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Malignant phyllodes tumour as an intracystic growth with osteosarcomatous differentiation

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INTRODUCTION
Phyllodes tumour is a rare fibroepithelial tumour of breast that represents 0.3-0.5% of breast neoplasms. Phyllodes tumour occurs commonly in women in their 5th or 6th decade and sometimes in younger than 20 years. The commonest clinical feature is a rapidly enlarging breast lump that often grows to remarkable size. Numerous studies have endeavoured to correlate the imaging features of benign and malignant phyllodes tumour on sonography and mammography.

A rare case of malignant phyllodes tumour with osteosarcomatous differentiation mimicking an intracystic tumour on sonography is presented with review of literature, pertinent imaging findings and histopathological diagnostic criteria.

CASE REPORT
A 36-year-old lady attended breast clinic for a large painful lump in the upper outer quadrant of right breast, since one month, which was gradually increasing in size. On examination of breast a well defined mass with smooth surface was palpable in the right breast, upper outer quadrant approximately measuring 9 x 7.5 cm. There were no skin or nipple changes. Axillary lymph nodes were not enlarged. There was no family history of breast malignancy and no risk factors.

On mammography, a large high density soft tissue density mass with well defined smooth margins was noted in the upper outer quadrant of right breast without micro-calcification or architectural distortion. There was no mammographic evidence of skin thickening, nipple retraction or axillary lymphadenopathy. On ultrasound, a large cystic lesion measuring 6.5 x 6.2 x 4.7 cm with internal echoes and an eccentric solid component measuring 4.4 x 4.8 cm was noted (Figure 1, 2). The solid component did not reveal flow on application of colour Doppler. Based on the appearance, the differential diagnosis of intracystic neoplastic lesion or a complex cyst with haemorrhage was made. Fine needle aspiration cytology was carried out initially using 21 G needle and 33 cc of thick dark red fluid was aspirated. The pathology report was inconclusive as it suggested acute inflammatory cells without any epithelial or malignant cells.

Within 4 weeks, the cyst refilled and excision biopsy was carried out under general anaesthesia. The resected specimen on gross examination measured 8 x 5.2 cm, within which was present a uniloculated cystic cavity measuring 5.5 x 3.5 cm. Inner surface of the cyst was tan to light brown and exhibited a single irregular solid area suggestive of papillary excrescences. Microscopic examination revealed biphasic neoplasm with prominent stromal overgrowth showing sheets of spindle cells with moderate atypia and pleomorphism. Occasional multinucleated tumour cells were also identified and the approximate mitotic activity was 19/10 HPF. Focal areas of chondroid differentiation were also seen. The histopathology confirmed the diagnosis of malignant phyllodes tumour with focal osteosarcomatous differentiation (Figure 3).

The patient’s recovery was uneventful. The metastatic workup was negative. There was no local recurrence on 6 month follow-up.

ABSTRACT
Phyllodes tumour is an uncommon fibroepithelial tumour of breast that often presents clinically as a rapidly enlarging lump. On mammography it appears as a soft tissue density well circumscribed mass. Ultrasound shows it to be a solid hypoechoic mass with small cystic spaces. A case of malignant phyllodes tumour with an atypical appearance of intracystic tumour on sonography is reported with pertinent imaging features and histopathological diagnostic criteria. The tumour had undergone osteosarcomatous differentiation.

Key words: Intracystic tumour. Malignant phyllodes tumour. Sonography. Osteosarcoma.
DISCUSSION

Intracystic tumours of the breast are not common and are mostly due to intracystic papilloma or intracystic breast cancer. Phyllodes tumour often grows rapidly and areas of necrosis may appear cystic. However, the phyllodes tumour showing intracystic growth is an unusual and rare presentation. Clinically, phyllodes tumour presents as a palpable rapidly enlarging breast mass that is usually sharply circumscribed, non-tender and not attached to the skin or chest wall. The size of the tumour ranges from 1 cm to greater than 40 cm. The case presented here presented clinically as a tender breast lump that was gradually increasing in size.

On mammographic imaging, the phyllodes tumour appears as a dense round or oval shaped mass with circumscribed or lobulated margins and rarely shows calcification. This case was no different on mammographic appearance and appeared as a well defined high density mass without any cluster or speck of calcification. Ultrasound findings are of an oval, round or lobulated circumscribed hypoechoic mass with variable posterior features and may have intratumoral cystic spaces. Increased vascularity is common on colour Doppler examination. On sonography, this case had an atypical presentation of a well defined cystic lesion with eccentric solid component without definite flow on Doppler study.

Buchberger et al. and Page et al. showed that at sonography these tumours are circumscribed masses with low level internal echoes and 3-10 mm cyst in 6 benign lesions. Phyllodes tumour showing intracystic growth is rare and the frequency of this entity similar to the present case is not clear. A case report by Jun et al. showed similar imaging features as in this case, but the pathological examination revealed a borderline case of phyllodes tumour. A case similar to this one radiologically and pathologically is reported by Matsuo et al., presenting a 64 years old lady with an enlarging mass in left breast, which on mammography appeared as an irregular high density lesion and intracystic papillary tumour on sonography. On excisional biopsy and histopathology, it was also diagnosed as malignant phyllodes with osteosarcomatous changes. Another case reported by Tomoharu et al. of a 54 years old woman with rapidly enlarging left breast, which on mammography appeared as high density mass with irregular margins and on sonography showed a cystic tumour similar to the present case. On histopathology, this proved to be a case of ductal carcinoma with squamous differentiation in malignant phyllodes tumour. Liberman et al. reported mammographic and sonographic findings between benign and malignant Phyllodes tumour and their data suggested that cystic areas on ultrasound were more common in malignant lesions as it was likely to undergo cystic degeneration. These findings are in keeping with the present case and the case reported by Matsuo et al. and Tomoharu et al.

Hidetake et al. described the MR features of phyllodes tumour of breast compared with histologic grade. He concluded that an irregular cyst wall, tumour signal intensity lower than or equal to normal tissue signal intensity on T2, weighted imaging and low apparent diffusion coefficient (ADC) correlated significantly with histologic grade and stromal hypercellularity.

Phyllodes tumour is pathologically classified as benign, borderline and malignant. The microscopic findings include hypercellularity of stroma, cellular atypia, margins and mitotic activity which is the most important prognostic factor. A mitotic activity of 4/10 PHF is suggestive of benign and more than 10/HPF suggestive of malignant subtype with intermediate microscopic features in borderline cases. The mitotic activity in our case was 19/10 PHF suggestive of malignant subtype. The percentage of malignant phyllodes tumour ranges from 23-50% and the prevalence of local recurrence is around 25%. It is the same for benign and malignant forms and is related to inadequate excision. Distant metastasis of tumour occurs in 10% of tumour and lungs are the commonest site with less common involvement of bone, liver and myocardium.

Phyllodes tumour is a rare breast lesion with an ability to metastasize and shows a high prevalence of local recurrence after surgery. Although the common imaging
feature on mammography is of a dense round or oval shaped mass with circumscribed or lobulated margins and on ultrasound of a circumscribed hypoechoic mass with a possible cleft like cystic spaces, the presence of an intracystic growth on sonography in a clinically palpable rapidly enlarging lump should include the differential of phyllodes tumour.

REFERENCES


