

Two relatives female patients with primary malignant phyllodes sarcoma and primary stromal sarcoma of breast - A clinical case

Strahil Strashilov¹, Angel Danchev Yordanov², Stanislav Slavchev³, Vasil Nanev⁴, Denislava Ivanova⁵, Momchil Ivanov⁶, and Ivan Ivanov⁷

1. Department of Plastic Restorative, Reconstructive and Aesthetic Surgery, Medical University Pleven, Bulgaria
2. Department of Gynecologic Oncology, Medical University Pleven, Bulgaria
3. Clinic of Gynaecology, University Hospital "St. Anna"-Varna, Bulgaria
4. Department of Surgical Oncology, Medical University Pleven, Bulgaria
5. Department of Obstetrics and Gynecology, University Hospital Sofamed, Sofia, Bulgaria
6. MHAT "Saint Paraskeva"-Pleven, Bulgaria
7. Department of Clinical Pathology, Medical University Pleven, Bulgaria

CASE STUDY

Please cite this paper as: Strashilov S, Yordanov A, Slavchev S, Nanev V, Ivanova D, Ivanov M, Ivanov I. Two relatives female patients with primary malignant phyllodes sarcoma and primary stromal sarcoma of breast - A clinical case. AMJ 2019;12(9):251–254.

<https://doi.org/10.35841/1836-1935.12.9.251-254>

Corresponding Author:

Angel Danchev Yordanov

Clinic of Gynecologic Oncology, University Hospital "Dr. Georgi Stranski", Medical University Pleven, Georgi Kochev 8A, Bulgaria

Email: angel.jordanov@gmail.com

ABSTRACT

The primary sarcoma of breast is a rare malignant tumour, which develops from the mesenchymal tissue of the mammary gland. It represents less than 1 per cent of all malignant diseases of the breast. The incidence is about 17 new cases per 1 000 000 women. The aetiology of that disease is unknown. The main method of treatment is the surgical excision with which includes the "safety-margin" of healthy-looking tissues.

We present a 57-year-old woman with probably familial, primary, malignant malignant phyllodes sarcoma. Her niece was diagnosed with periductal stromal sarcoma of the left breast five years ago. This evoked in us the idea that there is

probably a familial connection between the two diseases.

After radical surgery the patient was undergone to radiotherapy and she is without any signs of recurrence till now.

Key Words

Malignant phyllodes sarcoma, stromal sarcoma of breast, sarcoma of the breast

Implications for Practice:

1. What is known about this subject?

The primary sarcoma of breast is a rare malignant tumour.

2. What new information is offered in this case study?

We present a woman with probably familial, primary, malignant phyllodes sarcoma.

3. What are the implications for research, policy, or practice?

There probably exists genetical predisposition in certain families connected with the development of sarcoma of breast.

Background

The primary sarcoma of breast is a rare malignant tumour, which develops from the mesenchymal tissue of the mammary gland.¹⁻³ It represents less than 1 per cent of all malignant diseases of breast.⁴ The incidence is about 17 new cases per 1 000 000 women.⁵ The aetiology of that disease is unknown.⁶ The main method of treatment is the surgical excision with includes "safety-margin" of healthy-looking tissues, while in principle the axillary dissection is

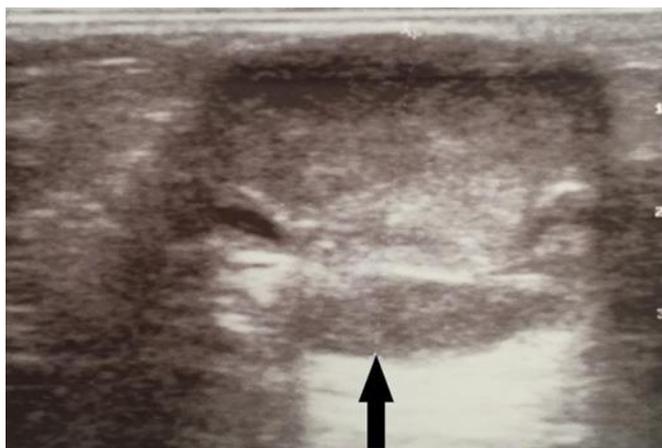
not indicated, apart from cases with lymph nodes clinically positive for metastases.⁷ Due to the low frequency of disease, data on that matter in the scientific literature are scanty and consist basically in case reports and small clinical series.⁸

We present a 57-year-old woman with probably familial, primary, malignant phyllodes sarcoma who has a niece diagnosed five years ago with stromal sarcoma of the left breast.

Case details

We present a 57-year-old woman with depressive disorder and chronic bronchitis as concomitant diseases, with no abnormalities ascertained by laboratory examinations, who were admitted at this Clinical Department of Surgery in March 2019, after signing of informed consent on the occasion of malignant phyllodes tumour of the left mammary gland. The patient palpated a hard as stone lump - one month before hospitalization - in the upper outer part of her left breast, with the size of a child's fist. Immediately after that, she underwent a physical examination, which proved the palpatory finding, and each mammography, which ascertained a neoplastic lesion with zones of lysis, with the total size of 5/4cm, localized in upper outer quadrant (Figure 1).

Figure 1: An echo mammographic image of a tumour of the left breast



During the examination of the patient, a fine-needle aspiration biopsy from the tumour was taken - under echographic control - the result of which, based on a cytological examination, was cells suspected for malignancy. Due to the ambiguous result from that examination, performing the open biopsy under local anaesthesia became necessary for precise histological verification of process, the result of which was malignant phyllodes

tumour.

A modified, radical mastectomy on the left side was undertaken at the Clinical Department of Surgery for complete excision of the malignant process. After the intervention, the breast - together with the lymph nodes - was sent for histological examination, which confirmed the primary diagnosis (Figures 2 and 3).

Figure 2: Macroscopic appearance of malignant phyllodes tumour

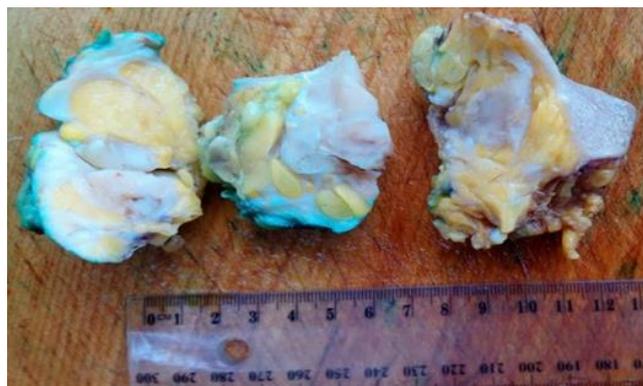
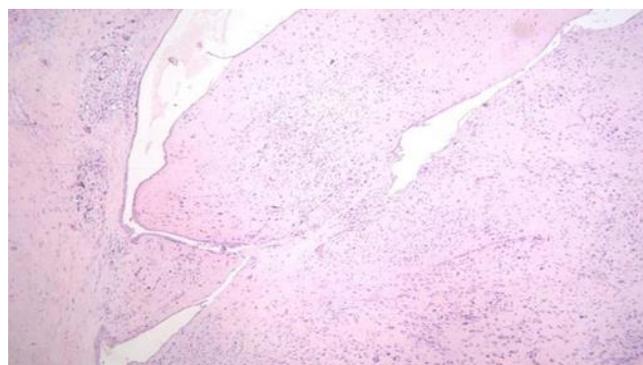


Figure 3: Histological picture of the malignant phyllodes tumour after staining with haematoxylin and eosin



No metastases in the axillary lymph nodes were found by histological examination.

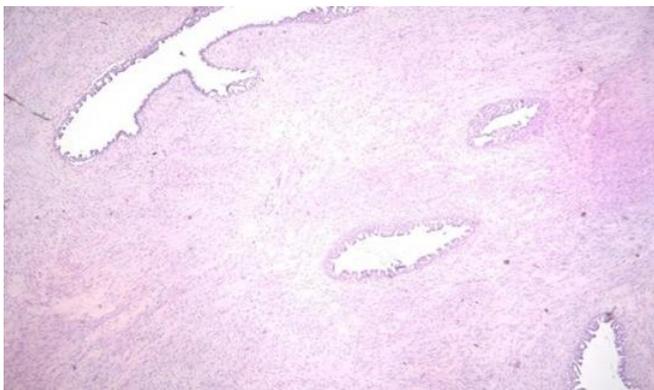
The chest radiograph and echography of the abdomen and lesser pelvis, which were performed, did not demonstrate any data on the dissemination of the process.

The patient was reported at the Local Oncological Committee, which appointed additionally only local radiotherapy. The latter therapy was timely performed and the patient is without signs of recurrence for five months.

During the patient's hospital stay at this Clinical Department, we carried out a thorough investigation of her

family history and found that her niece was also operated from a malignant disease of left breast five years ago. We explored the medical documents on the case after the informed consent of the relative. We found that this woman had an operated and histologically proved periductal stromal sarcoma of the left breast (Figure 4), which evoked in us the idea that there is probably a familial connection between the two diseases.

Figure 4: Histological picture of periductal stromal sarcoma in the patient's niece after staining with haematoxylin and eosin



Discussion

Malignant diseases of the breast are the most common oncological diseases in women.⁹ They have most frequently epithelial origin (carcinomas) and very rarely a mesenchymal one (sarcomas).¹⁰ The primary sarcoma of breast represents a very rare morbid entity, while several hundred cases of that kind were described in the world scientific literature until now.⁹ That disease most frequently affects women from 50–70 years of age, while in principle it may occur in people of all ages.¹¹ We must note that the primary sarcoma of breast is also observed in men, while its frequency is 5 per cent of all resemblant sarcomas.¹² Most of those sarcomas have secondary occurrence, while the most frequently develop after radiotherapy for the treatment of carcinoma of breast.¹⁰ This type of sarcoma originates in the mesenchymal part of the breast (the stroma), which comprises a big number of different types of cells as fibrocytes, and adipose, endothelial and muscular cells, etc. This is the reason the primary sarcoma of the breast to represent a heterogeneous group and to be named stromal sarcoma.^{7,8} The most common subtypes - based on anatomic pathology - are fibrous histiocytoma, fibrosarcoma, angiosarcoma, and spindle cell sarcoma.⁹ There are some immunohistochemical markers that can improve diagnose as CD10, CD29, vimentin, SMA p63, calponin, EGFR.¹¹

The risk factors for the development of sarcoma of the breast are not completely explicit, but the most frequent among them are the previous fibroadenoma of breast, external ionizing radiation, lymphedema of the breast, neurofibromatosis, etc.⁹

Those neoplasms represent clinically a rapidly growing tumour mass in one of the breasts, and they can reach gigantic sizes of up to 40cm in diameter.^{6,13} The skin above the tumour is most frequently without change, but it may be discoloured. They frequently produce local relapses and distant haematogenous metastases in the lung, bones, liver, etc.⁶

The group of breast sarcomas is reported as stromal sarcomas.⁷ The term stromal sarcoma means tumours arising from the intralobular stroma of the breast.¹⁴ These tumours contained specific type of stroma and they are different from those arising in usual soft tissue zones.

The diagnosis of primary sarcoma of breast is made using clinical examination, medical imaging (mammography, echomammography, magnetic resonance imaging), and biopsy. Due to the non-typical finding and rarity of the disease, it is frequently difficult to be diagnosed correctly and is confirmed mostly by histological examination.^{6,13} The early diagnostics of the disease are of particular significance because the survival rate depends directly on it.¹²

The sarcomatoid carcinoma, carcinosarcoma, fibromatosis, nodular fasciitis, and fibrous histiocytoma have to be considered in differential diagnostics.⁶

The therapy of primary sarcoma of breast is complex, while the surgical treatment plays a basic role in it. It includes most frequently the removal of the whole breast together with the tumour and pectoral fascia, while axillary lymph node dissection is not applied or is applied only in cases of lymphatic metastases. The rest of the therapy includes radiotherapy and chemotherapy.¹⁰

In our case, the sarcoma of breast presented as carcinoma based on the clinical picture, medical imaging, and cytological examination. The only aspect, which struck us, and was different from the situation with breast cancer, was the absence of metastases in the respective axilla despite the huge size of the primary tumour. Only after the excision was performed and histological examination of the whole tumour mass was carried out, was the final diagnosis made. Another interesting aspect that we found when investigating the family history of the patient was the

presence of primary sarcoma of the breast in a second-degree relative of the female sex. That suggested to us that there probably is a hereditary connection between the two diseases. This is because both of the tumours have similar provenance and origin from same tissue and they have some similarities in pathological findings. Of course we cannot prove it with sure. During the extensive investigation of scientific literature on the matter carried out by us, we did not find anywhere description of such a connection, but we consider that such one exists, and it should become an object of new genetical studies in families with sarcoma of the breast.

Conclusion

The primary sarcoma of breast is a very rare disease. The clinical, echomammographic, and cytological pictures of primary sarcoma of breast completely cover those on carcinoma of the breast, which makes virtually impossible its preoperative diagnosis. In the case of primary sarcoma of breast, regional lymphatic metastases are observed very rarely. There probably exists genetical predisposition in certain families connected with the development of sarcoma of the breast.

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PEER REVIEW

Not commissioned. Externally peer reviewed.

CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

FUNDING

None

PATIENT CONSENT

The authors, *Strashilov S, Yordanov A, Slavchev S, Nanev V, Ivanova D, Ivanov M, Ivanov I*, declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.