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Craniofacial Localization of Multiple Myeloma – A Case Report

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Abstract: ***Multiple myeloma is a malignant haematological disease characterized by proliferation of plasma cells in the bone marrow. Plsmacytoma may present clinically in different non-specific symptoms including bone pain, pathological fractures, fatigue, anaemia, renal failure etc. Oral or craniofacial manifestations may be the first sign of multiple myeloma. The diagnosis is established by blood, urine, imaging examinations and biopsy. This article presents a case of 63 years old patient with craniofacial localization of multiple myeloma***.

Keywords: multiple myeloma, plasma cells, plasmacytoma

1. Introduction

Multiple myeloma is a plasma cell tumour with multifocal infiltrations of the bone marrow and multiple osteolytic lesions of the bones with spongy structure - vertebrae, skull, pelvis, ribs, sternum etc.

This tumour is rarely found as a solitary plasmacytoma of bone or as an extramedullary plasmacytoma.

The first described case of plasma cell tumour was made by S. Solley in 1844. The term multiple myeloma was given by von Rustizky in 1873.

Multiple myeloma is a disease with unknown ethiology, characterized by proliferation of plasma cells in the bone marrow. It accounts about 1% of all types of cancers and more than 10% of hematologic malignancies [2, 5].

Plasmacytoma develops mainly in men aged 50–80 years, with a mean age of 60 years. It is more common in males, with a male to female ratio of 3:2 [7, 9].

There are approximately 3,000 new cases of myeloma reported in the UK each year representing 15% of blood cancers and 1% of all types of malignancies [2].

In multiple myeloma there is a diffuse neoplastic proliferation of plasma cells which produce immunoglobulins. This uncontrolled growth of plasma cells has many consequences including bone marrow failure, suppression of normal immunoglobulin production, skeletal destructions, renal insufficiency etc [2, 5].

Clinically the patient presents with bone pain, fatigue, renal failure, recurrent infections, pathological fractures etc. The involvement of mandible is infrequent, but is rarely affected as first bone. More than 30% of patients with multiple myeloma develop lesions in jaws [7]. The mandibular changes are more common in the posterior region of the mandible and oral manifestations may be the only initial symptoms of the disease [6, 10, 14].

Laboratory and haematological investigations show a certain anemia, leucopenia or thrombocytopenia, raised erythrocyte sedimentation rate. Bence-Jones proteins are detectable on urinalysis.

Radiographically most common finding has been punched out radiolucencies without sclerotic borders or periosteal reaction. Skull lesions are present in almost 50% of patients [17]. It has often been described as "moth eaten" appearance. The diagnosis is based upon the evaluation of bone marrow, plasma cell infiltrate, radiological changes, detection of monoclonal (M-) component by serum and urine protein electrophoresis [5, 9, 14].

2. Case Report

A 63year-old man entered the Department of Oral and Maxillofacial Surgery complaining of swelling in the left body of mandible and gingival bleeding which had appeared before four months. He gave medical history of having ache since 2 months in the second left molar which was extracted. He complained of permanent headache and fatigue.

A few days after the extraction patient noted a slow growing swelling. He complained of pain and had difficulty in chewing food. On extra oral examination no deformity of face was evident; there was no change of color of the overlying skin. Intraorally, a single swelling involving left body region of mandible extending from premolar region anteriorly to angle region posteriorly was evident.

On palpation swelling was firm, nontender, nonpulsative, non fluctuant with diffused margins. The buccal cortex was expanded bucco - lingually but more on buccal side. Bleeding on provocation was present.

The panoramic tomography (Fig 1.) showed an osteolytic lesion with defined margins along with cortical bone destruction in the left body of the mandible. It revealed multilobular radiolucency extending from left third molar region to the premolar area of the same side. Superioinferiorly radiolucency extended from just below the upper alveolar border to 1cm above the lower edge of the mandible; distally reaching the jaw angulus. Margins were distinct, well demarcated, no sclerotic border or periosteal reaction were present.There was no resorption of root apex of 38.

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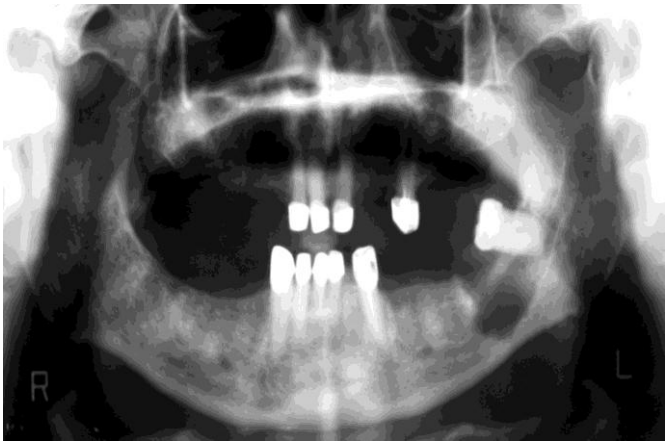

Figure 1: Panoramic tomography

Radiographic features of lateral skull showed the presence of multiple " punched out" and "moth-eaten" appearance radiolucencies of the entire skull with size ranging from 2 to 7 mm and with absence of sclerotic borders (Fig. 2.).

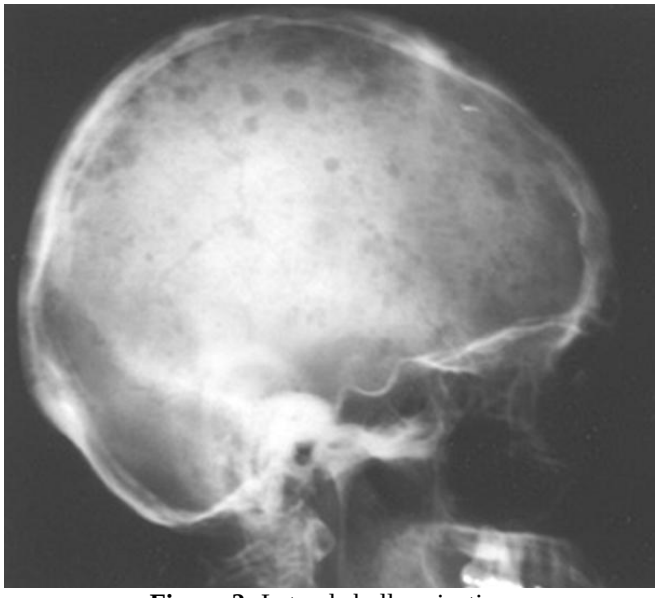

Figure 2: Lateral skull projection

No changes in other bones were found. Whole body bone scintigraphy demonstrated diffusely intense, increased and inhomogeneous tracer concentration only in the skull and left side of the mandible. Blood investigations revealed raised erythrocyte sedimentation rate, raised serum urea and serum creatinine levels. In the haemogram, a decrease in the number of erythrocytes, in the concentration of hemoglobin, in the hematocrit and in average corpuscular volume were found.

A urine test was positive for Bence-Jones proteins and increasement of the phosphate level in the urine was found. Incisional biopsy was performed. Histological analysis revealed plasmacytoid cells, with round, eccentric nuclei with fine granular chromatin and evident nucleolus, characteristics of a solid malignant haematopoetic neoplasm (Fig. 3).

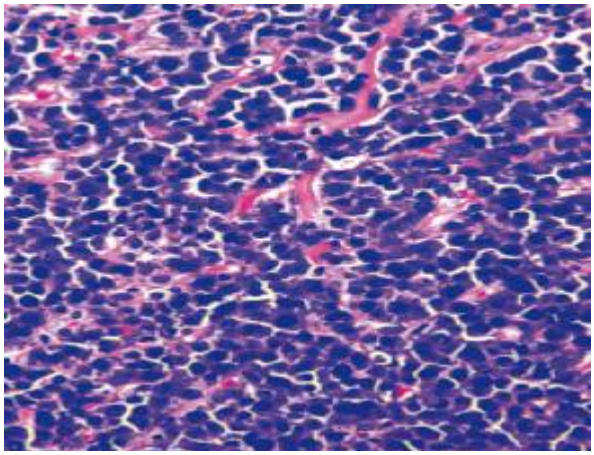

Figure 3: Haematoxylin and eosin stain; magnification 20 x

Based on the clinical, laboratory examinations radiographic findings and hystologycal result the diagnosis of multiple myeloma was established. The patient undergone chemotherapeutic treatment.

3. Discussion

Plasma cell myeloma (plasmacytoma) has three clinical variants: multiple myeloma, solitary plasmacytoma of bone and extramedullary plasmacytoma.

Multiple myeloma occurs in the disseminated form, affecting several bones. It is most common in patients older than 50 years of age with a mean age of 60 years and males are more affected than females. The common systemic manifestations include bone pain, pathologic fracture, renal failure, weight loss, fatigue, weakness, fever, infections etc.

Most common symptom of multiple myeloma is bone pain especially in the lumbal vertebral region. Approximately 70% of multiple myeloma patients complain of pain in bones [9]. Kyle et al. reported in his review of 1027 patients that bone pain, especially back pain (58%), fatigue and anemia (32%) were the most common symptoms [11].

Earlier, bone manifestations were explained with tumor cell infiltration and replacement of bone marrow substance. Recently was found that "osteoclastic activating factor", lymphokine, is responsible for the bone changes [8]. Myelomatous infiltrates frequently involve the vertebrae, calvaria, pelvis, sternum, mandible, clavicle and proximal portions of the humerus and femur - e. g. all the bones with spongy substance – places of haemopoesis.

In a review of 783 patients according to Epstein et al., approximately 14% of the patients had oral manifestations [8]. They include swelling, pain, gingival hemorrhage, odontalgia, paresthesia, tooth migration, mobility and loss, amyloid deposition in the tongue and other oral tissues [7, 13, 15].

The involvement of the oral cavity and jaws in multiple myeloma has been frequently reported in the literature [2, 8, 9, 11, 12, 13, 15, 17]. However, the initial manifestation of plasmacytoma in the mandible is rare; there are few reports in the literature [3, 6, 7, 14, 16].

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In our case the patient suffered from swelling in the oral cavity, headache and fatigue. Nowadays it's known that the plasma cells in multiple myeloma secrete a monoclonal immunoglobulin, a paraprotein. This is most commonly of the Ig G subtype, but production tends to be asynchronous which leads to incomplete immunoglobulin molecules and an excess of light chains of the Kappa or Lambda type. Whole immunoglobulin molecules cannot pass into urine through the glomerular filtration, but the light chains can. When present in the urine they are known as Bence-Jones proteins and are detectable on urinalysis. The plasma concentration of normal immunoglobulins is often reduced; this is known as immune paresis and results in frequent infections [2, 9].

In the case we present the Bence-Jones protein was present in urinalysis. The most common radiographic findings are "punched out" radiolucencies without sclerotic borders. In a retrospective study of 77 patients, Witt et al stated that skull manifestations were present in 46.7% and jaw manifestations in 15.6% of the cases [17].

The typical radiographic appearance is a well-defined "moth eaten" lytic defect, which may be solitary or multiple and often radiologically indistinguishable from bone metastases [7, 16]. In the presented case radiolucencies were found in the left mandibular body and in the skull. The changes in the skull were multiple, punched out, "moth eaten" osteolytic areas with distinct cortical margins. Radiolucency in the mandible was polycyclic without sclerotic borders or periosteal reaction.
Clinical and microscopic features may not be sufficient for distinguishing plasmacytoma from other malignancies arising in the oral cavity, such as poorly differentiated carcinoma, and other types of lymphoproliferative diseases [4, 5].

The differential diagnosis of small, multiple, separate, well-defined radiolucencies include multiple myeloma, multiple metastatic lesions and Langerhans' cell disease. Langerhans' cell disease and multiple metastatic lesions are the most probable diagnoses for multiple punched-out bony lesions in children. In adults, multiple myeloma and metastatic carcinoma could be diagnosis in cases of multiple bone involvement [15].

In the recent years chemotherapy with hematopoietic stem cell transplantation are the preferred treatment for multiple myeloma. Bisphosphonates reduce the risk of myeloma-related fracture; bisphosphonate related osteonecrosis of the jaws occurs in a small percentage of the patients [5].

Multiple diagnostic studies are performed to evaluate multiple myeloma. A complete blood cell count is used to assess anemia, thromhocytopenia, or leukopenia. An erythrocyte sedimentation rate is increased in cases of multiple myeloma. A comprehensive metabolic panel is performed to assess total protein, albumin, globulin, creatinine, and uric acid [4, 5, 15].
Conventional X-rays and computed tomography are used to confirm bone changes and pathological fractures.

In our case osteolytic changes were in the skull and the left part of the mandible. Cone-beam computed tomography is preferable for examinations in the craniofacial localization due to lower radiation [1].

Whole body scintigraphy is used to demonstrate disseminated multiple lesions. In the presented case the tracer concentration was expressed only in the skull and left side of the mandible. An MRT scan of the vertebrae of symptomatic patients is useful to see vertebral fractures and the spinal column [5]. Bone marrow aspirates and biopsy samples are used to diagnose and stage the disease and for prognostic information.

Treatment of multiple myeloma consists of chemotherapy and peripheral blood or bone marrow stem cell transplantation. Erythropoietin may be used to treat anemia resulting from myeloma itself or from the chemotherapy. Biphosphonates may be used to maintain bone strength, and antibiotics may he used to treat infections [5, 13].

4. Conclusion

A multidisciplinary approach toward the diagnosis should always be followed when multiple myeloma is suspected. Haematological examination with differential and total blood counts, biochemical assessment of renal function, calcium status, serum protein electrophoresis, quantification of immunoglobulins, immunoelectrophoresis, bone marrow biopsy and aspiration, urinalysis and a radiographic skeleton survey should be obtained. Knowledge for oral manifestations and craniofacial localization of multiple myeloma for the dentists is very important. This is essential for earlier diagnosis and treatment of these patients with multiple myeloma.

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