Synchronous/Concomittant Existence of Infiltrating Duct Carcinoma and Primary Extramedullary Solitary Plasmacytoma of Same (Left) Breast- Case Report of an Unusual Case

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Abstract: Extramedullary plasmacytomas are rare, especially when not associated with multiple myeloma. The co-occurrence of more than one oncologic illness in a patient can present a diagnostic challenge. Here we report an unusual case of synchronous/concomittant existence of solitary extramedullary plasmacytoma and infiltrating duct carcinoma in the same (left) breast. Synchronous Infiltrating Duct Carcinoma and primary extramedullary plasmacytoma has been previously reported only once. Careful histological examination of each disease is essential to make accurate diagnosis in order to maximize the therapeutic regimen to achieve cure/remission.

Keywords: Extramedullary plasmacytoma, Infiltrating Duct Carcinoma, Multiple Myeloma.

INTRODUCTION
Plasmacytoma is a malignant proliferation of plasma cells. It usually occurs as a component of systemic multiple myeloma, in association with elevation of serum paraprotein. Extramedullary plasmacytoma (EMP), accounting for approximately 3% of all plasma cell neoplasms, results from uncontrolled plasma cell proliferation and consists of monoclonal plasmacytic infiltration without bone marrow involvement [1]. Approximately 80%-90% of EMPs involve mucosa-associated lymphoid tissue of the upper airway and 75% of them involve the nasal and paranasal regions, while breast infiltration is very rare [2].

Extramedullary plasmacytoma is described most frequently in the upper respiratory tract but it may also be found in the oral cavity, gastrointestinal tract, lung, lymph nodes, skin, and subcutaneous tissue. Involvement of the breast is extremely rare [3-5]. It can occur as a primary isolated tumor or as an extramedullary manifestation in multiple myeloma [1, 5–9]. Published data regarding breast plasmacytoma comprise predominantly case reports and do not provide any statistical information.

While infiltrating ductal breast cancer is very common throughout the world, synchronous primary extramedullary plasmacytoma and infiltrating ductal carcinoma have been previously reported only once.

CASE PRESENTATION
A 67 year old female was referred to our hospital with a three months history of lump in upper outer quadrant of left breast. There was no associated breast pain, fatigue, weight loss or systemic symptoms except increase in the size of lump. She did not have any previous history of trauma or surgery to the breast. She had no family history of breast cancer. Her general condition was good. On physical examination, firm, irregular lump of 3 X 2 cm was noted in left breast. The lump was non-tender and there were no associated skin changes, nipple discharge or axillary lymphadenopathy. Laboratory tests including complete blood count, liver and renal function tests were normal. Ultrasound scans of the breast (left) showed a complex cystic lesion 18 X 10 mm on 9'o clock position (Fig.1). Mammography revealed well-defined mass without microcalcification. Fine needle aspiration was done and reported as suspicious for ductal malignancy. Subsequently lumpectomy was done. Grossly, there was single well circumscribed soft to firm mass. C/S showed a well-circumscribed mass with focal hemorrhagic areas, total measuring 2.5x2x1.5cm and an infiltrating firm white tumor mass msg. 2.2x1x1cm (Fig.2). Histopathological examination of the circumscribed mass revealed proliferation of the atypical plasma cells with pleomorphic nuclei and prominent nucleoli (Fig.3a). Sections from the irregular mass revealed infiltrating ductal carcinoma of no special type (IDC, NOS) showing neoplastic cells in
nests and cords invading into the desmoplastic stroma (Fig. 3b). Later on, further work-up done for the patient. Her serum immunoglobulins were within normal limits and no Bence-Jones proteins or other M components were detected. Serum calcium and phosphorus were also normal. Bone marrow biopsy was negative for plasma cells and lytic lesions were not seen on CT scan. Immunohistochemistry performed on the sections revealed nuclear positivity for ER, PR & negativity for HER2/neu in the infiltrating ductal carcinoma component. CD 138 positivity, Kappa light chain restriction, CK negativity was noted in plasmacytoma component, while the tumor cells were negative for lambda light chains. Simultaneously, a diagnosis of synchronous infiltrating duct carcinoma and plasmacytoma of left breast was given.

Fig-1: Ultrasonography showing complex cystic lesion

Fig-2: Showing a well circumscribed mass (Red Arrow) & an Infiltrating mass (Blue Arrow)

Fig-3a: No.1 area as seen in fig 2. Proliferating atypical plasma cells with pleomorphic nuclei and prominent nucleoli
Fig. 3b: No. 2 area as seen in fig. 2. Showing ductal arrangement of tumor cells. Proliferating Cells exhibit mild/moderate atypia, desmoplastic stroma.

Fig. 4: A. IDC showing ER positivity; B. Negative for HER2Neu; C. Kappa light chain restriction in the plasmacytoma cells; D. Negative for lambda light chains.

Fig. 5: Showing PR positivity in ductal cells.
Primary soft tissue extramedullary plasmacytoma (SEP) is uncommon and is defined as a malignant tumor of plasma cells arising in the soft tissue in the absence of bone involvement. It can occur in any organ as a solitary form of plasma cell neoplasm but especially in the head and neck region [10]. Although SEP can arise throughout the body, almost 80% to 90% of the cases arise in the head and neck areas [11, 18]. Approximately 70% occur in patients with multiple myeloma.

Occasional cases of extramedullary plasmacytomas are reported in the breast. They usually represent secondary involvement by a systemic process [12, 13] or develop after an extramedullary plasmacytoma has been discovered elsewhere in the body [14, 15].

Multiple primary malignant tumors are rarely observed in clinical practice; however, certain factors like environment and behavior including tobacco, occupation, pollution, and ultraviolet light have been described in the literature as their potential etiology. Genetic predisposition (Li-Fraumeni or Beckwith-Wiedemann syndromes), previous medical treatment (radiotherapy or chemotherapy) and complex interaction between all these factors also play a role [16].

Solitary plasmacytoma of the breast is extremely rare lesion [17]. Extramedullary plasmacytomas are seldom solitary and usually progress to diffuse myelomatosis as a first manifestation of multiple myeloma [18] or recurrence of multiple myeloma [19].

The majority of primary malignant tumors that occur in multiple organs are metachronous, while the presence of synchronous lesions is less common [17]. Only few case reports have been described in the literature with synchronous diagnosis of multiple myeloma and breast cancer. Cao et al. reported a case of synchronous infiltrating ductal carcinoma of the breast and breast plasmacytoma on initial presentation [16]. Khalbuss et al. reported a case of synchronous diagnosis of multiple myeloma and breast cancer with plasmacytid morphology on initial presentation [20]. This case illustrated the cytomorphologic similarities between an epithelial malignancy and multiple myeloma where anaplastic plasma cells can easily mimic the cytomorphology of adenocarcinoma and vice versa. Kherfani et al. reported a case of spinal cord compression from concurrent multiple myeloma and metastatic breast cancer in the same vertebra [21]. Al-Said Ali et al. reported synchronous diagnosis of multiple myeloma and breast cancer on initial presentation similar to our case in 2009 [22]. Although these are the examples of synchronous tumors, the case
of synchronous IDC, NOS with plasmacytoma was reported only once by Cao [16].

Clinically, BP manifests as palpable mass in most cases (82%). Other signs, such as breast pain, skin erythema or lymphadenopathy, are rare. Further the circulating paraproteins vary in type and include kappa and lambda light-chain patterns [19]. In this patient paraproteins were absent in serum and urine, but strongly positive for kappa light chains in the lesion. It is generally accepted that plasmacytomas are radiosensitive and excellent long-term results have been reported following radiotherapy [23, 24].

Our case demonstrated the clinical and histopathologic features 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; we believe that our initial therapeutic measures i.e. wide excision plus combination chemotherapy would have produced a satisfactory result. The significance of this case is that it is the second reported case of synchronous IDC, NOS & primary extramedullary plasmacytoma. The limitation of our report is that we could not follow up this patient in order to state the outcome of our management.

CONCLUSION
Since breast plasmacytoma usually does not have specific radiological or clinical features and can be misdiagnosed or easily missed, careful histological examination is essential to make the correct diagnosis and therapy of every patient. Thus, histopathological examination as well as immunohistochemistry was essential in correct diagnosis of such complicated lesions. And since the management of these coexisting malignancies is different, recognition of both the lesions at the time of presentation & before initiation of treatment is crucial.

REFERENCES

