

CASE REPORT

Extramedullary Plasmacytoma of Naso-oropharynx: A Rare Tumour

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Abstract:

Plasmacytoma is a monoclonal neoplasm derived from progenitor B lymphocyte. Extra-medullary plasmacytoma is proliferation of malignant plasma cells involving the soft tissues with the absence of malignant plasma cells in the bone marrow and with normal gammaglobulin in serum, which is a rarely occurring variant of solitary plasmacytoma (3-4% of all solitary plasmacytoma). Seventy two years old man, suffered from right side nasal blockage for last 6 months. Nasal endoscopy revealed ipsilateral complete choanal occlusion by an endophytic mass having smooth surface arising from the lateral wall of the nasopharynx with an extension to the same side of oropharynx. On computed tomography of paranasal sinus an endophytic homogenous enhancing soft tissue shadow arising predominantly from right postero-lateral nasopharyngeal wall with an extension to oropharynx and without any surrounding bony erosion. The lesion was completely excised endoscopically. Histopathology as well as immunohistochemistry confirmed plasmacytoma. Extra medullary plasmacytoma was diagnosed as per International Myeloma Working Group (IMWG) criteria. Patient was treated with radiotherapy in a total dose of 50Gy in 25 cycles. Follow up at 5 and 10 months showed no evidence of recurrence or progression to multiple myeloma. A rare tumourexta medullary plasmacytoma of naso-oropharynx has very good prognosis. The radiotherapy remains mainstay of treatment in extra-medullary plasmacytoma. The complete endoscopic excision before radiotherapy improves outcome of treatment as in this case.

Keywords: Endoscopy, Nasopharynx, Plasma cells

Introduction:

Plasmacytoma is a monoclonal neoplasm derived from progenitor B lymphocyte lineage. It presents commonly as multiple myeloma which are characterized by excess malignant plasma cells in bone marrow, multiple lytic bony lesion and presence of abnormal gammaglobulin (Bence-Jones protein) in serum and rarely occurs as solitary plasmacytoma (5-10% of all plasma cell tumors) [1-2]. Extra-medullary Plasmacytoma (EMP), which is a variant of solitary plasmacytoma, is even rarer (3-4% of all solitary plasmacytoma) [3]. The World Health Organization (WHO) classify plasmacytoma into Multiple Myelomas (MM) and solitary plasmacytoma [Solitary Bone Plasmacytoma (SBP) and Extramedullary/ Extraosseous Plasmacytoma (EMP)] [4]. However, the International Myeloma Working Group (IMWG) classification recognizes MM, SBP, and EMP as distinct entities [5]. Thus, EMP is characterized as a plasma cell tumor, along with SBP and MM, which is considered a more advanced stage of disease [6]. About 80-90% of EMP arise in the head and neck region, and the most common location is the upper aerodigestive tract especially the nasal cavity, paranasal sinuses, nasopharynx, oropharynx and larynx [7-9]. Other sites in the head and neck include orbit, palate, skin, skull base, salivary

glands, thyroid glands, tonsils, cervical lymph nodes[8]. Generally, symptoms are non-specific and depend on the site, local tumor mass effect and spread of the tumor by adjacent bone erosions. In the nasopharynx, symptoms of EMP include nasal congestion, anosmia, hyposmia, epistaxis, rhinorrhea, pain, and neck swelling. Obstruction of the aerodigestive passage may occur in advanced stage. Radiotherapy and surgery are the main treatment modalities for EMP in the head and neck [9]. We report our recent experience with EMP of the naso-oropharynx treated by complete excision using endoscope instead of intraoral approach before radiotherapy in the context of the limited literature on this topic.

Case Report:

Seventy two year old male presented with right sided nasal blockage for last 6 months. There was no history of loose teeth, facial swelling, diplopia or blurring of vision. There was no palpable cervical lymphadenopathy and no evidence of involvement of any cranial nerve. Complete blood count, renal function tests, serum electrolytes were within normal limits. Nasal endoscopy findings were suggestive of ipsilateral complete choanal occlusion by an endophytic mass having smooth surface arising from the lateral wall of the nasopharynx with an extension to the oropharynx on right side (Figs. 1 and 2). Computed tomography of paranasal sinus showed an endophytic homogenous enhancing soft tissue lesion with lobulated margins arising predominantly from right postero-lateral nasopharyngeal wall extending to oropharynx (Fig. 3). No associated bony

erosions were present. Endoscopic complete excision was done (Fig. 4) and histopathological examination revealed a nodular grayish tissue microscopically showing sheets of plasma cells seen underlying squamous mucosa (Fig. 5).

Russell bodies were also noted. Immunohistochemical studies revealed positivity towards CD138, CD38, Kappa and Lambda. Tumor cells negative for CD56, MUM1, CD79a, CD20. Mib1 shows almost no mitotic activity. Immunohistochemistry confirmed plasmacytoma. Following the histological diagnosis the serum electrophoresis and skeletal surveys were performed revealing neither the monoclonal band nor any lytic bony lesions respectively to rule out multiple myeloma. Also the bone marrow biopsy did not show clonal plasma cells in it. Extra medullary plasmacytoma was diagnosed as per IMWG criteria [10]. Patient was treated with radiotherapy in total dose of 50Gy in 25 cycles. After completion of RT and follow up till date, diagnostic nasal endoscopy and serum electrophoresis showed no evidence of local recurrence or development to multiple myeloma.



Fig. 1: Endoscopic View



Fig.2: Intraoral View Before and After 10 Months of Surgery

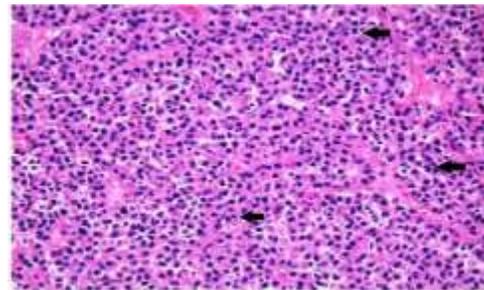


Fig. 5: Microscopic View after Excision

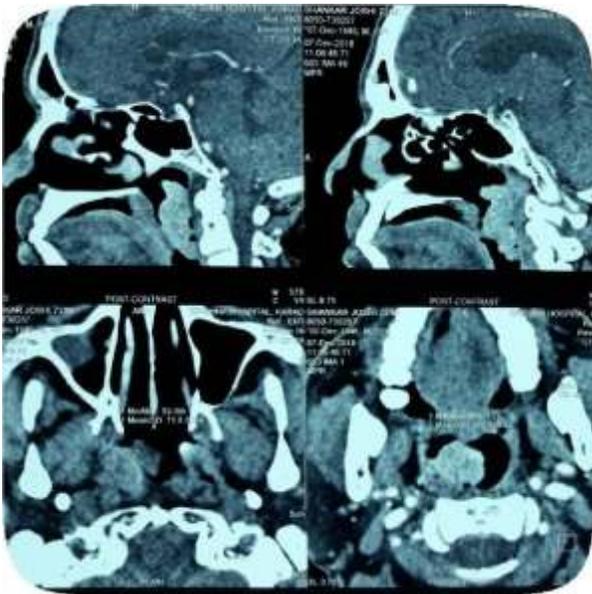


Fig.3: CT Scans Showing Extension of the Lesion



Fig. 4: Excised Specimen

Discussion:

It occurs between the 4th and 7th decade of life, the median age is 55 years although one-third of patients are under 50 year old [11]. There is a 3:1 male dominance [3]. This case report is of 72 years old male. The clinical staging of EMP is based on Wilshaw method, that classify soft tissue plasmacytoma into three clinical stages, namely Stage I- limited to an extramedullary site; Stage II- involvement of regional lymph node; Stage III- multiple metastases. In our case, the patient was in stage I since the tumor was confined to the primary site and as there was no cervical lymph node or obvious metastasis. Cervical lymph node metastasis is reported in 12-26% of cases at initial presentation [12]. Since these neoplasms may signal the presence of multiple myeloma, a full evaluation is required to exclude disseminated disease. Abemayor *et al.* [12] recommended a complete blood count with white blood cell count, platelet count, erythrocyte sedimentation rate, bone marrow biopsy, serum biochemistry including calcium, blood urea nitrogen, creatinine, uric acid, serum protein, serum, urine electrophoresis and a skeletal survey to rule out multiple myeloma [12] while Galleni *et al.* [8] recommended the

diagnostic criteria for EMP as: tissue biopsy showing monoclonal plasma cell on histology, bone marrow plasma cell infiltration not exceeding 5% of all nucleated cell, no evidence of myeloma or osteolytic lesion, absence of hypercalcemia or renal failure, absent or low serum or urinary M-protein level (Paraprotein concentration/monoclonal immunoglobulin <2 g/dL), even if present [8]. In less than 25% of patients, a monoclonal paraprotein can be detected in the blood or urine [13]. Our case satisfied all the criteria described above for the diagnosis of EMP. Immunohistochemistry eliminated other mimicking lesions, such as lymphoma, reactive plasmacytoma, and plasma cell granuloma. Immunohistochemical staining in this case showed strong positivity to lambda restriction. Generally for EMP, surgery or radiotherapy are acceptable treatment methods depending on the resectability of the lesion but combined therapy gives good outcome [13]. EMP is also highly radiosensitive, with local cure rates of up to 100% [7]. Multiple Myeloma (MM) is believed to be a disseminated form of EMP with conversion occurring in about 15-35%, usually in 2-3 years [6-8]. It is for this reason that long term follow-ups of patients to identify progression to MM early is of importance.

MM is indicated by the presence of Calcium increase, Renal insufficiency, Anemia, or Bony lesions (CRAB) and it is often associated with Polyneuropathy, Organomegaly, Endocrinopathy, Multiple Myeloma, and Skin (POEMS) changes [14].

Conclusion:

EMP of naso-oropharynx has very good prognosis with complete endoscopic excision prior to radiotherapy. Radiotherapy is the main stay of treatment in extra-medullary plasmacytoma. In spite of the difficulty in accessing the site of lesion during surgical excision, the endoscopic approach for complete excision before radiotherapy improves the outcome as in this case. The close collaboration between E.N.T. specialist, pathologist, and radiotherapist in management of extra-medullary plasmacytoma of naso-oropharynx is more desirable. In view of all possibilities of local recurrence or progression to multiple myeloma a long term follow up is essential.

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