ORIGINAL RESEARCH PAPER

Otolaryngology

KEY WORDS:

ENT MANIFESTATIONS OF MULTIPLE MYELOMA

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ABSTRACT

The modern Otolaryngologist is well aware that a systemic disease may manifest itself in our specialty. Multiple myeloma is a distinctive form of plasma cell dyscrasia with non-specific symptoms such as epistaxis, hoarseness of voice, asymmetry of the face, swelling of the palate and nasal obstruction. In this study, we describe a case of Multiple Myeloma with events leading to its diagnosis and its management.

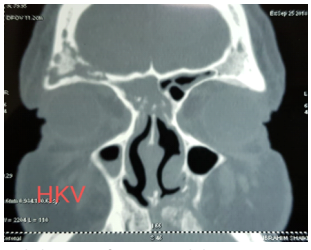
INTRODUCTION

Multiple myeloma (MM) is a type of neoplasm in which plasma cells accumulate in bone marrow and 10% of all haematologic malignancies is attributed to MM[1,2]. It is characterized by skeletal destruction, hypercalcaemia, renal failure and anaemia. In this article, we present a case of multiple myeloma, in order to create awareness about its manifestations in ENT.

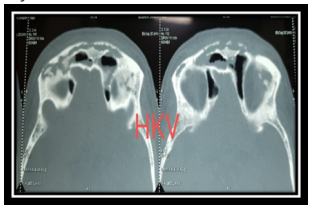
Case report

A 55-year-old male patient came to the ENT OPD with a history of headache, repeated epistaxis, nasal obstruction, anosmia, left sided facial pain and loosening of teeth since 3 months. He also had complaints of severe weakness and bone pain since 1 year. General examination was normal. On anterior rhinoscopy,mucoid crusts were present in the left nasal cavity obstructing it. There was severe tenderness over the left maxillary region and right frontal region. Oral cavity examination revealed loss of one upper incisor tooth on the left. Diagnostic nasal endoscopy showed debris and mucoid crusts along the floor of the left nasal cavity and lateral to the middle turbinate. Debris was sent for KOH mount and culture and sensitivity. CT PNS reported several bone lesions present in the right frontal bone and left maxilla. This raised our suspicions of Multiple Myeloma. On investigation ESR(147 mm/h), white blood cells(5.07×10^9/l) and S. Calcium (3.46 µmol/l)were raised and haemoglobin was decreased (7.2 g/l). Serum electrophoresis revealed a homogeneous spike in the gamma region identified as IgG. Nephelometry showed a higher IgL- level of 11.6g/l. with 2- microglobulin level at 3.76mg/l. Bone marrow aspirate morphology showed a diffuse infiltration of 35% atypical plasma cells. Immunohistochemical staining of a bone marrow trephine biopsy specimen revealed CD38- and CD138-positive cell infiltration (pathognomic of Multiple Myeloma).

The diagnosis of multiple myeloma was made, and the patient was treated with a combination chemotherapy of cyclophosphamide, methylprednisolone and thalidomide. Currently, the patient is free of epistaxis and has shown signs of improvement during the eight months following initial diagnosis.



Right frontal bone(A) and left maxilla(B) showing Lytic bone lesions seen in multiple myeloma



Multiple fractures noted over the frontal bone

DISCUSSION

Multiple myeloma is characterised by an increase in abnormal plasma cells (myeloma cells) in the bone marrow which form tumours in various regions of the body. These tumours may keep the bone marrow from making enough healthy blood cells. As the number of myeloma cells increase, fewer red blood cells, white blood cells, and platelets are made. The myeloma cells also damage and weaken the bone. Sometimes multiple myeloma does not cause any signs or symptoms. This is called smouldering multiple myeloma.

Our patient presented with complaints of headache, repeated episodes of epistaxis, nasal obstruction, anosmia, facial pain and loosening of teeth, bone pain and weakness. Other manifestations of multiple myeloma could be hoarseness of voice, asymmetry of face, swelling of the palate and enlargement of the salivary glands.

The following illustrates the diagnostic criteria for Multiple Myeloma

Disease entity	M-protein	Clonal Plasma Cells in Bone Marrow	End-organ damage	
Monoclonal gammopathy of undetermined significance	<30 g/L	<10%	No	No other B-cell ymphoproliferative disorder

Smouldering Myeloma	>30 g/L in serum Or >1 g/24hour in the urine	>10%	No	
Symptomatic Myeloma	Any value	>10% <10% if other criteria are met	Yes	
Solitary Plasmacytoma	All of the following: Biopsy proven solitary lesion No clonal plasma cells in the bone marrow An otherwise normal skeletal survey and MRI of the spine and pelvis No end organ damage			

In this era of novel agents, treatment varies depending on whether the patient is eligible for Autologous Stem Cell Transplantation or not. Age >65, poor performance status and significant organ dysfunction (significant liver disease, renal disease with creatinine >220 umol/L unless on stable chronic dialysis and/or NYHA class III-IV) render patients unsuitable for such intensive therapy. Thalidomide (alone or with prednisone) has been shown to improve prognosis post-ACST in young patients in six studies and to prolong prognosis in two trials in elderly patients.

Treatment for younger patients- The aim of therapy in these patients is to induce a rapid and deep response, reverse organ damage and decrease the burden of disease. This is done by the use of combinations of high dose dexamethasone with thalidomide (TD; Thalidomide+Dexamethasone),3,4 bortezomb (VD; Velcade i.e. bortezomib+Dexamethasone),5 or lenalidomide (Rd; Revlimid i.e. lenalidomide+low dose dexamethasone),6 or with more than one agent. VTD (velcade with thalidomide and dexamethasone),7 CTD (cyclophosphamide, thalidomide and dexamethasone),8 and CyBorD (cyclophosphamide, bortezomib and dexamethasone),9 seem to produce the highest response rates. Maintenance therapy- With the excellent results obtained with newer agents, it becomes necessary to use something to maintain these responses for as long as possible.

Treatment for patients who are transplant ineligible- The aim of therapy in this group is disease control with the least possible toxicity. Melphalan combined with prednisone has been the backbone of therapy for years producing 50% response rates. [10]

Differential diagnosis of multiple myeloma

MGUS differs from multiple myeloma in that the value of M protein is < 30g/L, whereas smouldering myeloma has M protein values >30g/L and is asymptomatic. Solitary Plasmacytoma presents with a single lesion, without evidence of systemic disease. Other conditions where M-protein may be present are AL-amyloidosis, B-cell non-Hodgkin lymphoma, Chronic lymphatic leukemia, Autoimmune diseases such as SLE.

CONCLUSION

As described, Multiple myeloma may present with vague symptoms. Therefore it is evident that a simple sign or symptom may represent one small facet of a generalized condition.

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