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Case Report

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Extramedullary Plasmacytoma of Nasopharynx and Maxillary Sinus: A Rare Metachronous Presentation

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Summary

A 52-year-old gentleman presented with two extramedullary plasmacytomas occuring within a period of 10 years. He first presented with 3 month history of nasal obstruction and snoring. Nasal endoscopy revealed a pale lobulated mass occupying the entire nasopharynx. He underwent excision of nasopharygeal mass via transnasal endoscopic-assisted approach. Post-operative histopathogical report revealed a diagnosis of extramedullary plasmacytoma. He received radiotherapy with a radiation dose of 45 Gy in 25 fractions over five weeks followed by regular post treatment surveillance .10 years later, he presented with similar complaints of nasal congestion. CT scan of the paranasal sinuses showed complete opacification of the right maxillary sinus obliterating the right osteomeatal complex with bony erosion adjacent medial wall of the maxillary sinus. Right endoscopic excision was performed therefore intraoperatively a mass covering the right maxillary ostia and wall of the maxillary sinus was removed completely. Histopathologic examination confirmed a diagnosis of extramedullary plasmacytoma. After multidisciplinary discussion, the patient was given further radiotherapy of 45 Gy in 25 fractions. Patient currently has recovered well in remission and under follow up for surveillance.

Background

Plasma cell neoplasms represent a heterogenous group of rare tumors characterized by the proliferation of a single B-lymphoid cell clone, producing a monoclonal immunoglobulin. They can appear as solitary lesions (plasmacytoma) or disseminated throughout the body-multiple myeloma (MM). Plasmacytomas may develop either as extramedullary (extraosseous) plasmacytoma (EMP), i.e., in the soft tissues, or as solitary bone plasmacytoma (SBP) [1]. The former arises from plasma cells located in mucosal surfaces. EMP is the rarest of all the three varieties with MM:SPB:EMP incidence ratio of approximately 40:2:1 [2]. of the three entities, EMP has the best prognosis, although within 10 years 11-30 % of cases can progress toward MM [3].

Although it is the rarest type of plasmacytoma, approximately 80-90% of EMP are found in the head and neck region with 40% occurring in the nasal cavity, 20% in nasopharynx and 18% in the oropharynx [4-6]. Other sites include salivary glands, thyroid gland, tonsils, cervical lymph nodes, larynx and skin [5, 6]. EMP typically presents between 55 to 60 years of age. Male preponderance is seen with incidence in males 4 times that of females, especially in African Americans [6]. EMP develops as a locally invasive submucosal tumour with a tendency to recur. Diagnosis depends upon biopsy and exclusion of disseminated disease (MM). Treatment of EMP includes radiotherapy and surgery.

Herein, we describe a case of nasopharyngeal EMP which was successfully treated into remission but had a metachronous EMP in the right maxillary sinus ten years after the initial presentation. To our knowledge, this is there is no other reported case of metachronous EMP in the head and neck regions after such a prolonged period.

Case Presentation

A 52-year-old Malay gentleman presented with two extramedullary plasmacytomas occuring within a period of ten years. He was first diagnosed with nasopharyngeal plasmacytoma

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when he presented with complaints of nasal obstruction and snoring duration. On examination, nasal endoscopy revealed a pale lobulated mass occupying the entire nasopharynx, suggestive of lymphoid hypertrophy. He underwent excision of nasopharyngeal lymphoid tissue via transnasal endoscopic-assisted microdebrider. Unexpectedly, post-operative histopathogical report revealed an extramedullary plasmacytoma. A bone marrow trephine biopsy ruled out multiple myeloma. He subsequently received radiotherapy (RT) with a radiation dose of 45 Gy in 25 fractions over five weeks followed by regular post treatment surveillance every three to six months by the otorhinolaryngology and hematology teams.

Ten years later, he developed similar complaints of nasal congestion duration of symptoms. CT scan of the paranasal sinuses showed complete opacification of the right maxillary sinus obliterating the right osteomeatal complex with adjacent medial maxillary sinus wall bony erosion. A right endoscopic excision surgery was performed under general anaesthesia. A right uncinectomy and middle meatal antrostomy revealed a mass covering the right maxillary ostia and wall of right maxillary sinus with pus drained. The mass was removed completely using angled endoscope and curved instruments. The postoperative period was uneventful, and patient reported no more nasal obstruction two weeks after ESS. Histopathologic examination confirmed the diagnosis of EMP of the right maxillary sinus. Bence-Jones protein was absent in urine. Renal function, serum calcium, serum protein electrophoresis, serum immunoglobulin free light chain (FLC) and skeletal survey were normal. After multidisciplinary discussion, the patient underwent further radiotherapy of 45 Gy in 25 fractions.

Discussion Include A Very Brief Review of Similar Published Cases

Extramedullary plasmacytoma (EMP) is an extremely rare disease and accounts for approximately 1 % of all head and neck malignant tumours and 4% of the non-epithelial sinonasal tumours [7]. Due to its predilection for the Mucosa-Associated-Lymphoid Tissue (MALT) in nasal cavity and paranasal sinuses, some authors have postulated that these could be due to chronic stimulation of the lymphoid tissue caused by inhaled irritants or viral infections and genetic alterations in the reticuloendothelial system which subsequently leads to neoplastic transformation [8, 9, 10].

The clinical presentation of EMP, particularly those arising in the head and neck regions, are indistinguishable from other malignant tumors. EMPs of the sinonasal region usually presents as progressive nasal obstruction and recurrent epistaxis [7, 11, 12]. Due to its nature of a locally destructive tumour, other possible symptoms include facial swelling, headache, proptosis, visual disturbances and pain [13, 14]. On imaging, CT may demonstrate local bony erosion and invasion in EMP of nasal cavity and maxillary sinus while MRI is superior in delineating the various soft tissue involvement. However, these imaging remain non-specific [15].

Histopathological examinations are of utmost importance in the diagnosis of an EMP. As the tumor is submucosal and the mucosa might be thickened as a result of chronic inflammation, a deep incisional or excisional biopsy is the gold standard for histological diagnosis [14]. Fine needle aspiration (FNA) is non-diagnostic and not recommended because of the limited tissue retrieved [8, 16]. Histological features include solid sheets, vague nest and diffuse infiltrates of immature plasma cells displaying round to oval nuclei with moderate pleomorphism. The presence of CD18 and CD338 markers are also checked in an immunohistological analysis. In addition to histopathological confirmation, a systemic work-up including complete blood count with white blood cell and platelet count, renal and liver function, serum and urinary protein electrophoresis, serum immunoglobulin level, serum calcium level, skeletal survey and bone marrow examination needs to be performed to rule out systemic disease such as multiple myeloma [17]. High levels of paraprotein in the serum or urine should raise the clinician's suspicion of a disseminated process, since paraprotein levels correlate directly with tumour burden. Histologic findings along with laboratory and radiographic findings showing the absence of disseminated disease (MM) confirms the diagnosis of extramedullary plasmacytoma.

The therapeutic strategy of EMP is varied and remains controversial due to lack of medical evidence for the optimal and effective treatment especially in recurrent EMPs. As EMP is a highly radiosensitive tumor, Radiotherapy (RT) is the most frequent form of treatment with a reported successful control rate of 80% [18-21]. Most authors agree on a 40-50 Gy irradiation to the paranasal sinuses as a safe regimen. However, Creach, et al. [12] reported a 11% risk of developing radiation-induced malignancy within 10 years of treatment while Alexiou, et al. [8] demonstrated a higher conversion rate to multiple myeloma following RT for EMP of the head and neck region. Recent studies demonstrate better long term outcomes through a combination of RT and surgical removal [8, 22-24]. Surgical removal of tumors may be for therapeutic or diagnostic purposes, such as in our patient. A combination therapy of surgery and RT provides better progression free survival. However, depending on the site of tumor, surgical resection can be highly invasive and surgery without RT shows increased rates of recurrence [18].

In conclusion, EMP is an extremely rare disease that occurs in the head and neck region, making this an important different diagnosis for head and neck malignancies. A comprehensive investigation is important to differentiate between relapse of extramedullary plasmacytoma and multiple myeloma. Long term multidisciplinary surveillance is crucial as EMP has a high risk of recurrence and developing to multiple myeloma despite many years after treatment, such as in our case. Patients should be assessed regularly by performing nasal endoscopy, fiberscopy , serum –free light chains and CT.

Learning Points/Take Home Messages 3-5 Bullet Points

- EMP is an extremely rare disease that occurs in the head and neck region, making this an important different diagnosis for head and neck malignancies.

•A comprehensive investigation is important to differentiate between relaspe of extramedullary plasmacytoma and multiple myeloma.

Acknowledgement

None.

Conflict of Interest

No conflict of interest.

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