Primary Systemic Amyloidosis Mimicking Rhinophyma

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A 55-year-old male presented with asymptomatic nodular growth over nose since 6 months. On examination it was ~5 cm, well-defined, waxy thick brownish plaque extending from left ala of nose to tip of nose with skin overlying showed ridging and dilated pilosebaceous openings [Figure 1a]. On further follow-up, we noticed periorbital ecchymosis, which was not present earlier, few waxy papules and nodules over scalp, lateral aspect of lower eyelids, purpura over neck and toe, finger and toe nails showing dystrophy, striations, and anonychia [Figure 1b], and tongue showing macroglossia. Histopathology showed eosinophilic deposits in the dermis [Figure 2a], which on special staining with Congo red showed apple green birefringence of fibrils under polarized light [Figure 2b]. On further investigation, 24 h urine protein levels were raised 793.80 mg/24 h and albumin and gamma band were present on electrophoresis. Therefore, final diagnosis of primary systemic amyloidosis was made.

Amyloidosis is a disease caused by extracellular deposition of abnormal insoluble protein fibrils with antiparallel β-pleated sheet. Amyloidosis is classified into systemic amyloidosis and primary cutaneous amyloidosis. The latter is more common than former. Systemic amyloidosis is classified as primary systemic amyloidosis, secondary systemic amyloidosis, hemodialysis associated amyloidosis, and familial form of amyloidosis. Primary cutaneous amyloidosis is classically divided into macular amyloidosis, lichen amyloidosis, and nodular amyloidosis. Primary systemic amyloidosis has various clinical presentations. Cutaneous finding appears early compared to systemic involvement in 40–50% of patients. Deposition of amyloid in dermis gives rise to waxy papules, nodules, and plaques. In our case, deposition of amyloid locally over nose gave rise to rhinophyma-like presentation. On going through the literature, we found head and neck amyloidosis is rare and usually of localized type.[1] Case reports of primary localized nodular amyloidosis over nose are present, but there are no case report of primary systemic amyloidosis.[2] Trachyonychia secondary to amyloid deposits around blood vessels and in the dermis of the nail bed has been reported, as present in our patient.[3] This case illustrates that rhinophyma as a presenting feature is mentioned for localized amyloidosis but not for systemic amyloidosis. It is unusual to think of amyloidosis when patient presents with rhinophyma-like growth in Indian skin, where periorbital ecchymoses may not always be noticed.

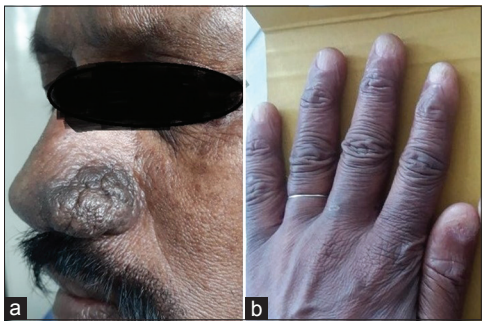

Figure 1: (a) Plaque over nose with dilated pilosebaceous openings simulating as rhinophyma. (b) Finger nails showing dystrophy, striations, and anonychia

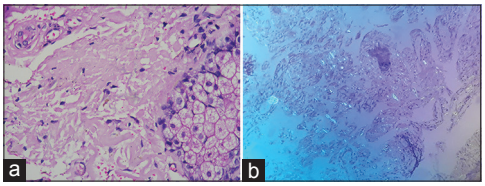

Figure 2: (a) Eosinophilic amyloid deposits in dermis (hematoxylin and eosin, ×40). (b) Apple green birefringence of amyloid fibrils with Congo red staining under polarized light (×40)

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

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