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Case report

Rare malignant tumours of the breast

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Background. Nonepithelial malignant tumours of the breast are rare. In this article 7 cases of the malignant phyllodes tumour and 2 cases of the primary breast lymphoma are presented.

Materials and methods. 1269 patients with breast malignancies were treated at the Department of Breast Diseases and Oncology of the Institute of Oncology, Vilnius University, during the period 1998–2000. There were only 7 cases (0.7%) of malignant phyllodes tumour (malignant cystosarcoma phyllodes) among all the breast tumours. All patients were female. Women's age ranged from 40 to 85 years and the mean age was 54 years. Two patients were in clinical stage II and five patients in clinical stage III of the disease. Five patients were treated by radical mastectomy, one patient by quandrantectomy. Sectoral resection was performed to one patient who suffered from heart disease. Two patients with primary breast lymphoma were during the 2000–2005 period. Patients were 80 years and 78 years old. Both patients were in 1st stage of the disease. One patient had a large diffuse B cell lymphoma; the second one had unspecified non-Hodgkin's lymphoma. Sectoral resections of the breast were performed for both patients followed by radiotherapy.

Results. Five patients with malignant phyllodes tumour are alive. One patient died from dissemination of the breast tumour after 3 months, and one patient died from another malignant tumour after 9 months. Two patients with breast lymphoma are alive.

Conclusions. 1. Clinical behaviour of malignant phyllodes tumour is benign in most cases, while rapid dissemination of the tumour appears in some cases. 2. Primary lymphoma of the breast has a benign clinical course.

Key words: malignant phyllodes tumour; primary lymphoma of the breast

INTRODUCTION

Nonepithelial malignant tumours of the breast are very rare. Phyllodes tumour of the breast (actually it is biphasic tumour, epithelial and nonepithelial) is the most frequent tumour among nonepithelial breast tumours. The incidence of phyllodes tumour is very low: 0.3% of all the breast tumours (1, 2). Average annual age-adjusted incidence of malignant phyllodes tumours was 2.1 per 1 million women during the period from 1972 to 1988 (3, 4). Phyllodes tumours typically present as palpable masses. Phyllodes tumours are divided into benign, borderline and malignant categories, depending on the histological findings of the tumour stroma. Malignant variant of phyllodes tumour is the most common among sarcomas of the breast. Classification of sarcomas of the breast is represented in Table 1. Malignant phyllodes tumours metastasize to viscera and rarely to axillary nodes (5). Wide local excision or simple mastectomy is recommended for benign cases, and radical mastectomy is recommended for those with a malignant stroma. Some authors identified stromal overgrowth as the essential characteristic for differentiating phyllodes tumour from fibroadenoma. Phyllodes tumours occur between the ages from 31 to 60 in 80% cases. The overall 5 and 10 years survival rates for malignant phyllodes tumours were 57% and 48%, respectively (6, 7). Other forms of the breast sarcomas are rare. For example, we have found only one report of malignant fibrous histiocytoma of the breast. The report deals with a 90 year old patient with ulcerated tumour of the left breast (8). Japanese authors (9) reported a case of 56 year old Japanese woman with a firm mass in the right breast. Breast-conserving surgery with axillary lymph nodes dissection was performed. Histological examination showed a primary giant cells malignant fibrous histiocytoma of the breast. No metastases were identified in the removed axillary lymph nodes.

Adjuvant therapy. In breast sarcoma cases with large or high grade tumour, with tumour close to surgical margins or in cases when tumour-free surgical margins cannot be obtained, postoperative radiation therapy is recommended. Information about adjuvant chemotherapy for sarcomas of the breast is scant.

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Table 1. **Classification of sarcomas of the breast**

Phyllodes tumour (cystosarcoma phyllodes)*
Stromal sarcoma
Malignant fibrous histiocytoma
Leiomyosarcoma
Rhabdomyosarcoma
Fibrosarcoma
Liposarcoma
Osteosarcoma
Spindle cell sarcoma
Chrondosarcoma
Carcinosarcoma
Lymphoma
Angiosarcoma
Lymphangiosarcoma (Stewart-Treves syndrome)

* Phyllodes tumours are unique in combining sarcoma with a benign epithelium.

Pure sarcomas derive solely from stromal (supporting) tissue. In carcinosarcomas both epithelial and stromal tissues are malignant. Angiosarcomas and lymphangiosarcomas arise from vascular endothelial cells. Primary lymphoma of the breast arises from intramammary lymphoid tissue or possibly from intramammary lymph nodes. Pure breast sarcomas are rare: Elson et al. (10) reported five cases of breast fibrosarcoma in women aged 48–79. Chondrosarcomas and osteogenic sarcomas of the breast are among the rarest of sarcomas of the breast. Osteogenic sarcoma of the breast arises from the stroma of long-standing fibroadenoma or possibly from progression of osseous metaplasia (11). Donegan (5) reported one case of malignant osteogenic sarcoma of breast. It was a 52 years old woman with a mass in the lower outer quadrant of the left breast. Carcinoma was suspected, and she was treated with a left radical mastectomy. The patient was well for 8 years, but then developed weakness and a chronic cough. An X-ray of the chest showed multiple metastases in both lungs, and subsequently metastases appeared in the subcutaneous tissues. Other forms of breast sarcomas, such as chondrosarcoma, leiomyosarcoma, liposarcoma, rhabdomyosarcoma are extremely rare among nonepithelial breast tumours. The literature reported one–two cases of patients with these sarcomas. Haematogenous dissemination is most frequent mode of spread of these tumours. Breast rhabdomyosarcoma metastasizing to regional nodes is relatively frequent. Mastectomy is the treatment of choice for these tumours.

Lymphoma. Primary breast lymphomas are considered as such in the absence of widespread disease only. Lymphoma represents 0.1% of breast malignancies. About 13% of patients have bilateral disease. Axillary nodes are involved in 30% to 40% of cases (12, 13). The vast majority of patients have non-Hodgkin's lymphoma. All histological types of lymphoma have been described, but diffuse intermediate B-cell non-Hodgkin's lymphomas are the most common (14). The treatment of breast lymphoma depends on tumour type and the stage of disease. After surgery (local excision) the patients must be treated by radiotherapy and chemotherapy. Mastectomy with lymph node dissection was reserved for local failures (15).

MATERIALS AND METHODS

1269 patients with breast malignancies were treated at the Department of Breast Diseases and Oncology of the Institute of Oncology, Vilnius University, in 1998–2000. In 7 cases (0.7%) histological examination revealed malignant phyllodes tumours. All the patients were female from 40 to 65 years old with the mean age 54 years. Two patients were in clinical stage II, five patients in stage III. Five patients were treated by radical mastectomy, one patient by quadrantectomy. Sectoral resection of breast was performed to one patient who suffered from heart disease. Three patients received radiotherapy after surgery. Two patients died during the follow-up period: one patient died from tumour dissemination 3 moths after surgery, while the second one died from another malignant tumour 9 months after the breast tumour surgery.

We present one case of malignant phyllodes tumour (Figs. 1–5): a 41 years old woman had a small tumour of the left breast 22 years ago, which began to grow rapidly one year ago. Radical mastectomy with axillary's lymph nodes dissection was performed. Histological examination of the tumour revealed malignant phyllodes tumour without metastases in 13 lymph nodes. Weakness and cough developed after 6 months. An X-ray examination of the chest showed multiple metastases in both

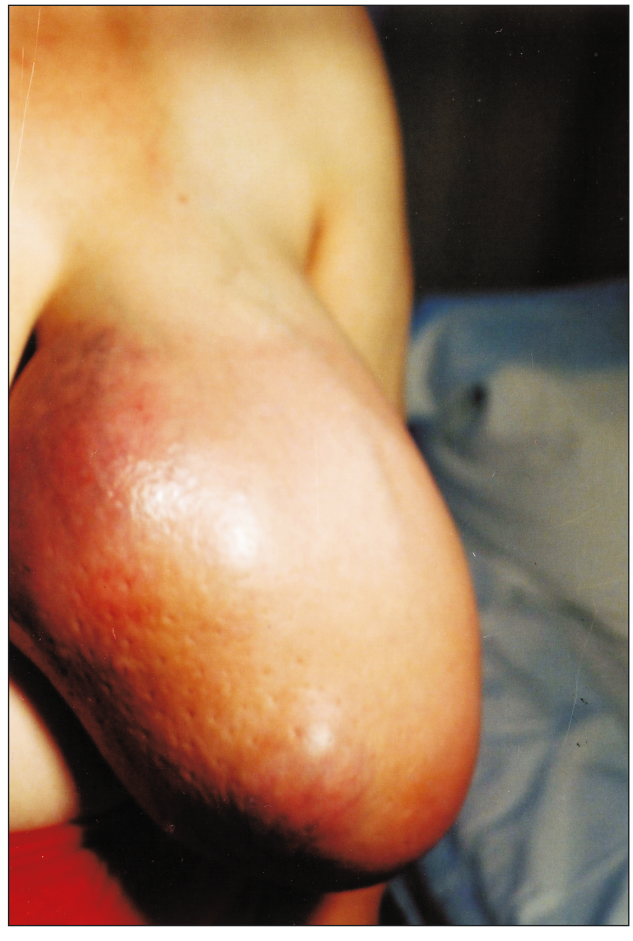

Fig.1. Gross appearance of the breast with large malignant phyllodes tumour in a 41 year old patient

lungs. The patient was treated by chemotherapy, but died after 3 months.

Other five patients are alive; there are no signs of progression of the disease.

Lymphoma of the breast. Two patients with primary lymphoma of the breast were treated at the Department of Breast Disease and Oncology of the Institute of Oncology, Vilnius University, in 2000–2005. One patient was 80; another one was 78 years old. The first patient had unilateral right nipple swelling (Fig. 6). Histology of the breast tissue infiltration by tumour cells is presented in the Figs. 7–9. There were no signs of tumour spreading to the other organs or lymph nodes, which represents stage I of the disease. The second patient had intraductal carcinoma of the left breast (pTisN0M0) 13 years ago. The patient was treated by quadrantectomy and received 42 Gy radiotherapy. Axillary lymph nodes dissection was performed 12 years after the surgery due to lymph nodes enlargement. Histological examination revealed diffuse large B cell lymphoma. The patient was treated by 39 Gy radiotherapy. Mammography of the right breast was performed one year later and 1.9 × 1 cm tumour was detected. Sectoral resection of the right breast was performed. The patient received 40 Gy radiotherapy after the surgery. Both patients are alive.

Fig. 2. Gross appearance of the breast tumour (radical mastectomy specimen): bilobed yellow pink tumour, measured 28 × 20 × 20 cm

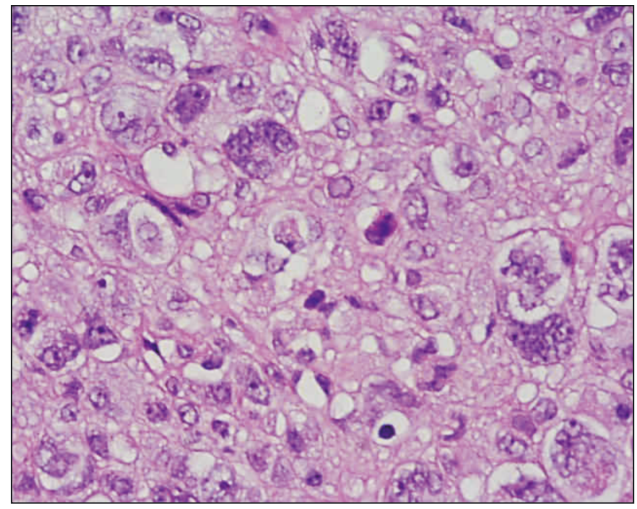

Fig. 4. Malignant phyllodes tumour: highly polymorphous atypical giant cells (HE × 400)

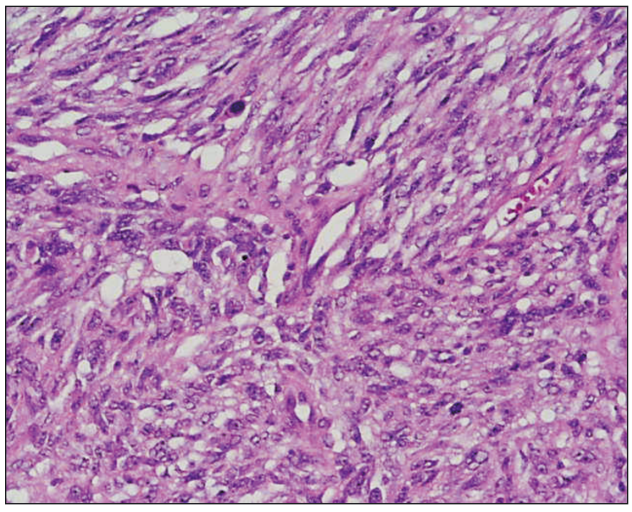

Fig. 3. Malignant phyllodes tumour: highly polymorphic stromal cells, mitotic figures (arrows) (HE × 100)

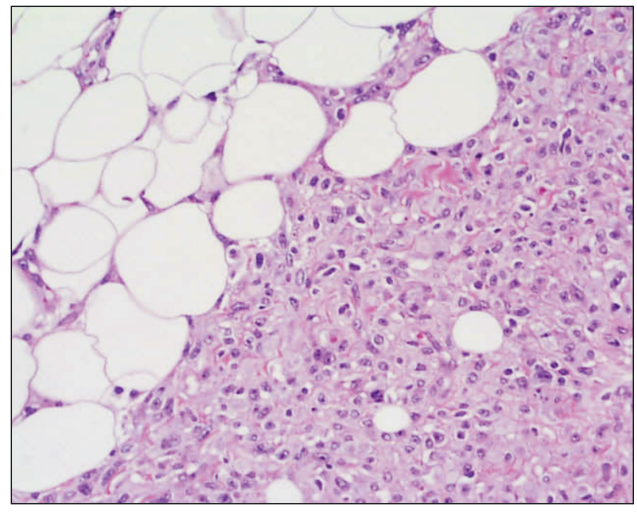

Fig. 5. Malignant phyllodes tumour with invasive growth: tumour cells spread between lipocytes (arrows) (HE × 100)

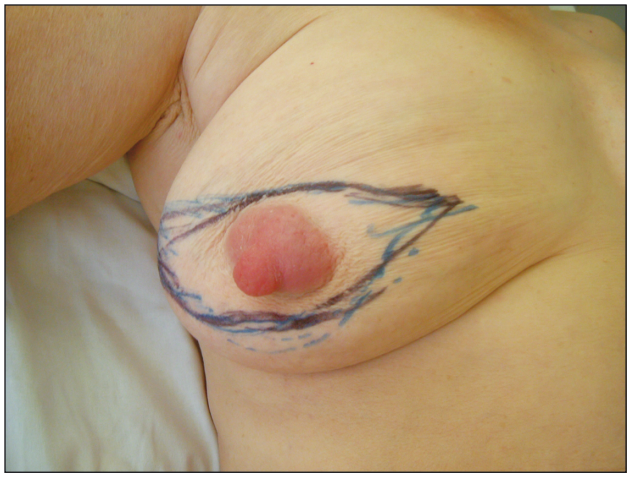

Fig. 6. An 80 years old woman had redness and an enlargement of the right breast nipple up to 0.9 × 2.3 cm 2 months ago. Resection of the central breast quadrant was performed. Histological examination of the tumour: non-Hodgkin's lymphoma, unspecified. The patient received 40 Gy radiotherapy after the surgery

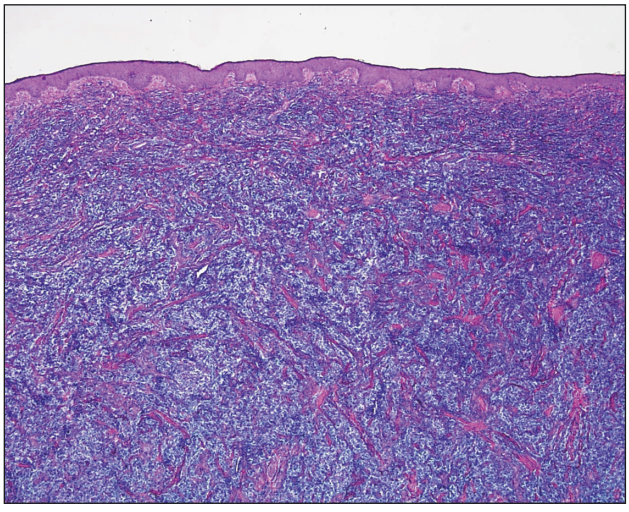

Fig. 7. Diffuse infiltrate in the reticular derma separated from the epidermis by undamaged papillary derma (HE × 20)

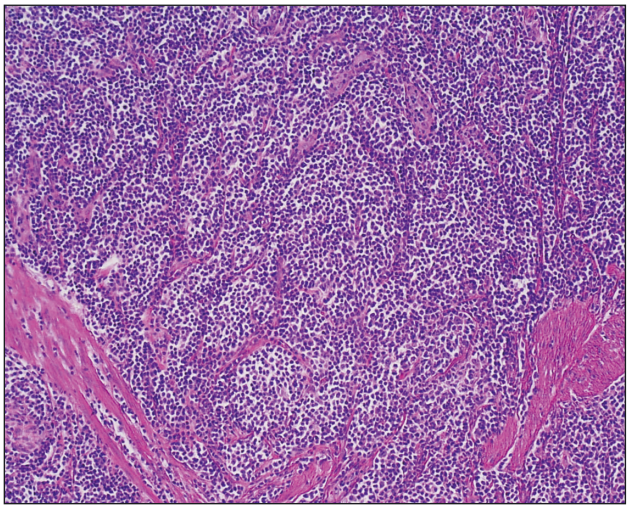

Fig. 8. Bright nodular pattern of the lymphoid infiltrate in the reticular derma (HE × 100)

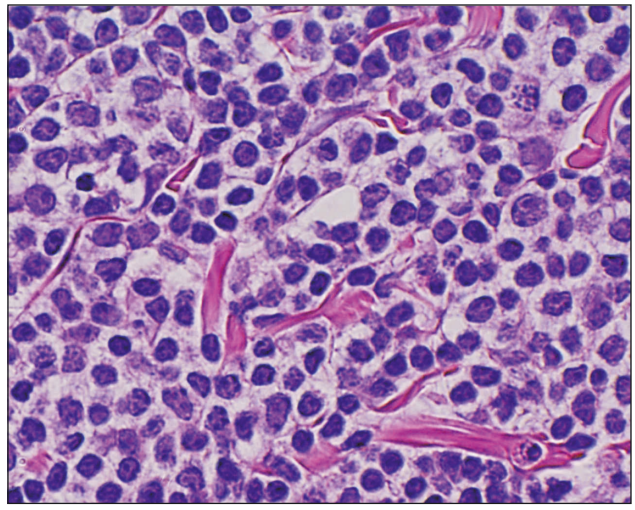

Fig. 9. Tumour composed of atypical lymphoid cells with relatively abundant clear cytoplasm and irregular nuclei (HE × 400)

DISCUSSION

Many authors reported that malignant phyllodes had stromal cells polymorphism, the incidence of malignancy of which was uncertain. Blichert-Toftet found a disappointing correlation between tumour grade and clinical course and noted that with recurrences, the character of tumours could become progressively more malignant (16). The patients generally present with breast mass. Some relate the presence of a small hard mass for many years, followed by the onset of rapid growth of tumour. Phyllodes tumours are highly variable in their gross appearance, but the majority display a solid, fleshy mass with cystic areas. It is possible to separate phyllodes tumours into two groups: the low-grade phyllodes tumour having a potential for local recurrence, and the high-grade tumours with a potential for metastases. Symptoms of primary lymphoma of the breast are different from those of malignant phyllodes tumour. The usual presenting symptom is a painless breast mass, and there is no specific mammographic abnormality that could predict lymphoma.

The treatment of primary lymphoma of the breast depends on the histological type of the tumour and on the stage of the disease. Results of the breast lymphomas treatment are similar to those for stage I and II lymphomas at other sites. Optimal therapy for patients with intermediate- and high-grade lymphoma localized to the breast (stage IE) includes chemotherapy and localized breast irradiation (15).

CONCLUSIONS

Clinical behaviour of malignant phyllodes tumours is benign in most cases, while rapid dissemination of the tumour appears in some cases. 2. Primary lymphoma of the breast had benign clinical course.

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RETI KRŪTIES PIKTYBINIAI NAVIKAI

S a n t r a u k a

Krūties piktybiniais neepiteliniais navikais sergama retai. Šiame straipsnyje aprašomi septyni krūties piktybinio filodinio naviko ir du pirminiai krūties limfomos atvejai.

Medžiaga ir metodai. Vilniaus universiteto Onkologijos instituto Krūtų ligų ir onkologijos skyriuje 1998–2000 m. buvo gydytos 1269 moterys, sirgusios krūties piktybiniais navikais, iš jų tik septynioms (0,7%) ligonėms buvo diagnozuotas piktybinis filodinis navikas (*cystosarcoma phyllodes malignum*). Moterų amžius buvo nuo 40 iki 85 metų, vidurkis – 54 metai. Dviem ligonėms buvo nustatyta antra ligos stadija, penkioms – trečia. Penkioms ligonėms atlikta radikali mastektomija, vienai – kvadrantektomija. Dėl širdies patologijos vienai ligonei padaryta sektorinė krūties rezekcija. 2000–2005 m. dviem ligonėms (80 ir 78 metų) diagnozuota pirmos stadijos krūties limfoma. Morfologiniu tyrimu vienai ligonei nustatyta B ląstelių difuzinė limfoma, kitai – ne Hodžkino tipo limfoma. Šioms ligonėms buvo atlikta sektorinė krūties rezekcija, po operacijos taikyta spindulinė terapija.

Rezultatai. Penkios ligonės, sirgusios piktybiniu filodiniu naviku, gyvena be ligos požymių. Viena ligonė mirė dėl naviko generalizacijos praėjus trims mėnesiams po gydymo, kita mirė po 9 mėnesių, susirgusi kita onkologine liga. Abi ligonės, sirgusios pirmine krūties limfoma, gyvena be ligos požymių.

Išvados. Sergant piktybiniu filodiniu naviku ligos klinikinė eiga dažniausiai yra gerybinė, tačiau kai kuriais atvejais gali įvykti greita navikinio proceso generalizacija. Sergančiųjų pirmine krūties limfoma ligos klinikinė eiga buvo gerybinė.

Raktažodžiai: piktybinis krūties filodinis navikas, pirminė krūties limfoma