Multiple myeloma presenting as dysphagia

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ABSTRACT

يعد ورم البلازماويات كتلة وحيدة منعزلة لخلايا الورم الأحادي في نقى العظم أو الأنسجة الرقيقة. أن ورم البلازميات خارج الخلية أو الورم النقوي المتعدد للحنجرة حالة نادرة نستعرض في هذا التقرير حالة مريض ذكر يبلغ من العمر 77 عاماً تم تشخيصه بورم نقوي متعدد مع صعوبة في البلع. قمنا بمناقشة ندرة الحالة وصعوبة تشخيصها. نستعرض هذه الحالة لزيادة وعي أخصائيو الأنف والأذن والحنجرة في تشخيص هذه الحالة والسيطرة عليها.

Plasmacytoma is a discrete solitary mass of neoplastic monoclonal plasma cells in either bone marrow or soft tissue sites. Extramedullary plasmacytoma or multiple myeloma of the larynx is extremely a rare condition. We report a 77-year-old male patient diagnosed with multiple myeloma and presented with dysphagia. The rarity of the disease incidence and difficulty of the diagnosis of this disease is discussed. We present this case to increase the awareness of the Otolaryngologists of this rare disease to expedite its diagnosis and management.

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Multiple myeloma (MM) also known as plasma cell myeloma is a bone marrow based multifocal autonomous proliferation of neoplastic plasma cell.[1,2] A single isolated lesion may occur either in bone (solitary plasmacytoma of bone) or soft tissue extramedullary plasmacytoma (EMP).[2] Multiple myeloma, solitary plasmacytoma, and EMP constitute a continuous disease spectrum of plasma cell neoplasm.[3] Multiple myeloma occasionally occurs as solitary extramedullary plasmacytoma.[4] Extramedullary plasmacytoma is reported to be in the submucosa of soft tissues of upper aerodigestive tract in 80% of cases.[5] Neoplasms originating from plasma cells are rare in the head and neck region.[6] Myeloma accounts for 1% of all malignancies and slightly more than 10% of all hematologic malignancies.[7] The annual incidence of newly diagnose cases in USA is 3-4 per 1,000,000 population.[8] Usually seen in age group 50-70 years with noted higher incidence in men.[9] We aim from this report to present our case of MM presented with absolute dysphagia, and points out few diagnostic challenges to alert treating physicians on this rare condition.

Case Report. A 77-year-old Indian male pilgrim presented to Otolaryngology - Head & Neck Surgery clinic during Hajj time at Al-Noor Specialist Hospital, Makkah, Saudi Arabia with 3 days history of severe dysphagia to solids and liquids. Flexible laryngoscopy showed diffuse supraglottic swelling with ill-defined epiglottic and hypopharyngeal mass. On initial assessment, hemoglobin was 7.3 gm/dl (normal 13-17), red blood cell 2.83 106/µl (normal 4.3-5.7), platelets 107 10^3/µl (normal 150-450), calcium 8.7 mg/dl, urea 167 mg/dl (normal 7-20), creatinine 2.2 mg/dl (normal 0.5-1.5), albumin 1.81 gm/dl (normal 3.2-5.0), total protein highly elevated 902 g/L (normal 60-85), erythrocyte sedimentation rate test >150 mm/hour (normal 1-25). The CT scan of the neck showed bulky epiglottis with ill-defined lesion in its right side and supraglottic larynx (**Figure 1**). Skull x-ray showed multiple lytic lesions (**Figure 2**). Multiple myeloma was suspected. Urinary Bence John protein was negative, serum immunoglobulin IgG was high 1894.4 mg/dl (normal 760-1550mg/dl), IgM and IgA were normal. Serum protein electrophoresis showed low albumen 28.6 (normal 53-65) and high Gamma globulin 45.5 (normal 12-22). Bone marrow biopsy showed a picture consistent with MM (**Figure 3**). He was started on IV steroids (Dexamethasone, IV, 8 mg, TID) and nasogastric tube feeding (Yenilmez medical, Istanbul, Turkey). His dysphagia responded well to therapy and was able to tolerate oral intake at time of discharge. As

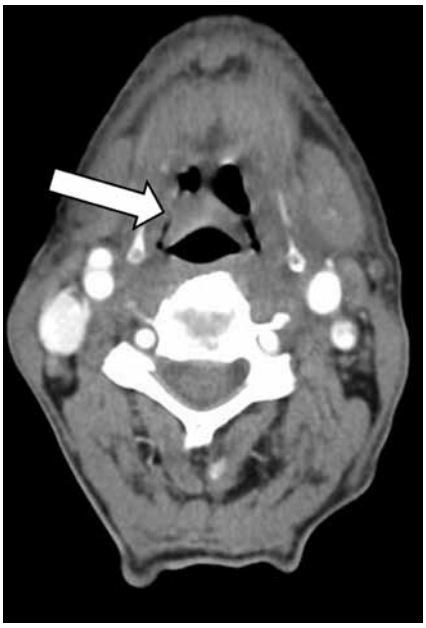

Figure 1 - Computed tomography scan of the neck showed bulky epiglottis with ill-defined lesion in its right aspect, ill-defined mass in the left side of supraglottic larynx.

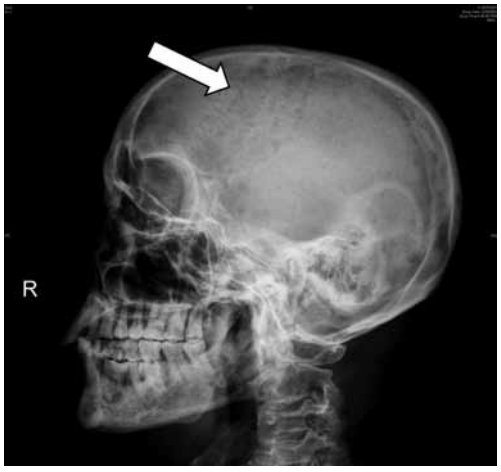

Figure 2 - Skull x-ray showed multiple lytic lesions.

the patient was a pilgrim he elected to go back to India with all relevant medical reports to continue his therapy and there was no further follow up.

Discussion. Malignant plasma cells in the bone marrow produce monoclonal antibodies which are typically found in urine and blood samples.[1,10] Multiple myeloma classically present with lytic lesions on bones, anemia, decreased renal function, and susceptibility to infections.[10] Those characteristic lytic lesions are due to the increased osteoclastic activity from bone

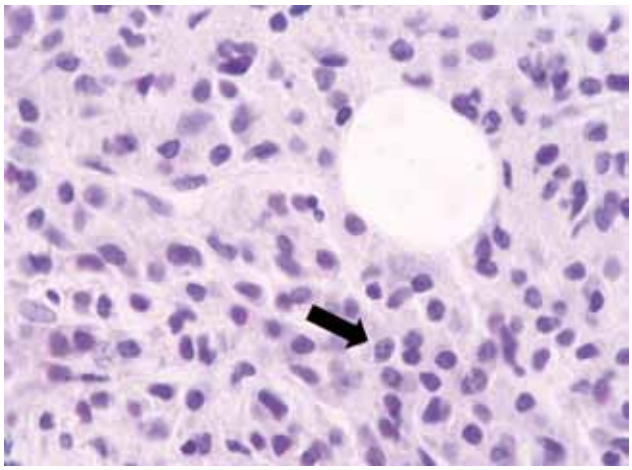

Figure 3 - Hematoxylin and eosin stain of bone marrow biopsy showing massively infiltrated bone marrow by plasma myeloma rounded tumor cells with large nuclei and nucleoli with prominent chromatin.

marrow infiltration.[10] Less than 100 cases of laryngeal EMP have been reported in the world literature.[2,4] Two reports from India reported a similar case like ours of laryngeal plasmacytoma.[2,9] Another report from Poland described a similar case where some therapeutic options were discussed.[4] Tesei et al from Italy reported 22 cases of EMP of the head and neck observed over 20 years period; of which only 2 cases were laryngeal.[5] The cause of MM is unknown. However, it was noted that exposure to ionizing irradiation might increase the incidence of MM.[1,10] Nickels, agricultural chemicals, petroleum products, aromatic hydrocarbon, benzene, and silicon have been also considered a potential risk factors.[10] Treatment options have increased significantly since the last 2 decades.[10] Melphalan plus prednisone has been the gold standards for the treatment of this condition for the last 40 years.[10] Salvage therapy for relapses or primary refractory disease usually undergo vincristine plus doxorubicin plus dexamethasone (VAD) regimen. This produces 40-50% response in relapses and 30% in primary refractory disease.[10] The most active agent in the combination therapy is known to be dexamethasone.[10] High dose dexamethasone pulse therapy alone induces responses in approximately 30-50% of patients, regardless of prior response.[10] The effectiveness of immunomodulatory agents such as thalidomide and lenalidamide, as well as bortizomib; a proteosome inhibitor, has greatly expanded the treatment options.[10] Localized external beam radiotherapy has also been used successfully.[8]

In conclusion, the incidence of extramedullary manifestation of MM in the larynx is rare, but many

therapeutic options exist. We recommend a high index of clinical suspicion to exclude this rare possibility while investigating a mass lesion in this region to avoid diagnostic and therapeutic delays.

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