

Phyllodes Tumour: An Update of 40 Cases

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Abstract

This is a retrospective study of 40 cases of phyllodes tumour treated at the National University Hospital since 1985. The mean age of presentation was 37.8 years, with a range of 15 to 58 years [standard deviation (SD) 11.22 years]. Ninety per cent of the patients were premenopausal. The mean size of tumours was 52 mm, with a range of 10 to 220 mm. Preoperative diagnosis was correct in only five patients, the lesion most commonly being mistaken as a fibroadenoma. Initial surgical treatment was by simple excision in 80% of cases. Histologically, the ratio of benign, intermediate and malignant tumours was 82.5%, 12.5% and 5% respectively. Recurrences occurred in only four patients (10%), all of whom were initially treated by simple excision.

The diagnosis of phyllodes tumour continues to be difficult even with newer modalities of preoperative investigations, resulting in simple excision as initial treatment of these tumours. Expectant follow-up should be continued for these patients, with wide excision of future recurrences.

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Key words: Benign breast disease, Breast sarcoma, Cystosarcoma phyllodes, Preoperative diagnosis, Surgical treatment

Introduction

First described by Johannes Muller in 1838 as a large, benign tumour of the breast, Phyllodes tumour is a rare pathological entity, accounting for no more than 1% to 3% of all breast lumps. Due partly to this rarity, questions regarding the preoperative diagnosis, clinicopathological correlation and optimal surgical treatment of initial as well as recurrent disease remain unanswered. This study reviews the clinical presentation, surgical treatment and histological features of all cases of phyllodes tumour seen at the National University Hospital from May 1985 to January 1995 and evaluates their surgical treatment.

Materials and Methods

Caserecords of all patients treated for phyllodes tumour at the National University Hospital from May 1985 to January 1995 were traced. All patients had phyllodes tumour confirmed histologically. Each patient was studied for her presenting complaints, clinical history, physical examination, initial treatment, recurrence and, if any, sarcomatous change. Patients who had not been reviewed for over a year were contacted by telephone and assessment made at the clinic for recurrence of disease.

Those who were lost to follow up were excluded from the study.

Results

There were 45 cases of phyllodes tumour in the interval studied. This represented about 2% of all breast lump excisions done in the department. Five of these 45 cases, all from the 1980s, were lost to follow-up and dropped from the study. Of the remaining 40 cases, the mean follow-up time was 23.4 months (range 2 to 82 months). All but one patient presented with a breast lump, the exceptional case being detected during routine examination by her family doctor. The mean size of the breast lump was 52 mm (range 10 to 220 mm). Patients had noticed their lumps for a mean duration of 32 months, with a range of two weeks to 30 years. None of the patients had complained of any breast pain or nipple discharge. Ninety per cent (36/40) of the patients were premenopausal. The mean age of presentation was 37.8 years (range 15 to 58 years). Five patients (12.5%) had a past history of fibroadenoma, while only one had previous fibrocystic disease. Twenty patients (50%) said their lumps had increased in size.

Twenty patients (50%) were subsequently treated with-

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out further preoperative investigations. Of the other 20 patients who were investigated further preoperatively, 16 had a fine needle aspiration, with a correct diagnosis of phyllodes in 25% (4). The other cytology reports were that of fibroadenoma (5), fibrocystic disease (3) and sarcoma (1). Three were considered to be inconclusive. Eight patients had a preoperative mammogram, only one being diagnosed as phyllodes tumour with this investigation. The preoperative diagnosis of phyllodes tumour was made in only five patients (12.5%). The most common preoperative misdiagnosis was that of fibroadenoma (75%), as well as fibrocystic disease (7.5%) and breast carcinoma (5%).

Thirty-two patients (80%) were initially treated by simple excision or enucleation procedure. Three (7.5%) had wide excision after preoperative diagnosis of phyllodes had been made and three (7.5%) had a partial mastectomy (two quadrantectomies and one segmentectomy) for large lumps. Total mastectomy was performed on two patients, one with a 22 cm mass involving the entire breast, the other with a malignant phyllodes diagnosed on trucut biopsy. Histological examination showed benign tumour in 33 patients (82.5%), malignancy in two patients (5%) and intermediate tumour in five patients (12.5%).

Recurrences occurred in four patients (10%). Review of the histological features of their initial tumours showed three benign and one intermediate grade phyllodes. All patients with recurrences were initially treated with simple excision. This represented 12.5% of all patients treated by simple excision (4/32). No patient initially treated with a more radical excision had local recurrence, regardless of histological grade. All recurrences occurred in the same quadrant of the breast as the initial tumours. The mean recurrence time was 21.5 months (range 8 to 33 months). Of the four recurrences, three were treated with simple re-excision again, while the fourth underwent segmentectomy. All three who were

initially treated with simple re-excision had a second recurrence. Subsequent treatment with wide excision and partial mastectomy in two of these patients resulted in cure. The third patient, who had undergone two simple excisions for recurrent benign phyllodes, developed sarcomatous transformation and underwent a total mastectomy. Despite systemic chemotherapy, she succumbed to her illness within 12 months of her mastectomy.

Two patients presented with *de novo* malignancy. One with a 30 cm tumour and metastatic disease to the lungs. She underwent total mastectomy but declined adjuvant therapy and survived 20 months. The second patient with malignant phyllodes had presented with a 10 cm breast lump. Partial mastectomy was performed and clear histological margins obtained. She is still well after four years follow-up.

Discussion

Since first described by Muller,¹ phyllodes tumour has remained an enigma both in clinicopathological correlation and optimal surgical treatment. This is partly due to its rarity, where phyllodes consisted of only 1% to 3% of all breast lumps. A recent review by McDaniel and Crichlow in 1989² noted only about 700 cases in the literature. While Muller first believed cystosarcoma phyllodes to be a large benign tumour of the breast, Cooper and Ackerman³ demonstrated its malignant potential. Even the term "cystosarcoma phyllodes" is a controversial one, the "sarcoma" suggesting malignancy when most of these tumours are benign. Some authors also refer to these tumours as "giant fibroadenomas", reserving the term cystosarcoma phyllodes for the malignant variant.⁴

Most Western series in the literature indicate a peak incidence of phyllodes tumour in the fourth and fifth decades of life. Our series has a mean age of presentation of 37.8 years, with 60% (24/40) of patients under 40 years of age.

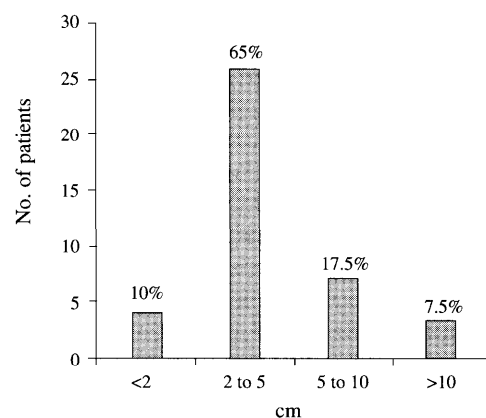
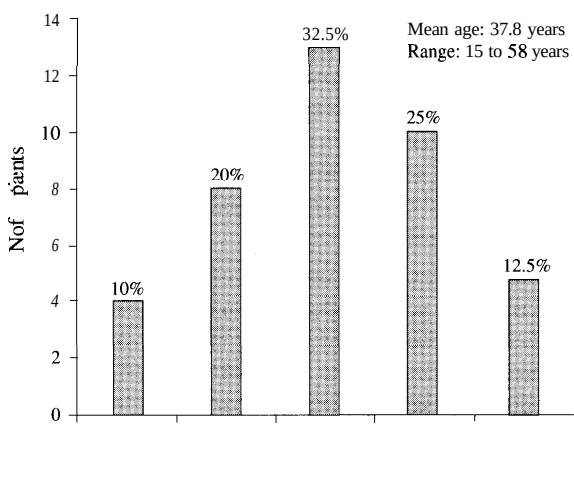


Fig. 2. Size of tumour (n = 40).

Preoperative diagnosis of phyllodes tumours remains difficult. In Western societies, where greater size and older age of onset may indicate something more than a fibroadenoma, the younger presentation and smaller size of these tumours found in the Oriental population makes the diagnosis even more difficult.⁵ In our series, 75% of the phyllodes tumours were under five cm in size, making the clinical feature unreliable for distinguishing these tumours from fibroadenomas. Based on clinical findings alone, half the tumours were mistaken as fibroadenomas and underwent simple excision without further preoperative investigations, especially in the younger age group. This high operative rate for suspected fibroadenomas reflects the reluctance of our women to have breast lumps treated conservatively.

Of the other half which were investigated further, mammograms, breast ultrasound and fine needle aspiration cytology have been used. Features suggestive of cystosarcoma phyllodes have been described using these modalities,^{6,9} although these have been based on retrospective studies of histologically proven tumours. On mammography, a “halo” effect seen around a well-defined lesion in phyllodes tumour has been thought to be due to compression of the surrounding stroma, while the demonstration of cystic structures within a well-circumscribed lesion with weak internal echoes has also been described in ultrasound. There appears to be no reliable indicator to distinguish benign from malignant phyllodes tumours based on these modalities. In our study, accuracy rates of mammograms and fine needle aspirations were 12.5% (1/8) and 25% (4/16) respectively. While we are increasing our use of breast ultrasonography in the assessment of breast lumps, only three patients in our series had breast ultrasound done, with a correct diagnosis made in one patient only.

Histology

The origin of cystosarcoma phyllodes still remains elusive, although its apparent correlation with fibroadenoma is often regarded as more than casual. A widely held view is that the stroma of phyllodes tumour is derived from the subepithelial connective tissue of the breast, similar to the intracanalicular fibroadenoma.¹⁰ Generally, neoplasias are regarded as being cystosarcoma phyllodes if they contain both epithelium and stromal components as part of the neoplasia associated with a greater degree of cellularity than fibroadenomas.” In our study, 12.5% (5) of patients had a previous history of fibroadenoma, although it is unclear if these arose from the same quadrant of the breast. Both light microscopy and electron microscopy have confirmed that only the stromal elements of the tumour have metastasizing potential.¹²

Both histologically benign as well as malignant cystosarcoma phyllodes have been known to metastasize. There is also as yet still no universally accepted criteria for malignancy. To illustrate this, Contarini et al¹³ had 23 slides of cystosarcoma phyllodes reviewed by separate pathologists and found a disagreement of 28.5%. While no one histological factor can determine the likelihood of recurrence or malignancy, prognostication has been attempted by scoring the combinations of histological features such as stromal overgrowth, mitotic figures, cytological atypism and character of the tumour margin, with good correlation to recurrence and malignancy.^{11,14,15} In our study, the patients with recurrence all underwent local excision initially. Of the four recurrent tumours, three were initially assessed to be benign while the other was intermediate. In the absence of histological predictors of malignancy, we should regard each case of cystosarcoma phyllodes as potentially malignant.

Treatment of cystosarcoma phyllodes should be based on its clinical behaviour, which is similar to that of other soft tissue sarcomas. Surgical treatment should be aimed at complete excision of the tumour, as the outlook for patients with metastatic disease is uniformly dismal, with no long-term survivors in any series.^{16,17} As tumour margins are usually indistinct, especially in higher histological grade tumours, wide excision with a margin of normal tissue is required.¹² In rarer cases where cystosarcoma phyllodes presents as a large breast mass, total mastectomy may be required. Axillary clearance is probably not necessary even for malignant cystosarcoma phyllodes as spread is primarily blood borne.”

The management of the patient where the tumour has been treated by simple excision under the impression that it is a fibroadenoma remains a dilemma. These patients make up the majority in our series. To subject them to a second operation when the diagnosis of phyllodes is made, is to perform needless operation on 90% of patients, many of whom are young. Therefore, a “wait and see” policy appears justified.** However, recurrence of even benign lesions can be expected in about 15% of cases, although figures of up to 40% have been reported after longer follow-up.⁷ It is debatable if some of these represent second tumours rather than true recurrences. Transformation of benign phyllode to malignant is thought to be rare.¹⁹ Surveillance should be provided for least two years as most recurrences would occur within this time.”

Metastatic disease has been reported to occur in 3% to 12% of patients,¹⁷ which usually leads to rapid deterioration and death in a matter of months. Numerous cytotoxic combinations have been attempted, few with progression-free intervals of over six months. Recently, Hawkins et al²⁰ have reported complete remission in two patients treated with ifosamide, alone or in combination with doxorubicin.

Conclusions

Cystosarcoma phyllodes is an uncommon breast tumour that in our population is clinically indistinguishable from fibroadenomas. Usual preoperative investigations are often unreliable and histological predictors of malignancy unsettled. Treatment is wide excision of the tumour although unsuspected cases treated with enucleation can be observed. Recurrences occur within two years and should be treated with wide excision. Metastatic disease has a dismal prognosis although newer chemotherapeutic regimes with ifosamide show promise.

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