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A case of isolated amyloidosis of tongue

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Objective: Atypical presentation of a case of amyloidosis
Background: Amyloidosis is a rare disease in which the protein, amyloid, accumulates in organs such as the kidneys, heart, liver, spleen, nervous system and digestive tract. (10). Amyloid fibrils are protein polymers that contain identical monomer units (homopolymers). Functionally, amyloids are important in physiological processes such as long-term memory formation, where stored peptide hormones are gradually released. Amyloidosis results from extracellular accumulation of pathogenic amyloid and intracellular aggregation of misfolded proteins.
Case report: A 53 year old male came with complaints of macroglossia and difficulty in articulation.A trucut biopsy of the tongue was sent for histopathological examination and on Congo red stain "Apple green birefringence" was noticed confirming diagnosis of amyloidosis and on further investigations patient was diagnosed with Multiple myeloma. Patient is now on anti-neoplastic agents (Tab. Bortezomib) and is improving.
Conclusion: Early diagnosis and early treatment are very important for early control and management of Multiple myeloma.

Amyloidosis is a rare metabolic storage disorder with an incidence of 5 million per year. Among which isolated tongue amyloidosis contributes 9% overall with slight male preponderance.(1). Amyloidosis can be primary or secondary. Primary disease has unclear pathology. (3) The usual signs and symptoms include severe fatigue and weakness , shortness of breath, numbness, tingling, or pain in the hands or feet ,swelling of the ankles and legs, diarrhea, possibly with blood, or constipation, Skin changes, such as thickening or easy bruising, and purplish patches around the eyes. Oral manifestations are an enlarged tongue, which sometimes looks rippled around its edge, waxy yellowish papules or nodular lesions over tongue or buccal mucosa (2)

Disease may be suspected when organ enlargement , proteinuria and problems with peripheral nerves are seen . Confirmatory test is tissue biopsy and Congo red stain.Due to the variable presentation, a diagnosis may be delayed.The localized and systemic forms of disease have vastly different prognoses and therapeutic management.(8)

A 53 year old male was referred with bilateral submandibular region swelling for evaluation and management. Patient presented with breathing difficulty and slurring of speech which has worsened gradually over a period of 2 years. Patient had complaints of snoring without arousal episodes or sleep disturbances. The swelling in the submandibular region was noticed for approximately 3 months.

On examination, the tongue was very bulky. Bilateral submandibular glands were palpable . Submandibular region fullness disappeared on protrusion of tongue. There was no fall in saturation or any signs of respiratory distress. Patient was advised Fine Needle aspiration cytology from the submandibular gland region but was inconclusive.

On analysing all sections of CT Neck there was suspicion of amyloid deposition in the tongue which warranted further evaluation. Patient did not have any purpuric patches anywhere in the body nor any nodular swelling along the lateral border of the tongue. The only presenting feature was macroglossia, which is enlargement of tongue disproportionate to size of jaws (10). The ideal method to diagnose would be a biopsy and histopathological analysis. CT Neck was suggestive of a deeper deposit and a deeper tissue for biopsy from the tongue , which is a very vascular region, would be very difficult. So a USG guided trucut biopsy with the help of an Interventional radiologist was performed from the suspected area of the tongue which is not easily accessible by routine biopsy.

Biopsy report confirmed features of amyloidosis. The Congo red stain showed focal congophilic deposits with " Apple green birefringence" under polarized light which disappeared after KMNo4 treatment.

Patient was referred to a hematologist for further management. Protein electrophoresis showed 'M protein' spike in Gamma globulin region and M protein was - 0.38 g/dl. Bone marrow

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trephine biopsy was also suggestive of Plasma cell dyscrasia and Serum levels of Free Kappa and Free Lambda were 36.39 and 143.93 respectively. Patient is under Anti-neoplastic agent, a proteasome inhibitor (Bortezomib) as advised by hematologist . On follow up, the patient improved symptomatically after initiation of treatment.

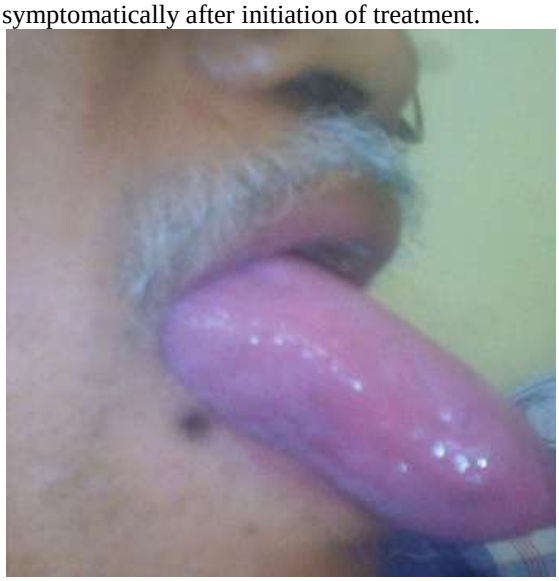

Figure 1 : Lateral view showing the bulky tongue

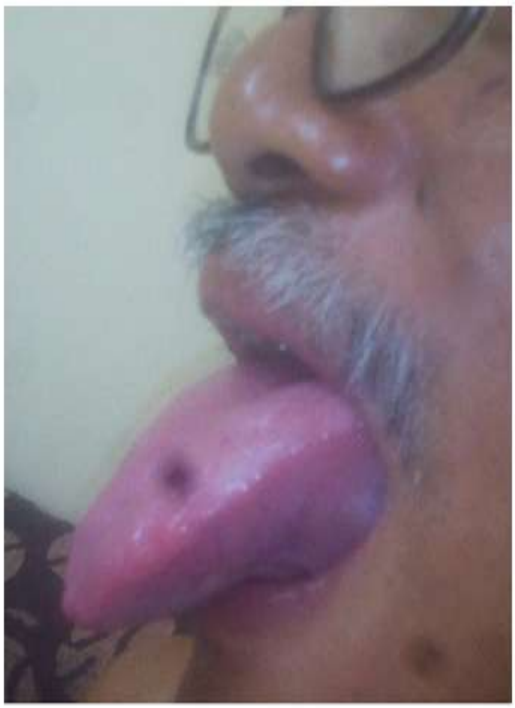

Figure 2: Lateral view depicting trucut biopsy site

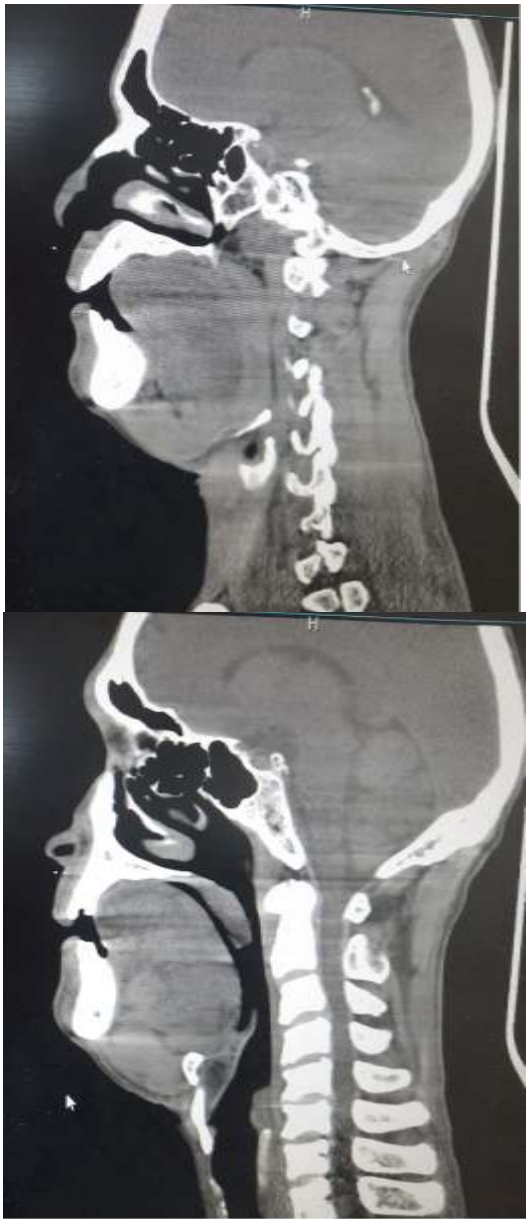

Figure 3: Sagittal sections of CT Neck (plain) showing the bulky tongue with suspicious amyloid deposits.

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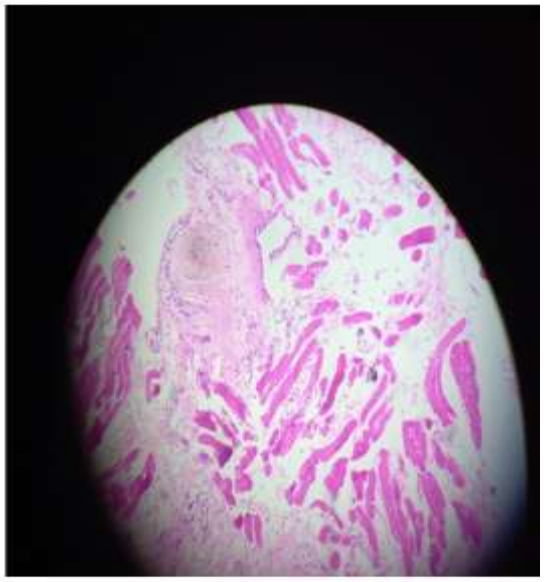

Figure 4: Amyloid showing eosinophilic staining on H&E

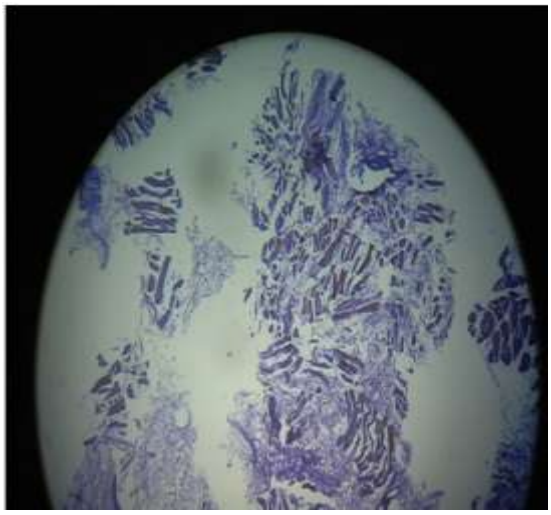

Figure 5 : Metachromatic staining wherein the blue colored crystal violet stains amyloid.

CONCLUSION:

Awareness and knowledge of amyloidosis and its atypical presentations are important for diagnosis. Early diagnosis and complete investigation of any case of amyloidosis is necessary in order to distinguish systemic from localized disease as well as to diagnose the underlying cause in each case.

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