Primary Osteosarcoma of Breast, A Rare Case.

ABSTRACT
Mammary sarcomas are very uncommon and make up less than 1% of all primary breast malignancies. Primary osteosarcoma of the breast is extremely rare and represents 12.5% of mammary sarcomas. A secondary lesion from a primary osteosarcoma of the bone should be considered in the differential diagnosis. In addition, the absence of a direct connection between the tumour and the underlying skeleton is mandatory for the diagnosis. We report a case of primary osteosarcoma of the breast occurring in a young patient with fatal evolution.

Key words: Osteosarcoma of breast, Stromal sarcoma

INTRODUCTION
Primary osteosarcoma of the breast is a very rare and an aggressive entity, which is histologically indistinguishable from the conventional osteosarcomas of the bone and other extra skeletal [1] ones. In this report, we have described a case with this uncommon tumour.

CASE REPORT
A 40-years-old woman presented with a mass which arose in the left breast without any pain, which was noticed two months back.

Examination and Investigation: The physical examination at admission revealed a mobile mass in her left breast. The mass was not attached to the sternum or the underlying rib. No axillary lymphadenopathy was detected on the physical examination. The rest of the breast was unremarkable. On mammography, the mass was found to be well demarcated and partially calcified. Liver echography, chest radiography and bone scan were normal. The patient underwent a simple mastectomy.

Histopathology: The gross examination of the specimen showed a firm mass with a variegated appearance, which measured 4cm x 3cm in the upper and the lateral quadrants of the breast and it was 1cm away from the deeper resection margin [Table/Fig-1].

The histological examination revealed abundant osteoid formation, areas of calcification and oval to spindle shaped moderately pleomorphic stromal cells. There were scattered multinucleated osteoclastic tumour giant cells [Table/Fig-2], H and E, X400. Multiple sections were taken to detect an epithelial component if any, thus ruling out the possibility of a carcinosarcoma.

Immunohistochemistry: An immunoreactivity was detected for vimentine but not for for AE1/AE3, CK7, CK20 and the oestrogen and the progesterone receptors. There was also no over expression of the HER-2/neu oncprotein.

DISCUSSION
The mammary sarcomas are very uncommon and they make up less than 1% of all the primary breast malignancies [1-3]. Angiosarcomas are rare, but they are the most common pure malignant stromal tumours of the breast. Primary osteosarcomas of the breast are extremely rare and they represent 12.5% of all the mammary sarcomas [1]. The rarity of this case prompted us to publish this case report. The histogenesis of primary osteosarcoma of the breast is not clear, but an origin from the totipotent mesenchymal cells of the breast stroma or a transformation from a pre-existing fibroadenoma or a phyllodes tumour has been suggested [2-4]. Primary breast osteosarcomas behave as highly aggressive tumours which are associated with an early recurrence and a tendency for spread via blood rather than through the lymphatics, commonly to the lungs [1,2].
Mammographically, these tumours often present as well circumscribed, dense lesions within the breast parenchyma, with focal or extensive coarse calcifications [1,5]. Osteosarcoma of the breast, presents as fibroblastic, osteoblastic, osteoclastic (giant cell-rich) and fibroblastic subtypes [4–6], as may be seen in its common bony counterpart. Most of the cases of undifferentiated sarcomas of the breast, after an extensive sampling, may show a focal neoplastic epithelial differentiation, and may thus be regarded as metaplastic carcinomas. The diagnosis of a metaplastic mammary carcinoma should be excluded before a primary breast osteosarcoma is diagnosed. The term, ‘sarcomatoid carcinoma’ has been adapted to reflect the appearance of a mesenchymal neoplasm in these epithelial malignancies. The term, ‘carcinosarcoma’ is usually used to describe the biphasic tumours which are composed of distinguishable malignant epithelial and sarcomatoid components with heterologous elements [4,5].

A secondary lesion from a primary osteosarcoma of the bone should be considered in the differential diagnosis. In addition, the diagnosis of a primary breast osteosarcoma, which is similar to those of other extra skeletal tumours, requires the absence of a direct connection between the tumour and the underlying skeleton [7,8]. In similarity to the above observation, no connecion was noted with the overlying bony ribs in this case. The first report of a malignant mammary neoplasm of the bone and cartilage was made by Bonet [9] in 1700. Jernstrom [10] observed one osteosarcoma among 3319 cases of mammary carcinomas. In a recent report, however, Carter et al., raised the question of benefit from an axillary dissection in the absence of adenopathy, in a subset of a metaplastic carcinoma with a predominant spindle component, given the rarity of the lymph node metastasis in their 29 cases [13]. Although an adjuvant combination chemotherapy has dramatically increased the survival of the patients with primary osteosarcomas of the bone, the responses of osteosarcomas of the breast are still unclear because of the limited data.

CONCLUSION
The awareness of this rare case and a proper documentation with a systematic follow up, shall in future, lead to the development of an appropriate management for such cases.

REFERENCES