Concurrent Extramedullary Plasmacytoma and Amyloidosis of the Waldeyer's Ring Extramedullary Plasmacytoma and Amyloidosis of the Waldeyer's Ring

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Abstract

Primary amyloidosis and extramedullary plasmacytoma (EMP) of the head and neck are rare conditions. Their occurrence in the same area is especially unusual, with only one case reported in Waldeyer's Ring (WR). We present a 61-year-old man with concurrent EMP and amyloidosis of the WR. He had been treated for obstructive sleep apnea (OSA) elsewhere and was referred to our clinic with progressive hoarseness, dysphagia, and right-sided hearing loss. Examination and imaging showed a mass in the nasopharynx that extended into the right nasal airway and palatine tonsil. A punch biopsy indicated amyloidosis. The patient then underwent partial nasopharyngectomy through a transpalatal approach, and the mass, including the right palatine tonsil, was completely removed. Final histopathology confirmed amyloidosis along with coexisting EMP. Systemic disease was ruled out, confirming localized pathology, and the patient received adjuvant radiotherapy. This case shows the very rare coexistence of amyloidosis and EMP in Waldeyer's Ring. It emphasizes the importance of thorough endoscopic and radiological evaluation in patients with OSA-like symptoms or unexplained head and neck masses. Although these conditions are uncommon, they should be included in the differential diagnosis.

Keywords: amyloidosis, Waldeyer's ring, extramedullary plasmacytoma, nasopharynx, tumor.

Introduction

Primary amyloidosis of the head and neck region is a very uncommon clinical entity. The most frequently involved site of the upper respiratory system is the larynx.[1]

Extramedullary plasmacytoma (EMP) is a localized monoclonal plasma cell tumor that arises in tissues other than the bone marrow and tends to occur during the fifth and seventh decades of life. These monoclonal plasma cells may sometimes secondarily lead to amyloid deposition.[2]

Waldeyer's Ring (WR) is composed of pharyngeal and tubal tonsils in the nasopharynx and palatine and lingual tonsils in the oropharynx.[3] EMP or amyloidosis in WR is rare, with only one reported case of both occurring together.[4] In this report, we present a 61-year-old patient with a concurrent EMP and amyloidosis of WR, which to our knowledge is the second case in English literature.

Case Report

A 61-year-old male initially presented to a sleep clinic with severe obstructive sleep apnea (OSA) symptoms in May 2023. He underwent polysomnography and was diagnosed with OSA and treated with continuous positive airway pressure (CPAP). One month after starting treatment, he developed new symptoms, including hoarseness, difficulty swallowing, and hearing loss in his right ear. The patient was then referred to our clinic for further investigation in June 2023.

On clinical examination, the right palatine tonsil was covered with an irregular yellowish polypoid mass. Flexible nasopharyngoscopy revealed the origin of the mass to be in the nasopharynx, showing a right-sided dominance, with inferior extension to the right lateral pharyngeal band and right palatine tonsil. The mass also extended anteriorly to the right posterior nasal airway (Figure 1). Otoscopy showed right otitis media with effusion. Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI) clearly revealed an inhomogeneous mass in the nasopharynx extending anteriorly to the right posterior nasal passage and inferiorly to the right side of the oropharynx (Figure 2). There was no lymphadenopathy at the cervical levels on CT and MRI.

A punch biopsy was performed endoscopically from the nasopharynx, and histopathological evaluation demonstrated the presence of deposits, stained by Congo red and identified as amyloidosis when the characteristic "apple-green" birefringence was obtained with polarized microscopy.

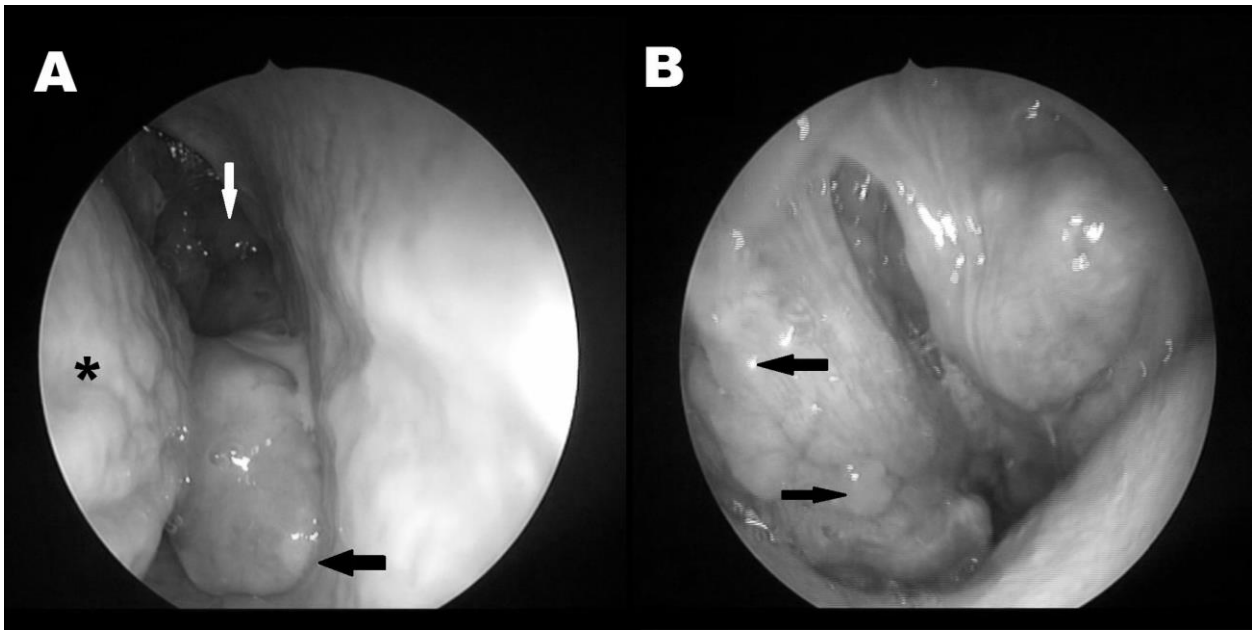

Figure 1: A) Endoscopic image of the nasopharynx. Black arrow: Anterior extension into the right posterior nasal passage. White arrow: Nasopharynx. Asterisks: Right inferior turbinate. B) Endoscopic image of right palatine tonsil. The enlargement of the right palatine tonsil is clearly visible, and the black arrows indicate amyloid deposits.

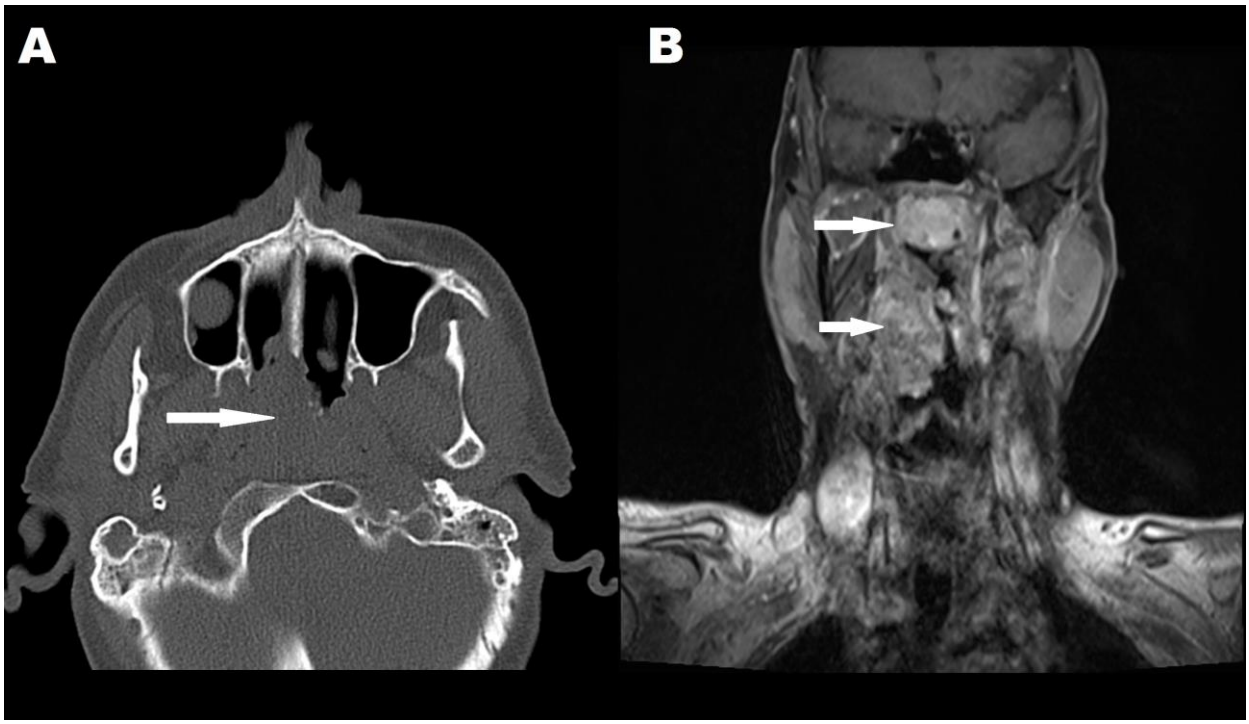

Figure 2: A) Axial CT scan shows an inhomogeneous mass located in the nasopharynx. White arrow indicates its anterior extension into the right posterior nasal airway. B) Contrast-enhanced coronal T1-weighted fat-sat MRI shows moderate-to-marked enhancement of the lesion. White arrows indicate the mass in the nasopharynx and right palatine tonsil.

A partial nasopharyngectomy via transpalatal approach was performed, and the mass, including the right palatine tonsil, was completely excised, the left palatine tonsil remained untouched. Postoperative histopathology revealed plasmacytoma in both nasopharyngeal and tonsillar parts of the mass, in addition to previously diagnosed amyloidosis. The histiocytes stained strongly for CD138 in addition to the presence of mature lambda monoclonal cells.

No complications occurred postoperatively, and the patient was referred to a hematology specialist to investigate for the presence of systemic amyloidosis and multiple myeloma. Bone marrow biopsy, urine/serum protein electrophoresis and immunofixation, and blood chemistry profiles showed no evidence of multiple myeloma. An abdominal fat biopsy was also performed for systemic amyloidosis and was negative.

The patient received postoperative radiotherapy with 50 Gy to the primary area and 45 Gy to the bilateral neck, for a period of 25 days. All symptoms, including sleep apnea, completely resolved after treatment and did not recur. At one-year follow-up, no residual disease was observed in the nasopharynx or tonsillar fossa.

Discussion

The larynx is the most common site of localized amyloidosis in the upper respiratory system; however, amyloidosis of the nasopharynx, oropharynx, and other sites in the head and neck have also been reported in different cases.[5] There are only three reported cases of amyloidosis of WR.[6]

Symptoms depend on the site of origin, but localized amyloidosis has a good prognosis and usually does not have systemic manifestations. Although usually seen in the 5th and 7th decades of life, there are some case reports of localized amyloidosis of the head and neck, seen in children.[7] EMP accounts for 4% of all non-epithelial tumors of the upper respiratory tract. They arise in the nasopharynx, sinuses, oropharynx, and tonsils. Symptoms vary from epistaxis, nasal discharge, sore throat, and hoarseness to dysphagia. The presence of multiple myeloma must be investigated.[8]

Amyloidosis and EMP are closely related through the abnormal production of monoclonal immunoglobulins by clonal plasma cells. EMP, like other plasma cell neoplasms, is often associated with the production of abnormal monoclonal immunoglobulins, either as intact molecules or as fragments such as light or heavy chains. These abnormal proteins may be detected in the blood or urine, or may accumulate within tissues, occasionally forming amyloid deposits.[9] In respiratory plasmacytomas, amyloid deposition is believed to happen more easily because of the high local levels of monoclonal immunoglobulins around tumor cells, providing a reasonable explanation for the development of amyloidosis.[4] Previous reports of head and neck EMP complicated by localized amyloidosis are exceedingly rare, with isolated cases described in the larynx and nasopharynx.[10] Michaels et al. observed amyloid deposits in 14 of 59 upper respiratory plasmacytomas, though only a single case involved WR. These observations reinforce the link between EMP and amyloidosis and highlight the importance of considering amyloid deposition in EMP lesions.[4]

Surgical removal is suggested to be the treatment of choice for localized amyloidosis.[6] Treatment for EMP, on the other hand, relies more on radiotherapy than surgery, since EMP is highly radiosensitive. That being said, a combination of radiotherapy and surgery seems to be the best logical treatment option.[8,11]

Our case is the second reported case of EMP and amyloidosis seen together in WR. However, the previous case was reported nearly 40 years ago, without our current radiological and endoscopic opportunities, so the details and extent of the lesion were not described in detail. So it would be fair to say that this is the first case that is diagnosed and discussed more thoroughly.

Conclusion

Concurrent EMP and amyloidosis is extremely rare. There are reports of single cases of this clinical concomitance, detected in the larynx and nasopharynx. This case highlights that patients with atypical OSA symptoms, that do not improve with standard treatment, should undergo an extensive endoscopic upper respiratory tract examination and, when indicated, radiological evaluation. Even though they are both extremely rare clinical conditions, EMP or amyloidosis should also be considered in the differential diagnosis of the head and neck masses.

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