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Extramedullary Plasmacytoma of the Gingiva

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Extramedullary plasmacytoma is defined as neoplastic proliferation of plasma cells in soft tissue. It accounts for up to 3% of all plasma cell tumors. Approximately, 90% of extramedullary plasmacytomas are found in the head and neck region commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity. Radiotherapy is the common modality of treatment with or without adjuvant chemotherapy. We report a case of extramedullary plasmacytoma of the gingiva.

Keywords: Extramedullary • gingiva • plasmacytoma

Introduction

Plasmacytoma is a malignant disease.1 It accounts for approximately 1 – 2% of human malignancies and occurs at a rate of about 3.5/100,000 per year.2 – 4 Plasmacytoma may present as one of the three distinct clinical entities: multiple myeloma (MM), solitary plasmacytoma of bone (SPB), and extramedullary plasmacytoma (EMP).5 – 7

EMP is defined as neoplastic proliferation of plasma cells in soft tissue.5, 8, 9 It accounts for up to 3% of all plasma cell tumors. These tumors are four times more common in men, in their sixth to eighth decades of life.10 Approximately, 90% of EMFs are found in the head and neck region commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity.10, 11 The etiology of this disease is unknown, but chronic stimulation, overdose irradiation, viruses, and gene interactions in the reticuloendothelial system have been suggested as etiologic factors.5

Martinella and Rulli, Webb et al, and Boozer et al reported cases of EMFs in the gingiva, tongue base, and soft palate.12 – 14

Case Report

A 54-year-old man came to Dr. Moshref’s Clinic in January of 2003 with the chief complaint of a mass in his mouth. He had noticed the mass 2 months earlier.

On physical examination, we observed a purple nodule in the vestibular area of left maxillary canine, similar to a peripheral giant cell granuloma, measuring about 6×4×3 cm. It had a polypoid surface and had filled the whole vestibular gutter of upper left canine. The tooth was completely mobile.

Radiographic examinations revealed cortical perforation, suggestive of a malignancy. Incisional biopsy was performed for him. The mass was friable and hemorrhagic. Biopsy was done from the surface mucosa and the deep lesion. Histopathologic features showed oral mucosa lined by nonkeratinizing stratified squamous epithelium. Areas of ulceration were seen. The underlying connective tissue was infiltrated by closely packed plasma cells arranged in sheets and islands with varying degrees of differentiation, some with perinuclear halo, occasionally 2 nuclei within a single cell. Russell bodies were seen. Our diagnosis was plasmacytoma (Figure 1).
His bone marrow biopsy was negative for malignancy and laboratory tests did not show anemia, hypercalcemia, or renal failure. Immunohistochemical study was carried out. Tumor cells were negative for LCA, CD3, CD20, CD45RO, CK, and Desmin. But they were rarely positive for CD138, CD38, and CD79. Ki-67 was positive in all tumoral cells. The immunohistochemical study was also consistent with plasmacytoma.

The patient received field radiotherapy to his gingiva (40 GY in 4 weeks) and was asymptomatic for more than 11 months.

Discussion

Multiple myeloma is a systemic disease and is characterized by neoplastic proliferation of monoclonal immunoglobulins (M protein). It is a disseminated disease involving many bones. The incidence of the MM is below 1% of all malignancies and 10% of all hematologic neoplasms. Solitary plasmacytoma is seen in 5 – 10% of all plasmacytoma cases. It can be classified into SPB and EMP. Plasmacytoma of bone is twice as common as extramedullary type.

Clinical manifestations of plasmacytoma of the oral cavity consist of jaw pain, tooth pain, paresthesia, swelling, tissue mass, mobility of teeth, migration of teeth, hemorrhage, and pathologic fracture of the involved bone.

According to Miller, EMP of the oral cavity can show a cauliflower growth. Solitary plasmacytoma differs from MM by lack of plasma cell infiltration in a random bone marrow biopsy. The affected patients may show no signs of anemia, hypercalcemia, or renal failure either. Majumdar et al reported that 15 – 38% of EMPs can express amyloid and IgG is the commonest immunoglobulin, which may be expressed by tumor cells.

On radiographs, EMP can erode bone making it difficult to be distinguished from SPB. Histopathologic features of EMP show a connective tissue greatly infiltrated by plasma cells which are like focal sheets, small islands, or plasmacytoid nodules.

The treatment of choice for EMP is radiotherapy because the disease is highly radiosensitive. Some reports have described long-term control following radiotherapy of the solitary lesion with a high likelihood of cure. Mendenhall et al have reported 94% local control rate in solitary plasmacytoma with doses exceeding 40 GY in four weeks while some other researchers have shown less favorable results. Adjuvant chemotherapy is sometimes indicated in an attempt to delay the conversion of the disease to myeloma. Surgical excision of the tumor is not a common practice.

The conversion rate of EMP to MM is 15 – 20%. The conversion rate of EMP to SPB is 48% and is associated with a poorer prognosis. Harwood et al reported the increased rate of conversion to MM if the EMP involved the adjacent bone. The conversion rate of EMP to MM is 15 – 20%. The conversion rate of EMP to SPB is 48% and is associated with a poorer prognosis. Harwood et al reported the increased rate of conversion to MM if the EMP involved the adjacent bone. Local recurrence has been reported to be up to 10%. Dissemination of the tumor takes place in 35 – 50% of EMPs.

The differential diagnoses of EMP are plasma cell granuloma, pseudolymphoma, and reactive plasmacytic hyperplasia. It is generally accepted that lesions consisting of monoclonal plasma cells are neoplastic, whereas lesions with multiclonal plasma cells are inflammatory.

Majumdar et al in 2002 reported two cases of EMP of paranasal sinuses and soft palate. Similar to our case, the patient who had soft palate lesion was a man and had no symptoms. But the patient with affected paranasal sinus was a female and complained of gradual loss of vision and protrusion of her right eye. Martinelli and Rulli in 1968 reported a case of EMP of the gingiva. Their patient had pain and gingivitis for about one year. Our case was found accidentally and had no symptoms except a mass for about 2 months.
References

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