CASE REPORT

Plasmacytoma of the Mandible

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Abstract

We describe a patient with an extramedullary plasmacytoma (EMP) of the mandible, which presented a diagnostic and therapeutic challenge on several levels. We discuss herein the clinical presentation surgery and the role of radiotherapy in this rare case.

Key Words

Plasmacytoma, Mandible

Introduction

Extramedullary plasmacytomas (EMPs) are rare tumours composed almost exclusively of plasma cells that arise outside the bone (1). Eighty percent of solitary EMPS occur in the head and neck region, although they represent less than 1% of all head and neck malignancies (2). These lesions may occur primarily without evidence of disease in other sites, or as part of systemic involvement during the course of multiple myeloma (MM). Although dissemination of these tumors is rare, they may be locally aggressive and demonstrate marked involvement and/or destruction of bones (3).

Case Report

In June 2000, a 57 year old female patient reported to the Department of Oral Health Sciences, Postgraduate Institute of Medical Education and Research, Chandigarh, with a complaint of swelling on left side of the face since 2 months, associated with a mild pain localized to the left side of the face only. On examination, the swelling was approximately 5 x 6 cm in size and extended from the mandibular left body to the ramus region. It was hard, tender and fixed while the overlying skin and mucosa were normal. Radiograph (Orthopantogram) revealed a destructive lesion, resulting in fracture of the mandible (Fig. 1).

A Fine Needle Aspiration Cytology was advised which showed features suggestive of plasmacytoma. An incisional biopsy was performed under local anaesthesia. Histopathology examination showed that the tumour was composed of sheets of plasma cells, many of which were binucleate. The cells had an eccentrically placed nucleus with 'cart-wheel' appearance of the nuclear chromatin and moderate amount of eosinophilic cytoplasm. Many immature plasma cells and abnormal mitosis were also noticed (Fig. 2,3). To rule out multiple myeloma, the patient was advised radiological skeletal survey, estimation and electrophoresis of the plasma proteins, urine analysis for Bence -Jones proteins and bone marrow examination. All of them proved to be normal or negative.

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A diagnosis of EMP of the Mandible was made. Management consisted of Radiation therapy in the form of 40 Gray in 20 fractions given over a period of 4 weeks. Patient was on regular follow up with 3 months interval. The patient recovered uneventfully, and one and half year later is well and free of disease. Radiograph shows healing of the mandibular fracture (Fig. 4).

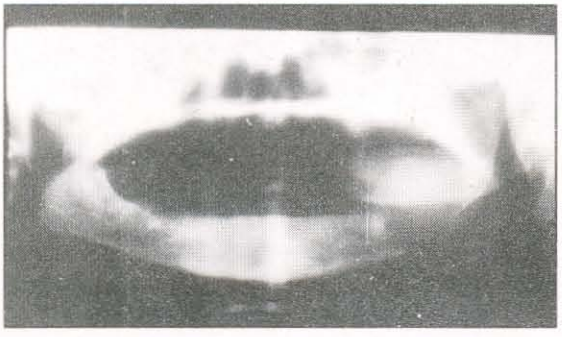

Fig 1. Orthopantogram showing lesion in left side of mandible before radiotherapy.

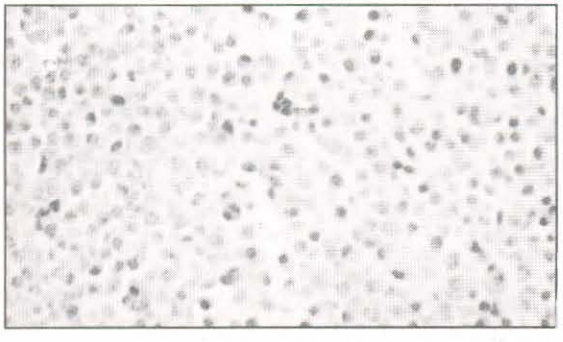

Fig 2. Photograph of tissue specimen showing features of Plasmacytoma under low power.

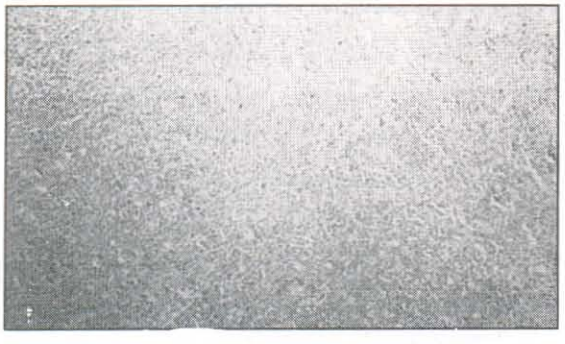

Fig 3. Photograph of tissue specimen showing features of Plasmacytoma under high power.

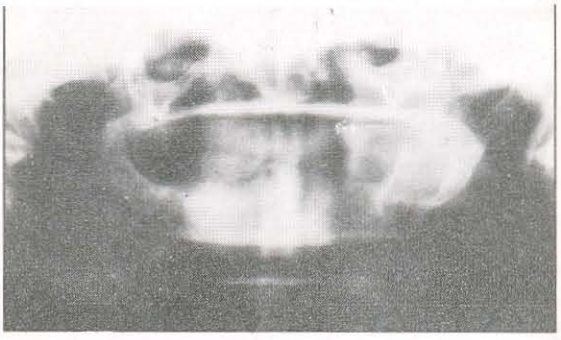

Fig 4. Orthopantogram showing healed lesion with sclerosis after radiotherapy.

Discussion

Plasmacytomas are a rare group of diseases with an overall incidence of 3 per 100,000 people per year. EMPs are rare tumors, accounting for less than 10% of all plasma cell malignancies. About 80% to 90% of EMPs occur in the head and neck region (4). EMPs are localized submucosally and predominate in the head and neck. Only 15% to 20% of cases progress to MM or develop regional disease. Local recurrence may occur in 6% to 10% of cases that have adequate initial treatment (3). A thorough search of the literature revealed only few previous case of a mandibular origin of this lesion (5) The presenting signs and symptoms are non specific and simply reflect the mass effect of the lesion. CT and MRI imaging define the soft tissue mass or associated bony destruction but cannot distinguish the Plasmacytoma from other tumours. Diagnosis depends on adequate tissue sample, usually by way of incisional or excisional biopsy depending on the location.

Histologically, Plasmacytomas are characterized by a diffuse or sheetlike proliferation of plasma cells with varying maturity and atypia. The nuclei are round to oval and eccentrically located, with a dispersed (clock face) nuclear chromatin pattern and a clear or halo area. Immunohistochemical staining demonstrates the monoclonal nature of the plasma cells and confirms the

neoplastic nature of the lesion. Because of its monoclonal nature, the neoplasm produces only one type of Ig light chain, either Kappa or Lambda.

Investigation of patients with suspected Plasma cell tumours should include a full work up to rule out multiple myeloma which includes urine analysis for Bence-Jones proteins,serum electrophoresis, bone marrow examination and a skeletal survey. Only when other signs of systemic disease have been ruled out can the diagnosis of a solitary plasmacytoma be made.

Several treatment options have been advocated including radiation therapy, chemotherapy, surgical excision and combined modalities. Plasmacytomas are highly radiosensitive, with regional control of over 80%. There is almost universal agreement that the treatment of choice for head and neck plasmacytomas is radiotherapy with surgery being limited to obtaining a biopsy or management of recurrence. The recommended tumour dose is 40-50 Gray/4-5 weeks/20-25 fractions with control rate of 80% (6). The radiation field should include the primary tumour and regional lymph nodes, when there is a clinical suspicion of their involvement. Prognosis for EMP is good with 70% survival beyond 10 years. Long-term follow up should be carried out to detect recurrence or manifestation of disseminated disease.

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