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Nasal Mass: Extramedullary plasmacytoma, A rare Entity: Case Report

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Abstract: ***Plasma cell tumour are by no mean uncommon tumour, however they are rarerly reported as extramedullary plasmacytoma(EMP) in nose and paranasal sinuses. We report a unusual case of isolated extramedullary plasmacytoma of bilateral nasal cavity in a 45 year old male, a few cases of which were reported previously. Pt was treated primaly with definitive radiotherapy at a dose of 5,000 cGy. Follow up of 6 month rvealed a complete reponse.***

Keywords: Extramedullary plasmacytoma, Paranasal sinuses, Radiotherapy.

1. Introduction

Extramedullary plasmacytomas are infrequent, typically solitary plasma cell tumour. EMPs of the head and neck region is a rare malignant tumour comprising approximately 3% of all plasma cell tumours. [1] They account for 1% of all tumors of the head and neck [2] and 4% of all nonepithelial tumors of the nasal tract. [3] Approximately 80–90% of EMPs involve the mucosa associated lymphoid tissue(MALT) of the upper airways, 75% of these involve the nasal and paranasal regions.[4]-[5] In this article we describing a case of EMP of the paranasal sinuses who was previously treated as chronic sinusitis but after futher investigation the correct diagnosis was made and treated accordingly.

2. Case Report

A 45 year old male patient presented to us with complains of bilateral nasal obstruction and nasal discharge since 3 months and few episodes of right side nasal bleed since 15 days. Before this patient was treated with oral antibiotics, oral antihistamincs, local nasal decongestant and local steroid spray by his family physician after an x-ray film of sinuses revealed opacification of bilateral maxillary sinuses with no improvement. Anterior Rhinoscopy revealed septum deviated to right side with a grayish, polypoidal, non-sensitive, friable mass in right nasal cavity covered with mucopurulent discharge. The medial wall of left maxillary sinus had expanded in to the nasal cavity and was eroded. Bilateral vision was 6/60. Rest of the head and neck examination was unremarkable.Nasal endoscopy revealed pale, soft and friable mass in right middle meatus. Biopsy was taken from the mass in right nasal cavity and sent for histopathological examination.

CECT scan of paranasal sinuses (fig.1) revealed bilateral sinonasal mass with destruction of part of bilateral maxillary sinuses(left>right), septum, middle and inferior turbinates and lamina paperacea with intraorbital involvement(right>left) however, no obvious infiltration of medial rectus or globe or optic nerve was seen.

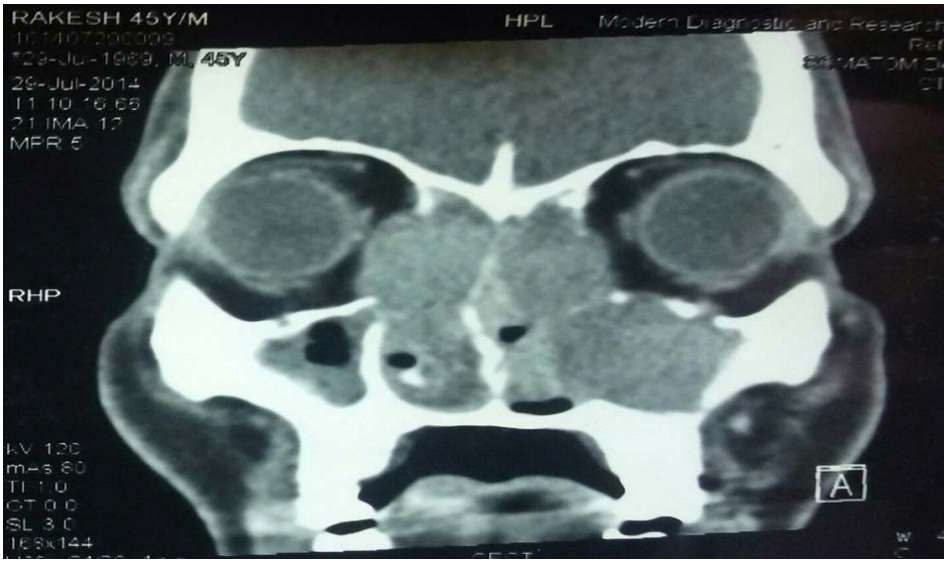

Figure 1: Bone window coronal CECT scan of the Paranasal sinuses demonstrating a mass in both nasal cavities & left maxillary sinus. The mass has destroyed the medial wall of the left maxillary sinus and bilateral lamina paperacea.

Histopathological examination revealed large numbers of mature and immature cells and proliferative capillaries. On immunohistochemistry Tumour cell expressed CD 138 +ve, while synaptophysin was –ve and CD 99 was non specific. The tumour cells were immunonegative for CD 20(pan B-cell marker), CD 3(pan T-cell marker) and ctyokeratin suggested the mass to be plasmacytoma. His laboratory reports revealed the following: Hb: 10.2g%, TLC: 8800/cc, DLC: P65, L30, M4, platelets 320 x 103 / L, ESR 120 mm/hr, Blood urea: 38mg%, S. creatinine 1.2mg%, S. uric acid 4.8 mg/dL, S. calcium 8.2 mg/dL, total protein 11.20 g/dL, S. albumin 3.20 g/dL, gamma globulin 7.1 g/dL, A:G ratio 0.45. Assay for Bences jones protein in urine and for serum myeloma protein were negative. Bone marrow biopsy and bone scan revealed no systemic lesions. The patient was sent for radiotherapy and given 50 Gy in 25 settings. Follow up at 6 months showed no recurrence.

3. Discussion

Plasma cell neoplasm constitute a group of disorders characterized by monoclonal proliferation of plasma cells and the presence of monoclonal immunoglobulins in serum. Plasma cell neoplasm depending on their site of development and clinical features, classified as multiple myelomas, medullary plasmacytoma or extramedullary plasmacytoma.[6] These are further classified as either localized(stage 1), localized including local lymph nodes(stage 2), or generalized (stage 3) according to clinical manifestations.[7] EMP is rare localized plasma cell tumour originating in soft tissues with no evidence of bone marrow disease and normal total body skeletal survey, however bone erosion adjacent to plasmacytoma may occur. [6] The presenting symptoms in sinonasal region are nasal obstruction, nasal discharge ,nasal mass,epistaxis and facial pain mimicking recurrent or chronic sinusitis [8] like in the present case. They are most commonly seen in patients between 50 and 60 years of age, more frequent in men[9] as was observed by us. EMPs have lower rate(20-30%) of conversion into disseminated multiple myeloma as comparison to solitary medullary plasmacytoma(58%) which are lytic bony lesion which raely involve head and neck region and usually affect long bones.[1] There is no sexual predilection in medullary plasmacytoma. EMP should be distinguished from non-neoplastic lesions like reactive plasmacytic hyperplasia, plasma cell granuloma, pseudolymphoma and from malignancies like haematopoeitic neoplasms, malignant melanoma, olfactory neuroblastoma, anaplastic carcinoma and metastases. Differentiation between plasmacytoma and polyclonal infiltrates of plasma cells requires immunohistochemical studies.[10] Plasma cell neoplasms are highly radiosensitive. Radiation therapy is currently the treatment of choice for EMPs and medullary plasmacytoma.[11] They respond well to radiation at doses of 4,500- 6,000 cGy, delivered as daily fractions of 125-200 cGy 5 days a week.[12] It is important to distinguish multiple myeloma from extramedullary plasmacytoma because the treatment and prognosis are different. Histopathological examination with immunohistochemistry of the excised tissue is a gold standard investigative procedure along with consultation with pathologist for correct diagnosis and to exclude any degree of malignant transformation in to multiple myeloma. It is essential to document new cases which should be followed carefully so that accurate, better treatment modalities could be formulated for future.

References

[1] Knowling M, Harwood A, Bergasagel D. Comparison of extramedullary plasmacytoma with soliatory and multiple plasma cell tumour of bone. J Clin Oncol. 1983;1:255-62
[2] Kost KM. Plamacytoma of the larynx. J Otolaryngol. 1990;19:141-6
[3] Grover N, Chary G, Makhija P, Rout P. Extramedullary plasmacytoma of the nasal cavity: treatment perspective in a developing nation. Ear Nose Throat J. 2006;85:434–6.
[4] Attanasio G, Viccaro M, Barbaro M, Seta E, Filipo R. Extramedullary plasmacytoma of paranasal sinuses. A combined therapeutic strategy. Acta Otorhinolaryngol Ital. 2006;26:118–20.
[5] Miller FR, Lavertu P, Wanamaker JR, et al. Plasmacytomas of the head and neck. Otolaryngol Head Neck Surg. 1998;119:614-8.
[6] Bataskis JG. Pathology consultation. Plasma cell tumour of the head and neck. Ann Otol Rhinol Laryngol. 1983;92:311-3.
[7] Wiltshaw E, The natural history of extramedullary plasmacytoma and its relation to solitary myeloma of bone and myleomatosis. Medicine. 1976;55:217-38.
[8] Castro EB, Lewis JS, Strong EW. Plasmacytoma of paranasal sinuses and nasal cavity. Arch Otolaryngol. 1973;97:326-9.
[9] Paris J, Dessi P, Moulin G, et al. Extramedullary plasmacytoma of the nasal cavity: A case report. Rev Laryngol Otol Rhinol. 1999; 120: 343-5.
[10] Kapadia SB, Desai U, Cheng VS. Extramedullary plasmacytoma of the head and neck. Medicine 1982;61(5):317–9. doi: 10.1097/00005792-198209000-00004.
[11] Nofsinger YC, Mirza N, Rowan PT, Lanza D, Weinstein G. Head and neck manifestations of plasma cell neoplasms.Laryngoscope. 1997;107:741-6.
[12] Wax MK, Yun J, Omar RA. Extramedullary plasmoytomas of the head and neck . Otolarngol Head Neck Surg. 1993;109:877-88.