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CASE REPORT

A rare case of a giant phyllodes tumor with degeneration and bleeding: diagnosis and treatment difficulties

Un caso raro de tumor filoides gigante con degeneración y hemorragia: dificultades de diagnóstico y tratamiento

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Abstract

One of the leading problems worldwide in oncology and surgery is breast cancer. There were 2,261,419 newly diagnosed cases of breast cancer in 2020, which is 11.7% of the total number of diagnosed cases of malignant tumors of various localizations and is one of the leading causes of mortality in women. The diagnosis of breast cancer is often a difficult case when there is no histological verification of the tumor or the results are inconclusive. In these setting proper exclusion of benign masses is essential. Among the benign neoplasms, a special place is occupied by an extremely rare type of tumor – a phyllodes tumor, which makes up 0.3-0.5% of all breast tumors. A phyllodes tumor is a fibroepithelial neoplasm characterized by an extreme rate of development and the ability to transform into a malignant tumor (sarcoma). The current case report describes a giant phyllodes tumor with degeneration and bleeding the required emergency treatment.

Key words: phyllodes tumor, mastectomy, breast cancer.

Resumen

Uno de los principales problemas a nivel mundial en oncología y cirugía es el cáncer de mama. En 2020 se diagnosticaron 2.261.419 nuevos casos de cáncer de mama, lo que supone el 11,7% del total de casos diagnosticados de tumores malignos de diversas localizaciones y es una de las principales causas de mortalidad en la mujer. El diagnóstico del cáncer de mama es a menudo un caso difícil cuando no hay verificación histológica del tumor o los resultados no son concluyentes. En estos casos es esencial excluir adecuadamente las masas benignas. Entre las neoplasias benignas, ocupa un lugar especial un tipo de tumor extremadamente raro: el tumor filodes, que constituye el 0,3-0,5% de todos los tumores de mama. Un tumor filodes es una neoplasia fibroepitelial caracterizada por una tasa de desarrollo extrema y la capacidad de transformarse en un tumor maligno (sarcoma). El presente caso describe un tumor filodes gigante con degeneración y hemorragia que requirió tratamiento de urgencia.

Palabras clave: Tumor filodes; mastectomía; cáncer de mama.

Introduction

One of the leading problems worldwide in oncology and surgery is breast cancer. Early diagnosis, prevention and treatment should be aimed at organ-preserving methods. Breast cancer is one of the most common malignancies among women[1]. According to GLOBOCAN[2], in 2020, 2,261,419 new cases of breast cancer were registered in the world, which is 11.7% of the total number of newly diagnosed cases of malignant tumors of various localizations. Mortality was 6.9% of the total number of deaths from malignant neoplasms, which corresponds to 684,996 cases. Therefore oncological screening and alertness among physicians in the diagnosis of neoplasms of various localization is essential.

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However, some of the mass lesions of the mammary gland are also benign neoplasms, which can be initially diagnosed as cancer. Among the benign neoplasms, a special place is occupied by an extremely rare type of tumor - a phyllodes tumor, which makes up 0.3-0.5% of all breast mass lesions[3]. A phyllodes tumor is a fibroepithelial neoplasm characterized by an extreme rate of development and the ability to transform into a malignant tumor (sarcoma). The etiology and pathogenesis of phyllodes tumors has not yet been clarified. There are many hypotheses of their origin, including the influence of genetic factors. Phylloid (leaf-shaped) tumor has a favorable prognosis and a 10-year survival rate of 87%[4,5].

This article presents a clinical case of management of a patient with a phyllodes tumor. Initially, after a comprehensive examination, she was suspected to have breast cancer. The patient was urgently hospitalized to the clinic due to the degeneration of the tumor that resulted in bleeding.

Case report

A 52-year-old woman, has been under observation for two years due to a mass of the left mammary gland, which she accidentally identified during palpation. The patient also had uterine myoma and iron deficiency anemia. Regarding the latter, she was consulted by a hematologist and underwent corrective treatment. Planned hospitalization and surgical treatment for breast formation were postponed several times due to difficult-to-correct anemia, with a decrease in hemoglobin level to 60 g/l and she was lost to follow-up on multiple occasions. In the period from 2020 to 2021, she noted an increase in size of the mass, and in May 2021, the patient was consulted by an oncologist. During examination there was a mass in the upper outer quadrant, that deformed the contour of the mammary gland. The skin was pale pink, the nipple and areola were without particularities but there was a positive symptom of “lemon peel” on the border of the outer quadrants of the left breast. She had a nodular mass in the upper outer quadrant of the left breast, on palpation with clear uneven contours, stiff, about 6 cm in size (negative progression compared to the data from 2020). The skin over the mass was hyperemic, not mobile; there were no discharge from the nipple. According to the ultrasound examination of the mammary glands from May 2021: at the border of the outer quadrants of the left mammary gland, there was a hypoechoic mass with a clear uneven contour of 4.5 * 5.5 * 4.1 cm. Mammography from May 2021 demonstrated a clustered microcalcifications and numerous areas of increased density, covered by surrounding tissues (**Figures 1A, 2A**).

Figure 1: Lateromedial mammography. **A** - 2021; **B** - 2022.

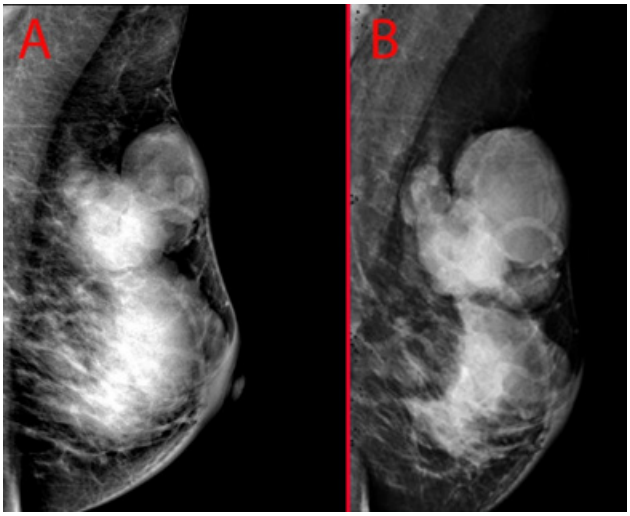


Figure 2: Craniocaudal mammography. **A** - 2021; **B** - 2022.

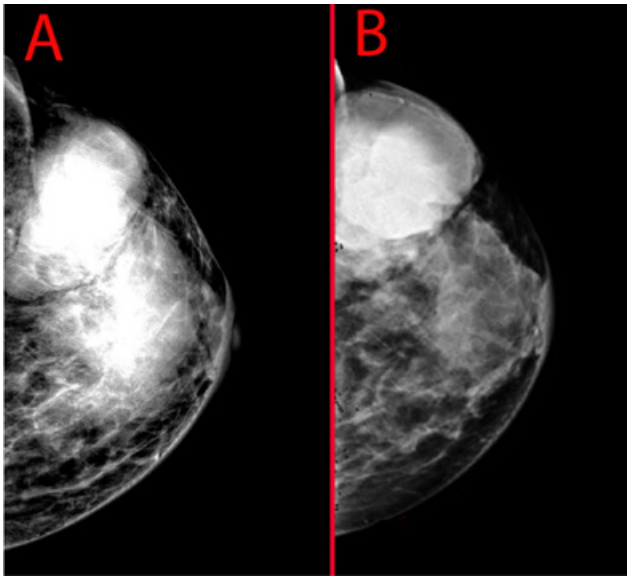
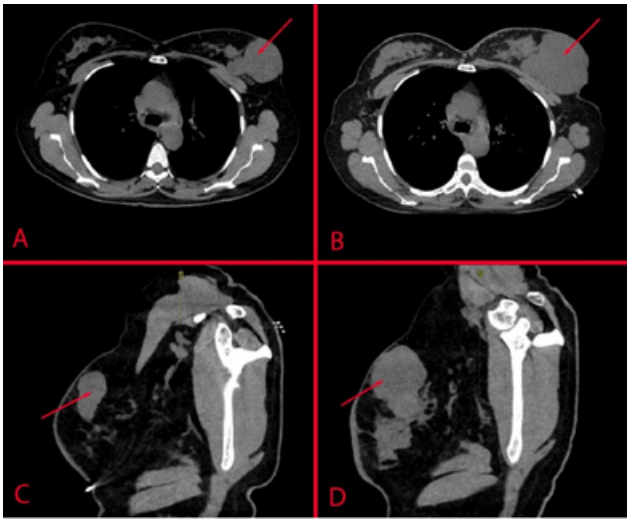


Figure 3: Mass of the mammary gland with a difference of one year (CT scan of the chest,). A - CT image from 2021 (axial section, the mass is indicated by an arrow); C - CT image from 2022 (axial section, the mass is indicated by an arrow); C - CT image from 2021 (sagittal section, the mass is indicated by an arrow); D - CT image from 2022 (sagittal section, the mass is indicated by an arrow).



The patient undergone a core-needle biopsy of the mass. According to the results of a histological study from May 2021 the specimen was presented by columns of breast tissue with fibrous and hyalinized stroma and proliferation foci with mild nuclear polymorphism. Mitoses were not reliably detected. The stroma showed compressed and dilated ducts with focal hyperplasia without atypia. The conclusion of the histological examination was that the mass was a fibroepithelial tumor of the mammary gland with an uncertain potential of malignancy. On CT scan of the chest from September 2021, the mass of the left mammary gland was determined as a group of nodes up to 72 mm, with a density of 13 HU. There were single subpleural foci up to 8 mm in size in both lungs (C8 segments), which did not allow to exclude secondary genesis (**Figures 3A, C**).

The patient was consulted by an oncologist due to suspected metastatic lung disease. However, the patient was lost to follow-up and came to the appointment only in October 2022. Repeated mammography showed an increase in the size of the mass (**Figures 1B, 2B**). During this period the patient was observed in the clinic, carried out the correction of anemia. A CT scan of the chest was performed in October 2022, which demonstrated that the lungs were without infiltrative changes only single dense small nodules of a post-inflammatory nature. There was an increase in the mass of the left breast (**Figures 3B, 3D**). An oncological consilium was held in October 2022. The consilium recommended PET-CT scan of the whole body in order to exclude secondary metastatic lesions. Due to the deterioration of the patients' condition, the degeneration of the tumor, its breakthrough to the surface of the skin and bleeding, she was hospitalized at the end of October 2022 on an emergency basis to our hospital.

During preoperative examination of the patient she had severe anemia (hemoglobin - 63 g/l). Objective examination the surface of the skin revealed an ulcerated protruding brown mass up to 10 cm in diameter that teared the skin, without involvement of the nipple (**Figures 4 A, B**). In the conditions of the oncological surgical department, as part of the preoperative preparation for surgery, anemia was corrected by transfusion of blood components (2 doses of erythrocyte suspension equal to 600 ml) and intravenous administration of iron (Likferr 20 mg/ml - 5 ml for 3 days). The bleeding was stopped by hemostatic sponges.

Ultrasound of the breast revealed fibrocystic mastopathy, multiple anechoic cysts, a mass with a lobulated contour, richly vascularized in the Doppler mapping mode (**Figure 5**).

Breast-sparing procedure was no possible due to the size of the mass, that occupied the majority of the breast. We performed a left mastectomy after preoperative stabilization and correction of anemia (hemoglobin 80 g/l). The postoperative period was uneventful, without complications and she received antibacterial treatment (1 g of cefotaxime 2 times a day), iron infusion (Likferr 20 mg/ml - 5 ml), symptomatic therapy (diclofenac 50 mg/ml - 3 ml and ketorol 20 mg/day). She was discharged on the 3rd day after surgery in a satisfactory condition.

Figure 4: Preoperative image of the mammary gland. A - anterior projection; B - lateral projection.

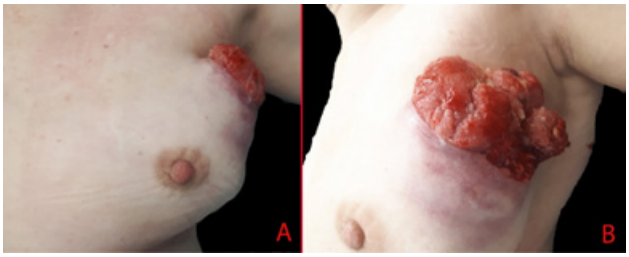
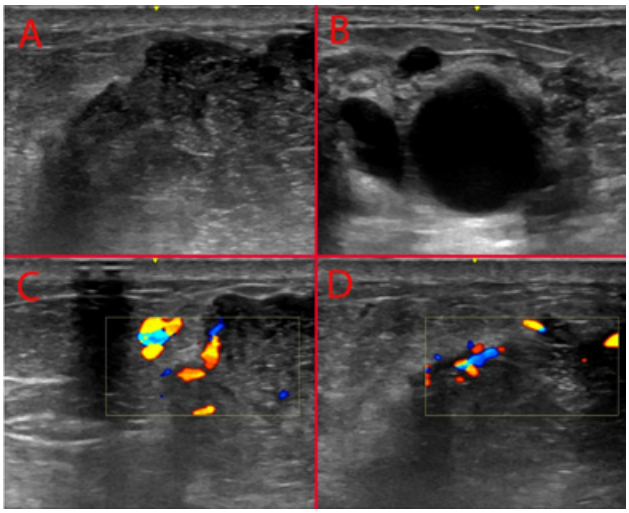


Figure 5: Ultrasound image of a mass of the mammary gland. A - The border of healthy and affected tissue; B - Multiple anechoic cysts of the mammary gland; C, D - the tumor is penetrated by large vessels.



According to histological examination the areas of breast tissue had growth of a leaf-shaped tumor with a typical epithelial lining of the ductal type, slightly increased cellularity of the stroma and numerous focal degenerative changes. The foci had multiple sites of degeneration and necrosis, lymphohistiocytic infiltrates, mycosidosis, hyalinosis, leukocyte infiltrates and ulceration of the skin over the tumor. There were no tumor emboli in blood and lymphatic vessels and intact resection margins (R0). According to the results of immunohistochemical studies an insignificant part of the tumor cells was positive for CD117 and p53, and Ki67 was 5%. Based on these results the tumor was classified as a benign phyllodes tumor of the mammary gland with degenerative and inflammatory changes. During the observation of the patient over a 6-month period there were no data for relapse after surgery and her hemoglobin level stabilized at the level of 90-100 g/l.

Discussion

Phylloid (leaf-shaped) tumor is a rare fibroepithelial tumor that can be benign, malignant or borderline. There is no definite algorithm for the management and treatment of patients with this type of tumor. There are mostly separate clinical observations and literature reviews on phyllodes tumors, but they are descriptive in nature and reveal the management of the patient only in a certain situation.

The standard for diagnosing both benign and malignant neoplasms of the breast are mammography and ultrasound of the mammary glands, but the sensitivity and accuracy of these methods for diagnosing a phyllodes tumor are minimal[6]. MRI of the mammary glands provides a more accurate result. According to a number of authors, there are features that may be more characteristic for phyllodes tumors when comparing patients with various tumors. The presence of a cystic component, strong lobulation (presence of septa) suggests a phyllodes tumor in 63%-74% of cases. Delayed phase contrast enhancement of T1WI demonstrates a heterogeneity of the structure of the mas that is more common in phyllodes tumors than in fibroadenomas and other benign neoplasms, while the homogeneity of the structure of the mass in 67% of cases was characteristic of fibroadenomas[7].

In most cases, these methods do not provide high accuracy in the diagnosis of phyllodes tumors, therefore histological verification is necessary. Fine-needle aspiration of a mass is uninformative taking into account that the sensitivity of the method is 22.7%. A core-needle biopsy followed by an immunohistochemical study is required for a morphological diagnosis. The sensitivity of the method is 66.2% and according to the available data, the diagnosis of a phyllodes tumor is established using a core biopsy in about half of the cases[5]. There are several grading systems, but the most common is classification as benign, borderline and malignant. This is based on the assessment of nuclear pleomorphism of stromal cells, stromal overgrowth, increased mitoses, increased stromal cellularity, which is usually diffuse, and infiltrative borders[3]. One study showed a correlation between the histological characteristics of a phyllodes tumor obtained by core-needle biopsy and the results of postoperative histological and immunohistochemical studies. Cases in which a phyllodes tumor was not ruled out were evaluated for cellular component, stromal distribution and outgrowth, nuclear atypia, and mitotic activity. The use of Ki67, topoisomerase IIa, CD34, CD117 and Bcl-2 may be helpful to properly classify the tumor. The obtained results correlated with the results of subsequent postoperative studies. In 58% of cases, the results of core-needle biopsy were identical with the results of postoperative histological studies. Hypercellularity and pseudoangiomatous hyperplasia of the stroma, moderate nuclear atypia, mitotic activity, and indistinct boundaries of the lesion, in 73-87% of cases, correlated with the postoperative pathomorphological diagnosis of a phylloid tumor[8,9]. Other immunohistochemical markers may include p53, EGFR, p16, and VEGF. Therefore, the abovementioned histological and immunohistochemical parameters can make it possible to predict the diagnosis of a phyllodes tumor based on core-needle biopsy data, but the accuracy of the method is far from ideal. The differential diagnosis should include: fibroadenoma, sarcomas, periductal stromal tumor, metaplastic carcinoma[4].

In the presented clinical case, the patient was not diagnosed based on core-needle biopsy. The patient did not undergo surgery for a long time due to a concomitant pathological condition (difficult to correct anemia), as well as suspicions of a malignant process and metastasis. During the year of observation, an intensive growth of the mas was noted (its increase by 2 times), as well as skin rupture, degeneration and bleeding, uncharacteristic for a benign mass. However, cancer tend to invade the surrounding tissue and grow through the skin, which was not noted in this case. In the literature, there are clinical cases of a giant phyllodes tumor, where the formation reached 45 cm, was accompanied by its decay, anemia, axillary lymphadenopathy, but without bleeding. The patient in the reported case underwent surgery in the amount of mastectomy and regional lymphadenectomy[10].

There is no data indicating bleeding from phyllodes tumors and skin rupture. In the reported clinical observations, the mass usually increased in size, the skin over them becomes thinner, acquires a bluish tint, but does not tear the skin under pressure.

The criteria (rapid growth of the lesion, skin rupture, degeneration of the lesion, bleeding), described in our clinical case, gave reason to differentiate this lesion from a malignant process. However, the management of such patient is difficult due to uncertain initial diagnosis. In an emergency situation that arose in the patient, it was important to quickly and efficiently provide medical care and, to the extent possible, to choose a radical method of surgical treatment.

Taking into account the large size of the tumor, we decided to perform a mastectomy after preoperative preparation. According to the literature, the treatment of benign phyllodes tumors is limited to surgery. For small tumors, a sectoral resection of the mammary gland is performed. Tumor enucleation is unacceptable due to almost 100% risk of tumor recurrence. The standard of care for phyllodes tumors is complete surgical excision with intact resection margins. In case of large bulky phyllodes tumors, mastectomy without lymphadenectomy is often the only possible

surgical procedure. Jae Hyuck Jang and coworkers in their retrospective study conducted from January 1995 to July 2009 reviewed 164 cases of surgical treatment of phyllodes tumors. A total of 148 (90.2%) patients underwent surgical excision of the tumor in the amount of sectoral resection. Mastectomy was performed in 16 patients (9.8%)[9]. The ultimate goal of such procedures should be breast preservation. Unfortunately, this is not always possible in case of large tumors with degeneration and bleeding. However, due to the rare incidence of such tumors diagnosis can be difficult. Based on SEER data registry (2000-2004) only 500 cases of phyllodes tumor are diagnosed in the US annually[11]. Breast reconstruction can be performed in these patients with other myocutaneous flaps or implant. However, they require a higher expertise level involving microsurgery and plastic surgery, a longer operative time and hospital stay[12-14]. In our case this was not possible due to emergency procedure, unstable condition and possible infectious complications after surgery.

Taking into account the size of the mass at admission, it was not possible to carry out an organ-preserving operation, because the tumor occupied most of the gland. The presented clinical case of a phyllodes tumor shows the complexity of diagnosis and the importance of a thorough history, examination of the patient and interpretation of the acquired data.

Conclusion

Phyllodes tumor is a rare benign tumor that can undergo transformation into sarcoma. In rare instances these tumors can grow to large size and can even breakthrough the skin requiring emergency treatment. In case of giant tumors, the optimal treatment option is excision with clear surgical margin when possible. This group of patients are rarely admitted in emergency settings with signs of tumor degeneration and bleeding that requires emergency mastectomy. However, the presented clinical case of a phyllodes tumor shows the complexity of diagnosis and the importance of a thorough history, examination of the patient and interpretation of the acquired data. Proper preoperative evaluation may lead to high index of suspicion that the tumor may actually be benign in its nature.

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Informed Consent Statement

Written informed consent has been obtained from the patient to publish this paper.

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Conflicts of Interest

The authors declare no conflict of interest.

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