VOL. 17|NO. 3|ISSUE 67|JULY-SEPT. 2019

Primary Laryngotracheal Amyloidosis Masquerading as Malignancy

Devkota K, Adhikari K, Poudel S

Department of Radiodiagnosis and Imaging

BP Koirala Institute of Health Sciences,

Dharan, Nepal.

Corresponding Author

Karun Devkota

Department of Radiodiagnosis and Imaging,

BP Koirala Institute of Health Sciences,

Dharan, Nepal.

E-mail: karundevkota@gmail.com

Citation

Devkota K, Adhikari K, Poudel S. Primary Laryngotracheal Amyloidosis Masquerading as Malignancy. *Kathmandu Univ Med J.* 2019;67(3):254-5.

ABSTRACT

Amyloidosis represents a heterogeneous group of disorders characterized by the deposition of amyloid protein in various target organs of the body. Amyloidosis is classified as systemic or local. Larynx is the common site for localized amyloid deposition and patient usually presents with hoarseness of voice. Because of the similar clinical and imaging features to malignant laryngeal mass, diagnostic confusion occurs and histopathological confirmation is usually needed. Although laryngeal tracheo-bronchial amyloidosis is rare, it should be considered among the differentials of benign laryngeal tumors.

KEY WORDS

Amyloidosis, Laryngotracheal, Malignancy

INTRODUCTION

Amyloidosis represents a heterogeneous group of disorders characterized by the deposition of amyloid protein in various target organs of the body.[1] Amyloid deposition in the larynx is rare, accounting for between 0.2 and 1.2% of benign tumors of the larynx.[2] Amyloidosis is classified as systemic or local. Localized amyloidosis affecting the respiratory tract is subdivided into laryngeal, tracheobronchial, and parenchymal categories.[3] Amyloid in the larynx, trachea and bronchus can be identified as subepithelial extracellular deposits of acellular, homogeneous and amorphous, eosinophilic material displaying apple-green birefringence with polarized light when stained with Congo red or that is metachromatic with crystal violet or methyl violet.[2] Laryngo-tracheobronchial amyloidosis can cause diagnostic confusion because of the similar clinical and imaging features to malignant mass and often needs histopathological examination for confirmation.

CASE REPORTS

A 50 year male presented with hoarseness of voice for which USG neck was suggested which showed an ill defined heteroechoic area in midline of neck at the level of vocal cord which was outlined by strap muscles anteriorly for which further imaging study was suggested. Patient subsequently underwent contrast enhanced CT examination which revealed irregular nodular enhancing soft tissue density lesion involving long airway segment extending from the level of C5 with transglottic involvement and reaching upto the level of carina in its inferior extent with few skip areas in between and causing luminal narrowing (fig. 1 and 2). Few coarse calcification were also noted in right lateral wall of trachea. On the basis of imaging, impression of laryngo-tracheal mass was given with advice for histopathology to rule out malignancy. Patient underwent tracheostomy under local anesthesia and biopsy was done under general anesthesia. The histological features were suggestive of laryngeal amyloidosis with apple green birefringence on congo red staining of deposits on polarized microscopy.

The patient was also investigated for signs of systemic amyloidosis. Haematological parameters , liver function tests, renal function tests were all within normal range for his age and gender.

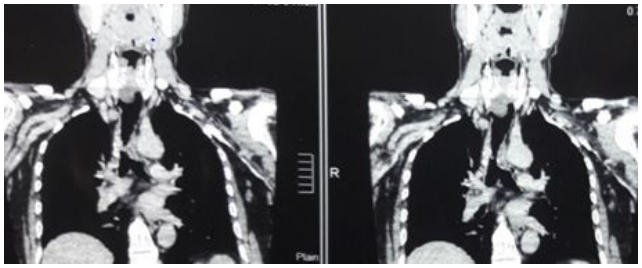

Figure 1. Coronal CT images show nodular thickening involving supraglottis, glottis, subglottis and trachea with skip area in between.

DISCUSSION

Virchow described amyloid in 1854 as starch like substance. In 1935, Reimann, Kouky and Ekund calssified amyloid as primary, secondary, tumor-forming and amyloidosis assosciated with myelomatosis.[4]

Though occurrence is rare in larynx, it is the most common site for isolated amyloid deposits to occur in the head and neck.[5] Laryngeal amyloidosis can present in a variety of ways. Patients may present with hoarseness, dysphagia, dyspnoea and stridor. Hoarseness has been reported as a frequent presentation.[1]

In localized amyloidosis, amyloid protein deposition is restricted to a single organ. Two theories account for the amyloid deposition in the target organ. First, the synthesis of amyloid within the target organ produces the proteins locally resulting in single organ involvement. Second, the

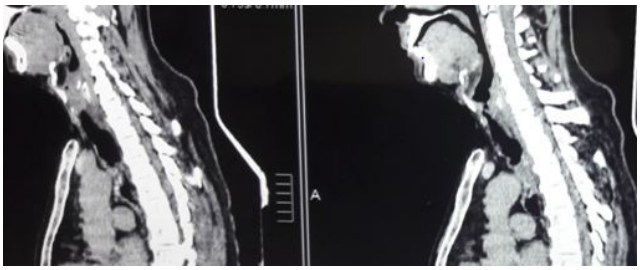

Figure 2. Sagittal CT images show nodular thickening involving supraglottis, glottis, subglottis and trachea with skip area in between.

distant or systemic light chains produce amyloid which gets deposited in the target organ due to a localized pathological process such as inflammation.[6]

On CT, it usually appears as well defined homogeneous soft tissue density submucosal mass with variable enhancement. Calcification may be seen. Bone erosion or destruction and lymphadenopathy are usually absent.[7] When it involves the trachea, the posterior membrane is also classically involved.[8]

MRI is considered to be a more specific technique than CT scanning since amyloid deposits present as an intermediate T1-weighted signal intensity and low T2-weighted signal intensity.[9] Unfortunately it was not done in our case as high strength MRI was unavailable in our institute.

Although laryngeal tracheo-bronchial amyloidosis is rare, it should be considered among the differentials of benign laryngeal tumors. Complete clinical examination and laboratory investigations are also of great value to exclude systemic involvement since treatment and prognosis differs markedly.

REFERENCES

1. Saleem T, Fatima S, Ghaffar S, Aftab K, Awan S. Two cases of primary laryngeal amyloidosis. *Journal of the Pakistan Medical Association*. 2009;59(11):789. https://www.ncbi.nlm.nih.gov/CBBresearch/Lu/Demo/PubTator/index.cgi?user=bc5cdr&searchtype=PubMed_Search&query=23914645[relatedto]&page=2&tax=
2. Thompson LD, Derringer GA, Wenig BM. Amyloidosis of the larynx: a clinicopathologic study of 11 cases. *Modern Pathology*. 2000 May;13(5):528. https://www.ncbi.nlm.nih.gov/pubmed/10824924
3. Ozyigit LP, Kiyan E, Okumus G, Yilmazbayhan D. Isolated laryngo-tracheal amyloidosis presenting as a refractory asthma and longstanding hoarseness. *Journal of Asthma*. 2009 Jan 1;46(3):314-7. https://www.ncbi.nlm.nih.gov/pubmed/19373642
4. Symmers WS. Primary amyloidosis: a review. *Journal of clinical pathology*. 1956 Aug;9(3):187. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1023944/
5. Behnoud F, Baghbanian N. Isolated laryngeal amyloidosis. *Iranian journal of otorhinolaryngology.* 2013;25(70):49. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3846235/
6. Chow V, Gardner K, Howlett D. Primary localized laryngeal amyloidosis presenting with dysphonia: a case report. *Journal of surgical case reports.* 2012 Nov 1;2012(11):rjs005. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3852952/
7. De Foer B, Hermans R, Feenstra L, Baert AL. CT and MRI of laryngeal amyloidosis. *InRöFo-Fortschritte auf dem Gebiet der Röntgenstrahlen und der bildgebenden Verfahren.* 1993 Nov;159(11):492-494). © Georg Thieme Verlag Stuttgart• New York. https://www.ncbi.nlm.nih.gov/pubmed/8219149
8. Yamamoto T, Maeda M, Mizuno S, Saga T, Itoh H. Primary diffuse tracheobronchial amyloidosis: radiologic findings. *Journal of thoracic imaging.* 2001 Jul 1;16(3):177-80. https://www.ncbi.nlm.nih.gov/pubmed/11428418
9. Yiotakis I, Georgolios A, Charalabopoulos A, Hatzipantelis P, Golias C, Charalabopoulos K, Manolopoulos L. Primary localized laryngeal amyloidosis presenting with hoarseness and dysphagia: a case report. *Journal of medical case reports.* 2009 Dec;3(1):9049. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2767149/