

# Recurrent Multiple Myeloma Presenting as a Breast Plasmacytoma

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### SUMMARY

We describe a patient with multiple myeloma, who initially responded to chemotherapy and went into remission. She presented 10 months later with a right breast lump which was confirmed by core biopsy to be a plasmacytoma. Further treatment with radiotherapy, thalidomide and later second line chemotherapy appeared unsuccessful and she showed rapid disease progression with rising paraproteins and new extramedullary plasmacytoma lesions in the forehead, supraclavicular region, nasopharynx, liver, spleen, pancreas and paraaortic lymph nodes.

### KEY WORDS:

*Multiple myeloma, Breast plasmacytoma, Extramedullary plasmacytoma*

### INTRODUCTION

Most patients with plasma cell neoplasia have generalized disease at diagnosis, fulfilling the diagnosis of multiple myeloma (MM). However, a minority of patients present with either a single bone lesion (solitary plasmacytoma of bone ;SBP), or less commonly, a soft tissue mass of monoclonal plasma cells (solitary extramedullary plasmacytoma; SEP). Breast involvement is a rare type of SEP and only a few cases have been reported<sup>1,2</sup>. However, in this patient, this was followed by multiple other extramedullary plasmacytomas.

### CASE REPORT

A 55-year-old woman presented with a firm, mobile mass measuring 9 X 8 cm in diameter in the upper outer quadrant of the right breast. She had presented one year earlier with general ill health and weight loss. Trepine biopsy of the iliac crest clinched a diagnosis of multiple myeloma (MM). The skull radiograph showed lytic lesions and the whole body scan showed increased tracer uptake in the fifth lumbar vertebra. The patient received chemotherapy ; vincristine, adriamycin, and dexamethasone; VAD. At the end of the fourth cycle, the serum paraprotein level which was initially 19.4g/L, became unquantifiable indicating disease remission. The patient was started on subcutaneous interferon injections three times per week to prolong the remission.

Seven months later, the patient noted a lump in her right breast which was rapidly increasing in size. Mammography showed a well circumscribed dense nodule which appeared benign. Fine needle aspiration cytology was unsatisfactory. Core biopsy revealed a plasmacytoma (Figure 1). The serum paraprotein level was noted to rise again (5.53g/L). Interferon

therapy was withheld. The patient was given radiotherapy to the right breast (30 Gy / 10 fractions) and to the L5 vertebra (20 Gy/5 fractions). There was significant reduction (>60%) in the size of the breast lump with good spinal pain relief. A month later, she developed two new lumps, one in the forehead and another in the left clavicular region indicating disease progression. She was started on thalidomide (100mg daily) and biphosphonates (Zoledronate 4 mg monthly infusion). She was also found to have hepatosplenomegaly and jaundice. Abdominal ultrasonography and CT scan located multiple hepatic and splenic nodules plus enlarged paraaortic lymph nodes and a pancreatic head swelling. Epistaxis developed a month later. A biopsy confirmed the development of nasopharyngeal plasmacytoma. She was started on a second course of chemotherapy (Cyclophosphamide 200mg daily for 5 days) with prednisolone. Soon after, she developed septicaemia with bronchopneumonia, right cardiac failure secondary to tricuspid incompetence and recurrent anaemia plus acute renal failure which led to her demise.

### DISCUSSION

Metastatic tumours of the breast are rare in comparison with primary breast cancer. They have been reported to constitute 0.4%-2.0% of all breast malignancies<sup>1,2</sup>. Plasmacytoma of the breast, either as a solitary extramedullary tumour (SEP), or as evidence of dissemination of MM, is exceedingly rare. To date, only 17 cases have been reported<sup>2</sup>. SEP are unusual tumours that may represent the initial manifestation of systemic MM or may herald a recurrence of previously treated or quiescent MM.

The majority of breast plasmacytomas are reported in women. The mean age at presentation was 53 years, with tumour sizes ranging from 1 to 7.5cm<sup>1,2</sup>. Distinguishing an extramedullary plasmacytoma of the breast from primary mammary adenocarcinoma is critical to avoid unnecessary surgery and guide therapy. FNAC or trucut biopsy has proven to be an excellent method in diagnosing primary and secondary malignancies<sup>1,2</sup>.

The Guidelines Working Group of the UK Myeloma Forum (UKMF), presented their guidelines on the diagnosis and management of solitary plasmacytoma of the bone and solitary extramedullary plasmacytoma (SEP) in 2003<sup>2</sup>, which remain unrevised in July 2010. They stated that SEP are less common than SBP and have a better prognosis and the majority can be cured with local radiotherapy.

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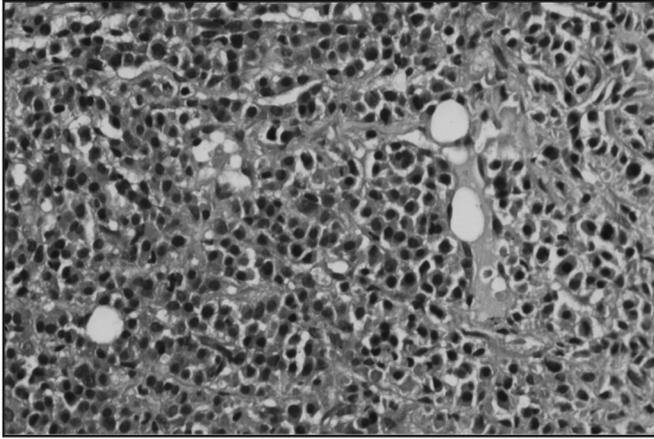


Fig. 1: Plasma cells with eccentric nuclei with clumped 'clockface' chromatin in a breast lump specimen.

However, they recommended that for SEP at sites other than in the head and neck region, complete surgical removal should be considered if feasible (Level III evidence). Patients with involved surgical margins should receive adjuvant radiotherapy (Level IV evidence). Adjuvant chemotherapy should be considered in patients with tumours greater than 5 cm and those with high grade tumours. Chemotherapy is indicated for patients with refractory and / or relapsed disease (Level IV evidence). Thalidomide has been used to treat SEP in the context of relapsed MM with variable results.

Post-radiotherapy, the local recurrence rate is less than 5% and the risk of distant relapse is less than 30%. Progressive disease may present as MM, SBP or soft tissue involvement of lymph nodes, skin or subcutaneous tissue. Distant metastasis tends to occur 2-3 years of initial diagnosis. At least two thirds of patients survive for more than 10 years<sup>2</sup>.

Plasmacytoma should be considered in a patient with history of multiple myeloma presenting with a breast lump. SEP when truly localized has a high cure rate with local treatment. The primary treatment for most patients with SEP is radiotherapy, but surgery may also be required. If underlying multiple myeloma is demonstrated, stem cell transplantation is indicated.

The role of interferon (IFN) in the treatment of multiple myeloma is based on the antitumour activity mediated by antiproliferative and immunomodulatory effects. It has been shown to prolong the plateau phase of the disease between four to 12 months but in general, only has a modest effect on the overall survival<sup>3</sup>. A close liaison between haematologist, radiotherapist and surgeon is crucial for planning optimal care for patients presenting with this condition.

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