

## CASE REPORT

# Extramedullary Plasmacytoma of the Mandible: Report of Two Rare Cases

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## ABSTRACT

Extramedullary plasmacytomas (EMP) are uncommon plasma cell tumors that arise from soft tissue and particularly, the submucosa of the upper respiratory tract. It affects males more than females and often presents in the 6th and 7th decade. Plasmacytomas in the oropharyngeal region have been reported in the gingival, tongue, buccal mucosa, and lips. We present two rare case reports of EMP occurring in the mandible.

**Keywords:** Mandible, Multiple myeloma, Plasmacytoma.

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## INTRODUCTION

Plasma cell neoplasms are clonal diseases of terminally differentiated B cells (monoclonal immunoglobulin secreting plasmocytes) that exist on a spectrum from the asymptomatic monoclonal gammopathy of undetermined significance to full-blown plasma cell neoplasms, or multiple myeloma (MM).<sup>1</sup> Plasmacytomas are rare tumors and are classified into MM, solitary myeloma of bone, plasma cell leukemia, and extramedullary plasmacytoma (EMP).<sup>2</sup>

Extramedullary plasmacytomas are uncommon tumors, with a worldwide annual incidence of 3 per 100,000 populations. Solitary EMPs account for less than 3 to 10% of all plasma cell neoplasms.<sup>3</sup> Extramedullary

plasmacytoma arises from soft tissue and particularly the submucosa of the upper respiratory tract (79%).<sup>4</sup> It affects males more than females and often presents in the 6th and 7th decade.<sup>5,6</sup>

Extramedullary plasmacytomas can occur either as an uncommon manifestation of MM, in approximately 4 to 7% of patients,<sup>7</sup> or as an even less common primary lesion. The latter has no systemic manifestation and a high propensity for the involvement of the upper respiratory tract, but can also occur in other sites and systems, including the gastrointestinal tract.<sup>8</sup> The increased incidence of EMPs in the head and neck is due to the predominance of lymph nodes and the reticuloendothelial system in the head and neck region.<sup>9</sup> Plasmacytomas in the oropharyngeal region have been reported in the gingival, tongue, buccal mucosa, and lips. Here, we present two rare case reports of EMP occurring in the mandible.

## CASE REPORTS

### Case 1

A 69-year-old female reported to the Department of Oral Medicine, Diagnosis and Radiology, Indore Modern Dental College and Research Center with a painful swelling on the right side of the lower jaw since 2 months. Initially, the swelling was the size of a lemon but increased suddenly in a short period of 15 days. The pain was severe, continuous, and lancinating type, and persisted throughout the day. Medical history was negative for bone pain, renal failure, or any other systemic manifestation. On extraoral examination, the swelling extended posteroanteriorly from the angle of the mandible to the retro-commissure of the mouth, and superoinferiorly from 2 cm below the ala-tragus line to the middle 3rd of the neck (Fig. 1). The swelling was 5 × 4 cm in size, showed no sign of inflammation, and on palpation was soft and tender. Mucosa overlying the swelling and surrounding mucosa appeared to be normal. Swelling was soft in consistency, tender, nonindurated, and nonpulsatile, with a tendency to bleed. No signs of anesthesia or paresthesia were present. The swelling had no intraoral counterpart and the regional lymph nodes were not palpable. Lateral oblique and posteroanterior skull radiograph showed no signs of bony destruction or any punched-out lesions

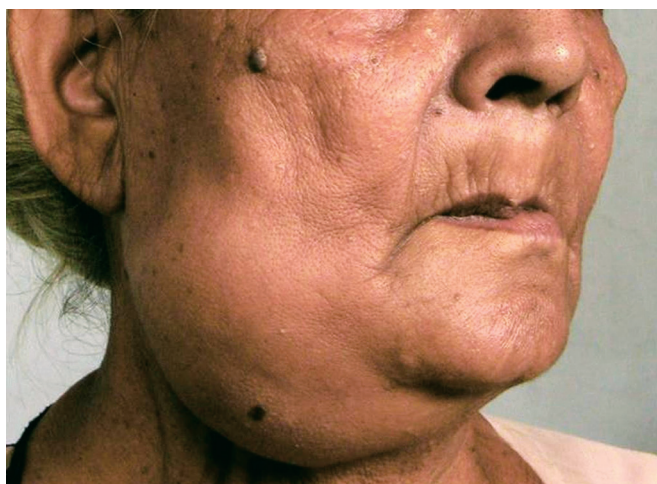
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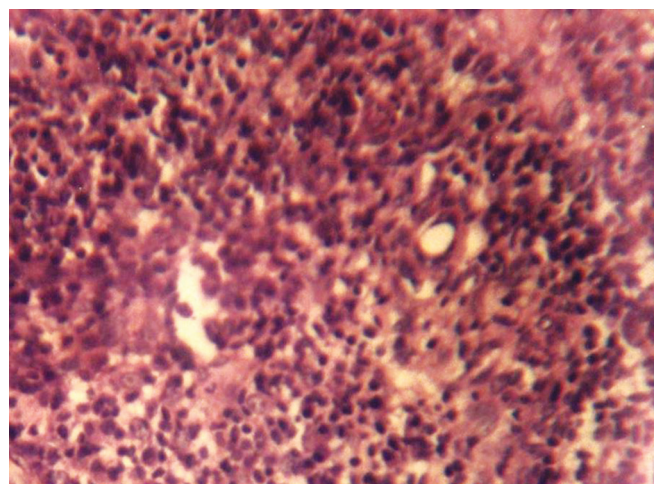
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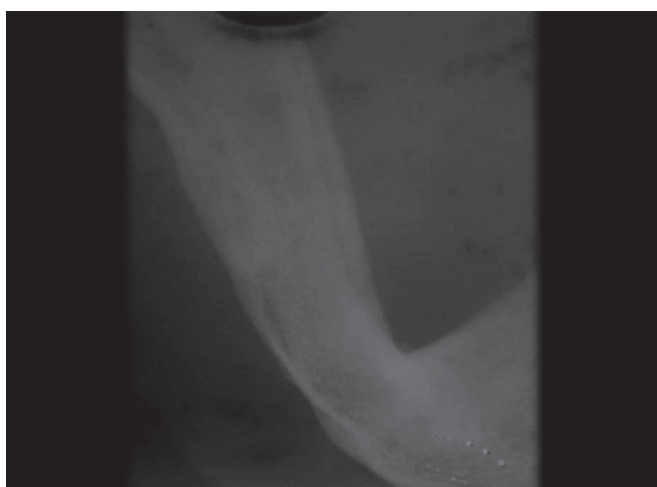
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**Fig. 1:** Extraoral swelling in the right mandibular body and submandibular region



**Fig. 2:** Sheets of plasma cells along with few lymphocytes and polymorphonuclear leukocytes suggestive of plasmacytoma



**Fig. 3:** Lateral oblique view of body of mandible shows no signs of bony destruction



**Fig. 4:** Posteroanterior skull radiograph shows no signs of bony destruction or any punched out lesions

(Figs 2 and 3). Incisional biopsy was performed and histopathology showed sheets of plasma cells along with few lymphocytes and polymorphonuclear leukocytes, characteristic of plasmacytoma (Fig. 4). The patient's full blood count and serum biochemistry measurements were normal. Serum electrophoresis did not show hyperglobulinemia or presence of abnormal M-proteins. The patient was advised surgery followed by radiotherapy.

## Case 2

A 40-year-old female reported to our department, with a swelling in the left posterior region of lower jaw since 4 months. The patient reported rapid, painless growth of the lesion and confirmed that she had been a tobacco chewer for 15 years. Her medical history was not significant. The patient had been medicated with antibiotics for 14 days without regression of the lesion. Intraoral examination revealed a single exophytic, pedunculated, nodular growth on the left mandibular alveolar mucosa,

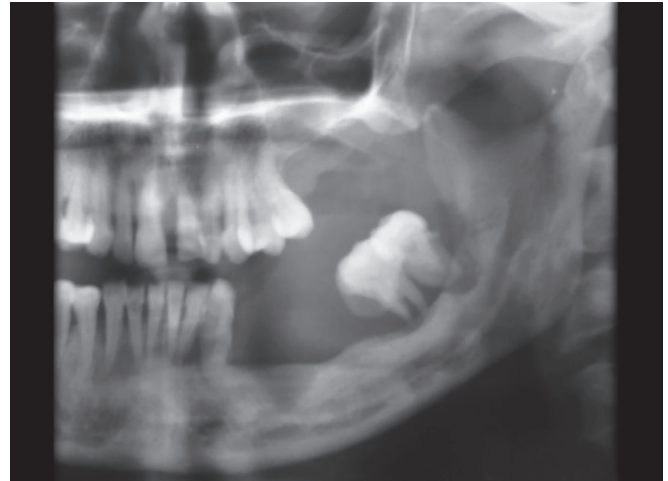
oval in shape, approximately 4×3 cm in size extending anteroposteriorly from the 34th to 38th region, and superoinferiorly from above the crown of 34 to 38 to a depth of mandibular left buccal vestibule (Fig. 5). Mucosa overlying the swelling and surrounding mucosa appeared to be normal. The swelling was soft in consistency, nontender, nonindurated, and nonpulsatile, with a tendency to bleed. No signs of anesthesia or paresthesia were present.

Panoramic radiograph showed severe alveolar bone loss in the mandibular posterior region and moderate alveolar bone loss in the mandibular anterior region and maxilla. Soft tissue shadow was seen in 35th, 36th, and 37th region (Fig. 6). The patient's full blood count and serum biochemistry measurements were normal. Incisional biopsy was performed under local anesthesia and tissue was sent for histopathological evaluation. Multiple bits of hematoxylin and eosin-stained section under microscopic examination showed severely diffuse focal aggregates predominantly of plasma cells and

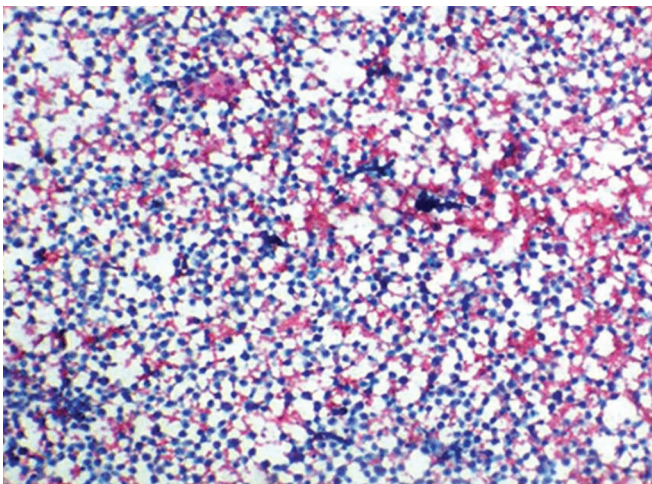




**Fig. 5:** Lesion in the posterior lower alveolar ridge



**Fig. 6:** Cropped panoramic radiograph shows severe alveolar bone loss and soft tissue shadow in 35, 36 and 37 region



**Fig. 7:** Hematoxylin and eosin-stained section shows severely diffuse focal aggregates predominantly of plasma cells and lymphocytes with numerous Russell bodies and mast cells in a loose to dense fibrocellular connective tissue stroma with rich vascularity

lymphocytes, with numerous Russell bodies and mast cells in a loose to dense fibrocellular connective tissue stroma with rich vascularity. Features were suggestive of plasmacytoma (Fig. 7). The MM screening included a full blood count, Bence-Jones protein, serum electrophoresis, skeletal survey, and bone marrow biopsies, all of which were negative. The tumor was excised surgically and was referred for radiotherapy.

## DISCUSSION

Dalrymple and Bence-Jones in 1846 first identified plasma cell dyscrasias when they described a condition with diffuse bone pain and marked proteinuria. However, Waldron and Mitchell in 1873 successfully recognized MM as a distinct histopathologic entity.<sup>10</sup> The EMP is a plasma cell tumor, which arises from soft tissues and located separately from the bone marrow.<sup>11</sup> Schridde in

1905 1st described EMP. Approximately, 90% of EMPs are found in the head and neck region commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity.<sup>12,13</sup> Extramedullary plasmacytoma also occurs in the skin, breast, lung, pleura, stomach, kidneys, small bowel, colon, ovary, uterus, and testes.<sup>14,15</sup> In our case, the tumor occurred on the mandible, which is a rare occurrence. Clinically, the EMP is present as a sessile or pedunculated exophytic neoformation, circumscribed or infiltrative, ranging in color from red-purple to gray or yellow-white.<sup>16-18</sup> Although it affects males more than females and often presents in the 6th and 7th decade,<sup>5,6</sup> in both our cases, it occurred in females, and it occurred in a 69-year-old female in our 1st case and in a 40-year-old female in the 2nd case.

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.<sup>19</sup> In our cases, chronic local irritation seems to be the most likely cause. Extramedullary plasmacytoma can be graded as low (grade 1), intermediate (grade 2), and high grades (grade 3), based on the cellular atypia. Based on the serum, urine electrophoresis, bone scan, bone marrow examination, and radiological assessment, EMP can be staged according to the spread of the disease. Stage I is the disease confined to one site. Stage II includes tumors with local extension of lymph node involvement. Stage III has metastatic spread.<sup>20</sup> The present cases are scored under grade 1 because of minimal evidence of cellular atypia with maximum resemblance to the parent cells and no evidence of local or distant spread.

The affected patients may show no signs of anemia, hypercalcemia, or renal failure.<sup>13</sup> However, 15 to 38% of EMPs express amyloid, and immunoglobulin G is the commonest immunoglobulin, expressed by tumor cells.<sup>12</sup>

Gammopathy (the presence of abnormal paraproteins) makes the diagnosis of primary EMP uncertain, as it is thought that EMPs do not produce abnormal paraproteins.<sup>2</sup> Other work, however, suggests that gammopathy may be consistent with EMP provided the abnormal immunoglobulin disappears after resection of the tumor.<sup>21</sup> Total serum protein level, monoclonal gammopathy, and size of lesion have been found to correlate with the risk of conversion to myeloma.<sup>22</sup> Serum beta-2-microglobulin may also be a useful prognostic indicator.<sup>23</sup> In addition, several immunophenotypic differences between EMP and MM have been reported, which may be useful for diagnostic utility. In particular, there is an absence of cyclin D1 and CD56 staining in EMP.<sup>24</sup>

The conversion rate of EMP to MM is 15 to 20%. The conversion rate of EMP to solitary plasmacytoma of bone (SPB) is 48% and is associated with a poor prognosis.<sup>12</sup> Extramedullary plasmacytoma is considered a low-grade malignancy. The median survival is 8.4 years as opposed to 2 years in MM.<sup>5</sup> Ozsahin et al<sup>25</sup> observed a 10-year rate of progression to MM of 36% for EMP and 72% for SPB. The primary modes of treatment for EMP are surgery and radiotherapy, both of which were received by our patients. Although EMP spreads through the lymphatics and elective regional lymph node irradiation has little influence on lymph node recurrence rates,<sup>26</sup> all patients should be followed up for life with repeated bone marrow aspiration and protein studies to detect the development of MM.

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