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CASE REPORT

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Wegener's granulomatosis of the breast: A case report

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Summary

Background: Differential diagnosis and detection of malignant lesions in mammography poses a challenge for many diagnosticians. Although mammography continues to be the best and cost-effective way to detect breast cancer, it has its limitations due in part to the radiological appearance of changes of a very rare condition. We are presented with changes that have met well-known radiologic criteria for the diagnosis of malignancy, yet they turn out to be benign, because they belong to a group of extraordinary lesions.

Case Report: In June 2010, a 56-year-old woman found a lump in her right breast. Mammography performed at another medical center, revealed a change 12 mm in diameter, rated as BI-RADS 4C. Physical examination revealed a palpable lesion, 15 mm in diameter. Ultrasonography revealed hypoechogenic change with ill-defined outlines, size 14×10 mm – BI-RADS 5. Fine needle aspiration biopsy was done and revealed no atypical cells. The patient underwent an open surgical biopsy and a histopathological diagnosis was suggested to be a type of Wegener's granulomatosis. Exclusion of cancer enabled the continuation of medical treatment of the underlying disease. There were no changes in the breast in follow-up studies.

Conclusions: The differential diagnosis of breast tumors should include rare conditions that can sometimes mimic breast cancer. One of them is Wegener's granulomatosis. Both, the diagnosis and treatment of the disease, require the cooperation of specialists from various fields, including clinicians, pathologists and radiologists.

Keywords: Breast Tumor • Wegener's Granulomatosis • Mammography

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Background

Differential diagnosis and detection of malignant lesions in mammography is still poses a challenge for many diagnosticians. Continuing education and experience of the clinician describing the lesion, play a significant role. In an attempt to maximize the rate of detection of malignant lesions various classifications of lesions visibile on mammography have been developed, e.g. de Gal classification of microcalcifications, BI- RADS classification for the assessment of the likely nature of the lesion. Although mammography continues to be the best and most cost-effective method for the detection of breast cancer, is has its limitations resulting from e.g. dense structure of the breast, location of lesions, their radiological appearance and the existence of rare lesions. Malignant lesions, that appear as benign, do occur. Other lesions meet the criteria for the diagnosis of malignancy, yet they are benign, because they are atypical. This paper will present one such case.

Case Report

In June 2010, a 56-y.o. patient presented to the breast disease clinic at an Oncology Center in Bydgoszcz with mammography results performed elsewhere. Test performed on 24 May 2010, revealed a lesion in the right breast, 12 mm diameter, tu malign? BI-RADS 4C. Clinical examination revealed a palpable lesion, 15 mm in diameter, somewhat hard, with irregular borders. An ultrasound performed on 17 June 2010 showed a hypoechogenic lesion in the right breast at 10 o'clock, with ill-defined borders and measuring 14×10 mm – BI-RADS 5. On 22 June 2013, fine needle aspiration biopsy was performed and revealed no atypical cells. The aspirate contained multinucleated giant cells, exudate cells and fragments of fat

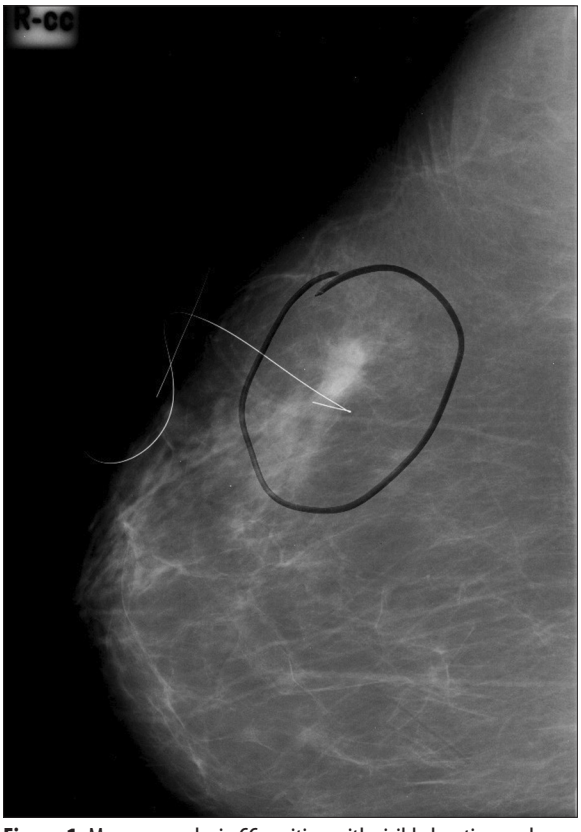

Figure 1. Mammography in CC position with visible location anchor around the lesion, blurred outlines and strong saturation.

tissue with features of fibrosis. The patient was qualified for surgery with suspicion of right breast cancer. Open surgical biopsy was performed (Figures 1 and 2). Histopathological examination dated 23 August 2010, described a necrotic nodule 1.8×1.0 cm with necrotic center and periphery with abundant lymphocytic infiltration, fibrosis, focal presence of inflammatory multinucleated giant cells. In the remaining material, 10×8 cm and 5×5 cm fat atrophy with focal mastopathy were found. Immunohistochemical studies were negative for epithelial tumor marker – cytokeratin 7 (CK7), but a positive result was obtained for vimentin (marker for tumors of mesenchymal origin) and leukocyte common antigen (LCA) encountered in lymphomas and inflammatory processes [1]. As a result of a 2008 surgery for an inflammatory tumor in the upper lobe of the right lung and a diagnosis of Wegener's Granulomatosis, tissue samples were sent for a consultation. The patient suffered from Wegener's granulomatosis since 2002, with involvement of the skin, kidneys and respiratory system. Consultation results, dated 6 September 2010, stated: "After studying the microscopic appearance of granulomatous lesions in the lung and after re-reviewing the samples of the left breast lump, there is a high similarity between both processes, due to the presence of markers of vasculitis, necrosis in granulomatous histologic differentiation with scattered giant cells of histiocytic origin and lymphatic suppurative focal inflammatory exudate. Changes observed in the breast may correspond to a process of Wegener's granulomatosis type".

Exclusion of cancer would have enabled the continuation of pharmacological treatment of the underlying disease.

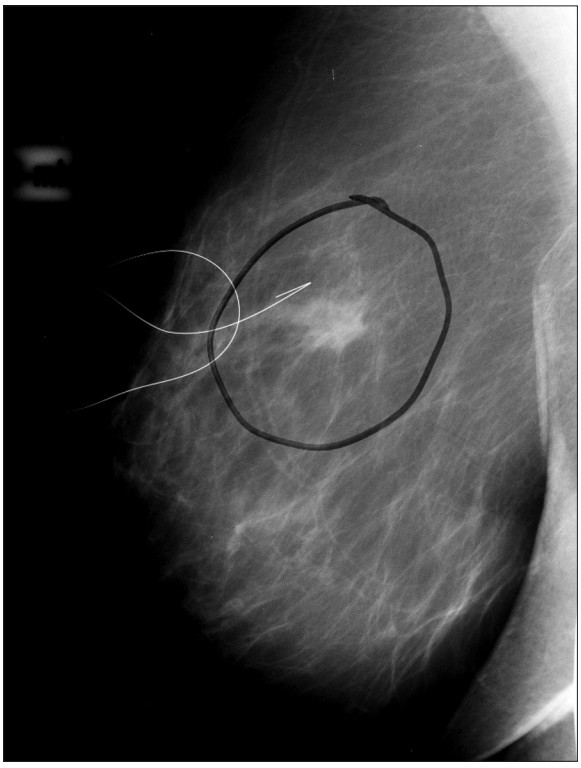

Figure 2. Mammography in a lateral position with visible location anchor around the lesion, blurred outlines and strong saturation.

The patient remains under the management of a rheumatologist and reports for follow-up breast examinations. Mammography performed on 20 August 2013, no pathologic lesions were found.

Discussion

Wegener's granulomatosis belongs to a group of diseases characterized by necrotizing inflammation of small and medium-sized blood vessels, mainly in the upper and lower respiratory tract and the kidneys. Its incidence is estimated to be about 5 per 100,000 persons. Men are affected more frequently. Etiology is unknown. Pathogenesis suggests a role of antibodies that can activate immune cells (neutrophils) and damage the endothelium, respiratory system, kidneys and other organs. Non-specific symptoms occur, such as weakness, fever, weight loss and lack of appetite [2]. The onset of the disease is dominated by the usual symptoms of upper airway disease, such as obstruction of the nasal passages, nasal secretions of various types, including bleeding, pain and tenderness in the vicinity of lateral sinuses and otitis media (hearing loss may occur). As the disease progresses, destruction of cartilage and nasal bones may occur [3]. Commonly, symptoms of lower respiratory tract involvement occur, i.e. cough, bronchitis, hemoptysis, pulmonary hemorrhage and shortness of breath. Kidney involvement is initially asymptomatic, but with time renal failure develops. Muscle and joint pain is rare. Establishment of a correct diagnosis depends on proper history and physical examination, as well as additional diagnostic tests (detection of C-ANCA antibodies, urine tests, pulmonary imaging studies). Peripheral blood reveals

neutrophilia, thrombocytosis, elevated ESR, normocytic anemia. However, final confirmation of diagnosis is possible only on the basis of a characteristic histopathologic appearance of the specimen collected during biopsy of the affected organ (usually the mucosa of the upper respiratory tract, lung or kidney). Treatment depends on the duration and form of the disease. Cyclophosphamide in combination with an oral glucocorticoid in a gradually tapered dose is used most frequently in the systemic form of the disease. Treatment should be continued for about a year after the symptoms have disappeared. Eight-year survival is 80%. The earlier the treatment is started, the better the prognosis. The prognosis is much worse if renal involvement occurs at the time of the diagnosis. Approximately 10% of patients suffer from drug-resistant form of the disease.

Wegener's granulomatosis may also present in unusual locations, such as the gastrointestinal tract, heart or nervous system [2]. Wegener's granulomatosis belongs to group of rare disorders of granulomatous inflammation of the breast. The underlying process can be related to infectious agents (tuberculosis, candidiasis), autoimmune diseases (sarcoidosis, rheumatoid nodules, Wegener's granulomatosis), but it can also be idiopathic [2]. In some cases, granulomatous inflammation occurs in the form of a tumor, which requires complete diagnostic workup to exclude breast cancer. This is the form of the disease described in our case. Previous history of Wegener's granulomatosis with systemic involvement and fulfillment of serological and histological criteria, have helped establish a proper diagnosis in this patient. Breast tissue involvement in the course of Wegener's granulomatosis occurs very rarely, as reflected in the literature [4,5]. In 2009, Allende and Booth performed a review of the English-language literature containing 28 patients with breast involvement in the course of Wegener's granulomatosis [6]. That same year, Dimitropoulos et al. reported another case [6,7]. Changes in the breast may occur as one of many manifestations in patients with a typical course of the disease, or rarely, may be the first symptom. Particularly challenging are cases with breast tumor in medical history. Most of the reported cases involved women, but in 1997, the first report of breast involvement in a man was published [7,8]. Most changes were localized to one side, but may affect both breasts. Most of the patients described received immunosuppressive therapy (cyclophosphamide and prednisone) with good outcome. Some patients underwent surgery.

Conclusions

This case confirms that the differential diagnosis of breast tumors should include rare conditions that can sometimes mimic breast cancer. Wegener's granulomatosis is a disease of multiple presentations, whose course cannot be predicted. Both, the diagnosis and treatment of the disease, require the cooperation of specialists from various fields. Oftentimes, histopathological examination is necessary to explain the changes observed in clinical imaging. Correct interpretation of microscopic findings should be based on the results of consultation by a clinician, pathologist and radiologist.

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