University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2018, Vol. 5(4)

ROLE OF CT AND MRI IN SOLITARY EXTRAMEDULLARY PLASMACYTOMA OF THE NASAL CAVITY

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Abstract : Extramedullary plasmacytoma is a rare neoplasm characterized by monoclonal proliferation of plasma cells. Most lesions occur in the head and neck, primarily in the upper aerodigestive tract. The nasal cavity and nasal septum is the common sites of occurrence in head and neck. In this case report one patient of age 48 admitted in our hospital with history of nasal obstruction and anosmia. Patient underwent investigations including CT, MRI and diagnosed to have extramedullary plasmacytoma and confirmed with histopathology and immunohistochemistry. And the patient is on combined radiotherapy and chemotherapy. The clinical and histopathologic findings of plasmacytoma are discussed. In order to exclude systemic involvement, systematic approach using clinical, laboratory, and radiologic investigations was performed. Extramedullary plasmacytoma of the nasal cavity is very rare, it should be considered as one of the differential diagnosis in nasal cavity mass lesion especially in adult age group.

Keyword :extramedullary plasmacytoma(EMP), anosmia , nasal cavity, CT, MRI, radiosensitive.

INTRODUCTION: Extramedullary plasmacytoma is a rare soft -tissue malignancy composed of plasma cells. Eighty percent of these tumors occur in the head and neck, 28% occur in the nasal cavity, and 22% occur in the paranasal sinuses **. They represent 3% to 4% of all sinonasal cavity tumors**. About 20% of head and neck extramedullary plasmacytoma are initially associated with multiple myeloma. The tumors are four times more likely to occur in males than in females, 95% of the tumors occur over the age of 40 years. Imaging modalities CT, MRI is very useful diagnosis however demonstration of plasma cells by immunoglobulin clonality via **immunohistochemistry is the mainstay**. **On CT, extramedullary plasmacytoma of the sinonasal cavities are homogeneous, enhancing polypoid masses that remodel surrounding bone. On MR imaging they have an intermediate signal intensity on all imaging sequences, they enhance well**, and because they are highly vascular, they may have vascular flow voids. Radiation therapy and surgery are the treatments of choice. Alkylating agents and steroids help patients with painful bone lesions and in patients with systemic disease.

CASE REPORT:48yr old male patient admitted with the complaints of anosmia , right sided nasal obstruction, mild epistaxis on and off for four month duration. Clinically patient had mild exophthalmos of right eye.

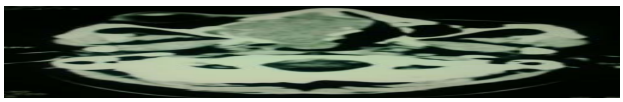
Nasal endoscopy revealed a soft, friable, polypoid mass occupying the right nasal cavity. Clinicians were suspecting carcinoma of nasal cavity, olfactory neuroblastoma, etc. And patient was referred to our radiology department for diagnosis, further characterisation and to look for the extent of the lesion.

***RADIOGRAPH*:**Skull frontal radiograph shows homogenous radio opacity in the right nasal cavity, bilateral frontal and ethmoidal sinus.

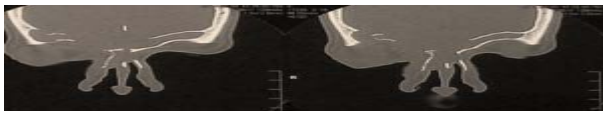
SKULL RADIOGRAPH

***CT*:** It showed a well defined iso to hyperdense soft tissue density mass lesion of size 4.9(cc) x 3.2(ts) x 3.6(ap) cm noted occupying the entire right nasal cavity causing expansion of right asal cavity with destruction of inferior and middle turbinate, lesion also extends to right maxillary sinus. The lesion extends superiorly in to bilateral ethmoidal and frontal sinus, medially in to extraconal compartment of right eye casuing mild proptosis, extends intracranialy in to anterior cranial fossa and posterioinferiorly in to choana. There is bony erosions noted in the walls of bilateral ethmoid sinus , frontal sinus, cribriform plate, right lamina papyracea and crist a galli. The lesion shows homogenous enhancement with intravenous iohexol contrast medium.

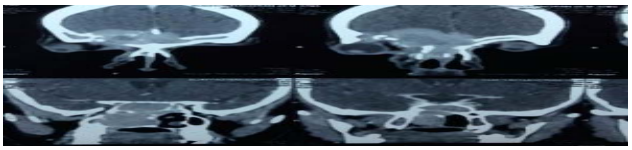
CT PNS- PLAIN



CT PNS - PLAIN

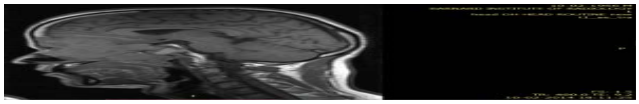


CT BONE WINDOW



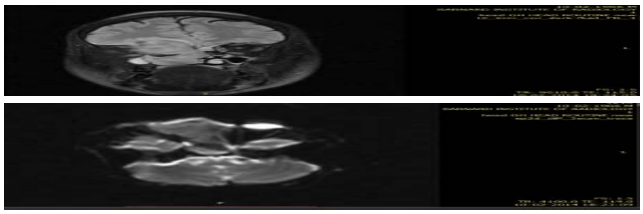
CT CONTRAST STUDY: MASS LESION SHOWS HOMOGENOUS ENHANCEMENT AND LESION EXTENSION IN TO ANTERIOR CRANIAL FOSSA IN THE RIGHT SIDE

MRI: Mass lesion in the right nasal cavity appears iso intense in both T1 and T2 weighted image and extends superiorly in to bilateral ethmoidal and frontal sinus, medially in to extraconal compartment of right eye casuing mild proptosis, extends intracranialy in to anterior cranial fossa and posteriorly in to choana. spenoid sinus mucocele noted due to obstruction by the huge mass lesion. The lesion appears hyperintense in FLAIR images showing mild diffusion restriction and low ADC value in diffusion weighted and ADC images suggesting a cellular nature of the tumor. The lesion shows intense homogenous enhancement following intravenous injection of gadolinium.

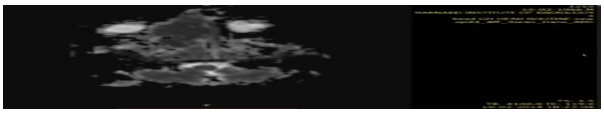


MRI TI WEIHTED SAGITTAL IMAGE - ISO INTENSE LESION MRI T2 WEIGHTED AXIAL IMAGE - LESION EXTENDS FROM NASAL CAVITY TO ETHMOID SINUS ANDEXTRACONAL COMPARTMENT IN TO RIGHT EYE.

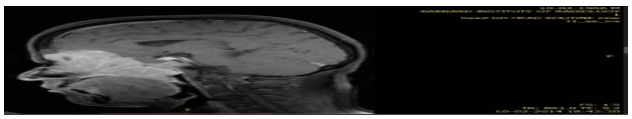
MRI FLAIR CORONAL



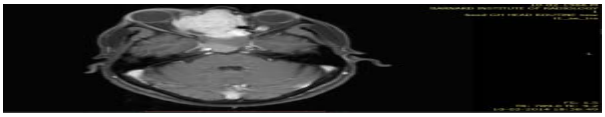
MRI DIFFUSION WEIGHTED IMAGE



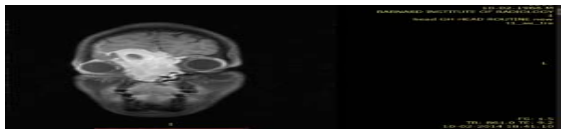
MRI ADC IMAGE



MRI CONTRAST TI SAGITTAL IMAGE



MRI CONTRAST T2 AXIAL IMAGE



MRI CONTRAST FLAIR CORONAL IMAGE

IMMUNOHICTOCHEMISTRY POSITIVE FOR CD38

Thus with above imaging findings we were considering a differential diagnosis of olfactory nueroblastoma, nasal cavity meningioma, carcinoma, lymphoma and a rare possibility of plasmacytoma of nasal cavity. Diagnostic nasal endoscopic guided tissue biopsy of the mass lesion done. Tissue histopathology showed a sheet of discohesive neoplastic plasma cytoid cells with eccentric hyperchromatic nuclei with irregular chromatin distribution. The immunohistochemical study also showed sheets of plasma cells which **decorates intensely for CD 138 which is highly**

suggestive of plasmacytoma. systemic workup was done to rule out multiple myeloma. Results of serum electrophoresis and urine tests were negative for myeloma component or Bence-Jones protein. Bone marrow needle biopsy and skeletal survey were negative for the patient. Thus it is case of **solitary extramedullary plasmacytoma of the nasal cavity**. No nodes involved. As lesion is highly radio sensitive and falls under stage 1(E) locally extended, Patient was put on combined chemotherapy (CVP regimen) and radiotherapy (40 Gray 20 fractions).

DISCUSSION:

Plasma cell tumours include the disseminated form, multiple myeloma, and the medullary (solitary plasmacytoma of the bone) and the extramedullary localized form. Approximately 80-90% ofextramedullary plasmacytoma involve the Mucosa - Associated Lymphoid Tissue of the upper airways, 75% of these involve the nasal and paranasal regions.. The most common clinical findings are: blocked nose, soft tissue mass (fleshy, yellowish grey to dark red sessile, polypoid, or pedunculated), epistaxis, nasal discharge, pain, more rarely cranial nerve palsy and neck lymphadenopathy. In our case, a systemic work-up including blood profile, renal and liver function, serum and urinary protein electrophoresis, serum immunoglobulin level, skeletal survey and bone marrow examination has been performed to exclude a systemic disease such as multiple myeloma. RT is the treatment of choice in EMP localized in the head and n eck, not extending through the floor of the anterior and middle cranial fossae and into the orbit, whereas the role of surgery is usually limited to biopsy and to excision of residual disease. EMPs are most often observed in the head and neck, regional lymph nodes draining the site should be investigated because they can be involved in up to 25% of cases. EMPs can be associated with systemic presentation:

Wiltshaw and Woodroof proposed, in 1979, a staging system for EMPs:

Stage I: tumour confined to primary site;
Stage II: involvement of drainage lymph nodes;
Stage III: evidence of metastatic spread (Multiple Myeloma).

Our case patient is categorised in stage I E (extended) and as the extramedullary plasmacytoma is highly radiosensitive, radiotherapy (40 Gray in 20 fractions) is given as treatment of choice. Chemotheraphy also given (CVP regimen). Six month follow up of the patient showed significant decrease in the size of the lesion.

CONCLUSION :

Thus CT play important role in assessing extent of the lesion, bony destructions and involvement of sinuses. MRI gives a very good soft tissue characterisation and also extent of the lesion. Thus imaging plays important role in the diagnosis of extramedullary plasmacytoma though histopathology and immunohistochemistry is mandatory. Even though extramedullary plasmacytoma is rare tumor of nasal cavity accounting for 3-4%, extramedullary plasmacytoma should be considered as one of the differentials in the nasal cavity mass lesion especially in adultS in fourth to fifth decade. And systemic work is must to rule out multiple myeloma, and long term follow up is also needed to look for if any progression of solitary extramedullary plasmacytoma to multiple myeloma.

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