CASE REPORT

Plasmacytoma of the Breast: A Report of a Rare Disease

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ABSTRACT

BACKGROUND: Extramedullary plasma cells tumours are rare. Much more rarer is their occurance in the breast tissue. Our aim is to report a single case of this very rare lesion (at least from an African perspective) that we incidentally diagnosed histopathologically as a primary extramedullary lesion in a 53 year old woman.

CASE DEATILS: Clinical records of a 53 year old postmenopausal woman was referred from a secondary health centre to our clinic with a three weeks' history of right breast lump were reviewed. There was no associated pain, nipple discharge, weight loss or systemic symptoms nor was there a previous history of trauma or surgery to the breast. On examination: two discrete lumps measuring 3x2 and 2 x 1.5cm in the upper medial quadrant of the right breast were identified. The lumps were firm, irregular in shape, not attached to the skin or underlying tissues. Tentative diagnosis of adenocarcinoma of the breast was made, with a differential as fat necrosis. A wide excision biopsy was done four days later for histology, after an inconclusive cytological examination of smear of which the result revealed plasmacytosis. The liver function test, Plasma proteins electrophoresis, electrolytes, urea, creatinine, bicarbonate and pelvic X-rays, and abdomino-pelvic ultrasonography were normal. Bence Jones proteins were negative in urine. Histology of bone marrow aspirate revealed scanty plasma cells.

RESULTS: She received 20mg dexamethasone, 20mg adramycin, and 2mg vincristine intravenously and 200mg of alloperinol daily by mouth for three days before leaving by the 4th treatment day against medical advice for personal reasons.

CONCLUSION: This rare lesion should sometimes be considered as a differential diagnosis of a breast lump, as it does not differ from the common lesions clinically, especially in older women.

KEYWORDS: Extramedullary, plasmacytoma, primary, breast, lesion

***DOI:*http://dx.doi.org/10.4314/ejhs.v25i4.12**

INTRODUCTION

Plasmacytoma is a malignant proliferation of plasma cells (1-3). It usually occurs as a component of systemic multiple myeloma, in association with elevation of serum paraprotein (4-6). When located outside the bone marrow, it is rightly referred to as extramedullary Plasmacytoma (1-6). The most common site of occurrence is the upper airways (3,5,6). Occasionally, an extramedullary Plasmacytoma may occur as an isolated tumour without an associated elevation of serum paraprotein or multiple myeloma in areas such as the lungs, gastrointestinal tract, spleen, pancreas and, very rarely, in the breast (7-10). When it occurs in the breast, it usually manifest as a lump, in a fashion similar to many lumpy lesions of the breast (8-10).

The possibility of diagnosing a breast extramedullary plasmacytoma is approximately 0.01% (7,8,9). The clinical diagnosis of a breast plasmacytoma is difficult, if not impossible, because presentation mimics common breast lesions such as adenocarcinoma or a fibroadenoma (3,4). However, a history of breast lump,

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confirmed by physical examination assisted by the histology of excised specimen will in most cases lead to a diagnosis of breast plasmacytoma (1-3). Bone marrow biopsy is required to confirm or rule out the presence of plasma cells in the marrow, and a computerized tomography scan of the upper airway is required to rule out the possibility of synchronous lesions. Assay for serum paraprotein is equally essential (7,9,10).

The possibility of diagnosing a breast extramedullary plasmacytoma is approximately 0.01% (7,8,9). The clinical diagnosis of a breast plasmacytoma is difficult, if not impossible, because presentation mimics common breast lesions such as adenocarcinoma or a fibroadenoma (3,4). However, a history of breast lump, confirmed by physical examination assisted by the histology of excised specimen will in most cases lead to a diagnosis of breast plasmacytoma (1-3). Bone marrow biopsy is required to confirm or rule out the presence of plasma cells in the marrow, and a computerized tomography scan of the upper airway is required to rule out the possibility of synchronous lesions. Assay for serum paraprotein is equally essential (7,9,10).

While an adenocarcinoma of the breast will often present with ill defined edges and palpable axillary nodes (10,11), especially in stages two and above; a fibroadenoma often has smooth well defined edges without axillary adenopathy: a breast plasmacytoma as demonstrated in our patient, was not smooth, but without axillary adenopathy. In most part of Africa where many breast cancer cases present at an advanced stage, the differentiation will usually not be difficult.

Experience in the treatment of this tumour is limited because of the rarity of the disease, but total excision plus radiotherapy with or without chemotherapy is known to achieve satisfactory disease control (12, 13, 14, 15). However, the patient must be followed up for years, because many of the patients subsequently develop multiple myeloma requiring further systemic treatment and continuing care.

We report a case of histologically diagnosed solitary plasmacytoma of the right breast in a 52year old menopausal woman, who initially presented with a breast lump suspicious of breast cancer. This is probably the first reported case of such a tumour from our country and perhaps from this part of the world, based on our literature search.

CASE REPORT

A 53 year old postmenopausal woman was referred from a secondary health centre to our clinic with a three week history of a lump in her right breast. The lump was discovered while taking her bath. There was no associated pain, nipple discharge, weight loss, or systemic symptoms nor a previous history of trauma or surgery to the breast. She has no family history of breast cancer. She does not smoke cigarette nor take alcohol. She had her menarche at the age of 14 years, and was 6 years postmenopausal at the time of presentation .She was Para 10+4, breast fed all her six surviving children for at least one year and has never been on birth control pills.

On examination, she was in satisfactory physical health. The important physically findings were: two discrete lumps measuring 3x2 and 2 x 1.5cm in the upper medial quadrant of the right breast. The lumps were firm, irregular in shape, not attached to skin or underlying tissues. They were not tender to palpation, and the axillary lymph nodes were not palpable. Regional findings elsewhere were normal. A tentative diagnosis of adenocarcinoma of the breast was made, with a differential as fat necrosis. A wide excision biopsy was done four days later for histology[after an inconclusive cytological examination of a smear taking by fine needle aspiration]; the result revealed plasmacytosis.

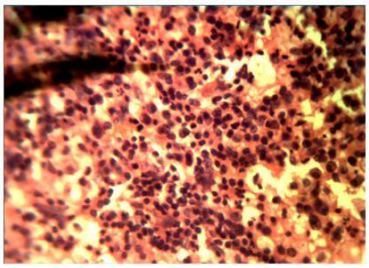

Figure 1*: Histopathologic slide of the specimen showing plasmacytoma cells*

There are several atypical plasma cells containing irregular nuclei, prominent nucleoli, in association with scattered mature plasma cells.

The liver function test, Plasma proteins electrophoresis, electrolytes, urea, creatinine, bicarbonate and pelvic X-rays, and abdomino-pelvic ultrasonography, were normal, Bence Jones proteins were negative in urine. Histology of bone marrow aspirate revealed scanty plasma cells. She was then referred to the clinical hematologist to be managed as a case of solitary plasmacytoma of the breast. She received daily, 20mg dexamethasone, 20mg adramycin, and 2mg vincristine intravenously and 200mg of alloperinol by mouth for three days. She left by the 4th treatment day against medical advice, for personal reasons.

DISCUSSION

A Solitary Plasmacytoma of the breast is extremely rare lesion (3, 8). From our literature search, only twelve cases of extramedullary plasmacytoma of the breast have been. One of the recent reports was by Mehmet et al (16) in the Turkish Journal of Hematology, of a bilateral extramedullary breast plasmacytoma. In the eleven previous reports (17) of breast plasmacytoma, three of the patients had synchronous occurrence of breast tumors and multiple myeloma, 2 presented after a previous diagnosis of multiple myeloma, and only 1 patient initially presented with breast tumour, but subsequently developed systemic disease. In the remaining 5 patients, the disease remained limited to the breast even after more than four years of follow up. The case reported by Mehmet et al, and Bloomberg et al (3) were unique in that they were bilateral diseases.

In our index patient, marrow biopsy demonstrated scanty plasma cells, abdominal ultrasonography did not reveal any lesion in the liver, spleen, nor the pancreas. Although we did not do computerized tomography scan of the upper airways, Otorhinolaryngological examination did not reveal any lesion, suggesting that the breast lesion was probably primary.

Ross et al (18) reviewed ten cases, and noted that multiple Myeloma may precede or occur synchronously with or become evident soon after the diagnosis of a breast plasmacytoma. However, the clinical course of a breast plasmacytoma depends on whether the tumour is truly solitary or just a manifestation of disseminated Myeloma (3, 12, 16). In a truly solitary disease, Luca et al (17) stated that wide local excision plus radiotherapy is usually satisfactory. The majority of patients remain free of local recurrence or systemic manifestation of Myeloma for years. In a few, however, there is an early transformation into multiple myeloma, necessitating further treatment, usually with poor comparative outcomes (14, 18).

It is critical to distinguish a breast plasmacytoma from an adenocarcinoma of the breast in order to avoid unnecessary surgery and cyclic cytotoxic chemotherapy (9, 19). This can only be done histologically, with the histologic hallmark of a plasmacytoma being the presence of several atypical plasma cells containing irregular nuclei; prominent nucleoli, in association with mature plasma cells. Marrow plasma cells are often scanty, usually less than 5% of marrow cells.

Our case demonstrated the clinical and histologic feature of this rare breast lesion. While clinical experience in the management of this lesion is limited basically because of the rarity of the disease itself; however, we believe that our initial therapeutic measures [wide excision plus combination chemotherapy] would have produced a satisfactory result had the patient been able to complete the treatment and subsequent follow-up period. The significance of this case is that it is the first reported case of primary extramedullary breast plasmacytoma, from our region of world, and most probably the 13th reported case worldwide.

The limitation of our report is that we did not follow up this patient in order to state the outcome of our management.

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