Case Report

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Solitary Plasmacytoma Of Mandible: A Rare Case Report

B.V. Harish[1*] Manoj Kumar Kanta[2] T. Surya kiran[3] Sowmya Mantha[4] K. Anant Nag[5] Soundarya Mopidevi[6] Nvvsk Chaitanya[7]

[1]Senior Lecturer, Department of Oral and Maxillofacial Surgery, St Joseph Dental College, Duggirala, Eluru, India.
[2]Reader, Department of Oral and Maxillofacial Surgery, GSL Dental College and Hospital, Rajahmundry, India.
[3]Senior Lecturer, Department of Oral and Maxillofacial Surgery, GSL Dental College and Hospital, Rajahmundry, India.
[4]BDS, i dentist, Eluru, India.
[5]BDS, i dentist, Eluru, India.
[6]BDS, i dentist, Eluru, India.
[6]Krishna Chaitanya Dental Care, Kakinada, Andhra Pradesh, India.

ABSTRACT

Background: Plasmacytoma found as a mixed radiopaque radiolucent lesion around an impacted molar is uncommon, however it is essential to rule out for its counterpart i.e., multiple myeloma. Solitary bone plasmacytoma, typified by clonal proliferation of plasma cells, belongs to a group of lymphoproliferative clinical disorders. It most typically involves a single bone or extramedullary site. It is a dyscrasia of plasma cells that involves the jaw bones very rarely, and which is commonly seen in the retromolar area, angle, and ramus of the mandible. They must be differentiated from multiple myeloma as prognosis between both vary considerably. It accounts for 3% of all plasma cell neoplasms, with approximately 50% of the cases transforming into MM over 3-4 years. This report describes a case of a mixed radiopaque-radiolucent lesion in the mandible, later diagnosed as plasmacytoma in an old-aged male patient, treated surgically, and systemic workup was done to rule out multiple myeloma. Despite the rarity of plasmacytoma in the jaws, there is an incidence of 0.15% of cases of multiple myeloma, the first manifestation of the disease appears in the jawbones and oral cavity.

Keywords: Solitary plasmacytoma, plasma cells, multiple myeloma.

INTRODUCTION

Uncontrolled proliferation of plasma cells is the characteristic of plasma cell neoplasms. Solitary bone plasmacytomas (SBPs) and extramedullary plasmacytomas (EMPs) are the localized forms of the disease. In contrast, multiple myeloma (MM) is a systemic clonal proliferation of plasma cells based in the bone marrow. (1) SBP usually manifests as a spinal disease most frequently affects the thoracic vertebrae. (2) Other common bones that may be involved are the sternum, clavicle, rib, and humerus, but that of the mandible is a rarity. (3)

In the maxillofacial area, plasmacytoma may be classified into different clinical forms: a manifestation of the disseminated form (MM), a solitary lesion in the soft tissues, or a solitary bone lesion with or without invasion of local structures. (3) SBP is observed as centrally localized in bones. The prognosis of SBP is poorer than EMP, and approximately 50% of them will transform to MM (4) been observed to occur within three years in as much as two-thirds of cases. Patients may usually present with pain, tooth mobility, severe bleeding, or pathological bone fractures. With its non-specific clinical and radiological features, biopsy becomes essential in order to establish the diagnosis. (5)

Although the histopathological features of solitary plasmacytoma are characteristic, few clinical conditions of the oral cavity as poorly differentiated carcinoma and lymphoproliferative disorders may also exhibit similar microscopic features to those of in plasmacytoma. Hence, the diagnosis of solitary bone plasmacytoma (SBP) requires a solitary bone

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*Correspondence Dr. B.V. Harish.
iDentist Multispecialty Dental Clinic and Implant Centre, RR Pet, Eluru, Andhra Pradesh, India.
Email: Not Disclosed

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lesion, with both confirmatory histopathological and Immuno-histo Chemistry (IHC) analysis, with the definite support of the hematological investigation. (6) Investigations must be undertaken to determine the absence or presence of multiple myeloma. (5)

Regardless of its course, it has a significant effect on the patients' quality of life, and mortality rates are still significant. (7) In this article, we report clinical, radiological, histological, as well as immunohistochemical features in a 60-year- old male patient with solitary plasmacytoma of the mandible.

CASE REPORT

A 60-year-old patient reported to our outpatient department in December 2019 with a chief complaint of pain and red-colored fluid discharge from the lower right back tooth region of jaw since five days. Pain was gradual, moderate, lancinating and continuous, radiating towards right ear with no history of paresthesia. Patient gives medical history of diabetes since five years and is under medication. Non-hypertensive.

On clinical examination, the maximal mouth opening was 4 cm, without deviation or clicking on the temporomandibular joint. There was no evidence of submandibular, submental, or cervical lymphadenopathy. There was a small painful mucosal opening in the right lower back tooth region. (figure 1) Aspiration of the lesion was negative, excluding the possibility of vascular and cystic lesions.

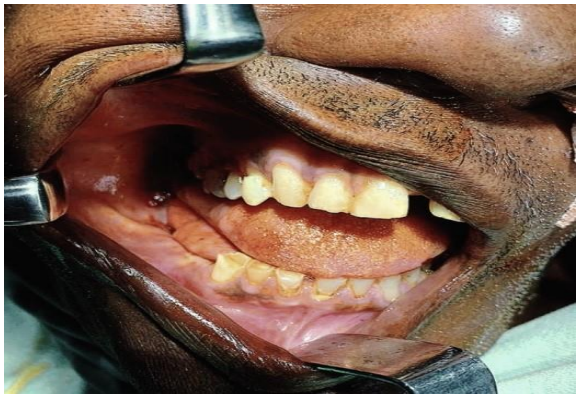

Fig 1: Mucosal opening in the right lower back tooth region.

Panoramic radiograph revealed mixed radiopaque-radiolucent lesion surrounding an impacted molar tooth extending from lower right central incisor up to the right sigmoid notch. (figure 2) The extent of the lesion was confirmed through cone- beam computed tomography (CBCT).

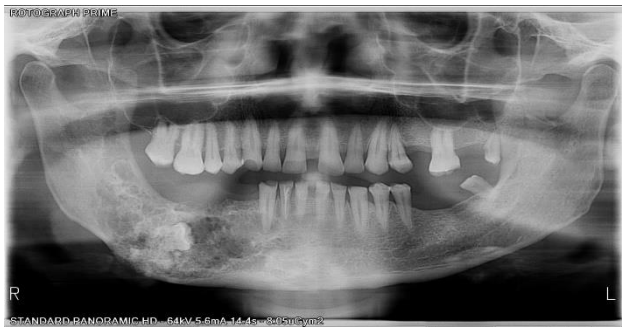

Fig 2: Panoramic radiograph

The preoperative complete blood picture was normal. An incisional biopsy of the bony lesion was performed by general dentist under local anesthesia and was sent to general pathologist, Histopathology features were suggestive of "benign odontogenic tumor" most possibly calcifying epithelial odontogenic tumor (pindborg tumor).

TREATMENT:

Resection was planned under general anesthesia. The procedure along with complications, were explained to the patient, and informed surgical consent was taken. Under general anesthesia, lip split incision with chevrons modification was given and then continued up to submandibular and post ramal regions combined with intraoral vestibular incision up to the coronoid notch, layer by layer dissection was carefully done, the facial artery was ligated in submandibular region, the marginal mandibular nerve was identified and preserved. The lesion was exposed. (fig: 3)

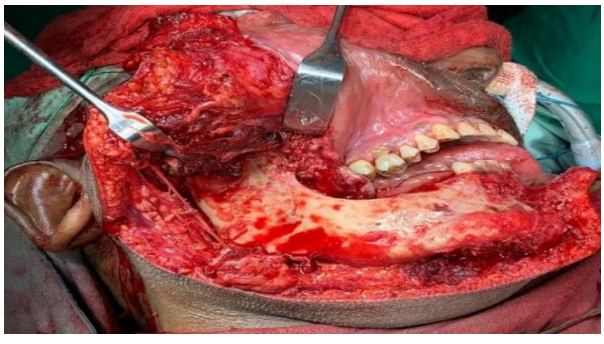

Fig 3: Surgical exposure of lesion

The bony cut was made with a safety margin of 1cm away from the lesion up to the region of left mandibular central incisor (31), and bony cut was extended from sigmoid notch to posterior border of ramus sparing the unaffected condylar stump. (figure 4,5)

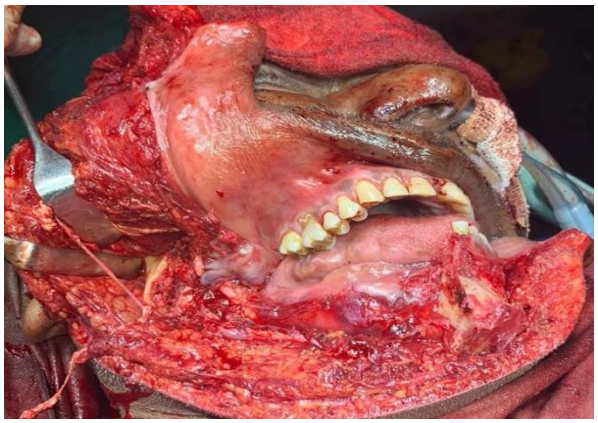

Fig 4: Resection

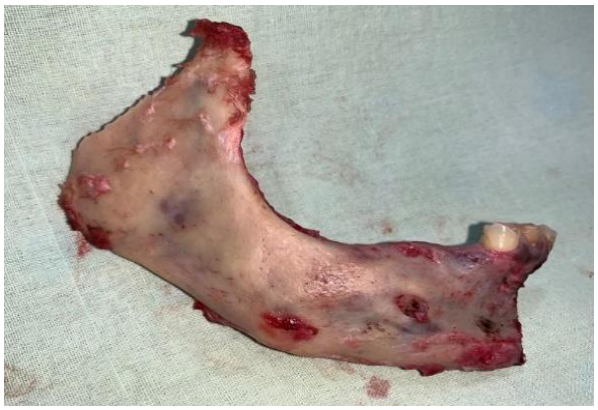

Fig 5: Resected specimen

Initial reconstruction was done with a 20-hole 2.5 mm long reconstruction plate. The reconstruction plate was adapted to the remaining mandible of left side and to the remaining condylar stump on right side and fixed with screws and also additional reinforcement by suturing surrounding musculature to holes of reconstruction plate. (figure 6) A drain was placed in position followed by layer-by-layer closure, and the specimen was sent for histopathological analysis.

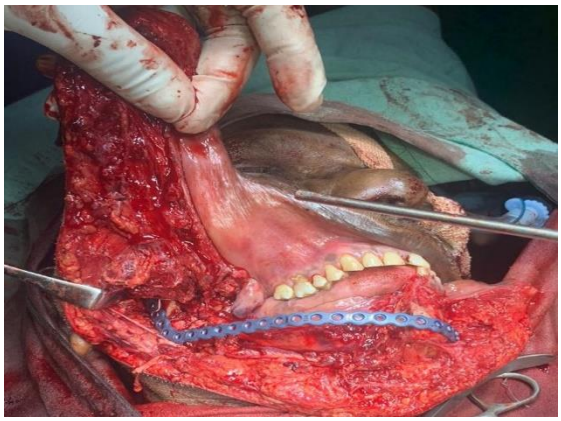

Fig 6: Reconstruction with recon plate

On the second postoperative day, there was a sudden fall of Hb% {7%} that was taken care with blood transfusion. On the fourth postoperative day, the patient complained of urinary retention, shortness of breath with swelling of legs there were increased levels of fasting, and postprandial blood sugars along with serum creatinine and urea indicating acute renal failure, Immediate referral to nephrologist was carried out to address acute renal failure and dialysis was carried regularly for one week, renal function tests along with sugar levels and Hb% were normal. The nephrologist commented that the altered levels of various parameters are due to surgical stress and post-operative medication. The Patient's overall health improved and he was discharged with the advice of monthly reviews for six months.

To our surprise, the histopathological report revealed the postoperative specimen as a **plasmacytoma.** The sections revealed fibro edematous connective tissue with inflammatory infiltrate, interspersed among the connective tissue are sheets of plasma cells exhibiting nuclear and cellular pleomorphism and increased abnormal mitotic figures. Aggregations of immunoglobulins were seen in the stroma. (figure 7) The specimen was further sent to immune histochemistry studies for confirmation

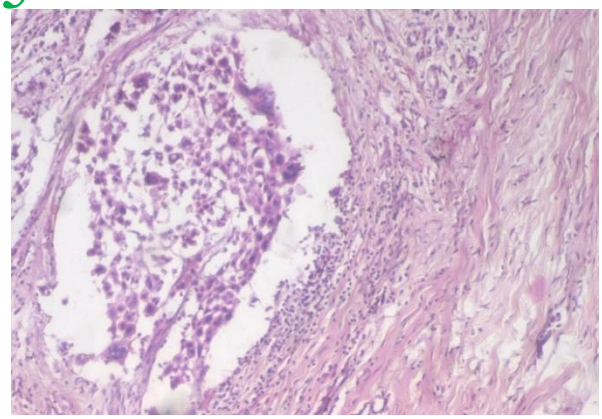

Fig 7: Histopathology picture

Immunohistochemical staining for kappa and lambda antibodies showed positivity for kappa protein in the tumor cell membrane, and tumor secretions were positive for kappa protein and faintly positive for lambda protein. These features suggest plasmacytoma.

To rule out multiple myeloma, we performed systemic workup, a skeletal survey with plain radiographs of chest (figure8), long bones(figure9,10), and skull (figure 11) was done and showed no other osteolytic lesions, urine analysis was negative for Bence Jones proteins . Bone marrow aspiration revealed-features suggestive of plasmacytosis.

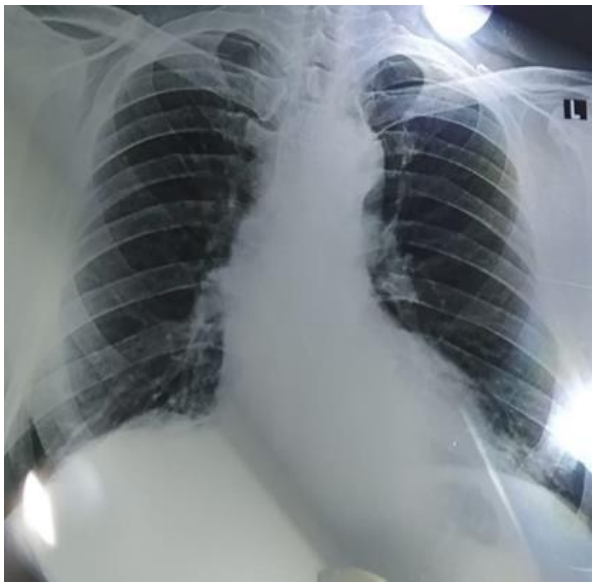

Fig 8: Plain radiographs of chest.

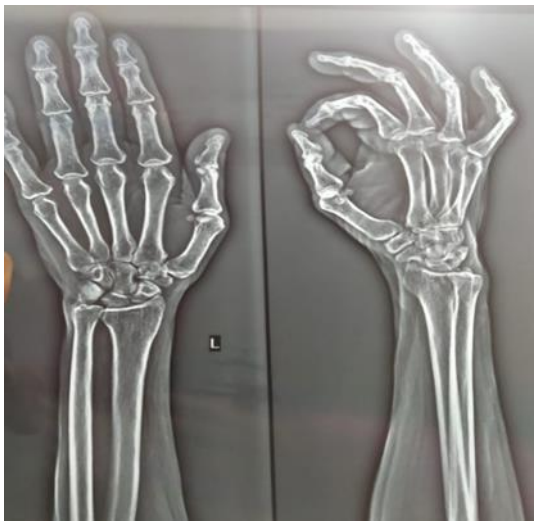

Fig 9: Radiograph of long bones.

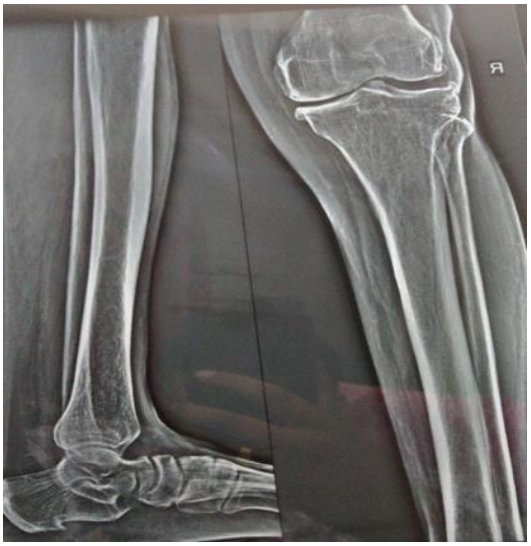

Fig 10: Radiograph of long bones.

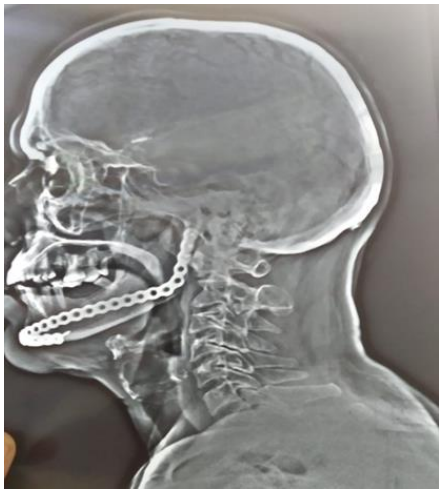

Fig 11: Radiograph of skull.

As there were post-operative anemia, acute renal failure, bone marrow plasmacytosis and anticipating the conversion of solitary plasmacytoma to multiple myeloma as evident from the literature, the patient was referred to an oncologist for necessary treatment. In 2nd and 9th month post-operative review the wound healing was satisfactory and no signs of recurrence were observed. (figure 12)

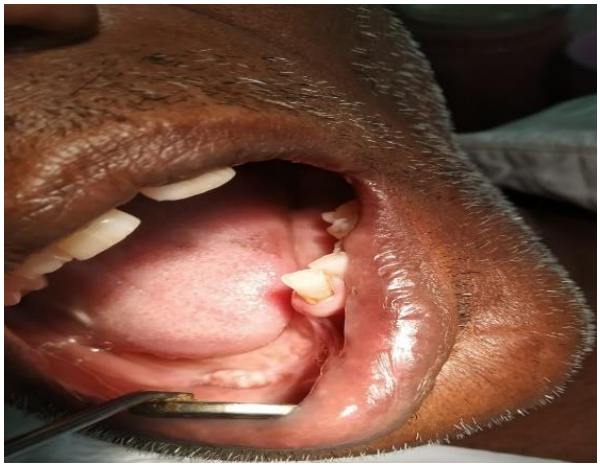

Fig 12: 9th month post-op picture.

DISCUSSION

There are different forms of plasma cell (PC)neoplasms. The general location of multiple myeloma is in the bone marrow and shows a wide range of laboratory, clinical, and radiographic findings (8). On the other hand, solitary plasmacytoma (SP) is characterized by an absence or minimum plasmacytosis of bone marrow and an isolated mass of clonal plasma cells, with symptoms of principal lesion. It is present either as a soft tissue variety called extraosseous or extramedullary type or a single lesion, i.e., solitary bone plasmacytoma. The solitary variety is a rare type with an incidence of 0.15% (8).

Normally Plasma cells generate a spectrum of antibodies with different heavy chains (IgM, IgG, IgA, IgD, and IgE) and light chain (kappa and lambda) characteristics. Except for a very few non-secreting cases, a common early finding in early Plasma Cell Disease is the presence of a monoclonal heavy, and light chain restricted antibody, referred to as a paraprotein or M protein. (9)

SPB occurs in a slightly younger age group than MM, with an average age of diagnosis being 55 years and is more prevalent in males than females with the ratio being. The present case shows SBP in a male of 60yrs which is in harmony with the literature.

The most common clinical symptoms of SPB are pain in the jaws and teeth with paraesthesia/anesthesia, mobility and migration of the teeth, hemorrhage, swelling in hard and soft tissues, and pathological fractures. But plasmacytoma does not show any characteristic sign related to the tumor (10). In our case, the patient presented with a chief complaint of red colored fluid discharge from the lower right back tooth region of the jaw since five days, with no significant medical history. On examination, there was a small painful mucosal opening in the right lower back tooth region.

Based on the multilocular radiolucent lesion of the mandible on orthopantomograph, the differential diagnosis for this lesion included ameloblastoma, odontogenic myxoma, keratocystic odontogenic tumor; central giant cell granuloma; aneurysmal bone cyst and metastatic carcinoma. On plain radiographs, SBP appears like odontogenic tumor with multilocular or unilocular appearance with well-defined borders. (11)

In our case, there was well defined “punched out” lytic lesion extending from parasymphysis to ramus region sparing condyle on the right side of the mandible.

For a diagnosis of SP, the International Myeloma Working Group (IMWG) requires all four of the following criteria be met:

- biopsy-proven lesion,
- normal bone marrow,
- normal skeletal survey and
- absence of end-organ damage.

Table 1: Diagnostic criteria have been enumerated by Bataille and coworkers(12)

Diagnostic criteria for SP of bone (12):
Typically, no M protein in serum/urine (a small M component is present in 50%) solitary area of bone destruction due to clonal plasma cells Bone marrow not consistent with MM Normal skeletal survey

No end-organ damage other than solitary bone lesion, presence of a solitary bone tumor, a biopsy showing plasma cell histology, absence of myeloma cells in bone marrow examinations, absence of anemia, hypercalcemia or renal involvement, absence of or a low monoclonal component on serum electrophoresis, and normal levels of immunoglobulin after treatment.

Even though there is a clear difference between solitary plasmacytoma of bone and extramedullary plasmacytoma, it is important to mark the updated criteria given by International Myeloma Working Group (IMWG) for the diagnosis of multiple myeloma (13)

As per the update given by WHO, there are two subtypes of SPB : one which does not have clonal marrow plasma cells distant from plasmacytoma and thus, shows a normal bone marrow. In 10% of cases this variety may progress to multiple myeloma in 3, conversely there is 60% progression rate in a SPB subtype which shows less than 10% clonal marrow cells (13).

The International Myeloma Working Group (IMWG) did not adopt to the clear distinction made by the WHO update between SPB and EMP. Instead it has blended the two varieties into a single lesion i.e., solitary plasmacytoma (13), which could be described as "not otherwise specified" (NOS) type. This represents a single type lesion, showing variable differentiation of clonal plasma cells and this lesion is of either soft tissue or bone marrow. The bone marrow of iliac crest does not show any clonal plasma cells in these patients. When isolated primary tumor is excluded pelvis and spine show normal bones under CT and magnetic resonance imaging (MRI). Moreover, there is no evidence of end organ damage relating directly to disorder of plasma cells, such as the characteristic symptoms of Multiple myeloma viz., hypercalcemia, renal insufficiency, anemia, bone injury (CRAB). There are chances of multiple myeloma within 3years in 10% of these patients (13).

From the above group of lesions IMWG study has further segregated, solitary plasmacytomas with less than 10% clonal bone marrow plasma cells (BMPC)as a separate entity, which is the only difference between this sub type and the NOS sub type of solitary plasmacytoma (SP-NOS) i.e., presence of <10% clonal bone marrow. But, SPB of this subset has a markedly different course with 60% of cases advancing to PCM in 3 years. In contrast, the EMP of this subset will progress to PCM in 3 years, in 20% of cases only (13)

Table 2: Definition and course of plasmacytoma adopted from (13)

Solitary plasmacytoma, Not otherwise specified
1. Solitary disorder of bone or soft tissue, composed of monoclonal plasma cells (by biopsy). 2. Bone marrow biopsy, remote from the plasmacytoma, is normal. 3. MRI or CT show normal skeleton, except for plasmacytoma. 4. No evidence of anemia, hypercalcemia, renal insufficiency or bone lesions, due to plasma cell neoplasm (CRAB).
Solitary plasmacytoma with < 10% of bone marrow commitment.
1. As above mentioned with additional monoclonal plasmacytes < 10% in a sporadic bone marrow biopsy 2. Of the above lesions, 60% of solitary plasmacytoma of bone will develop into multiple myeloma 3. 20% of Extramedullary plasmacytoma will transform into Multiple Myeloma

Recently IMWG has given a new updated definition of SP with the advent of more sensitive techniques to assess BM plasmacytosis (8), as per which SPB can defined as a presence of solitary lytic lesion due to infiltration of monoclonal plasma cells with or without soft-tissue extension, and EMP consists of a soft-tissue mass that is not in contact with bone (8)

In the present case initially, an incisional biopsy was performed which revealed it to be a “benign odontogenic tumor most possibly “pindborg tumor”. Based on the radiographic and histopathological report, aggressive resection of the lesion with healthy bony margins was planned under general anaesthesia. The post-operative resected specimen was sent for histopathology, which revealed fibro edematous connective tissue with inflammatory infiltrate. interspread among the connective were sheets of plasma cells exhibiting nuclear and cellular pleomorphism and increased abnormal

mitotic figures, aggregations of immunoglobulins were also seen in the stroma. Immunohistochemical staining for kappa and lambda antibodies showed positive for kappa protein in tumor cell membrane and tumor secretions were positive for kappa proteins with faintly positive for lambda proteins. These features suggested it to be a plasmacytoma.

To rule out other plasma cell disorders. Radiographs for long bone, cranium and thorax to identify lytic bony lesions if any were advised. Urine examination for monoclonal antibodies was performed and a bone marrow biopsy was advised. Urine examination for Bence jones protein which are a hall mark for plasma cell disorders was negative. Radiograph of long bones, cranium and thorax showed no evidence of lytic lesions.

Bone marrow aspiration showed 8% of marrow nucleated cells suggesting it as a marrow plasmacytosis with megakaryocytic dysplasia, which hints it to be a second subtype as suggested by WHO where in 60% of the cases turn into PCM in 3years.

Modalities of treatment are radiation, chemotherapy, surgery, while combined radiation and surgical approach gave good outcomes with a low rate of local recurrence. The course of solitary bone plasmacytoma is relatively benign, 80% of patients do not show recurrence after five years while 20% may progress to multiple myeloma (14).

In the present case depending upon the histopathological diagnosis resection was planned with a healthy margin of 1mm.The post-operative excised specimen revealed it as SPB. Presently the patient is under follow up and advised for radiotherapy.

CONCLUSION

It is uncommon to suspect a mixed radiopaque-radiolucent lesion surrounding an impacted molar tooth of the mandible as a plasmacytoma. However, if it gets diagnosed on histological and IHC studies, it should be taken into account to rule out its disseminated form, i.e., multiple myeloma. Therefore, clinicians must have a high index of suspicion and meticulously evaluate patients for optimal treatment. The continued surveillance of the patient is essential even after the three-year disease-free period, for infrequently, multiple myeloma can still develop.

CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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