردی در تدوین و چاپ مقاله
Malignant Mesenchymal Tumor of Male Breast: Primary Chondrosarcoma

Puneet Kumar Bagri¹, Surendra Beniwal², Ajay Sharma¹

Abstract
Breast Sarcomas have relatively been rare and accounted for 1% of all primary malignant tumors of the breast. Pure and primary chondrosarcoma of the male breast would be an extremely rare tumor. It might arise either from the breast stroma itself, or from underlying bone or cartilage. A 65-year-old man has presented with a rapidly growing breast mass since 5 months. Physical examination has established a large firm to hard mass with regular margins in the region of right breast. There was no axillary lymphadenopathy. Contrast enhanced MRI of breasts has shown a mixed-signal intensity multi lobulated cystic-solid mass (10.4 cm × 10.3 cm × 9.9 cm) appearing predominantly hyper intense on T2W and hypo intense on T1W. The tumor has diagnosed as a low-grade chondrosarcoma of the breast by histopathological and immunohistochemistry analysis. Right sided radical mastectomy with grafting has done. It has seemed to be very important to identify the mammary primary sarcomas as entity separated from the carcinomas of the breast.

Keywords: Chondrosarcoma; Male breast; Radical mastectomy

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Introduction
Malignant tumors which originated from mesenchymal tissue, has occurred very rarely in the breast. The metaplastic carcinomas which have characterized by a combination of mesenchymal and epithelial components would be uncommon malignancies of the breast [1]. Pure primary sarcomas have been the rarest malignancies in mammary tissue. The chondrosarcoma has been a typical example of these rarest tumors of the mesenchymal tissue. Less than 10 cases have published in the literature [2–4].

Case Report
A 65 years old man has presented in our department with a complaint of a right sided breast mass which has rapidly grown for 5 months. Breast palpation has revealed a painless, less mobile, and firm to hard mass of 10×10cm with regular margins in area of right breast. The axilla was clinically negative.

Contrast enhanced MRI (Figures 1A and B) of breasts has revealed huge mixed signal intensity multi lobulated cystic-solid extra-pulmonary mass lesion involving the right anterior chest wall extending from infra clavicular region to the level of xiphisternum measuring 10.4 cm (SI) × 10.3 cm (AP) × 9.9 cm (Trans) appearing predominantly hyper intense on T2W and hypo intense on T1W. Heterogeneous predominantly peripheral solid tumoral enhancement has observed with multiple large loculated central areas of necrosis and intensely enhancing septae. Pre contrast T1W hyper intensity has indicated internal hemorrhage. There was no axillary lymphadenopathy.

Histopathology has revealed the overall appearances of low-grade chondrosarcoma with a tumor composed of lobules of cartilage of varying size separated by fibrous tissue. The tumor has seen to focally infiltrate into surrounding skeletal muscle.

Immunohistochemical studies have performed by standard revealed that chondrosarcomatous elements were positive for S-100 and vimentin, but negative for cytokeratin and also for estrogen and progesterone receptors.
After routine investigations, right sided radical mastectomy with grafting has done. Patient has discharged in well condition postoperatively. But he has lost follow-up after three months.

Discussion

This paper has discussed one of the rarest mesenchymal malignancies of the breast, primary chondrosarcoma. As a primary breast tumor, chondrosarcoma might occur in three different forms: as a pure neoplasm (pure chondrosarcoma), as the stromal component of a histologically malignant phyllodes tumor, or as chondrosarcomatous differentiation in a metaplastic carcinoma. In this report we have reported a case of histologically pure chondrosarcoma.

The primary chondrosarcoma of the breast has been an extremely rare entity. It has contained chondrosarcomatous sectors which resulted from some mammary tissue.

Prognosis of chondrosarcomatous breast tumors has not fully known, because many of the reported cases were difficult to analyze (owing to lack of detailed clinical or morphologic information). These tumors were usually large-sized that have occurred in more than 40 year old woman. Axillary lymph nodes have found in 14-29% of the cases, most of which were reactive hyperplasia. The present case substantiates the clinical findings of previously reported cases [5].

To diagnose a primary chondrosarcoma of the breast, a non-mammary site has been excluded clinically and histologically. Other types of pure sarcoma, such as spindle cell sarcoma, neuroectodermal tumor, angiosarcoma etc, have also been reported [6, 7]. The history of the patient had an important feature as the mass had grown rapidly without systemic and deleterious effect on the patient’s health status. These tumors have tended to grow rapidly and present themselves as a mass for a short duration [2, 3, 7]. A large multilobular mass with regular margins and without a sign of regional invasion has detected by palpation. These findings have appeared as important features of this large tumor. Previous reports have also pointed out that these large tumors have not generally invaded skin and regional lymph nodes despite their locally advanced nature [2–4, 7]. A relatively circumscribed, well demarcated mass, as in our patient, was an imaging characteristic of a pure mesenchymal or a metaplastic carcinoma with sarcomatoid differentiation. It has been reported that imaging studies seldom led to the diagnosis of sarcoma in a suspected benign lesion. The mass was complex echoic on ultrasound and has shown round hyperdense opacity on mammography [4, 7, 8].

The very limited number of such cases has not permitted us to establish an appropriate therapeutic approach. Chondrosarcoma in common locations has generally known as refractory to all types of conventional chemotherapy and radiotherapy. Surgery has remained the only effective treatment [5, 9, 10]. Pure chondrosarcoma and metaplastic cancer of the breast has rarely invaded axillary lymph nodes and would be generally hormone receptor-negative [1, 2, 4, 11]. This extremely rare tumor has tended to grow rapidly, and it was usually large at first physical examination. A large, hyper dense and complex echoic mass with regular margins has given the impression of a benign tumor on mammography and ultrasound. Despite its large size, it has not invaded local and regional structures.

Figure 1. Contrast enhanced MRI of breasts reveals huge mixed signal intensity multilobulated cystic-solid extra-pulmonary mass lesion involving the right anterior chest wall appearing predominantly hyperintense on T2W and hypointense on T1W. Heterogeneous predominantly peripheral solid tumoral enhancement is observed with multiple large loculated central areas of necrosis and intensely enhancing septae.
Regarding systemic management, there was no standard treatment protocol, and a large variety of chemotherapy protocols have been employed in treating this disease.

Systemic therapy principles have been derived from small retrospective case reviews of primary breast chondrosarcomas and extrapolated from studies of non-breast chondrosarcomas, since the clinical behavior and histology were similar. The tumor was negative for any of the hormonal receptors. This fact has been supported the theory that adjuvant therapy with estrogen antagonists and other hormone manipulations had no role in treatment of mammary sarcomas. The adjuvant treatment could decrease the rates of local and systematic recurrences, but the results were not significant because of the rarity of this pathological entity and the small numbers of cases have reported, which made the evaluation of the role of the chemotherapy and the radiotherapy in the primary breast chondrosarcoma more difficult [12].

Conclusion
It has seemed to be very important to identify the mammary primary sarcomas as entity separated from the carcinomas of the breast. The primitive chondrosarcoma has remained a rare pathology, among which the therapeutic modalities and the forecast were credibly identical to those of the sarcomas of the same type arising in the other localizations.

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Conflict of Interest
The authors had no conflict of interest.

Authors’ Contribution
Puneet Kumar Bagri has participated in all care of the patient and has written the draft of manuscript. All authors have made contributions by making diagnosis and intellectual input in the case and have read the manuscript.

References
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