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A case report



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Phyllodes tumours (PTs) are rare fibroepithelial neoplasms representing about 0,2% to 2% of all breast tumors with an incidence of about 2.1 per million. The classification proposed by the World Health Organization for PTs into benign, borderline, and malignant is based on a combination of several histologic features. High-grade malignant phyllodes tumors may spread by hematogenous route. While smaller and moderate size malignant phyllodes may typically be seen, gigantic ones with larger than 10 cm in diameter are very rare.

We report an unusual case of a giant malignant phyllodes tumor with metastases that grew over a 6 years period causing significant ulceration, body disfigurement and physical transformation. Our experience indicated that surgical treatment of malignant phyllodes tumor might be an option for improving patients' quality of life, regardless of the extremely poor prognosis.

KEY WORDS: Breast, Malignant phyllodes tumor, Surgical treatment

Background

Phyllodes tumor is a rare fibroepithelial breast tumor accounting for 0.2-2% of breast tumors in women worldwide.¹⁻³ Phyllodes tumor types are pathologically classified as benign, borderline and malignant based on histologic tumor characteristics: tumor margins, stromal overgrowth, tumor necrosis, cellular atypia, mitotic count⁴. Approximately 50% of this tumor is benign and 25% is borderline^{4,5}; 16-30% is malignant⁶. The incidence of metastatic disease among patients with malignant phyllodes tumor is estimated to be approximately 20-25%^{7,8}. Most frequently, metastases develop to the lung, bones, brain and liver^{7,9,10}.

Approximately 20% of phyllodes tumors are considered giant with a size greater than 10 cm in diameter^{10, 11}. We report a rare and unusual case of a giant phyllodes tumor with metastases to the lung and liver that grew over a 6 years period causing ulceration, disfigurement and altered body image.

Case Report

A 60-year-old woman came to the outpatient clinic of our breast center with a 6 year history of an important mass of right breast. She reported a self-examination of a nodular mass of the inner quadrant of right breast (about 2 cm) 6-years before. The patient had never undertaken a clinical or radiological breast check. The mass had gradually increased by size, and in the three month period before her visit, three skin ulcerations had appeared, with exudation and occasional bleeding. The patient presented body disfigurement and physical transformation. The patient had no fever or anemia. The only symptoms reported were due to faulty posture linked to the weight of the tumor. The worsening symp-

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toms (increased bleeding and exudation and appearance of foul smell) had prompted her to seek medical care. She had family history of breast cancer (mother at the age of 85-year). Menarche was at the age of 14, one pregnancy at the age of 33 (by caesarean section), she breast-fed for 1 year, and started her menopause at the age of 50. She had followed an estrogen-progestin therapy for short periods in the past. The only significant comorbidity was found to be a GERD in combined therapy with PPI.

Laboratory data were within normal limits except for hemoglobin 10.5 g/dl (range 12-15 g/dl). Physical examination showed a large exophytic lumpy tumor of 43x40 cm fully occupying the right breast, with three areas of skin ulceration (about 3x3 cm, 8x7 cm and 12x9 cm) of the outer quadrant.

Her left breast and the rest of her clinical examination were normal. Due to pain and size of the right breast mass, the patient was unable to have a mammography performed. There were no suspicious findings in the left breast or axilla (Fig. 1).

The computed tomography showed a giant breast mass with multinodular confluent aspect, inhomogeneous enhancement (due to the presence of necrotic-colliquative components) and the evidence of multiple calcifications in the most caudal portion. The mass affected the soft tissues of the breast full thickness, with infiltration of large and small ipsilateral pectoral muscles. The soft tissues of the intercostal spaces did not always show a cleavage plane with the adipose mass (Fig. 2A). It revealed no axillary pathological lymph nodes and nor of the internal mammary chain. The computed tomography scans also showed multiple pulmonary micro-

nodular images of non-specific meaning and a solid non-calcified nodule (5 mm) in the lateral basal segment of the left inferior lobe, suspected to be of repetitive nature. After administration of a contrast agent, the left lobe of the liver, segment II, showed an unevenly hypodense nodule (48 mm x 40 mm x 45 mm) associated to a minimum ectasia of some bile ducts in upstream of the injury, therefore suggesting the mass was suspected for metastases (Fig. 2B).

It was not possible to perform a preoperative biopsy because the patient refused the procedure.

She underwent a right mastectomy with partial resection of the pectoral muscles. The tumor did not appear macroscopically to be invading the chest wall. The tumor was characterized by important angiogenesis with several centimeter-sized vessels.

The wound was closed with no need of skin grafting and the total blood loss was under 100cc (Fig. 3).

The resected tumor was 41x32x22 cm and weighed 14200 gr (Fig. 4) and appeared as a fleshy, multinodular confluent neo-formation with large necrotic, colliquative and calcified areas. Microscopic findings showed a malignancy spindle cell with moderate atypia and high mitotic activity (up to 28x10 HPF). Additional section showed a residual epithelial structure sometimes cystic and distorted without heterologous areas. The margin of the resected tumor showed a focal and partial infiltration of the muscle included into the resection. The final histopathological diagnosis was malignant phyllodes tumor.

The patient was discharged on the second postoperative day in a good condition with no wound complications. The patient performed oncological examination with a significant improvement in quality of life.



Fig. 1: Pre-operative photograph showing giant right breast mass of 43x40 cm with skin ulceration and severe disfigurement.

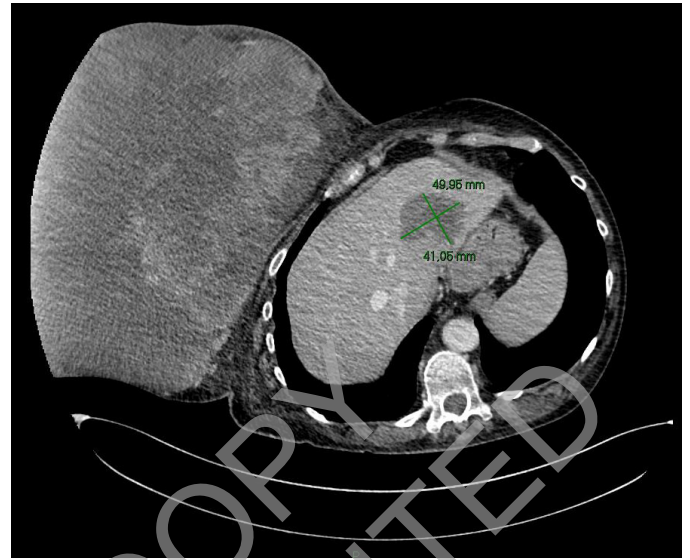


Fig. 2: A) The computed tomography shows a giant breast mass only partially included in the FOV due to its size; B) The computed tomography shows suspected liver metastases of 48 mm x 40 mm x 45 mm.



Fig. 3: Final result after mastectomy.

Discussion

Phyllodes tumor is a rare fibroepithelial tumor accounting for 0.2-2% of breast tumors in women worldwide ¹⁻³. Phyllodes tumors types are pathologically classified as benign, borderline and malignant according to the following standard criteria (4,9):

- a) Benign: 0-4 mitosis / 10x magnification area, minimal stromal hypercellularity and atypia, minimal or moderate stromal overgrowth, and negative surgical margins;
- b) Borderline: 5-9 mitosis / 10x magnification area, moderate stromal hypercellularity, atypia and stromal overgrowth, negative or permeative surgical margins;
- c) Malignant: >10 mitosis / 10x magnification area, moderate or marked stromal hypercellularity, atypia and stromal overgrowth, permeative surgical margins.



Fig. 4: The excised mass, 43 x 40 x 40 cm.

Approximately 50% of this tumor is benign and 25% is borderline ⁵; 16-30% is malignant ⁶. The treatment for phyllodes tumors is wide local excision with sufficient margin of normal breast tissue or

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mastectomy. Mastectomy is often required in the case of giant phyllodes ⁶⁻¹¹.

The role of adjuvant radiotherapy and chemotherapy remains uncertain, but consideration can be given for their use in cases of malignant phyllodes tumours ¹².

Malignant phyllodes tumors may spread by hematogenous route. Distant metastases are most common in the lungs (66%), bone (28%), brain (9%), followed by the liver and the heart ⁴. In a review of 67 patients who had metastatic phyllodes tumors, Kessinger and colleagues described the frequency of metastasis in each site: lungs (66%), bones (28%), heart (9%), and liver (6%). Lymph node metastases occur in less than 1%, therefore axillary dissection is not routinely recommended ¹³⁻¹⁷ unless the lymph nodes are pathologic on clinical examination ¹⁸⁻²³. Once patients with malignant phyllodes tumor developed metastasis, their prognosis is extremely poor. After the development of metastases, the mean overall survival is 30 months ⁵. In the patients who experiences a systemic recurrence treatment should be as recommended in the NCCN guidelines for soft tissue sarcoma ²⁴.

Conclusion

We experienced a rare case of giant malignant phyllodes tumor of the breast with high malignant characteristics causing significant ulceration, body deformity and physical transformation.

Our experience indicated that surgical treatment of malignant phyllodes tumor might be an option for improving patients' quality of life, regardless of the extremely poor prognosis.

We hope that the description of this particular and unusual case may emphasize a proper breast health education and underline the negative consequences of alternative or delayed treatment.

Authors' contributions

All authors participated in the conception and design of the study. DDG and SDA drafted the manuscript and performed the revision of the final document. GF assisted with the literature research and drafted the manuscript. RM drafted the manuscript and performed the revision of the final document. All authors read and approved the final manuscript.

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I tumori filloidi sono rare neoplasie fibroepiteliali che rappresentano circa lo 0,2-2% di tutti i tumori al seno. La classificazione proposta dalla Organizzazione Mondiale della Sanità divide i tumori filloidi in benigni, borderline e maligni in base ad una combinazione di varie caratteristiche istopatologiche. I tumori filloidi maligni giganti, con un diametro maggiore di 10 cm, sono molto rari. Noi presentiamo un caso insolito di tumore filloide maligno gigante con metastasi, che nell'arco di 6 anni è cresciuto così tanto da determinare una significativa deformazione del corpo con trasformazione del fisico della paziente. Abbiamo pertanto eseguito un intervento chirurgico di mastectomia con asportazione della neoplasia con l'obiettivo di migliorare la qualità di vita della paziente, indipendentemente dalla prognosi estremamente sfavorevole. Ci auguriamo che la descrizione di questo particolare ed insolito caso possa essere l'occasione per rimarcare ancora una volta l'importanza di una corretta educazione alla salute del seno e sottolineare le conseguenze negative di trattamenti alternativi o ritardati.

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