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Original Research Paper

Primary amyloidosis of vocal cord masquerading as malignancy- a rare presentation

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ABSTRACT-

Objective- To study role of histopathological examination for confirmation of Primary Amyloidosis of vocal cord and to differentiate the same from malignancy. **Case report**- Here, we present a case of isolated laryngeal amyloidosis in adult female which mimics malignancy clinically. Amyloidosis is gradual in onset and has unpredictable manner. It is a benign, heterogeneous, multisystem organ disorder affecting many tissues and organs. More commonly it affects kidney, Spleen, Liver, Heart, tongue along with many endocrine organs. However, Primary involvement of larynx is a very rare phenomenon in clinical settings. Isolated amyloidosis of vocal cord is a rare entity in clinical practice with only 0.2 to 1.2% of benign tumors of larynx. **Conclusion-**Amyloidosis commonly mimics malignancy in clinical settings. Hence, it is mandatory to diagnose it accurately as it affects the treatment.

Key Words- Primary Amyloidosis, Vocal cord, Congo red, Isolated

INTRODUCTION:-Amyloid is a proteinaceous substance and its deposition within extracellular space is pathological and termed as Amyloidosis. It is deposited in various tissues and organs in a wide spectrum of clinical settings. Amyloid deposition has insidious onset and in an unpredictable manner. Hence, it's difficult to clench its diagnosis clinically and ultimately Histopathological examination with special stains is the mainstay for its accurate diagnosis. Isolated amyloidosis of vocal cord is a rare entity in clinical practice with only 0.2 to 1.2% of benign tumors of larynx.The patient usually present with change of voice and dysphonia. Rarely, stridor and even dysphagia can occur.

CASE REPORT:- A 26year old female presented to ENT OPD with chief complaints of Hoarseness of voice since past few years which recently got deteriorated. She underwent fibre optic laryngoscopy which revealed polyp on left sided vocal cord. The clinical suspicion of malignancy was raised. However, the mobility of vocal cords was maintained. The systemic examination was normal. The biopsy from polyp was taken. The histopathological examination was done which revealed massive stromal deposition of amorphous, acellular, pale, eosinophilic pluffy material giving cotton candy appearance (**Fig-1A&1B**). To confirm the nature of substance special stains were put. The substance show characteristic apple green birefringence with Congo red **(Fig-2)** on polarized microscopy which confirms it for Amyloidosis.(1) However, PAS stain (**Fig-3**) and silver methanamine (**Fig-4)** were negative. No evidence of invasion and dysplasia was seen.

DISCUSSION:–The amyloid material consists 95% of fibril proteins, with remaining 5% being the P component and other glycoproteins. Amyloidosis is a rare, benign entity with gradually progressive clinical course. It is characterised by deposition of abnormally folded fibrillar protein within extracellular space resulting in pressure atrophy of parenchyma. Its pathogenesis involves immunological mechanisms due to abnormal protein folding. Amyloidosis is classified as primary and secondary with further characterisation of Primary form into localised disease involving single organ and generalised disease with multiple organ and

tissue involvement. [2, 3] The isolated laryngeal involvement is very rare. The clinical presentation is usually with Hoarseness of voice, stridor and sometimes with dysphagia and compromised airway. The generalised form is associated with splenomegaly, lymphadenopathy and bone tenderness. [4, 5] Histopathological examination along with special stain is the gold standard for accurate diagnosis as it is difficult to diagnose clinically. Amyloidosis is a rare phenomenon. Isolated involvement of vocal cord is even rarer. [6, 7] Therefore complete work up to rule out secondary association is must. It can mimic malignancy in clinical scenario. Hence, Histopathological examination along with confirmation by special stains is mandatory in such cases for accurate diagnosis.

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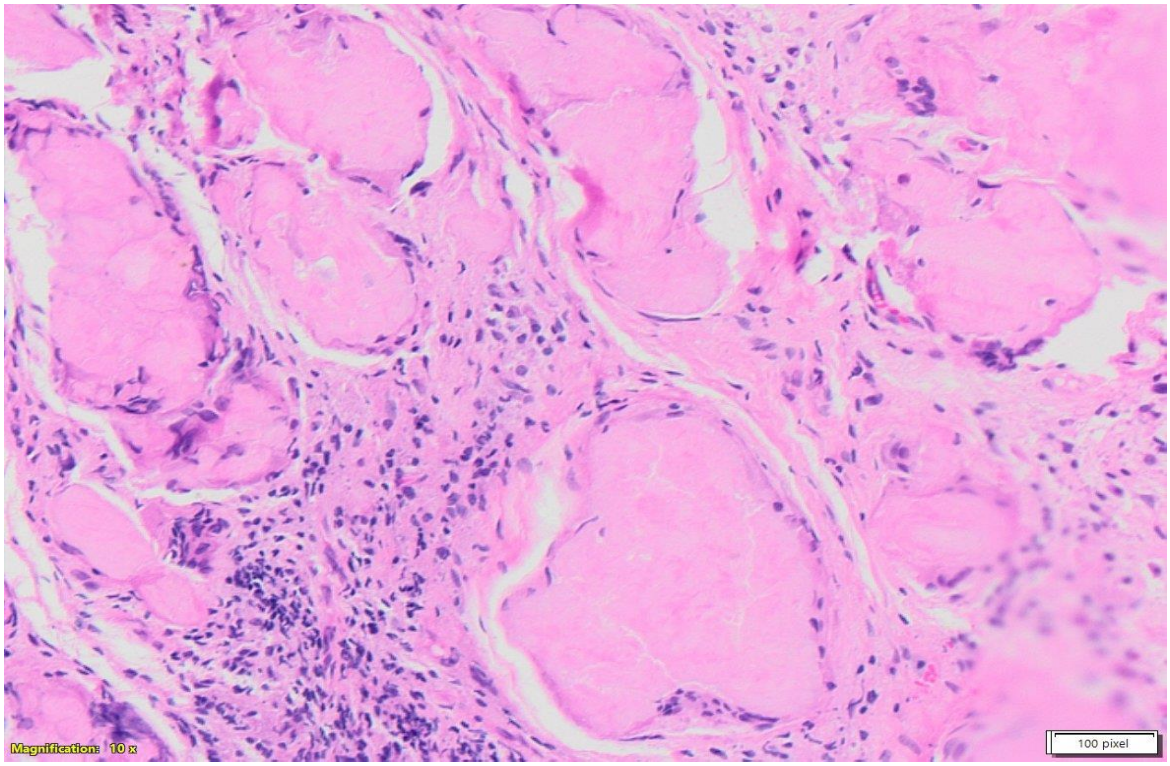
Figure Legends-

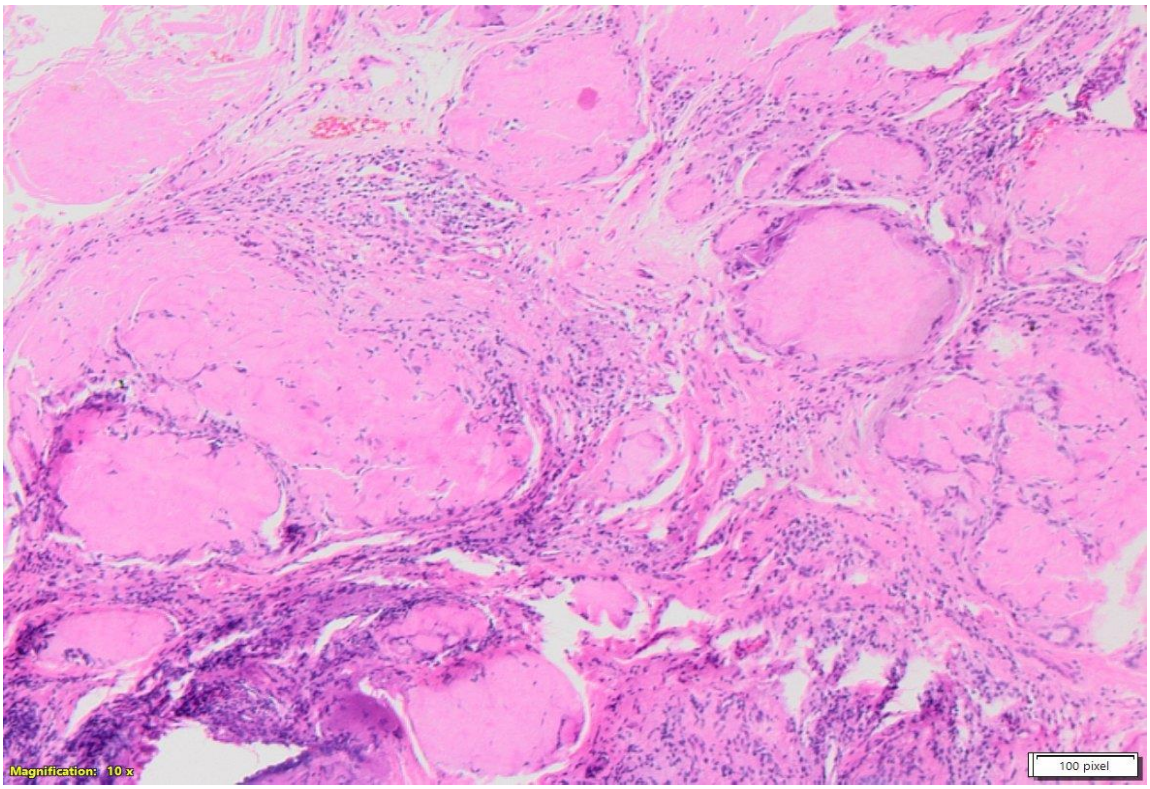
Figure 1: A) Amyloid deposition as eosinophilic, homogenous, Pluffy material (H and E, 10X)
And B) (H and E, 40X)

Figure 2: Congo red Staining showing apple green birefringence (40X)

Figure 3: PAS Staining Negative for Amyloid (10X)

Figure 4: Silver Methanamine Negative for Amyloid (10X)

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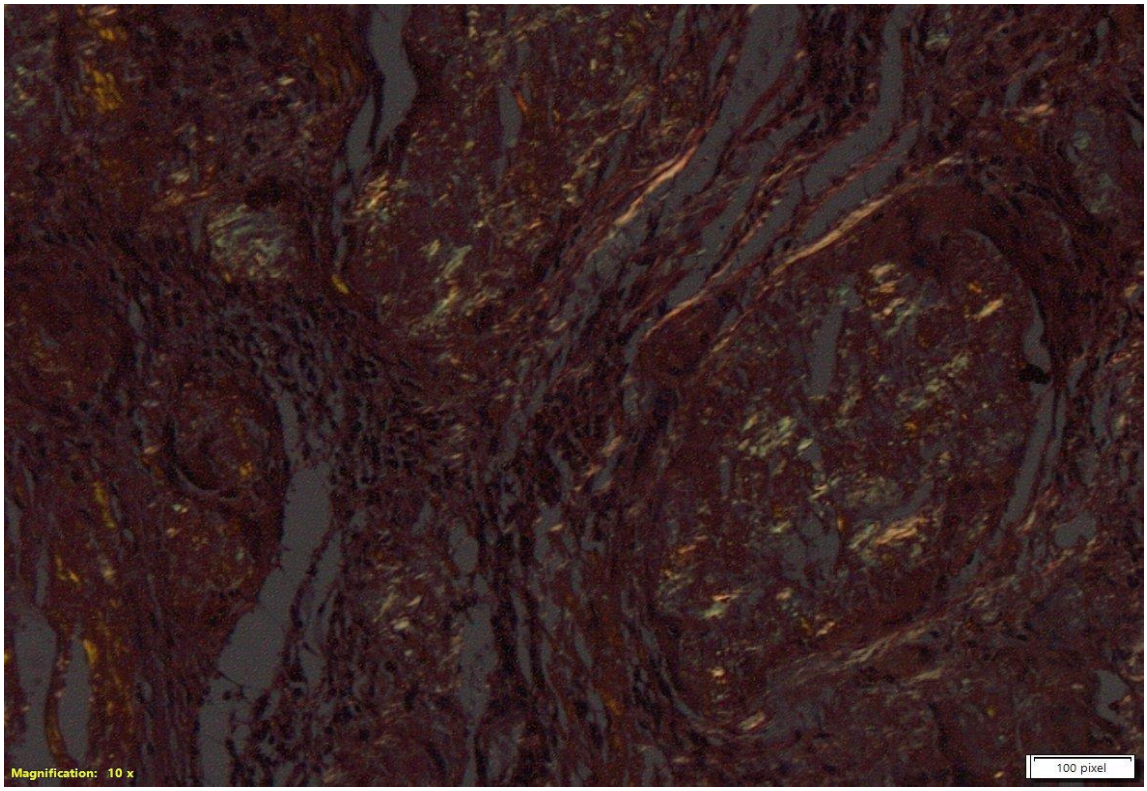
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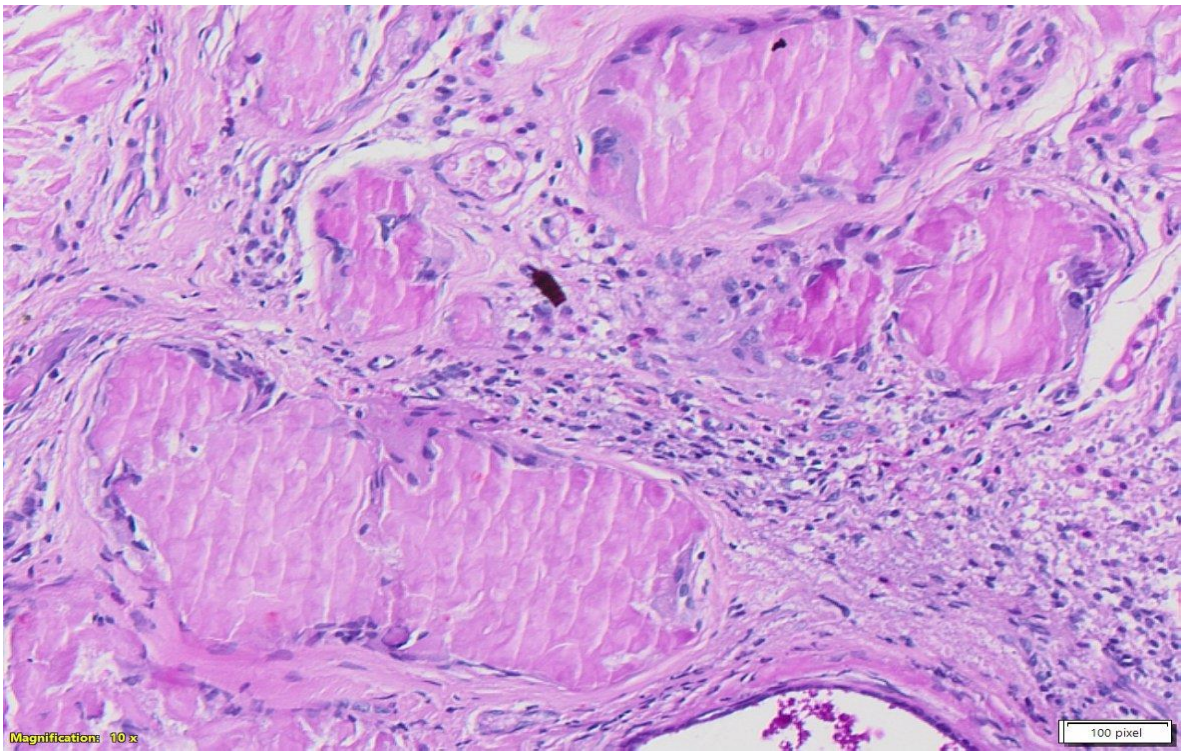
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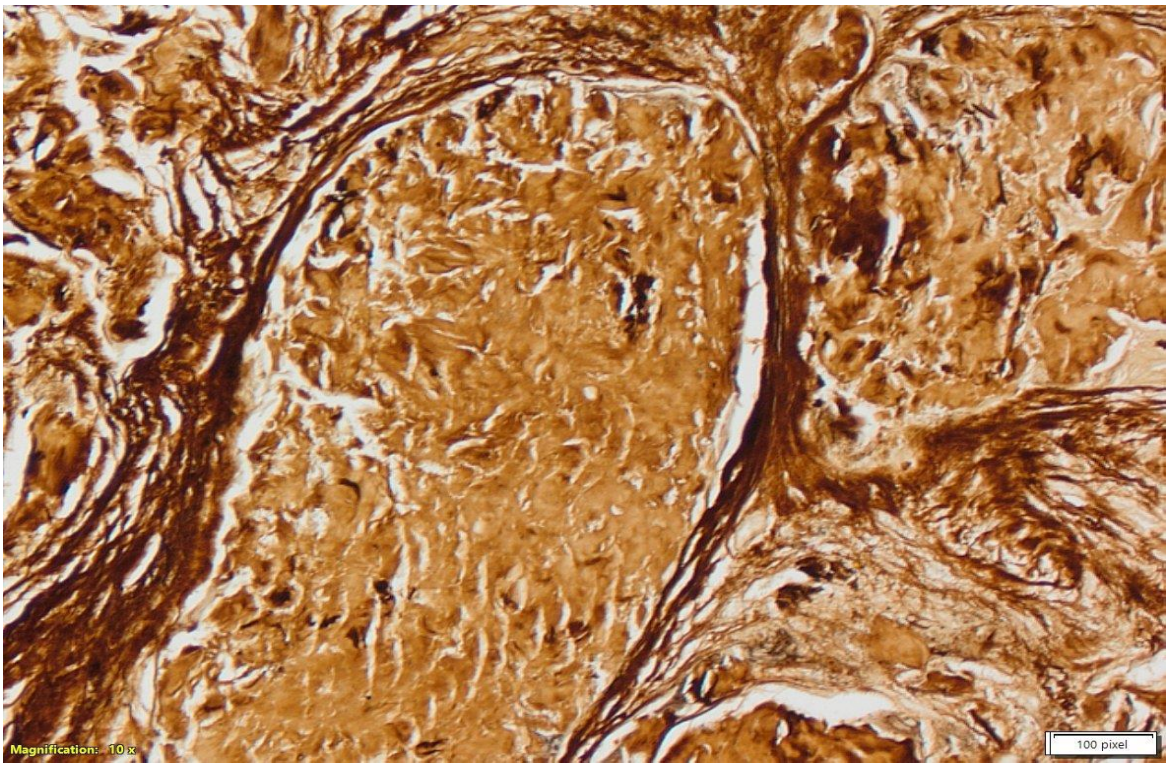
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