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Brief report

Extramedullary plasmacytoma of the breast simulating breast cancer

C. van Nieuwkoop[a,*], R.W.M. Giard[b], H.F. Veen[c], A. Dees[a]

[a]*Department of Internal Medicine, Ikazia Hospital, Montessoriweg 1, 3083 AN Rotterdam, The Netherlands*
[b]*Department of Clinical Pathology, Ikazia Hospital, Montessoriweg 1, 3083 AN Rotterdam, The Netherlands*
[c]*Department of Surgery, Ikazia Hospital, Montessoriweg 1, 3083 AN Rotterdam, The Netherlands*

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Abstract

A patient with a palpable mass in the breast suggestive of carcinoma underwent radical modified mastectomy. Surprisingly, histology of the tumor revealed an extramedullary plasmacytoma. Further diagnostic work up showed no evidence of underlying multiple myeloma. Among neoplastic lesions of the breast, although rare, malignancy of mesenchymal or lymphoproliferative origin should always be considered.

Keywords: Breast cancer; Plasmacytoma; Multiple myeloma

Introduction

Extramedullary plasmacytoma is defined as a malignant tumor of plasma cells arising in soft tissue in the absence of skeletal involvement. It may appear in any organ, either as a rare solitary form of plasma cell neoplasm, or as a (first) manifestation of disseminated multiple myeloma. It is described most frequently in the upper respiratory tract but it may also be found in the gastro-intestinal tract, lung, lymph nodes, skin and subcutaneous tissue [1,2]. Involvement of the breast is extremely rare with, to our knowledge, only 17 patients described [2–7]. We describe a patient operated upon for suspected carcinoma of the breast that subsequently turned out to be a first manifestation of multiple myeloma.

Case report

A 75-year-old woman had been seen for years by a cardiologist with aortic and mitral valve prosthesis and tricuspid regurgitation after rheumatic valve disease. As a result, chronic hemolytic anemia and atrial fibrillation occurred requiring frequent admissions for either blood transfusion or treatment of congestive heart failure. In February 1999 she was referred to a surgeon with a 4-month history of a slowly growing mass in her right breast. She denied associated pain, skin changes, inflammation or nipple discharge. There was no history of weight loss, bone pain, night sweats or recurrent infections. On exami-

*Corresponding author. Tel.: +31-10-297-5000; fax: +31-10-485-9959.

E-mail address: ikazint@knmg.nl (C. van Nieuwkoop).

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nation, both breasts showed symmetrical nipple inversion with normal skin. Palpation of the left breast was normal. In the right breast a 2×2 cm mass was palpated in the lateral upper quadrant. The tumor was firm and mobile and not attached to underlying tissue or overlying skin. There was no axillary or general lymphadenopathy and apart from a total irregular pulse rate consistent with atrial fibrillation and heart murmurs there were no abnormal findings on general physical examination.

Mammography revealed a lobulated mass measuring 2.1 cm by 1.4 cm. Cytology of fine needle aspiration suggested poorly differentiated carcinoma and a modified radical right mastectomy was performed.

Surprisingly, histology of resected tissue turned out to be subcutaneous plasmacytoma of the breast without involvement of axillary lymph nodes (Fig. 1). An immunohistochemical stain was positive for kappa and lambda light chains. There was also reaction with keratines.

Laboratory tests revealed an ESR of 50 mm in the first hour, hemoglobin 7.3 mmol/l (normal 7.5–10 mmol/l), white blood count 6.5×10^9/l ($4.0–10.0 \times 10^9$/l) with normal differential, platelet count 119×10^9/l ($130–350 \times 10^9$/l) and a reticulocyte count of 38‰ (normal 3–15‰). Serum chemistry showed: urea 8.4 mmol/l (2.7–7.5 mmol/l), creatinine 112 mmol/l (normal 55–95 mmol/l), total protein 84 g/l (64–80 g/l), calcium 2.31 mmol/l (normal 2.20–2.60 mmol/l), albumin 53 g/l (35–50 g/l/), total bilirubin 35 μmol/l (1–20), direct bilirubin 17 μmol/l (0–5), alanine aminotransaminase 26 μ/l (0–45), aspartate aminotransaminase 41 (0–40) and lactate dehydrogenase 1281 μ/l (300–620). The haptoglobin was < 0.1 g/l (0.4–2.2 g/l) and direct Coombs' test was negative. Serum protein electrophoresis, immuno-electrophoresis and immunofixation were normal. No Bence Jones or other M components were detected in the urine. A skeletal survey showed two non-specific osteolytic lesions of the skull. The patient refused a bone marrow aspirate.

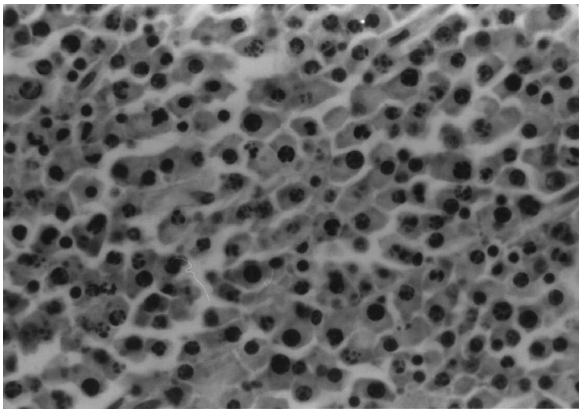

Fig. 1. High magnification of the plasmacytoma, showing sheets of plasma cells with nuclei varying in size and density (H&E stained, $\times 400$).

Discussion

Plasmacytomas usually arise from the bone marrow, known as myeloma, which can be divided into an uncommon solitary and a common multiple entity (multiple myeloma, myelomatosis). In approximately 70% of patients with multiple myeloma, autopsy findings show microscopic neoplastic plasma cell infiltration outside the skeletal system that may occur in almost any organ [1].

However, extramedullary plasmacytomas are rarely detected macroscopically and thus are seldom noted before death. Again there is a distinction between a solitary form and a disseminated form. The latter is indistinguishable from disseminated multiple myeloma and can be considered end-stage advanced disease. When present in a solitary form, the lesion is usually associated with a relatively mild behavior and long survival, suggesting that it concerns a truly different disease entity within the group of plasma cell tumors [1]. The upper respiratory tract predominates as the presenting site, but it may appear in any organ [1].

In the breast, plasmacytoma is very unusual and was first described by Cutler in 1934 [8]. Since then, several case reports have been published with reviews of the literature [2–7]. Six of these cases were solitary lesions with a maximal disease-free follow-up to 46 months after initial diagnosis. Local treatment was administered by either radiotherapy or surgery [9]. Eleven cases were associated with multiple myeloma. In six of these, the diagnosis of plasmacytoma of the breast led to the diagnosis of

multiple myeloma, while in five cases the patients were known with multiple myeloma. In these 11 cases, standard chemotherapy depending on the stage of multiple myeloma was given and prognosis depended on the stage of multiple myeloma.

In all but two cases, histology of an (excisional) biopsy or cytology with a fine needle aspirate revealed the diagnosis plasmacytoma of the breast. In one case, the biopsy specimen was initially interpreted as medullary carcinoma. A radical mastectomy was subsequently performed making the true diagnosis: solitary plasmacytoma of the breast [10]. In another case, a frozen section of incision biopsy material showed evidence of a poorly differentiated malignant tumor. Radical mastectomy was performed and eventually led to the diagnosis of plasmacytoma of the breast; which was a manifestation of recurrent multiple myeloma [2].

In our patient, physical examination and mammography suggested breast carcinoma, which was supported by cytology of fine needle aspirate showing evidence of a primary poorly differentiated adenocarcinoma of the breast. A modified radical mastectomy was performed and, surprisingly, histological examination of resected tumor tissue diagnosed a plasmacytoma of the breast. Further diagnostic work-up was complicated by the patient's attitude, refusing complete co-operation; in particular, a bone marrow aspirate. All other tests did not support the diagnosis of underlying multiple myeloma. No further treatment was therefore performed.

Curious whether progression to multiple myeloma would develop we repeated laboratory tests including complete blood count with differential, serum protein- and immuno-electrophoresis. During 18 months follow-up the results of these tests remained normal while the patient was still asymptomatic. We concluded that our patient had a solitary plasmacytoma of the breast and thus the patient belongs to the group with reasonably positive prognosis. Follow-up will be further continued, as this could be a first manifestation of multiple myeloma.

Based on our review of the literature, we conclude that plasmacytoma of the breast is exceedingly rare and, when seen, is frequently associated with multiple myeloma. Among neoplastic lesions of the breast, primary epithelial malignancies are by far the most common, but malignancies of mesenchymal or lymphoproliferative origin should not be forgotten, especially in those patients with a history of hematological malignancy. However, as seen in our patient, such considerations usually do not lead to a different approach or treatment.

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