CaseReport

An Unexpected Cause of Hoarseness of Voice in a Healthy Teenager

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Abstract

Laryngeal amyloidosis is a rare cause of stridor in a healthy young adult. We report a case of localised laryngeal amyloidosis, including our MRI findings, which included a necrotic centre that has not previously been described. This case also highlights the need for a high index of clinical suspicion to achieve the correct histopathological interpretation.

Keywords: *medical sciences*

Introduction

Primary amyloidosis is an unusual entity, occurring without any pre-existing primary disease. Isolated laryngeal amyloidosis is even more uncommon, accounting for less than 1% of all benign laryngeal tumours. Only two hundred such cases are reported in the literature. Descriptions of the MRI appearance of primary laryngeal amyloidosis are scarce in the literature, and the presence of a necrotic centre has not previously been described.

Case Report

RM, an 18-year-old Malay boy, presented with hoarseness of voice for a duration of one year prior to presentation. It was also associated with stridor, dysphagia, and intermittent fever. Indirect laryngoscopy revealed a polypoid mass arising from the left side of the hypopharynx and oropharynx. The lesion was excised by laser knife surgery guided by laryngoscopy under general anaesthesia. The histopathological examination results were interpreted as respiratory papillomatosis.

The patient remained asymptomatic for a year before similar symptoms recurred. During the second presentation, an MRI was done, revealing lobulated submucosal lesions involving the posterior wall of the right oropharynx, right aryepiglottic fold, vallecula, and paralaryngeal spaces bilaterally down to the hypopharynx at the level of false cord. The lesions were isointense to the muscle on T1WI and slightly hyperintense compared to the surrounding muscle on T2WI (Figure 1). Following contrast administration, there was avid but patchy enhancement of these lesions (Figure 2). There was a cystic area in the right pyriform fossa that did not show enhancement. The appearance of the non enhancing cystic paralaryngeal lesion most likely represents a necrotic centre (Figure 3). True cord and subglottic larynx were normal. Based on MRI analysis, the probable diagnosis of recurrent juvenile papillomatosis in the right oro- and hypopharynx was made which has caused localised narrowing of the laryngeal vestibule.

A chest X-ray done during the second presentation did not show any evidence of luminal narrowing involving the tracheobronchial tree.

The initial diagnosis of laryngeal papillomatosis was doubted by the primary managing team, as the age of presentation was atypical. The pathologist reviewed the slides from the original biopsy sample, which showed the presence of amorphous eosinophilic material in the stroma that was positive for amyloid after Congo red staining, consistent with a diagnosis of amyloidosis.

Discussion

Amyloid is derived from the Greek words amylon, meaning “starch”, and eidos, meaning “resemblance”. The clinical presentation encompasses a wide spectrum, ranging from a potentially lethal condition or merely an incidental finding. The symptoms depend on the site affected and the magnitude of the amyloid deposition (1). Amyloid is composed of pathological proteinaceous material deposited in between cells of various tissues. It can occur in variety of clinical conditions (1). Consequently,

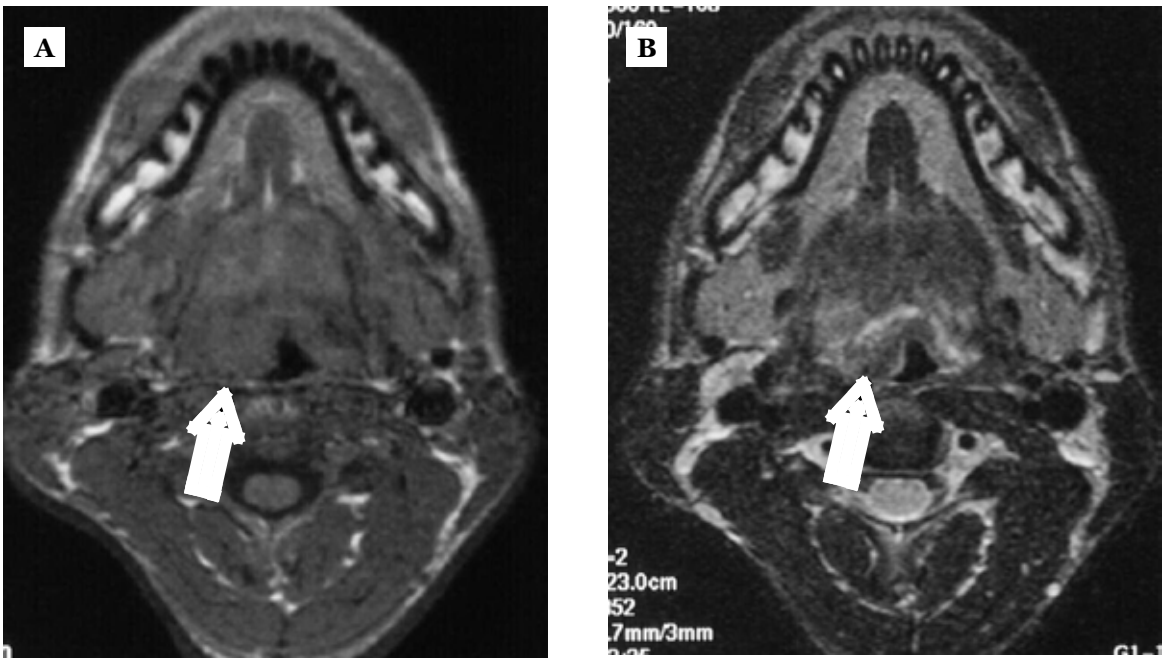

Figure 1: Axial images showing a submucosal lesion involving predominantly the right aryepiglottic fold, which is isointense on T1WI (A) and has intermediate signal intensity on T2WI (B).

amyloidosis should be regarded not as a single disease but as a group of diseases that exhibit tissue deposition of protein that displays a similar histological appearance (1). Under light microscopy with standard tissue staining, amyloid deposits will appear as an amorphous, hyaline, eosinophilic extracellular material. The material may appear to be encroaching on or producing pressure effects on the adjacent cells (1).

Amyloidosis can be categorised based on its biochemical-clinical manifestation (1). In this classification scheme, amyloidosis is divided into systemic amyloidosis and localised amyloidosis. Clinically, systemic amyloidosis can be subclassified as a primary or secondary lesion (1). Primary amyloidosis is associated with immunocyte dyscrasia, whereas secondary amyloidosis is associated with chronic inflammation or tissue destruction (1). Localised amyloidosis is limited to a single organ without involvement of other sites. This case is an example of localised amyloidosis occurring at the larynx. No amyloid-associated disease was detected in this patient.

The respiratory tract may be affected diffusely or focally from the larynx to the smallest bronchioles (1). In the larynx, amyloidosis may appear as a deposition of acellular eosinophilic material infiltrating the connective tissue stroma (in a haematoxylin- and eosin-stained section) and forming a nodular lesion. With increasing amount of amyloid deposition, pressure effect exerted may render the seromucinuous glands to disappear while the covering epithelium remains intact (2). Frequently, mononuclear inflammatory cells can be seen at the periphery of the amyloid material. This may correspond to a localised form of immunocyte-derived amyloid (1).

The diagnosis ultimately depends on identification of this proteinaceous material in appropriate biopsy specimens (1). The amyloid deposits may be confused with other hyaline material, such as collagen or fibrin (1). Histological diagnosis of amyloidosis is best supported by Congo red immunochemical staining, which imparts a pink or red colour to amyloid deposits under ordinary light and produces a green birefringence under crossed polarised light. This appearance is shared by all types of amyloid material due to the cross-β-pleated configuration of the amyloid fibrils (1). In the present case, the histopathological evaluation during the first presentation may have missed the amyloid material in view of the low clinical suspicion of amyloidosis and scanty presence of amyloid material. As amyloid deposition in the upper respiratory tract is generally submucosal in nature, it may form a polypoid lesion. On histopathological examination, the polypoidal lesion caused by amyloid deposition is lined by stratified squamous epithelium, resembling the epithelial lining seen in polyps due to papillomatosis.

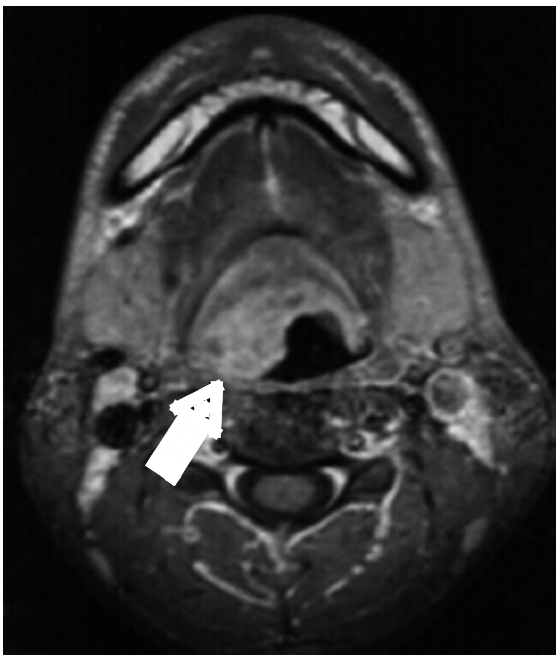

Figure 2: Axial T1WI with gadolinium showing avid but patchy enhancement of the lesions involving the aryepiglottic folds.

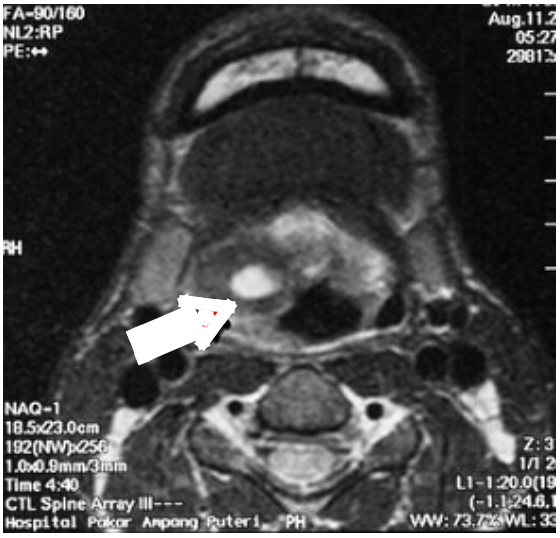

Figure 3: T2WI showing the focal area of high signal intensity within the lesion in the right paralaryngeal space; this is likely to represent necrotic tissue.

Laryngeal amyloidosis was first documented in 1875 as reported by Fernandes et al. (3). It is rare, accounting for less than 1% of all benign laryngeal tumours. Only over two hundred cases have been reported in the literature. Laryngeal amyloidosis occurs most commonly in the fifth decade of life with male predominance (4,6). In laryngeal amyloidosis, disease is usually localised to the larynx, and the aetiology is usually primary (4). The ventricles and false vocal folds are affected most frequently (4,5).

Few descriptions are available in the literature regarding the MRI features of laryngeal amyloidosis (4). To our knowledge, there are also no previous reports of laryngeal amyloidosis with necrotic centre. On T2-weighted images (T2WIs), the lesion may appear to have a slightly higher signal than the surrounding muscles, as seen in this case, but it may also appear hypointense relative to the surrounding muscles (4). On T1WI, lesions are similar in intensity to the peripheral muscles, showing marked contrast enhancement (4,6). Therefore, imaging is non-specific, and its role is mainly to evaluate the extent of the disease and to rule out other differential diagnoses (6).

The therapy of choice is local excision (i.e., CO_2 laser resection) (4). Amyloidosis is a benign process. Nonetheless, it can be progressive or can recur after treatment. Regular follow-up with MRI is thus recommended, as its recurrent nature may require repeated surgical interventions (7).

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