Plasmacytoma of the Bilateral Breasts: A Case Report

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AUTHORS' CONTRIBUTIONS
All authors contributed to this case report.

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None

CONSENT
No (patient remains anonymous)

ETHICAL STATEMENT/HUMAN AND ANIMAL RIGHTS
Not applicable

ABSTRACT

Extramedullary plasma cell tumors in the breast are extremely rare neoplasms which may often be misdiagnosed as primary breast carcinoma. In this case, we discuss a 76-year-old woman who was initially diagnosed with solitary extramedullary plasmacytoma of the retroperitoneum and was subsequently found to have recurrent plasmacytomas of the bilateral breasts. This report describes characteristics of plasmacytomas of breast and the characteristics on imaging.

CASE REPORT

BACKGROUND

This case describes a rare presentation of plasma cell neoplasm of the bilateral female breast. Breast plasmacytoma can mimic primary breast cancer or other lesions on imaging. This case emphasizes the importance of distinguishing plasmacytomas of the breast from primary breast tumors to provide appropriate management and therapy.

CASE REPORT

The patient is a 76 year old female with a history of treated extramedullary plasmacytoma who was found to have bilateral breast masses, which were determined to be recurrent plasmacytoma. She was diagnosed with solitary extramedullary plasmacytoma on an initial CT scan of the abdomen and pelvis for abdominal pain. Initial imaging revealed an enhancing soft tissue retroperitoneal mass measuring 5cm along the anterior L3 and L4 vertebrae and posterior to the IVC. This mass was further characterized on MRI to be homogeneously enhancing, measuring 4.6 x 5.0 x 4.5 cm located along the anterior L3 and L4 vertebrae and posterior to the IVC (Figure 1). The patient then underwent PET-CT scan, which revealed the mass to be metabolically active and without other sites of metastasis or intraosseous involvement. The retroperitoneal mass was biopsied and determined to be a plasmacytoma. She was treated with radiotherapy and completed a total dose of 45Gy, with resolution of the retroperitoneal mass.

Two years following her diagnosis, on surveillance imaging of the initial retroperitoneal mass, a CT scan revealed a small focus of locally recurrent disease as a lucent osseous lesion at L2, which raised concern for myeloma. MRI also revealed progression of disease with an enhancing retroperitoneal soft tissue mass at the level of L4-5. As a result, the patient completed a course of stereotactic body radiotherapy (SBRT) to a dose of 30Gy/5fx.

Six months later, mammography revealed new worrisome complex cystic nodules in the bilateral breasts (Figure 2,3). These complex masses were then biopsied under ultrasound guidance. Pathology revealed the breast biopsies demonstrated immunoreactivity using plasma cell markers, CD138 (Syndecan-1) and MUM1 (Figure 4,5). PET/CT was

then obtained revealed hypermetabolically active right breast nodule with SUV max of 2.2 and a hypermetabolic left breast nodule with SUV max of 3.1 (Figure 6). Restaging MRI was consistent with disease progression, including masses in the left parietal calvarium, right nasal cavity, parapharyngeal space and bilateral lacrimal glands. At this time, the patient also presented with symptoms of right sided sinus pressure and blurred vision of the right eye. She then underwent systemic therapy with lenalidomide (Revlimid), bortezomid (Velcade) and denosumab (Xgeva) with decrease in size of left parietal calvarial mass and resolution of other previously noted masses. A week later, repeat breast mammography and PET/CT were performed and showed resolution of the bilateral breast masses and breast FDG avid lesions seen previously (Figure 7,8).

DISCUSSION

Etiology & demographics

Plasmacytomas, a rare manifestation of plasma cell dyscrasias, are exceptionally uncommon within breast tissue, with recent literature primarily documenting unilateral occurrences often associated with multiple myeloma [1]. This contributes to the diagnostic challenges posed by extramedullary breast plasmacytomas, frequently misidentified as primary breast tumors or other lesions. Plasmacytomas are rarely found within breast tissue [2]. Recent literature has described the lesions to be most often unilateral and often occurring in the setting of multiple myeloma [1]. Breast plasmacytomas can be diagnosed through histological analysis, revealing atypical plasma cells containing irregular nuclei and prominent nucleoli [2]. The mean age at presentation is 53 years, with usual clinical findings of a palpable breast mass without other ancillary findings [3].

Clinical & imaging findings (Differential Diagnoses)

Distinguishing primary breast plasmacytomas from other neoplasms of the breast is essential to guiding further treatment and assessing overall prognosis. **A** key differentiator is the absence of clinical or imaging features suggestive of multiple myeloma, which helps distinguish solitary tumors from disseminated disease. Unlike multiple myeloma, primary plasmacytomas are not associated with hypercalcemia, renal insufficiency and anemia commonly seen in multiple myeloma patients. However, 20% of cases present with small M-protein levels, typically with serum or urinary paraprotein levels below 2g/dL. The differential diagnosis also includes invasive ductal carcinoma, which express epithelial markers such as estrogen receptors and cytokeratin, which are absent in plasmacytoma. Immunohistochemical staining is crucial in distinguishing plasmacytoma from other plasma cell-rich or plasmacytoid neoplasms, including IgG4-related sclerosing disease, metastatic melanoma, and extramedullary myeloid tumors, all of which exhibit distinct immunohistochemical profiles not characteristic of plasmacytoma [6].

Primary and secondary plasmacytomas do not have significant differentiating radiological characteristics. On mammography, they may present as hyperdense, round or oval masses with well- or ill-defined margins and may be associated with microcalcifications. On ultrasonography, breast plasmacytomas may be hypo- or hyperechoic with well- or ill-defined margins with hypervascularity and variable posterior acoustic features. Lastly, on MRI breast plasmacytomas manifest as intermediate/low signal on T1/T2-weighted images. High signal on DWI and low ADC values with early strong and fast/delayed washout enhancement kinetics have been shown to be suggestive of malignancy [4]. Large tumors have been associated with necrosis and infiltration or encasement of surrounding structures [5]. PET/CT can be used to detect metastatic lesions.

Treatment & prognosis

Primary breast plasmacytomas has an improved prognosis compared to that of secondary involvement of the breast by multiple myeloma. In the case of solitary disease, treatment involves wide local excision and adjuvant radiotherapy with 70% of patients remaining disease-free at 10 years [3]. Early transformation into multiple myeloma has been recorded, necessitating further treatment and is associated with poor comparative outcomes [2].

TEACHING POINT

In our index patient's case, she had a history of solitary extramedullary plasmacytoma of the retroperitoneum and then was found to have bilateral breast involvement in the setting of metastatic disease. This case emphasizes the importance of distinguishing plasmacytomas of the breast from primary breast tumors to provide appropriate management and therapy.

QUESTIONS

Question 1: Histological analysis of plasmacytoma of the breast reveals which of the following?

1. Atypical plasma cells containing irregular nuclei and prominent nucleoli (Applies)
2. Epithelial elements
3. Fascicles of spindle cells interrupted by broad bands of collagen
4. Adipocytes with a plexiform capillary network
5. None of the above

Explanation: Breast plasmacytomas can be diagnosed through histological analysis, revealing atypical plasma cells containing irregular nuclei and prominent nucleoli [2].

Question 2: True or False: The mean age of presentation of plasmacytoma is 53 years, with patients usually presenting with palpable breast mass.

1. True (Applies)
2. False

Explanation: The mean age at presentation is 53 years, with usual clinical findings of a palpable breast mass without other ancillary findings [3].

Question 3: True or False: On mammography, plasmacytoma of the breast may present as hyperdense, round or oval masses

with well- or ill-defined margins and may be associated with microcalcifications.

1. True (Applies)
2. False

Explanation: On mammography, plasmacytomas may present as hyperdense, round or oval masses with well- or ill-defined margins and may be associated with microcalcifications. Primary and secondary plasmacytomas do not have significant differentiating radiological characteristics.

Question 4: After establishing diagnosis of plasmacytoma, what is the best next step?

1. No action needed
2. 3 Month Follow Up
3. 6 Month Follow Up
4. 1 Year Follow Up
5. Wide local excision and adjuvant radiotherapy (applies)

Explanation: In the case of solitary disease, treatment involves wide local excision and adjuvant radiotherapy with 70% of patients remaining disease-free at 10 years.

Question 5: True or False: Patients with plasmacytoma of the breast are not at risk of early transformation into multiple myeloma.

1. True
2. False (applies)

Explanation: Early transformation into multiple myeloma has been recorded, necessitating further treatment and is associated with poor comparative outcomes [2].

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FIGURES

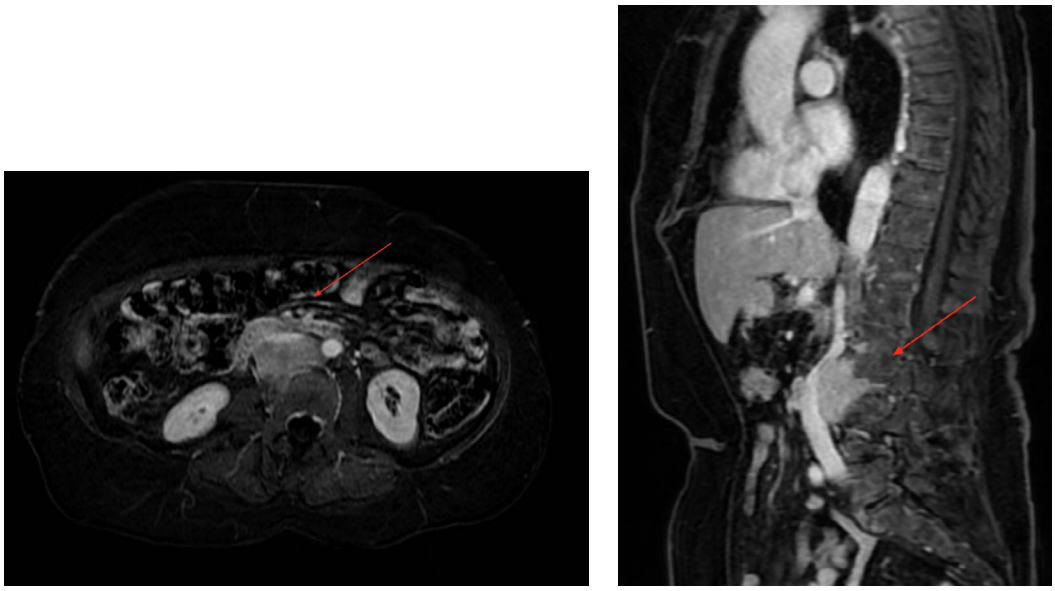

Figure 1: Contrast enhanced MRI images demonstrate a homogeneously enhancing mass corresponding to solitary extramedullary plasmacytoma of the retroperitoneum as indicated by the red arrows.

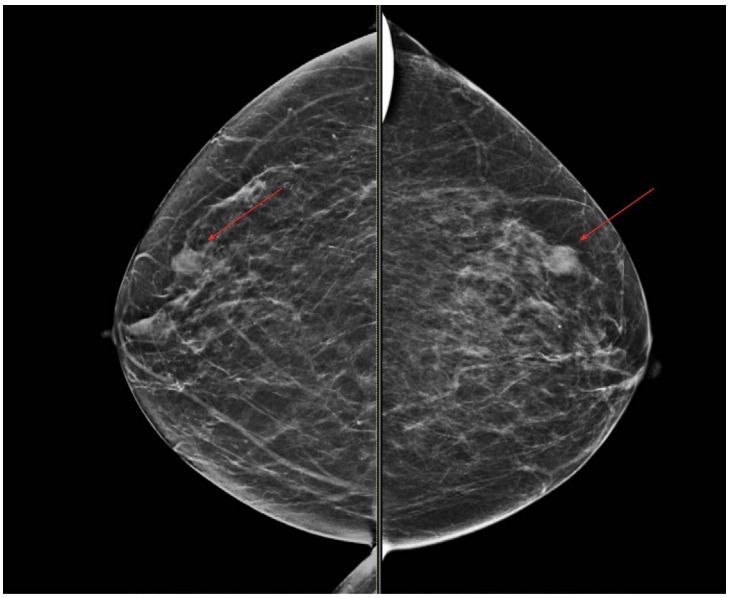

Figure 2: Bilateral craniocaudal mammographic views demonstrate bilateral breast nodules as indicated by red arrows.

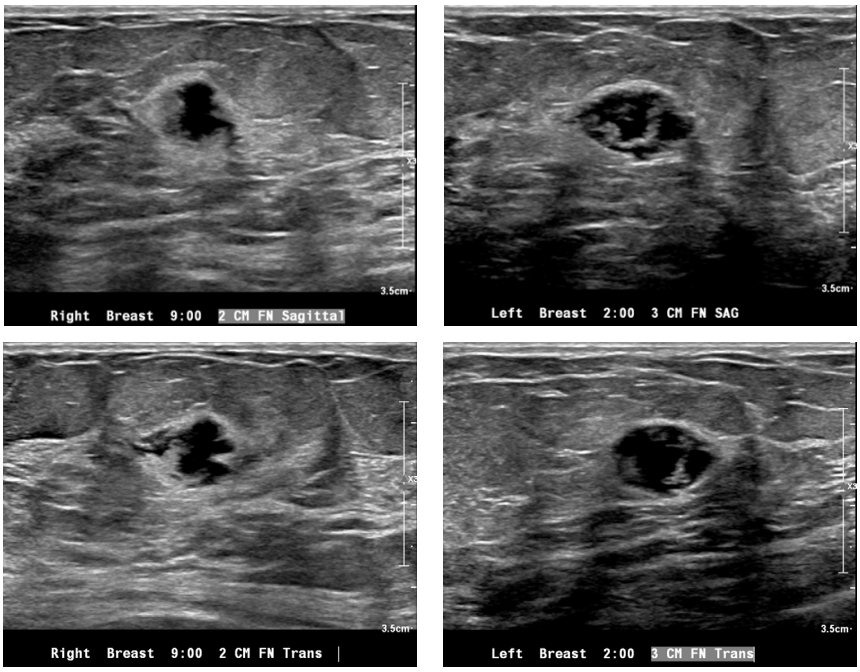

Figure 3: Sagittal and transverse ultrasound images of complex bilateral breast nodules

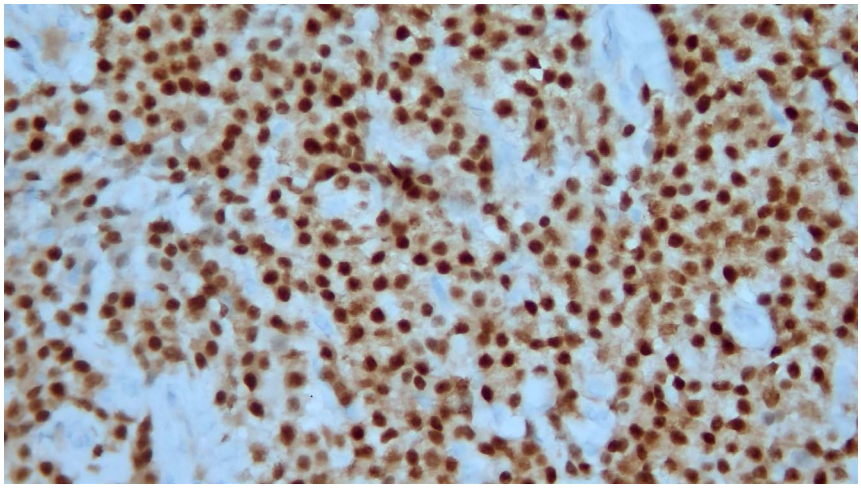

Figure 4: Breast biopsy section showing MUM1 immunoreactivity

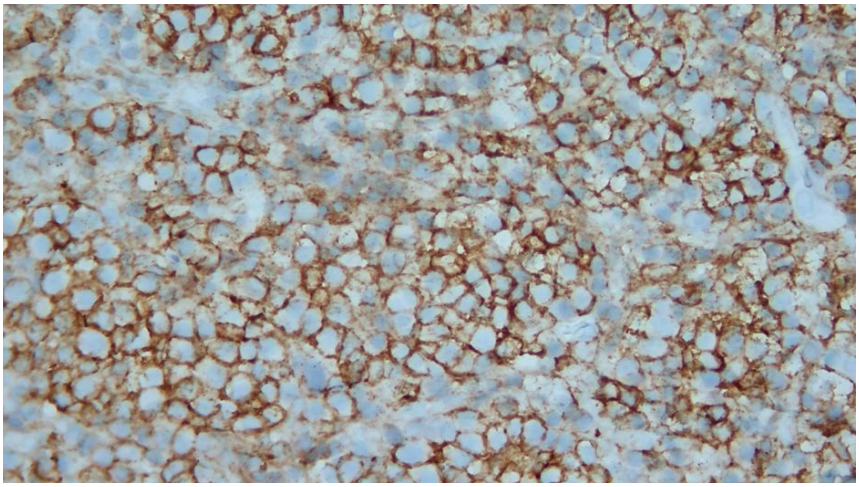

Figure 5: Breast biopsy section showing CD138 immunoreactivity

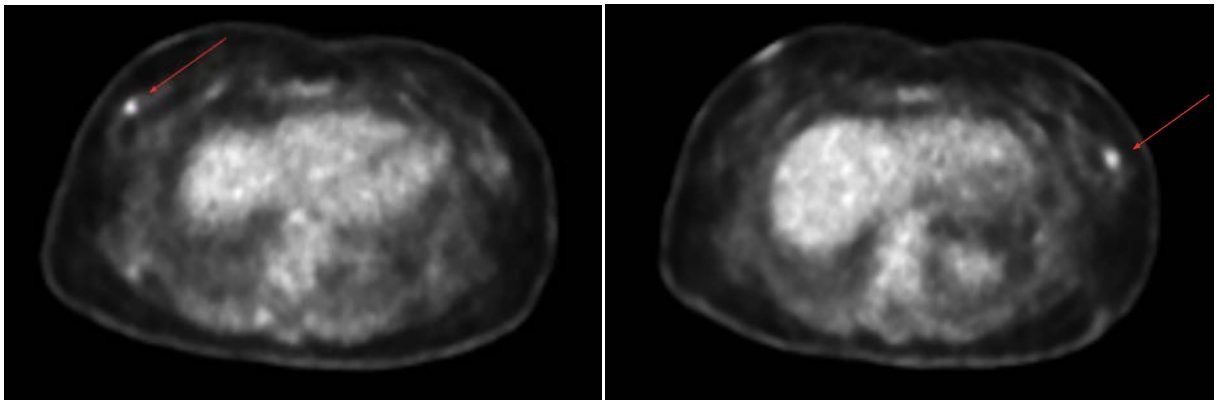

Figure 6: PET/CT images of FDG-avid bilateral breast nodules as indicated by red arrows.

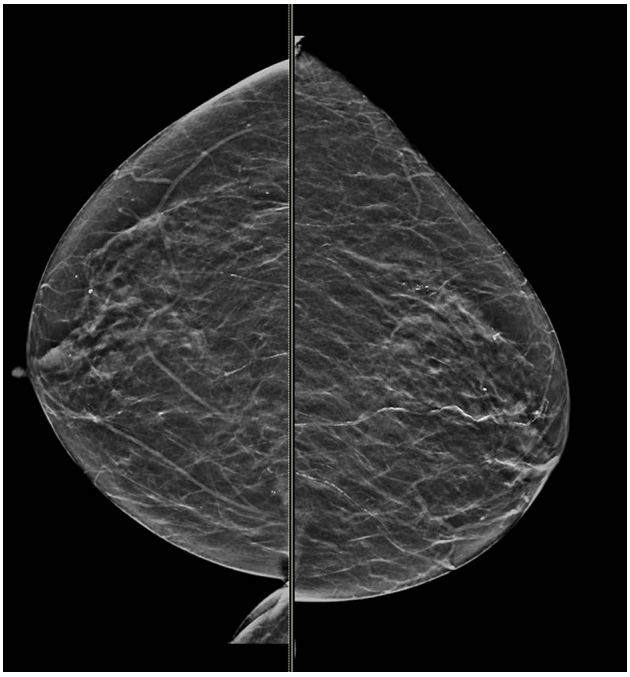

Figure 7: Craniocaudal mammographic views revealing resolution of bilateral breast nodules.

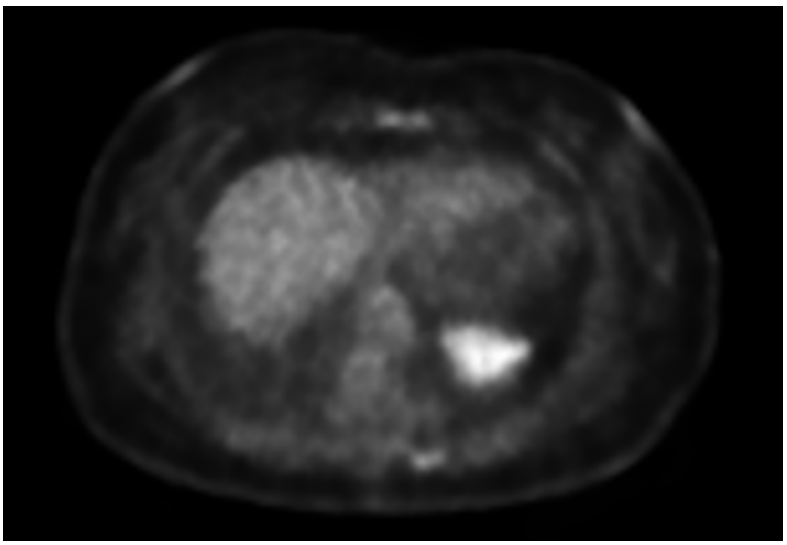

Figure 8: PET/CT images revealing resolution of bilateral FDG-avid breast nodules

KEYWORDS

Plasmacytoma; Breast; Mammography; Multiple Myeloma