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Case Report

Extramedullary Plasmacytoma Maxilla - A Case Report

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Abstract

Plasma cells are normally found in Bone Marrow, it is responsible for Immunity response by making antibodies to fight infections. A localised buildup of abnormal plasma cells either inside the bone or outside the bones causes Solitary plasmacytoma [1]. Solitary bone plasmacytoma is a rare malignancy in head and neck region.

It usually occurs in adult male with average age of diagnosis. A 49 year old presented in our outdoor patient department with complain of pain and swelling in Left Maxillary area for last one months. Bone Marrow Biopsy shown Mildly hypercellular marrow spaces with granulocyte hyperplasia. Megakaryocyte and erythroid series of cells are proportionally rated.

Biopsy from local area shown poorly differentiated malignant neoplasm. CECT-FACE findings were a large enhancing necrotic mass lesion, PET Scan report shows tumour invades the anterior wall to involve the pre- maxillary soft tissue.

Presence of an Isolated region of Bony destruction due to clonal plasma cells with morphological normal bone marrow examination with very low clonal plasma cell infiltration <10% are diagnosed criteria of solitary plasmacytoma other criteria are normal kidney function, occurrence of anaemia and hypercalcemia lower serum or urine level of monoclonal antibodies [7].

Keywords: Solitary Plasmacytoma; Malignant Neoplasm; Extramedullary Plasmacytoma

Introduction

Plasma cells are normally found in Bone Marrow, it is responsible for Immunity response by making antibodies to fight infections. A localised buildup of abnormal plasma cells either inside the bone or outside the bones causes Solitary plasmacytoma [1]. It is adjacent solitary mass of proliferating malignant monoclonal plasma cells in the bone called as myelomatous type, outside bone in soft bone called extra medullary type. It may be solitary or multifocal [1-3]. Solitary bone plasmacytoma are localized in bone marrow as a single focus of plasma cells, While Extra-medullary plasmacytoma originates from plasma cells located in mucosal surface. Solitary bone plasmacytoma is a rare malignancy in head and neck region. It usually occurs in adult male with average age of diagnosis of 55 years [3-5]. It contributed 3% of all plasma tumor [6].

Case Report

A 49 year old presented in our outdoor patient department with complain of pain and swelling in Left Maxillary area for last one months. He was having single episode of epistaxis 1 year back. There was no history of fever, loss of appetite, nasal blockage, orbital symptoms. There was no clinical lymphadenopathy,no proptosis or any sign of any orbital involvement. He consulted to private doctor one month back.

O/E – poor oral hygiene

- No growth seen protruding from upper hard palate.
- H/O pain in left upper jaw while chewing.
- No H/O nasal obstruction.

He has been advised for routine blood examination, Bone Marrow Aspiration, Bone Marrow Biopsy, Contrast Enhanced Computed Tomography Head and Neck, PET Scan, Biopsy from local area. Peripheral Blood smear report shown RBCs with mild anisopoikilocytosis with macrocyte and normochromic cells, no rouleax formation noted.

Biopsy from local area shown poorly differentiated malignant neoplasm. Repeat biopsy shown plasmoblastic myeloma. On Immunohistochemistry examination plasmoblastic myeloma remain the same. Serum electrophoresis showed hypoalbuminemia and hypergammaglobulinemia and B2 macroglobulin level was raised (3.08 mg/l).

CECT-FACE findings were a large enhancing necrotic mass lesion, measuring size ~ 42×23×40 mm centered on the left maxillary sinus causing gross destruction of the roof, anterior, lateral and medial wall of the left maxillary sinus. The left inferior orbital wall is destroyed. The mass extending anteriorly into the premaxillary soft tissue, superiorly through the orbital floor to the intraconal space. The lesion also seen causing significant destruction of the left alveolar ridge and infiltrating the posterior aspect of superior gingivobuccal sulcus. The lesion seen extending medially into the choana and ethmoid sinus, through destruction of left osteomeatal complex, middle, and inferior turbinate and causing near complete blockage of the left choana.

PET SCAN report shown

PET Scan report shows tumor invades the anterior wall to involve the pre- maxillary soft tissue. Posteriorly, the mass appears to invade the left pterygopalatine fossa. Mild non FDG avid mucosal thickening seen in the right maxillary sinus. Rest of the bilateral paranasal sinuses, nasopharynx, hypopharynx, including larynx and the parapharyngeal and paralaryngeal soft tissue appears normal. There is no significant FDG avid cervical or supraclavicular lymphadenopathy seen.

Bone marrow biopsy report shows mildly hypercellular marrow spaces with granulocytic hyperplasia. Megakaryocytic and erythroid series cells are proportionately noted.

Impression

Mildly hypercellular marrow with granulocytic hyperplasia.

Peripheral Smear Report shown, RBCs show mild anisopoikilocytosis with macrocytic and normochromic cells.

No rouleaux formation noted.

Bone Marrow Aspiration shown Cellularity: Aspirate smears are particulate and hypercellular for age.

Hematology report

- Blasts 01%, Promyelocytes 04%, Myelocytes 13%, Metamyelocytes 08%,
- Neutrophils 46%, Lymphocytes 06%, Monocytes 01%, Eosinophils 00%,
- Basophils 00%, Erythroid series 19%, Plasma cells 02%.
- Hematology report shown Impression of Hypercellular marrow with granulocytic hyperplasia.

Histopathology report

Figure 1: CECT-FACE findings were a large enhancing necrotic mass lesion, measuring size ~ 42×23×40 mm.

Figure 2: CECT Image showing lesion also seen causing significant destruction of the left alveolar ridge and infiltrating the posterior aspect of superior gingivobuccal sulcus.

Figure 3: Contouring of disease area.

Figure 4: Tumor cells are found positive for CD 138(EP201).

Figure 5: Tumor cells are found positive for Ki67(Bh360).

Figure 6: Tumor cells are found positive for H&E.

Figure 7: Poorly differentiated Malignant Neoplasm. (Microscopic picture of Biopsy), left posterior alveolar region biopsy.

Discussion

Presence of an Isolated region of Bony destruction due to clonal plasma cells with morphological normal bone marrow examination with very low clonal plasma cell infiltration <10% are diagnosed criteria of solitary plasmacytoma other criteria are normal kidney function, occurrence of anemia and hypercalcemia lower serum or urine level of monoclonal antibodies [7].

Allegra., *et al*. [8] reported a 43 years old male patient who presented with maxillary swelling, diagnosed abscess initially. After pathological diagnosis finally reported as solitary plasmacytoma Kamal., *et al*. [9] reported a swelling in (R) Lower Submandibular swelling which was not painful, after pathological examination it was diagnosed as solitary plasmacytoma.

Chao MW., *et al*. [10] reported a patient with multiple plasmacytoma without progression into multiple myeloma in 14 years period evaluation. Early treatment can reduce the local complication of disease [11,12].

A case report of 50 patients of solitary plasmacytoma of few showed that usually tumour presented as a single osteolytic lesion with plasmacytosis in bone marrow. Half of the cases programmed to multiple melanoma, poor prognosis reported in that report [11].

Rodriguez-Caballero B., *et al*. [12] reported a 64-year-old male with tumour in (L) mandibular angle with exterior to (L) Mandibular area Radiological Investigationrevealed an osteolytic lesion in (L) mandibular area and diagnosis confirmed as solitary plasmacytoma.

It is a highly radio sensitive tumour [13]. Radiotherapy is the treatment of choice for Extra-medullary plasmacytoma and solitary plasmacytoma with dose of 45-60 greys, 2gy per fractions, 5 fractions per week schedule [14,15]. Excellent tumour control shown [16,17].

For Residual disease these cases in which disease has progressed to multiple myeloma or has chance to progress, adjuvant chemotherapy with radiotherapy or surgical excision may be suggested [18-21].

In this patient lesion is solitary without any metastasis, fulfilling the exterior of solitary plasmacytoma we planned him for radiotherapy with 60gy/30 fraction, 200 cgy per fraction 5 fraction per week.

Conclusion

The maxillary solitary plasmacytoma is an uncommon disease that only affects myleomatous tissue and does not spread to other parts of the skeleton. It is a radiosensitive tumor so it can be treated with radiotherapy. Chemotherapy can be suggested for tumors progressing towards multiple myeloma. Surgery can be advised for residual tumors.

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