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A Case of Plasma Cell Myeloma Presenting with a Huge Ulcerating Breast Mass

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ABSTRACT

Plasma cell myeloma **(**PCM) of the breast is extremely unusual. It probably constitutes 0.7% of extramedullary plasmacytomas**,** and 0.2% of all breast malignancies. Clinically and radiologically, PCM of the breast is indistinguishable from breast carcinoma or other primary breast disorders. Therefore, histomorphological examination with immunohistochemistry are considered as gold standards for its diagnosis in tissue samples. We report a case of a 30-year-old female presented with a progressive huge ulcerating left breast mass, despite chemotherapy. She had a history of an initially misdiagnosed humeral lytic lesion, and recurrent lytic boney lesions for the past 7 years. Positive CD138, CD56 and Kappa immunostaining has confirmed PC differentiation and monoclonality of the tumor cells. Subsequently, PCM of breast was diagnosed and the patient was transferred for chemotherapy and follow-up.

Keywords Plasma cell myeloma, Breast, CD138, CD56

Introduction

Extramedullary plasmacytomas (EMP) are unusual tumors that may present as a solitary lesion or as manifestation of systemic multiple myeloma (MM) in patients who remained free of symptoms for many years after initial diagnosis **(1)**. EMP is most often located in the head and neck region, mainly in the upper aero-digestive tract **(2)**. PCM of the breast is extremely unusual **(3)**, with approximately 83 cases reported worldwide between 1928 and 2019 **(4)**. Therefore, the prevalence of PCM in the breast is unknown, however, among 869 cases of EMP that had been reported in 400 literatures, breast PCM constituted 0.7% of cases **(5)**, and it was found to account for 0.2% of all breast malignancies in another series in which 15% of PCM were primary lesions, while 85% arose in the setting of MM **(6)**. Thus, PCM of the breast can occur in the absence of MM, in active MM, or in relapsing MM **(4)**. The existing case represents a relapsing MM as the patient had a misdiagnosed lytic humeral lesion 7 years before, a sphenoid mass, and recently detected multiple lytic boney lesions.

Case Report

Case referral and history

On the 18th of April 2021, we received transferred microscopic slides and paraffin blocks at the Pathology Laboratory of our institution's Oncology Center for pathological consultation. The sent material was prepared from a left simple mastectomy specimen of a 30-year-old female patient who was operated for a breast mass on the 6th of March 2021 at another private center. Prior to examining the pathology slides, we sought the available patient's previous medical/surgical history and the available data extracted from reports are shown chronologically in Table 1.

Table 1. The patient's previous medical/surgical history extracted chronologically from reports

Date	Finding/approach	Diagnosis/outcome
26/04/2014	Proximal humerus lytic lesion, debridement.	Malignant round cell tumor of bone. Picture in favor of Ewing's sarcoma.
02/07/2017	Magnetic resonance imaging (MRI) on the brain.	Well-defined mass involving the sphenoid and clivus and displacing the carotid artery and the pituitary gland, mostly metastatic.
02/02/2020	Bone survey.	Multiple lytic boney lesions in all scanned bones.
02/02/2021	Developed an upper outer quadrant left breast mass. Tru-cut biopsy was taken for histopathology.	Picture consistent with invasive lobular carcinoma of the breast. Cytokeratin, LCA and CD138, immunohistochemical staining were recommended to exclude a lymphoid or plasmacytoid neoplasm.
04/02/2021	Immunohistochemistry on the breast tru-cut biopsy.	Positive stains: LCA, CD138. Negative stains: Cytokeratin, ER, PR and Her2/neu. Diagnosis: Plasma cell myeloma.
Over the next month	Received Cyclophosphamide, bortezomib and dexamethasone (CyBOR-D); Lenalidomide; and Dexamethasone, cyclophosphamide, etoposide and cisplatin (DECP) chemotherapeutic regimens.	Progressive lesion, not responding to all lines of chemotherapy.
11/03/2021	Left simple mastectomy. Specimen is 27x20x15 cm, covered with an ulcerated skin ellipse. Cut section: offensive, soft to rubbery, nodular, necrotic, hemorrhagic greyish-white mass (18x15x12 cm). Axillary fat, containing 2 lymph nodes (the largest is 5x4x4 cm).	Microscopically: Picture suggestive of large cell non-Hodgkin's lymphoma involving the breast and the 2 sampled axillary lymph nodes, for immunophenotyping.
23/03/2021	Immunophenotyping on the breast mass.	Positive stains: Diffuse strong CD43 and myeloperoxidase, focal CD30, and Ki-67 in 60% of tumor cells. Negative stains: Cytokeratin, CD20 and CD3. Diagnosis: Myeloid sarcoma.

CD; cluster designation, LCA; leukocyte common antigen; ER; estrogen receptor, PR; progesterone receptor; Her2/neu; epidermal growth factor receptor 2

Approach to diagnosis

Examination of the referred hematoxylin and eosin-stained slides revealed diffuse infiltration of the breast tissue by a densely-packed, dyscohesive, round to oval neoplastic cells set in a delicate fibrovascular stroma. The cells had eccentric round or oval nuclei, relatively abundant dense eosinophilic cytoplasm, and distinct cell borders. Cellular pleomorphism, frequent large, bizarre and multinucleated cells, and mitotic figures were noticed. The tumor cells infiltrated the overlying ulcerated epidermis, replaced the full-thickness breast tissue and encroached on the underlying skeletal muscle without its infiltration. Upon evaluation of the IHC satins performed in our laboratory, the tumor cells showed diffuse strong positivity for CD138, CD56, and CD43. Ki-67 labelled 40-50% of tumor cell nuclei **(**Figure 1). CD117, CD34, myeloperoxidase, CD3, CD30, CD20 and cytokeratin all were completely negative. The second ordered IHC panel revealed diffuse and strong positivity for Kappa light chain and negativity for Lambda light chain, confirming the light chain restriction. The diagnosis of "plasma cell myeloma (PCM) of the breast" was confirmed and the final histopathology report was signed out and uploaded to the Medical Oncology Department via our center's electronic database to start appropriate management of the case.

Discussion

As reported in our case, patients with breast PCM usually present with a palpable mass that may be associated with pain, skin manifestations as erythema, peau d'orange or ulceration, or axillary lymphadenopathy in a minority of cases. In most of the previous reports, the mass was solitary (66%) and unilateral, but less likely multiple or bilateral. The reported masses were found to range from 0.7 to 15 cm.in size, and the patients ranged in age from 32 to 85 years **(3,6-10)**. Notable, our patient was slightly younger and had a huger mass than previously reported, and likewise was associated with axillary lymphadenopathy.

Radiologically, PCM of the breast shows non-specific findings mimicking breast cancer or other primary breast disorders, producing diagnostic errors **(3,11)**. They appear as round or oval masses, with high density in mammograms showing either well-defined or poorly-defined boundaries. Ultrasound reveals hypoechogenic lesions with minimal shadowing or

heterogeneous masses, while MRI signal tends to be hypointense on T2 and intermediate on T1, with signs of hypervascularization that can also be detected with color doppler study **(3,8)**. Nonetheless, all of these features seem to be non-pathognomonic. Unfortunately, we were unable to come across the radiologic findings for the presented case.

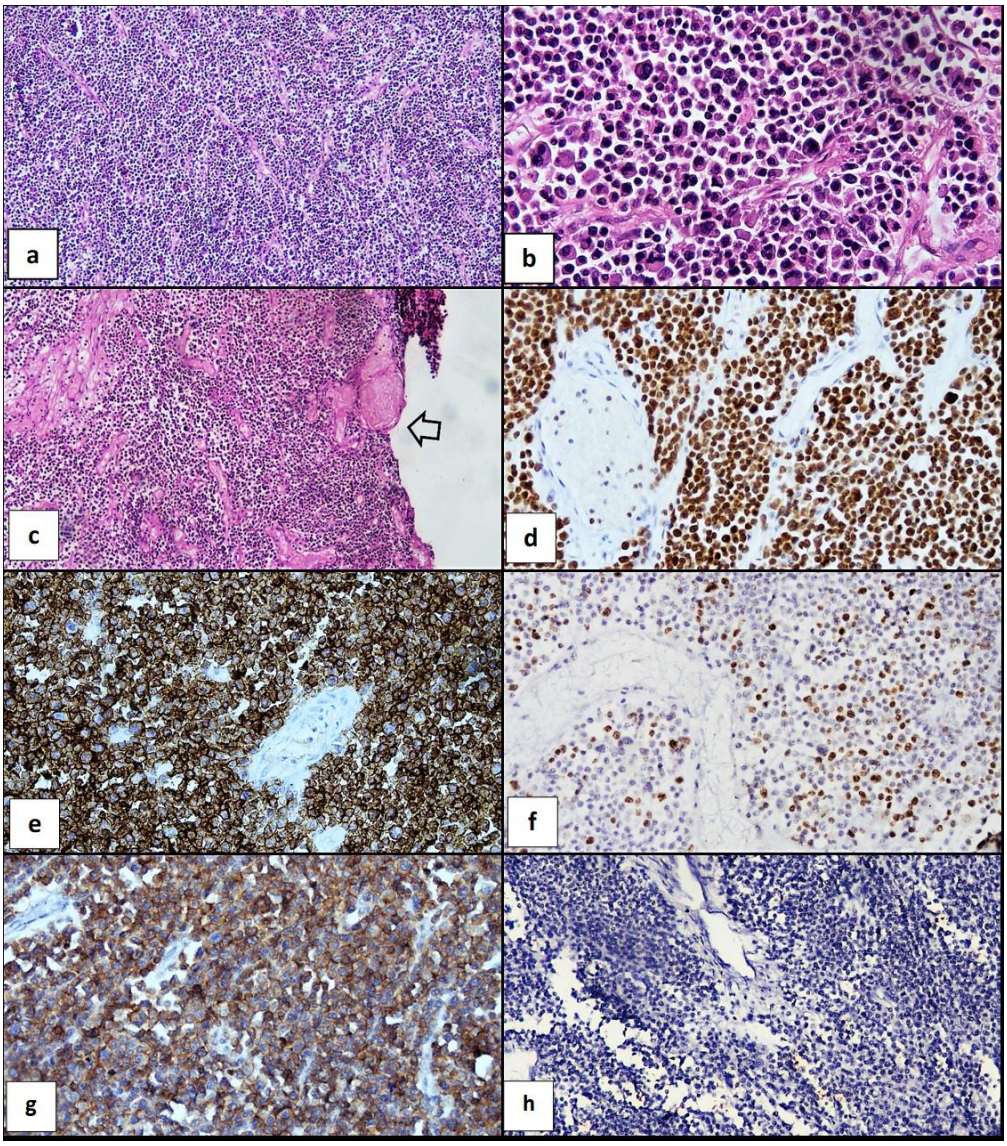

Figure 1. Breast mass with plasma cell myeloma. Appearance of the tumor pattern (a; H&E, x100) and cell characters (a; H&E, x250). Epidermal infiltration, ulceration (arrow) and granulation tissue (c; H&E, x100). Immunohistochemistry shows diffuse positivity for cytoplasmic CD138 (d), membranous CD56 (e), 40-50% nuclear positivity for Ki-67 (f) and Kappa light chain restriction (g) with negative lambda chain (h) (d-h; diaminobenzidine, x250)

Table 2. The differential diagnosis of plasma cell myeloma in the breast

Differential diagnosis	Recommended immunohistochemical panel
Pleomorphic lobular breast carcinoma	CK, ER, PR, Her2/neu, GCDFP-15
Plasma cell mastitis and pseudolymphoma	Polyclonal Kappa and Lambda light chains, CD20, CD3 (with evidence of pleomorphic inflammatory infiltrate)
Malignant melanoma	SOX10, HMB45, S100, MelanA / MART1, MiTF
Granulocytic (myeloid) sarcoma	CD117, CD34, Myeloperoxidase.
Anaplastic large cell lymphoma and diffuse large B-cell lymphoma	LCA, CD20, CD3, CD30, ALK

CD; cluster designation, LCA; leukocyte common antigen; ER; estrogen receptor, PR; progesterone receptor; Her2/neu; epidermal growth factor receptor 2, GCDFP-15; gross cystic disease fibrillary protein 15, SOX10; SRY-related HMG-box 10, HMB45; Human Melanoma Black 45, MiTF; Microphthalmia Transcription Factor, ALK; anaplastic lymphoma kinase

Thus far, the histomorphological examination with immunohistochemistry are considered as gold standards for the diagnosis of PCM in tissue samples. As demonstrated in our case, the histopathologic differential diagnosis of PCM in the breast may include: pleomorphic lobular breast carcinoma, plasma cell mastitis, pseudolymphoma, malignant melanoma, granulocytic (myeloid) sarcoma, anaplastic large cell lymphoma and diffuse large B-cell lymphoma (Table 2). Likewise, a case of PCM of the breast mimicking an inflammatory carcinoma or a breast abscess has been reported **(7)**. In our experience, the diffuse strong immunohistochemical expression of at least 2 of the plasma cell differentiation markers such as CD138 and CD56 plus the determination of monoclonality are mandatory to confirm diagnosis in conjunction with the microscopic features of the tumor cells which entails plasma cells at different grades of maturation. In most of the recently reported breast PCM cases, the confirmed diagnosis was based on diffuse cytoplasmic CD138 reactivity and lambda chain restriction of the neoplastic plasma cells **(3,4,8,11)**. Virtually, CD138 should be diffusely expressed in all cases, while CD56 is expressed in a range from 43-71.7% of neoplastic plasma cells, likewise, LCA and CD20 are expressed in small percentages, while CD30 should be completely negative in all cases **(12)**. Regarding CD43, its expressed in extramedullary myeloid tumors as well as in a wide range of B-cell hematologic neoplasms including plasmacytoma **(13)**, thus having no differentiation utility.

Historically, the therapeutic strategies to treat EMP included: radiation therapy alone, surgery alone or combined surgery and radiation if complete tumor resection is doubtful or impossible and/or if lymph nodes are affected **(5)**. Based on risk stratification, the current therapies for MM include induction chemotherapy with triple regimens, autologous stem cell transplant, and maintenance chemotherapy. With the addition of chemotherapy, some authors reported regional recurrences in up to 25% of patients **(6)**, while others reported significant regression of breast lesions following chemotherapy using vincristine, doxorubicin and dexamethasone **(8)**. In striking disparity, the breast lesion presented herein progressed despite using different lines of myeloma-specific chemotherapy.

From prognostic perspective, the specific outcomes for breast PCM are unknown, but generally EMP in the setting of MM has a poor prognosis. A recent comprehensive study demonstrated that the overall survival is better for para-osseous involvement compared to EMP (not reached *vs*. 46.5 months) and that EMP at relapse had the worst prognosis with an overall survival of 11.4 months despite appropriate therapy **(14)**. For this reason, we intend to follow-up this presented case in order to obtain insights into the possible prognosis for such unusual PCM presentation.

Conclusion

Despite its extreme rarity, PCM should be included in the differential diagnosis of a palpable breast mass in female patients with a history of MM. As there are no pathognomonic clinical or radiological findings for PCM in the breast, histopathological examination and immunohistochemistry remain the gold standards for diagnosis. Good correlation between the patient's past medical and surgical history and the current presenting lesions are mandatory to make a correct differential diagnosis of breast masses.

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Conflict of Interest

The author declares no conflict of interest.

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Ethical Approval

Inapplicable according to the rules of local ethical committee due to the retrospective nature of the study and the inability to identify the patient from the presented data and figures. Informed consent was obtained from the patient.

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