Localized Amyloidosis of Palate

Sir,

Amyloidosis is a metabolic disease characterized by the extracellular deposition of abnormal insoluble proteinaceous fibrillar material in one or more organs. These deposits are derived from misfolded proteins that aggregate to form linear fibrils with an approximate diameter of 7.5–10 nm and a cross β-pleated sheet conformation, evidenced by X-ray diffraction.[1] Amyloidosis may be systemic or localized to a single organ, such as lungs, brain, or skin. According to some authors, the localized form has a better prognosis, particularly when the head and neck area is affected.[2] The most common sites of involvement in the head and neck are the thyroid, larynx, and subglottis. Amyloidosis of the oral cavity is less frequent and usually tends to involve the tongue, lip, and buccal mucosa.[3] To the best of our knowledge, isolated involvement of the palate has been reported in less than ten cases in worldwide literature.[3-9]

A 46-year-old Indian male smoker presented with painless swellings on the palate, which he had first noticed about 2 months earlier. His past medical and family history was unremarkable. Clinical examination revealed multiple smooth nodules ranging from 1 to 2 cm in diameter, located bilaterally on the hard palate, with one of the lesions arising at the junction of the hard and soft palate [Figure 1]. The mucosa overlying the lesions had a yellowish discoloration. On palpation, the lesions were soft and nontender. The rest of the oral cavity was normal apart from nicotine staining of the teeth. Cervical lymph nodes were not palpable. No abnormality was noted on further cutaneous and systemic examination. Based on these findings, xanthogranuloma, lipoma, pleomorphic adenoma, amyloidosis, mucoepidermoid carcinoma, and lymphoma were considered as the differential diagnoses.

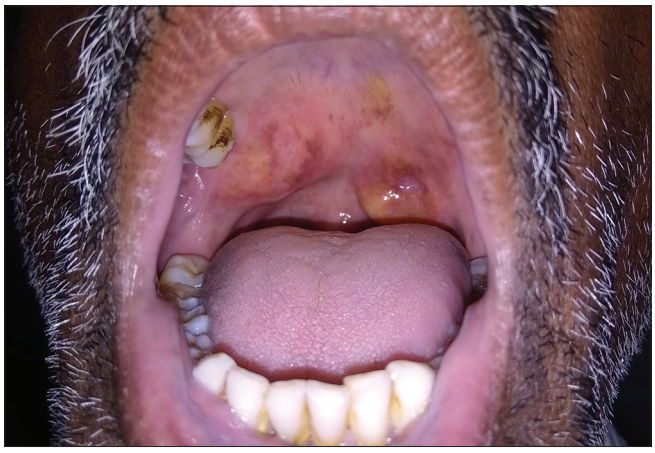

Figure 1: Oral cavity showing multiple nodules bilaterally on the palate with overlying mucosa showing yellow discoloration

An incisional biopsy was taken from one of the lesions under local anesthesia and sent for histopathological examination. The section showed tissue lined by mature squamous epithelium and homogenous eosinophilic amorphous material deposited in the subepithelial region with interspersed connective tissue cells and capillaries [Figure 2a]. There was no evidence of granuloma or malignancy. The amorphous material had positive staining for Congo red, taking a reddish-orange color under light microscopy [Figure 2b]. Apple green birefringence was noted under polarized light. The patient was evaluated further to identify associated systemic diseases or possible involvement of other organs. A complete hemogram, liver and renal function tests were within normal limits. Serum protein electrophoresis was normal, and urinalysis did not reveal any proteinuria. X-ray of the palate and maxillary sinuses was unremarkable. Chest X-ray, echocardiography, and abdominal ultrasonography excluded any organomegaly or cardiac dysfunction. On the basis of the

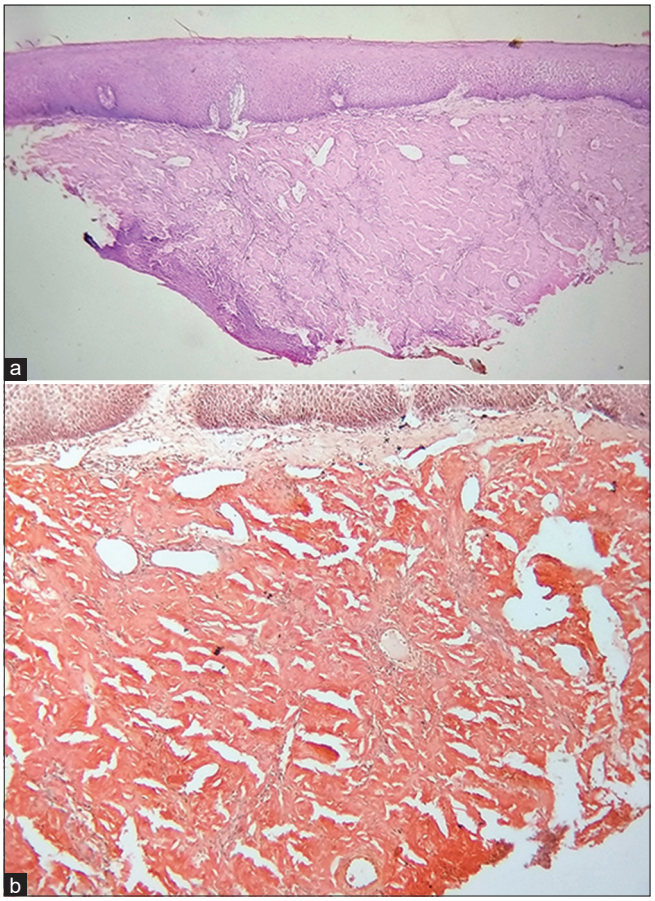

Figure 2: (a) Homogenous eosinophilic amorphous material in the subepithelial region (hematoxylin and eosin, ×40); (b) Reddish orange staining of the amorphous material under light microscopy (Congo red, ×100)

clinical, laboratory, and histological findings, a diagnosis of localized oral amyloidosis was made.

The patient was kept under observation with periodic monitoring. No evidence of local progression or development of systemic disease was noted after 1 year of follow-up.

The clinical presentation in previously reported cases of localized amyloidosis of the palate range from bleeding and fullness of the ear and nasopharynx due to lesions on the soft palate, to ulceration, soreness, and painless nodules on the hard palate. The characteristic histological finding in our case helped to establish the diagnosis of amyloidosis. Amyloid stained with hematoxylin and eosin appears as a homogeneous eosinophilic amorphous substance. When stained with Congo red, it takes up a red color under light microscopy and shows apple-green birefringence under polarized light. The specific type of amyloid fibril can be further determined by immunohistochemical staining or immunoelectron microscopy.[1]

Localized amyloidosis of the head and neck region has been reported not to be generally linked to any systemic disease.[2,3] Nevertheless, the site of involvement in the oral cavity may have diagnostic importance. Although none of the reported cases affecting the palate had a systemic association, amyloidosis of the tongue has been suggested as a clinical sign associated with plasma cell dyscrasia or dialysis-related systemic amyloidosis.[3] Therefore, further investigations are mandatory for the evaluation of systemic involvement in cases of oral amyloidosis.

There is no consensus on the management of localized oral amyloidosis. Treatment is mostly symptomatic, and surgical or laser excision is indicated only in case of functional impairment.[3] In our patient, the lesions being nonprogressive and asymptomatic, a conservative approach was preferred.

To conclude, dermatologists should be aware of this rare form of amyloidosis and consider it as a differential in the case of palatal nodules. Once the diagnosis is confirmed by biopsy, the patient should be evaluated thoroughly and followed up regularly for systemic involvement.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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