SOLITARY PLASMACYTOMA OF MAXILLA IN A HBSAG POSITIVE PATIENT: A CASE REPORT

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ABSTRACT: Plasma cell neoplasm's constitute a group of disorders characterized by monoclonal proliferation of plasma cells and presence of monoclonal immunoglobulin in the serum. We report a case of solitary bone plasmacytoma of maxilla, a variant of PCNs. Because solitary bone plasmacytoma are quite rare in the oral cavity, they are often not commonly included in the differential diagnosis of lesions occurring in the maxillary palatal region. The definitive diagnosis requires computed tomography scan, histopathological examination and immunoelectrophoresis.

KEYWORDS: Plasmacytoma, solitary, plasma cells, HBsAg, extramedullary.

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INTRODUCTION: Plasmacytoma is defined as any discrete, solitary mass of neoplastic plasma cells either in bone marrow or in various soft tissue sites.[1-3] PCNs occurring in head and neck region are classified by Batsakis into three types: Extra medullary plasmacytoma, solitary plasmacytoma of bone and manifestation of multiple myeloma.[4] They account for approximately 3% of plasma cell tumors, and for less than 1% of head and neck tumors.[5,6] The disease is more common in males, with male: female ratio of 2:1frequently presents during the fifth and sixth decades of life. Solitary bone plasmacytoma has a male: female ratio of 2:1, with a median age of 55 years and primarily affects the axial skeleton especially the vertebrae[7] (Dimopoulos et al, 2000). Osseous lesions constitute approximately 70% of all plasmacytomas. They involve primarily marrow-containing bones, with a predilection for the vertebrae, femur, and pelvis[8] (Bolek et al., 1996). International Myeloma Working Group has set up diagnostic criteria for solitary plasmacytoma of bone, extramedullary plasmacytoma and multiple solitary plasmacytomas (table 1)**.** We report a case of solitary plasmacytoma occurring in maxillary palatal region.

CASE REPORT: A 55 year old previously healthy lady with no comorbidities presented to us with history of swelling and pain in the right hard palate region of maxilla since 3 months. History of loosening and fall of tooth were present. On clinical examination the swelling on right hard palate was firm, tender, fixed 4x6 cm extending from 1st pre molar to soft palate, not crossing mid line, no active bleed on touch, with free overlying mucosa. Right upper gingivobuccal sulcus was normal. There was no proptosis or abnormality of vision. No cervical lymph node palpable clinically. [Figure 1]

On anterior rhinoscopy, no growth was seen in the nasal cavity. Posterior rhinoscopy was normal. Nasal endoscopy with a 0 degrees rigid endoscope showed no abnormality.

The paranasal sinuses radiograph showing opacity of the right maxillary sinus. Paranasal sinus CT scan showed heterogenous mass involving right half of hard palate with destruction of postero lateral wall of right maxilla. [Figure 2].

Incisional biopsy was done and Histopathology of mass showed features of plasmocytoma with amyloid deposition, cells with nucleus seen place eccentrically and with prominent nucleus [Figure3]. Immuno staining showed neoplastic cells strongly positive for CD 138 and cells positive for leucocyte common antigen and creatinine kinase. [Figure 4]

Serum electrophoresis showed sharp monoclonal paraprotein measuring 1.5g/dl in gamma globulin region. Pattern suggestive of monoclonal gammopathy with rise in Ig-G and beta 2 micro globulin seen. [Figure 5]

Blood investigation reports were normal except for positive hepatitis B serology.

Rest of axial skeleton imaging and bone marrow aspiration were normal.

DISCUSSION: Plasma cell neoplasms (PCNs) are rare lymphoid disorder arising due to monoclonal proliferation of β cells that undergoes malignant transformation into plasmacytoid cells. Manifests as generalised or localised form. The generalised form involving multiple bones is called multiple myeloma (MM), with incidence of approximately 95%.[9,10] The localised form may involve a single bone – solitary bone plasmacytoma (SBP) or only soft tissues - extramedullary plasmacytoma (EMP).

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Plasmacytomas account for approximately 3% of plasma cell tumors and less than 1% of head and neck tumors. MM is the most important symptomatic monoclonal gammopathy and represents approximately 1% of all cancers, and 10% of all hematological neoplasm's.[11] It is characterized by numerous abnormal plasma cells permeating the bone marrow and overproduction of monoclonal light-chain or heavy-chain immunoglobulin's that are identifiable in serum or urine.[12] The exact etiology is not known, exposure to certain chemicals, irradiation, viruses and genetic factors have been suggested.[13] The diagnostic criteria for SBP and EMP are mentioned in Table. 1.[14] The diagnosed by histologic confirmation of a malignant proliferation of plasma cells, hematologic and biochemical findings, urine analysis and skeletal radiographic survey.[15] Several case reports with SBP and HIV have been reported but there is no literature of SBP occurring in a hepatitis B positive patient. We could not find any cause or association of SBP in hepatitis patient.

*A small M-component may sometimes be present in blood or urine.

SPB is uncommon tumor of plasma cells, which manifests itself as a single osteolytic lesion without plasmocytosis of bone marrow not affecting other parts of skeleton and constitutes approximately 3% of all plasma cell neoplasms. SPB is more common in males, with male: female ratio is 2:1 with a median age of 55 years. The most common sites of SPB are vertebrae and long bones. SBP rarely involves head and neck region. 30-60% of SBP progress to MM and 10 year survival rate is 16%.[16]

Its presence in maxilla is extremely rare. The presenting symptom may be very vague, non-specific depending on site or pain in the hard palate and teeth, paraesthesia, anesthesia, mobility and migration of the teeth, hemorrhage, swelling in hard and soft tissues and pathological fractures.[17-22] In our case the main complaint was pain and swelling in hard palate.

20-30% of EMP progress to MM. 10 year survival is 70%.[23] While treating these patients one must be careful to differentiate MM from SPB or EMP because treatment and prognosis differ. 5-10% of SPB or EMP may be misdiagnosed as MM in early stage. The prognosis of SPB is poorer than EMP and approximately 50% of SPB will transform to MM.[21,22,24]

The association of plasmacytomas along with hepatitis B virus is a rare entity but association with hepatitis C virus have been established, though not very clear, immunoglobulins produced by plasmacytoma may be in soluble at cold temperature, which causes a cryoglobulinemia, in particular if a chronic C viral hepatitis is associated with the plasmacytoma.[25]

Workup of PCNs includes complete blood count, serum calcium, bone marrow biopsy, whole body skeletal evaluation with x-ray, serum and urine immunoelectro-phoresis and CT or MRI. Fine needle aspiration is not diagnostic; tissue biopsy±IHC confirms the diagnosis. Microscopically plasmacytomas appear as diffuse/ sheet like proliferation of plasma cells with atypia. The cells have cart-wheel nucleus with basophilic cytoplasm, occasionally multinucleated giant cells are also seen. The nuclei are oval to round, eccentrically located with dispersed (clock faced) nuclear chromatin and halo area. The presence of associated inflammation and plasma cell infiltration makes diagnosis difficult. Differential diagnosis of plasmacytomas includes benign reactive plasmacytosis, undifferentiated carcinoma, NHL, malignant melanoma and esthesioneuro-blastoma. Hence immunohistochemistry plays vital role in making diagnosis.

PCN are highly radio sensitive. Radiotherapy is treatment of choice for SPB or EMP.[21] RT given at dose of 4000 - 6000 c GY in daily fractions over 4-6 weeks is treatment of choice and provides excellent loco regional control.[26] Surgery is rarely indicated in cases of RT refractory cases and localised disease confined to parotid, thyroid gland or cervical lymph nodes. Chemotherapy has no role to play.

CONCLUSION: PCN s presenting in head and neck region is rare and may be part of a systemic disease like multiple myeloma. A high index of suspicion and thorough initial histopathological.

Work-up may help in establishing a definitive diagnosis and providing optimum treatment.

MM must be differentiated from EMP and SPB as treatment and prognosis differ. Early diagnosis and effective treatment is essential in the management of solitary bone plasmacytoma because of its rarity and late stage of presentation. The diagnosis is confirmed by single lytic bony lesion in patient with normal skeletal survey and normal bone marrow biopsy. RT is treatment of choice. Long term follow up is necessary to identify recurrence or multiple myeloma development.

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DIAGNOSIS	CRITERIA
Solitary plasmacytoma of bone	No M-protein in serum and/or urine* Single area of bone destruction due to clonal plasma cells Bone marrow not consistent with multiple myeloma (plasma cells <5%) Normal skeletal survey (and MRI of spine and pelvis if done) No related organ or tissue impairment
Extramedullary plasmacytoma	No M-protein in serum and/or urine* Extramedullary tumour of clonal plasma cells Normal bone marrow

	Normal skeletal survey No related organ or tissue impairment
Multiple solitary plasmacytomas (+/- recurrent)	No M-protein in serum and/or urine* More than one localize- area of bone destruction or extramedullary tumour of clonal plasma cells which may be recurrent

Table 1: International Myeloma Working Group diagnostic criteria of solitary plasmacytoma of bone, extramedullary plasmacytoma and multiple solitary plasmacytomas (+/-recurrent)

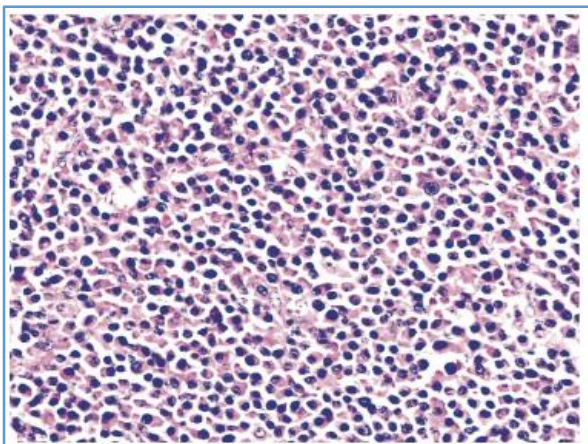

Figure 1

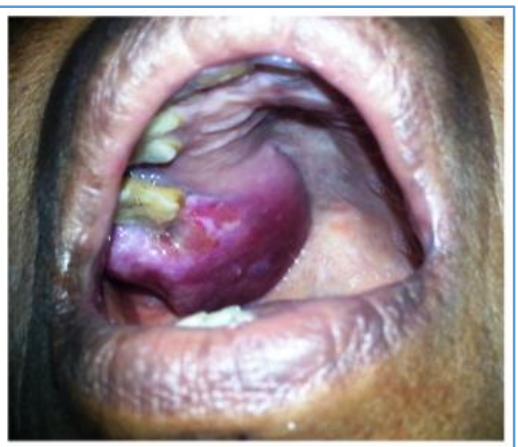

Figure 2

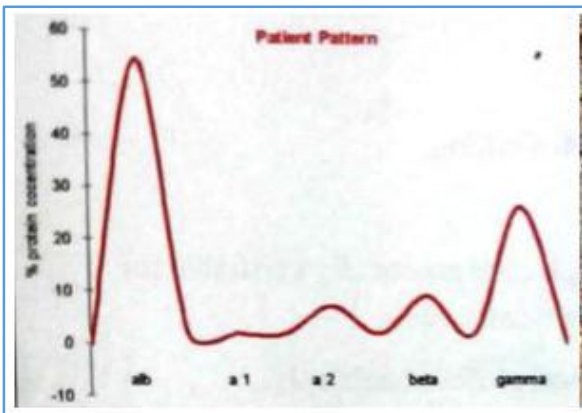

Figure 3

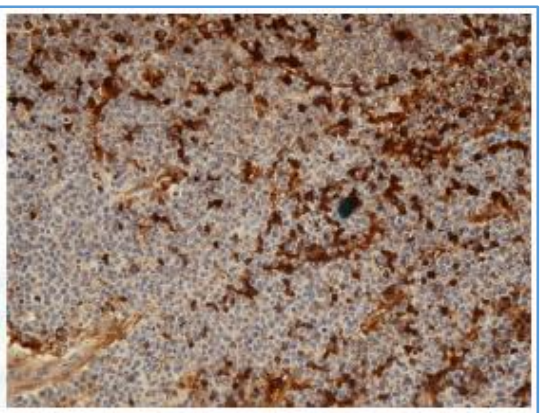

Figure 4

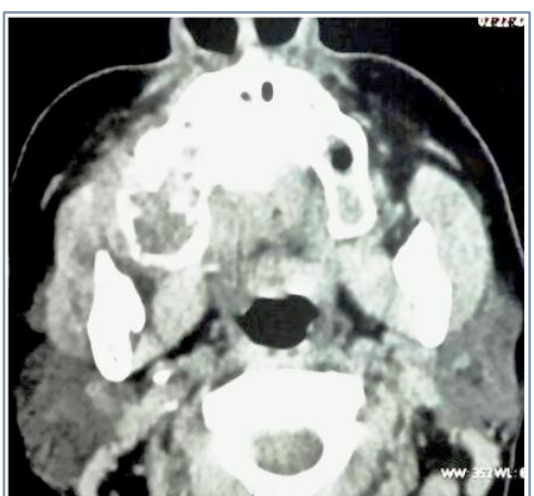

Figure 5