



Case Report

Extramedullary Plasmacytoma of Soft Tissues and Gingiva

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Abstract: Extramedullary plasmacytoma (EMP) is a rare plasma cell neoplasm of soft tissue without bone marrow involvement or other systemic characteristics of multiple myeloma. It accounts for 3% of all plasma cell tumors. Multiple extramedullary plasmacytoma is defined when there is more than one extramedullary tumor of clonal plasma cells and such presentation has not been described earlier. We report such rare case of multiple extramedullary plasmacytoma involving multiple soft tissues in chest, abdomen, mandible, maxilla, and gingiva.

Key Words: Extramedullary plasmacytoma; Gingiva; Myeloma

Introduction:

Extramedullary Plasmacytoma is defined as neoplastic proliferation of plasma cells in soft tissues, as a sole lesion without bone marrow plasmacytosis or related organ or tissue impairment (ROTI).¹ It accounts for upto 1-2% of human malignancies, 3% of myeloma cases and is usually localized in submucosal lymphoid tissue of nasopharyngeal and paranasal sinuses.^{2,3} Multiple plasmacytomas involve more than one localized area of bone destruction or extramedullary tumor of clonal plasma cells without bone marrow metastasis or ROTI.¹ Multiple Soft tissue localization elsewhere is extremely uncommon. We present a case of Multiple extramedullary plasmacytoma presenting as multiple subcutaneous soft tissue nodules of chest, abdomen, gingiva, mandible and maxilla, confirmed by pathology report and followed up until 1 year later.

Case Report

A 65 year old woman presented with a swelling in the posterior triangle of the neck near the thoracic outlet of 4 months duration. CT scan revealed a mass below the 1st Rib. A core biopsy was performed and the diagnosis of Plasmacytoma was made on histopathology. The bone marrow aspiration was negative for

myeloma cells and the skeletal X-ray did not show any lytic lesion.

Two months later, she again presented with a swelling in the lower left anterior gingival region below the upper maxillary canine. An incisional biopsy was performed on the lesion. Grossly, the lesion was soft and gelatinous. Microscopy showed oral mucosa lined by nonkeratinizing stratified squamous epithelium. The underlying connective tissue showed closely packed plasma cells with varying degree of differentiation, some with perinuclear halo, occasionally two nuclei within one cell. Dutcher Bodies were appreciated. A diagnosis of plasmacytoma was given. A bone Marrow aspiration was performed which was negative for neoplastic proliferation of plasma cells. Serum Electrophoresis showed normal levels of IgG and IgA but a mild increase in IgM-78 and B2-microglobulin-2.5mg/dl. Lab Tests did not show any signs of anemia, hypocalcaemia and renal failure. The patient refused to take any treatment. One year later, the patient developed multiple subcutaneous soft tissue swelling on the surface of chest and abdomen. The face was grossly distorted because of swellings on mandible and maxilla. A FNAC was performed on the multiple nodules, which showed plasmacytoid cells arranged in sheets and scattered singly. The nuclei were round to oval with a dispersed nuclear chromatin pattern and a clear halo. Few of them showed prominent nucleoli. Bone marrow was repeated again which showed sheets of plasma cells (mature and immature) involving more than 20% of the bone marrow cells. She finally died due to complications 3 months later.

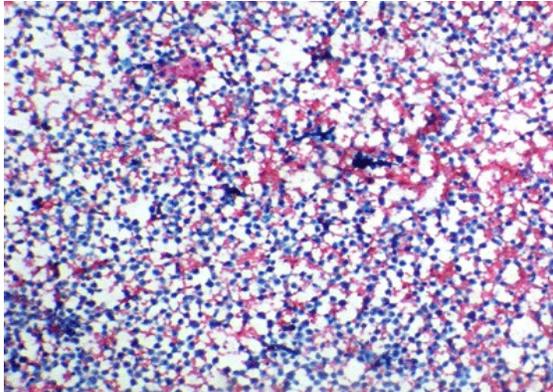


Fig 1 : Low Power 10X : FNAC shows sheets of plasma cells

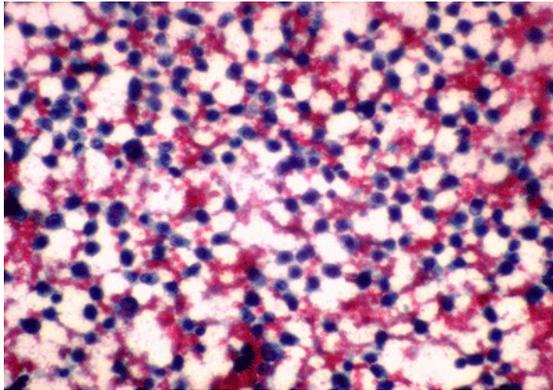


Fig 2: High Power 40X : Plasma cells with eccentrically shaped nuclei

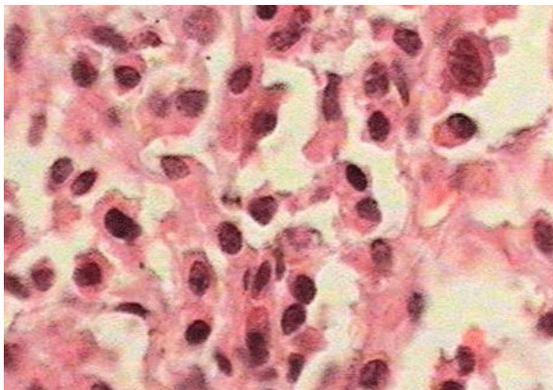


Fig 3: Biopsy Showed plasma cells with varying degree of differentiation.

Discussion

Plasma cell neoplasm can be classified into the following types: multiple myeloma (bone marrow and other systemic involvements), solitary myeloma (bone plasmacytoma), extramedullary (soft tissue) plasmacytoma, and plasmablastic sarcoma.⁴ Primary plasmacytoma, whether osseous or non osseous, is distinguished from multiple myeloma by absence of hypercalcemia, renal insufficiency and anemia, normal skeletal survey, absence of bone marrow plasmacytosis, and serum or urinary paraprotein < 2g/dl.⁵

Primary extramedullary plasmacytoma can be solitary or multiple.⁶ The International Myeloma Working Group in 2003 has recognized a separate classification of plasmacytomas that occur as multiple sites of disease in soft tissue, bone, or both soft tissue and bone as multiple solitary plasmacytoma.¹ Our patient

had multiple subcutaneous soft tissue involvement in chest, abdomen and also on mandible, maxilla and gingiva which fits into the classification of primary multiple EMP.

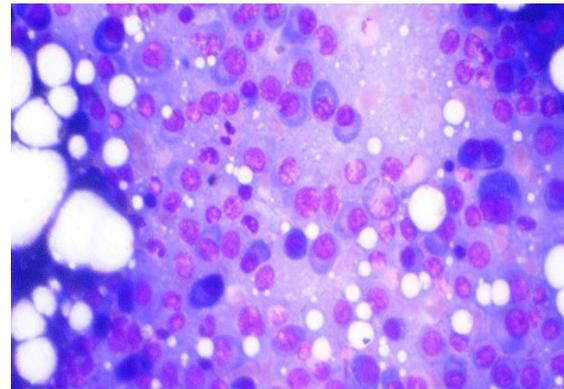


Fig 4: Bone marrow: The patient finally progressed to multiple myeloma.

EMP affects males three to four times more often than females, with an average age of 55. However, one third of patients with EMP are under 50 years old.⁶ In a comprehensive literature search reviewing over 700 patients with EMPs, the EMPs developed predominately in head and neck region, especially in the upper respiratory tract such as nose, paranasal sinuses, nasopharynx and tonsils. Infrequent sites of involvement include the gastrointestinal tract, liver, spleen, pancreas, lungs, thyroid, breast, testis or skin.⁷

EMP affecting the gingiva as a soft and gelatinous growth in the left upper canine is a very rare. It was first described by Martinelli in 1968 as a sessile neoplasm from left to right canine, easily to be confused with chronic gingivitis.⁸ Moshreff et al reported a polypoidal growth in left upper canine clinically similar to peripheral giant cell granuloma.⁹ EMP presenting as multiple subcutaneous soft tissue masses on the chest and abdomen and also on mandible and maxilla is very rare event and sequential involvement of these subcutaneous tissues has been not described earlier.

EMP should be differentiated microscopically from other types of plasma cell tumors, such as reactive plasmacytoma and plasma cell granuloma, or plasmablastic lymphoma. The presence of Dutcher bodies and monoclonal expression of plasma cells on histopathology easily distinguishes plasmacytoma from plasma cell granuloma and reactive plasmacytosis. The cells in plasmablastic lymphoma are more single cell type without maturation and not associated with monoclonal gammopathy as in plasmacytoma.

Like other plasma cell tumors, EMPs are highly radiosensitive with 80 to 100% of patients successfully achieving local control and with a 50 to 65% ten-year disease-free survival rate. The risk of distant metastasis or conversion to multiple myeloma, is <30%.¹⁰ Distant metastases tend to appear within 2-3 years of the initial diagnosis and our patient was finally diagnosed with multiple myeloma and died because of the complications later.

Conclusion

We present a rare case of primary multiple extramedullary plasmacytoma involving subcutaneous tissues of chest, abdomen, mandible, maxilla and also involving the gingiva. The patient finally progressed to multiple myeloma.

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