ETHMOIDAL SINUS PLASMACYTOMA WITH INTRACRANIAL EXTENSION: A CASE REPORT WITH CLINICAL AND RADIOLOGICAL FEATURES

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ABSTRACT: Plasmacytoma is a neoplastic proliferation of plasma cells that may manifest as multiple myeloma, primary amyloidosis, or monoclonal gammapathy of unknown significance. Plasmacytoma may be primary or secondary to disseminated multiple myeloma and may arise from osseous (medullary) or non-osseous (extramedullary) sites. Primary extramedullary plasmacytoma can be solitary or multiple[1]. The International Myeloma Working Group in 2003 recognized a separate classification of plasmacytomas that occur as multiple sites of disease in soft tissue, bone, or both soft tissue and bone as multiple solitary plasmacytoma. Primary extramedullary plasmacytoma is rare, accounting for only 4% of all plasma cell tumors. Here we report a rare case of extra medullary ethmoidal sinus plasmacytoma with intracranial extension with its clinical, radiological and histological features.

KEY WORDS: Extramedullary Plasmacytoma, Ethmoidal, Intracranial extension.

INTRODUCTION: Plasmacytoma is a mass of neoplastic monoclonal plasma cells. It is classified into medullary (arising in bone) or extramedullary (arising in soft tissue) depending on its origin. Extramedullary plasmacytoma (EMP) represents 4% of all plasma cell tumors[2], 1% of all head and neck malignancies and 0.4% of upper respiratory tract malignancies[3]. Eighty percent of them affect the head and neck[4]. The main sites of involvement are the nasal cavity, paranasal sinuses, nasopharynx and the oral cavity. Men are affected 3 times more than women[5] with average age presentation of 60-70 years[6]. Local lymph node affection occurs in 10 to 20 % of extramedullary plasmacytoma[7]. Metastasis occurs in 35 to 50% of cases[8]. It should be differentiated from other destructive diseases in the maxillary sinus such as olfactory neuroblastoma, lymphoma, anaplastic carcinoma and metastatic tumors[12]. Extramedullary plasmacytomas are highly radiosensitive, so radiotherapy is the best choice of treatment[2]. Long - term follow up is mandatory, as local recurrence and dissemination can occur many years after the original lesion has been treated[13]. Chemotherapy may be added to treatment if there is recurrence of metastasis. Here we report a rare case of solitary EMP in the Ethmoid sinus with intracranial extension.

CASE REPORT: A 60 years old patient presented in medicine OPD of our institute with history of single episode of generalized tonic clonic seizure and nasal bleeding since last 15 days. He also had history of left sided facial swelling and upper eye lid partial closure for three months. On examination his general condition was good, vitals were stable. Respiratory and cardiovascular systems were clinically normal.

CT scan of Head and PNS showed 6 x 5cm sized, soft tissue density, heterogeneously enhancing mass, occupying left ethmoid sinus extending in to the frontal sinus, with destruction of lamina papyracea and cribriform plate on both sides with intra-orbital and intracranial extension. A

non enhancing white matter hypodensity in left frontal lobe was also seen, suspicious of brain parenchymal involvement in the frontal region.

MRI of PNS and Brain was performed which revealed 6 x 5 x 3cm sized, well defined mass lesion of altered signal intensity appearing heterogeneously hyperintense on T2W and FLAIR images, involving the ethmoid and frontal sinuses with intracranial extension and involvement of frontal lobe parenchyma with perilesional edema.

MR Spectroscopy revealed raised Choline peak at the site of lesion in ethmoid sinus and intracranially indicating malignant neoplasm.

Histopathological examination revealed; cells positive for CD138 and EMA but negative for CD19. Morphology favours plasma cell neoplasm with very low KI67. Patient underwent multiple cycles of chemotherapy after which there was resolution of the mass lesion.

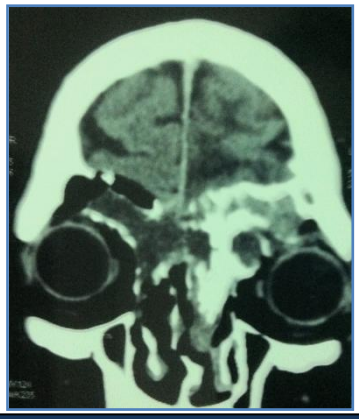
DISCUSSION: Dalrymple and Bence-Jones first identified plasma dyscrasias in 1846 when they described a condition with diffuse bone pain and marked proteinuria. However, it was not until 1873 when Rustizky et al successfully recognized it as a distinct histopathologic entity: multiple myeloma. Since then, this plasma cell neoplasm has been classified into one of three categories: the disseminated form, multiple myeloma, and the localized forms of medullary and extramedullary plasmacytomas. The last variant, solitary extramedullary plasmacytoma (SEP), accounts for less than 2% of all neoplastic plasma dyscrasias and occurs in any part of the body, especially in the head and neck[15]. Nevertheless, this rare tumour represents only less than 1% of all malignancies in the head and neck region[16]. About 75–80% of these tumours originate in the submucosa of the upper aerodigestive tract; of these, 75% involve the nasal tract[15-17]. Solitary extramedullary plasmacytoma of the paranasal sinuses are uncommon and are of B lymphocyte origin[4]. The lesions are characteristically multiple, punched out and osteolytic. Localised growth of multiple myeloma occurs in 5% of cases[18]. Intracranial growth has rarely occurred; the incidence given in the literature is 0.03% to 0.7%[18,19].

The clinical presentation includes asymptomatic meningeal involvement[20], encephalopathy[21,22], cranial nerve palsies[23,24,25,26,27], neuropathy[28,29], convulsions or hemiparesis[30], obstruction of the superior sagittal sinus[31] or features of increased intracranial pressure and possibly uncal herniation[26,31]. Spontaneous haemorrhage into the tumour has been reported, may be related to increased vascularity of the lesion, or the bleeding diathesis associated with the disease[32,29].

Its diagnosis depends on histology and by immunocytochemistry[9]. EMP microscopically consists of sheets of plasma cells which may be monomorphous or pleomorphic[10]. By immuno-histochemical demonstration of one light chain monoclonal staining and one heavy chain class, most EMP can be differentiated form reactive plasma cell infiltrates with polyclonal staining[10]. It is divided into 3 grades according to its cellular atypia into low, intermediate and high grade[11]. Multiple myeloma should be excluded by serum and urine protein electrophoresis and immuno-electrophoresis, skeletal survey, bone scan and marrow biopsy[5].

Radiologically, the most characteristic features include osteolytic lesions on plain skull X-ray, without evidence of bone erosion[34]. On CT scan the lesion is slightly hyperdense with homogenous enhancement following injection of contrast material[19,32,27,33]. Angiographic studies show a highly

vascular lesion[19,27]. Treatment of multiple myeloma has included chemotherapy[25,34,32], radiotherapy[23,24], the administration of alpha interferon[20], and surgical removal[19,27].



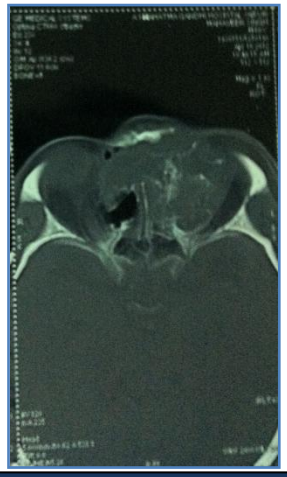
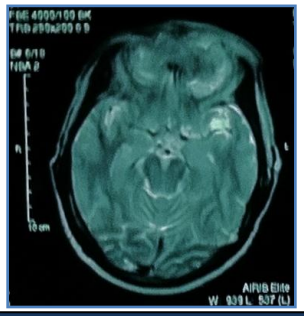


Fig. 1 & 2: Coronal CT image in brain window and Axial image in bone window shows ethmoido-nasal, heterogeneous, soft tissue density mass with destruction of lamina papyracea and cribriform plate on both sides with intra-orbital and intracranial extension.



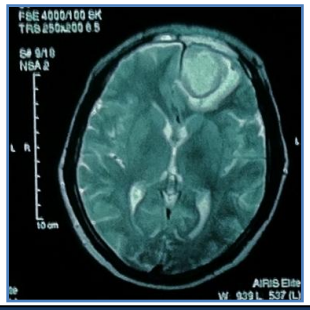
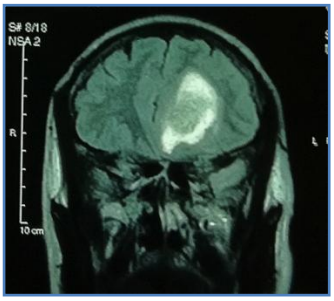


Fig. 3 & 4: Axial T2W images reveal ethmoido-nasal lesion of altered signal intensity appearing heterogeneously hyperintense with intracranial extension and involvement of frontal lobe parenchyma with perilesional edema.



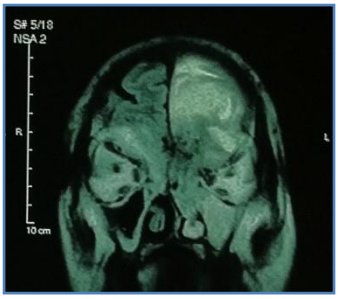


Fig. 5 & 6: Coronal FLAIR images show heterogeneous area of altered signal intensity seen involving the ethmoid sinus and nasopharynx with intracranial extension involving frontal lobe parenchyma on left side with perilesional edema.

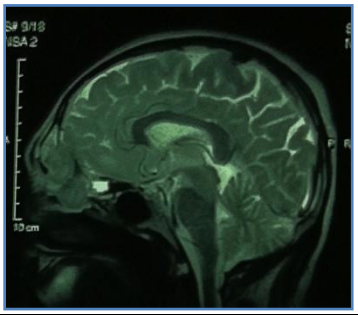
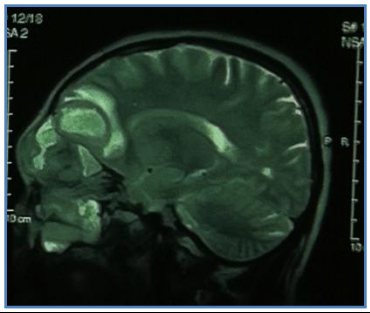
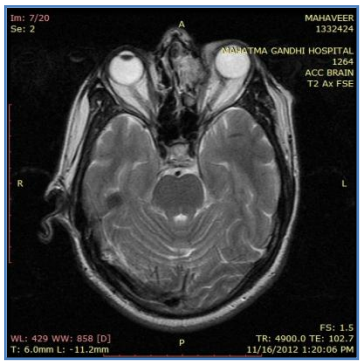
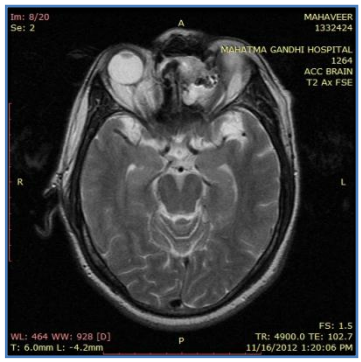



Fig. 7 & 8: Sagittal T2W images reveal ethmoido-nasal lesion of altered signal intensity appearing heterogeneously hyperintense with intracranial extension and involvement of frontal lobe parenchyma with perilesional edema.





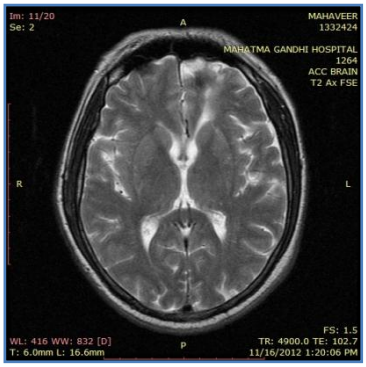


Fig. 9, 10 & 11: Post Chemotherapy, Axial T2W images reveal significant reduction in size of the ethmoido-nasal-intracranial lesion.

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