Malignant Phyllodes Tumour: A Case Report

M Sani, V Leow, Z Zaidi, Y Mazuwin, S Hassan, M Zainal

Citation

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Abstract

"Cystosarcoma phylloides", or generally known as phyllodes tumour, is a rare breast tumour accounting for less than 1% of all breast neoplasms. Phyllodes tumours form a spectrum from benign tumours, similar to fibroadenomas, to malignant tumours with a propensity for rapid growth and metastatic spread. High-grade malignant phyllodes tumour (MPT) is a very rare but aggressive breast malignancy and forms approximately 25% of all phyllodes tumours[1]. We report a case of a 44-year-old lady who presented with a painless right breast lump for 3 months duration. The mass was initially 3cm in diameter and firm in consistency. Fine-needle aspiration cytology showed a benign breast lesion. In view of the rapidly growing mass, from 3 to 12cm diameter in 3 months, a decision for a frozen section intraoperatively was made. The results came back as malignant phyllodes tumour. The patient underwent right mastectomy with axillary exploration.

CASE SUMMARY

A 44-year-old lady presented to us with a painless right breast lump for 3 months duration. She had no significant family history of breast carcinoma or of any other past medical illnesses. The mass was initially 3cm in diameter and firm in consistency. Fine-needle aspiration cytology (FNAC) during the first visit to the clinic showed a benign breast lesion. Mammography revealed a benign breast lesion with multiple cysts. In view of the rapidly growing mass, from 3 to 12cm diameter in 3 months, a decision for a frozen section intraoperatively was made. The results came back as malignant phyllodes tumour. The patient underwent right mastectomy with axillary exploration. The postoperative course was uneventful and she was discharged at day 4 postoperatively. Computed tomography scan postoperatively showed no liver and lung metastasis.

Figure 1

Figure 1: Right breast specimen with area where initial tissue biopsy for frozen section was taken (arrow)

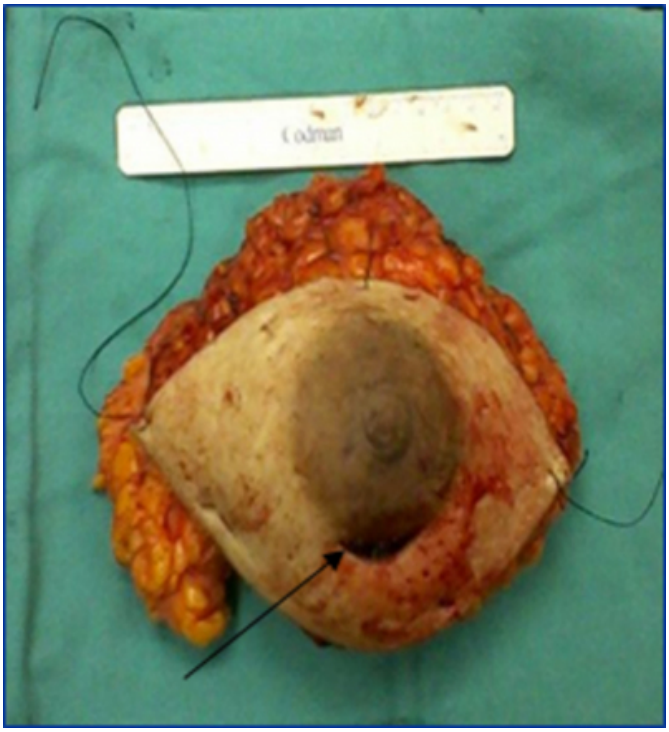


Figure 2

Figure 2: Lateral view of mastectomy specimen showing the underlying tumour

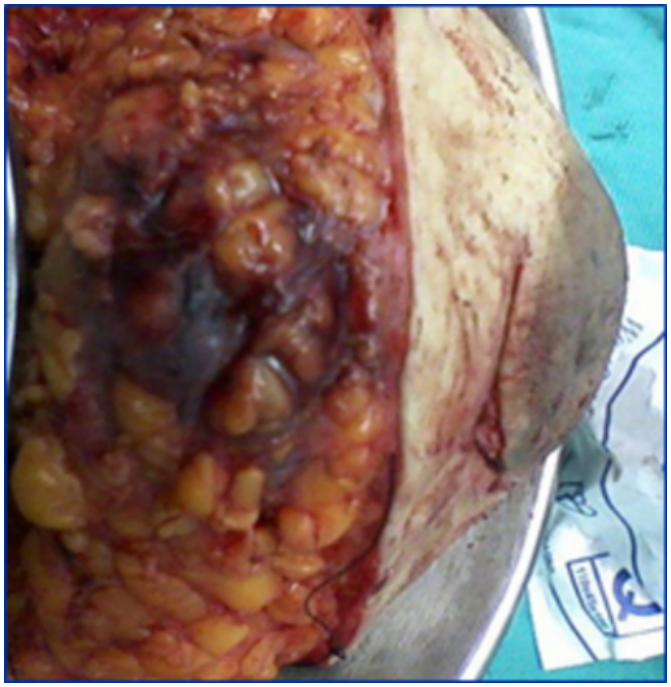
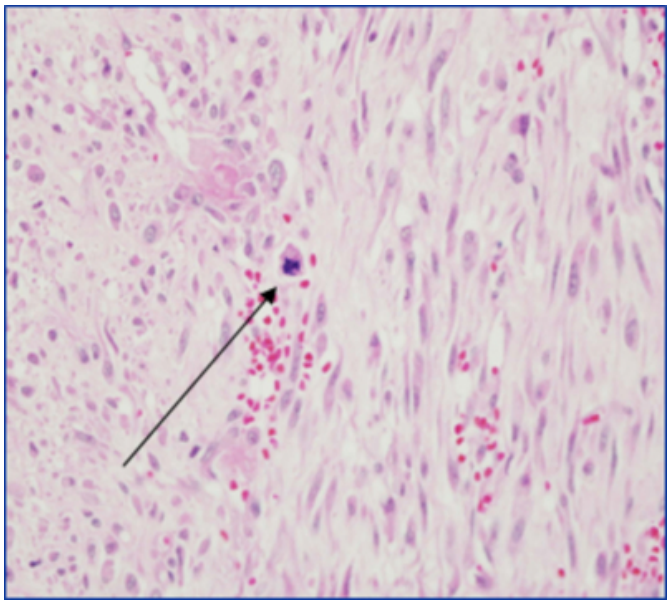


Figure 3

Figure 3: High power view shows pleomorphic spindle-shape tumour cells with abnormal mitosis (arrow)



DISCUSSION

‘‘Cystosarcoma phylloides’‘, now generally known as phyllodes tumour, is a rare breast tumour accounting for less than 1% of all breast neoplasms. The terminology ‘‘phyllodes tumour’‘ given by the World Health Organisation (WHO) classifies it into benign, borderline and malignant tumours according to histopathological features. High-grade malignant phyllodes tumour (MPT) is a very rare but aggressive breast malignancy and forms approximately 25% of all phyllodes tumours[1].

Although most phyllodes tumors occur in women between the ages of 35 and 55 years, adolescent and elderly women are also affected. The etiology of this rare disease is still unknown. However, its incidence is higher in whites in general, in Latina whites and East Asians in particular. Mangi et al. observed an 18% incidence of fibroadenomas in their study, suggesting a possible overlap and misdiagnosis between benign phyllodes tumour and fibroadenoma[2].

Clinically it appears as a round, mobile and painless mass. In most patients, axillary lymph nodes are not palpable at presentation, because metastatic spread of these tumors is primarily hematogenous (lungs, pleura, and bone have been the most common sites of metastasis. Metastasis to axillary lymph nodes occurred in only 2%[3].

The current classification of phyllodes tumour is given as benign, borderline or malignant (25-30% of cases). Histologically, the phyllodes tumour consists of epithelial cells and connective tissue with more stromal proliferation than that of fibroadenoma, often accompanied by cellular atypia. For malignant phyllodes tumour, they are further devided into borderline, low-grade, and high-grade on the basis of the following histological criteria: tumor borders, mitotic activity, stromal atypia, and stromal overgrowth. Only the stromal cells have the potential to metastasise. The malignant character of the phyllodes tumour is therefore confirmed by the microscopic appearance of the stroma[4].

Phyllodes tumours are clinically similar to fibroadenomas and they have both mammographic and sonographic characteristics in common. On ultrasound the tumours were lobulated in most of the cases. Heterogeneous internal echoes and ntramural cysts are also said to be suggestive of phyllodes tumours[5]. Phyllodes tumour on mammography is described as a sharply defined round or oval mass with lobulation[5].

Difficulties with diagnosis of phyllodes tumour by FNAC have been reported. The cytologist reported phyllodes tumour in only 23% of cases where FNAC was done at the time of diagnosis. In cases where core biopsy was done at the visit where the diagnosis was made, the core biopsy correctly diagnosed 65% of phyllodes tumours[5].

Surgical treatment is generally accepted as the most

important and primary therapy for phyllodes tumours, regardless of its histological type. A lumpectomy with clear margins (more than 1 to 2cm) is recommended for small lesions (up to a size of 3cm). A segment resection also with clear margins is recommended for larger lesions (over 3cm in size). A mastectomy should be performed in the case of even more extensive, borderline or infiltrative areas. An axillary lymphadenectomy should only be performed in clinically conspicuous cases, as phyllodes tumour has predominantly haematogenous metastases and axillary metastases occur in less than 10% of cases[2,4].

The role of radiotherapy remains unclear from published reports because of small patient numbers and a lack of controlled data. Pandey et al. suggested that adjuvant radiotherapy also improved the disease-free survival. August and Kearney recommended that adjuvant radiotherapy be considered for high-risk phyllodes tumors, including those >5cm, with stromal overgrowth, with >10 mitoses/high-power field, or with infiltrating margins[3].

Different chemotherapy regimens have been applied in malignant phyllodes tumours. Doxorubicin and ifosfamide-based chemotherapies have shown some efficacy in women with metastatic spread of phyllodes tumours. In one study of 101 patients, 4 patients were treated with chemotherapy and a role for adjuvant chemotherapy in patients with stromal overgrowth was considered. This recommendation has so far not been confirmed in literature. Altogether, there is no clear indication for adjuvant chemotherapy for patients with phyllodes tumours[2].

Locar recurrences (LR) are a common complication of high-grade lesions with a reported frequency of approximately 26% (12-65%)[1]. Stromal overgrowth, larger tumor size, and involved margin were all significantly correlated with LR. Stromal overgrowth increased the probability of LR 7-fold, whereas if the margin was <1cm, the risk of LR increased 5-fold, and if tumor size was >10 cm, then the prevalence of LR was four times greater than for smaller tumors[3].

The 5- and 10-year survival rates for malignant phyllodes tumour range from 54% to 82% and from 23% to 42%, respectively[2,3].

CONCLUSION

High-grade malignant phyllodes tumour is a very rare but aggressive breast malignancy. Stromal overgrowth, larger tumor size, and involved margin were all significantly correlated with local recurrences. Stromal overgrowth carries a grave prognosis. Either wide local excision with adequate margins or mastectomy is an appropriate treatment for patients with malignant phyllodes tumour.

References

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