The Journal of Laryngology and Otology
December 1988. Vol. **102.** pp. 1153–1155

Nasal involvement in systemic amyloidosis

S. P. DUBEY,* M.S., Y. N. MEHRA,† F.R.C.S., D.A.B.O. (U.S.A.), A. K. BANERJEE,‡ M.D.,
S. B. S. MANN,§ M.S., K. GARG,¶ M.D. and V. I. NEHRU,‖ M.S. (Chandigirh, India)

Abstract
A localized amyloid deposit in the nose is extremely rare and involvement of the nose in systemic amyloidosis has not yet been reported in the literature. Here a case of nasal involvement in systemic amyloidosis is presented.

Introduction

Amyloidosis may be defined as a group of diseases of unknown aetiology characterised by the extracellular deposition of an eosinophilic, relatively insoluble, fibrous protein in one or more sites of the body (Cohen and Rubinow, 1983).

Though amyloid deposits occur in a variety of locations in the head and neck, upper aerodigestive tract, and lower respiratory tract, nasal involvement is extremely rare in the localized form and has not yet been reported in the systemic form of the disease.

Case report

A 64-year-old female, a known case of primary amyloidosis with systemic involvement, was referred to the ENT Department in August 1987, complaining of gradually increasing nasal obstruction with occasional nose-bleeds for the past two to three years, and widening of the nose for the last 18 months. There was no history of trauma.



FIG. 1
Clinical photograph.

On ENT examination, the patient had widening of the nasal dorsum (Fig. 1). Anterior rhinoscopy showed a pale, polypoid, fleshy, friable mass at the level of the middle turbinate in both nasal cavities. The masses did no bleed on touch, nor was there any shrinkage with vasoconstrictor. The findings on posterior rhinoscopy were within normal limits. The oral cavity, oropharynx, hypopharynx, larynx and ears were normal on clinical examination.

X-rays of the paranasal sinuses showed widening of nasal dorsum and a soft tissue opacity in both nasal cavities. Subsequently, the patient underwent a CT scan which showed a non-enhancing soft tissue opacity in both nasal cavities with fragmentation of the bony nasal dorsum (Figs. 2 and 3). The paranasal sinuses were normal.

Systemically, the patient had histologically proved amyloid deposits in the heart, rectum, and subcutaneous tissue adjoining the wrist, elbow and the ankle joints.

In order to give an adequate nasal airway, surgical excision of the mass was performed six weeks after presentation. After orotracheal intubation and hypopharyngeal packing, a 4 cm long midline vertical incision, beginning at the root of the nose was made. The subcutaneous tissue and periosteum of the bony nasal dorsum was thickened and friable. The nasal bones and the frontal processes of the maxillae were eroded by the mass. Both nasal bones and frontal processes of the maxilla were also softened. The fleshy friable mass was scooped out from both nasal cavities. There was brisk haemorrhage throughout the procedure, for which one unit of blood was transfused during operation. After haemostasis was achieved, the wound was sutured in two layers and the nasal cavities packed with medicated ribbon gauze.

Histopathological examination of the specimen removed at operation showed multiple fragments of soft tissue lined on one surface by nasal mucosa and fragments of bone and nodular masses of eosinophilic hyaline material reacting positively to stains for amyloid (Fig. 4). Plasma cells, macrophages and foreign body giant cells were present around some of these areas (Fig. 5). Some of the amyloid nodules showed focal

* Senior Resident, Department of Otolaryngology, † Professor and Head, Department of Otolaryngology, ‡ Professor, Department of Pathology, § Associate Professor, Department of Otolaryngology, ¶ Lecturer, Department of Radiodiagnosis, ‖ Senior Resident, Department of Otolaryngology, Nehru Hospital, Postgraduate Institute of Medical Education and Research, (PGIMER), Chandigarh—160012, India.

https://doi.org/10.1017/S0022215100107571 Published online by Cambridge University Press

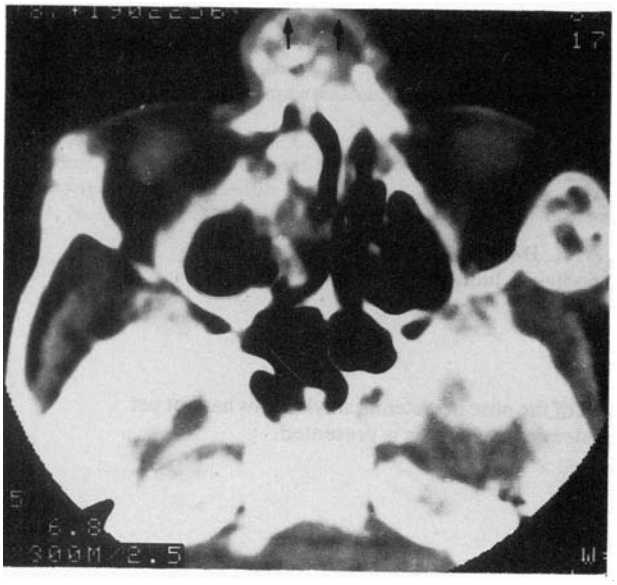


FIG. 2

Axial scan at the level of the lower part of the orbit showing destruction and fragmentation of the bony nasal dorsum with thickened periosteum (arrows), haziness of the anterior ethmoid cells. Middle and posterior ethmoid cells, and sphenoid sinus are normal.

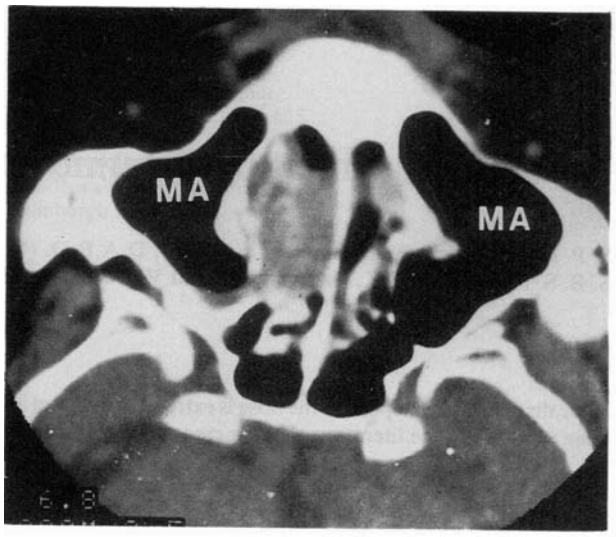


FIG. 3

Coronal section through the anterior part of the nasal cavity showing soft tissue opacity in both the nasal cavities (more marked on left side), maxillary antrum (MA) are normal on both sides.

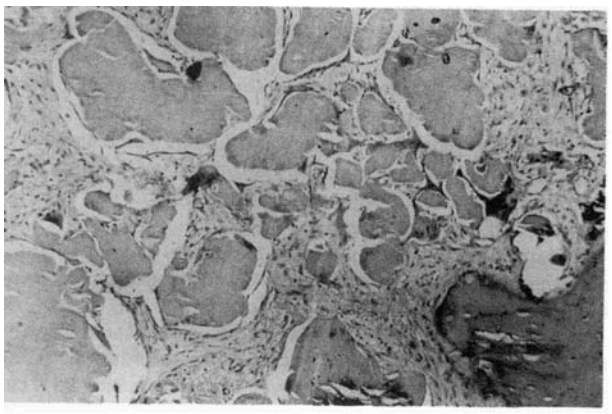


FIG. 4

From biopsy of nasal mass showing nodular deposits of amyloid in the submucous tissue and bone. (Hematoxylin & eosin, ×120).

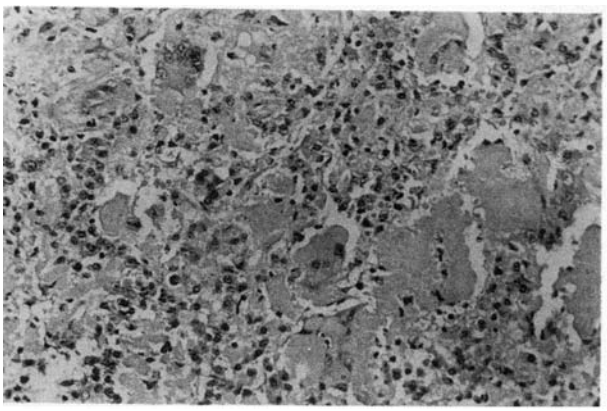


FIG. 5

Another area showing plasma cell and multinucleated giant cells in relation to the amyloid deposits. (Hematoxylin & eosin, ×120).

calcification. Amyloid constituted the major part of the tissue and the cell reaction was relatively weaker.

Discussion

Rokitansky (1842) was the first to describe systemic amyloidosis. Virchow (1860) coined the word amyloid to describe the starch-like waxy degeneration of organs and described its characteristic reaction with iodine to form a characteristic yellowish-red or reddish-violet colour. Symmers (1956) first proposed a comprehensive classification of amyloidosis. The most frequent site of involvement in the upper air passage is the larynx (Holinger *et al* . 1959). The next most common locations are the base of the tongue, trachea, nasal cavity, and pharynx. Localized deposits of amyloid can also occur in the head and neck, aerodigestive tract and lower respiratory tract in the absence of systemic involvement, in the eye, the major and minor salivary glands, and submucosal deposits within the nose, paranasal sinuses, nasopharynx, oral cavity, oral pharynx, larynx, bronchial tree and the lung (Simpson II *et al.*, 1984). Symptoms, when present, are caused by the physical presence, size and location of the mass as happened in our case.

Asymptomatic involvement of upper aerodigestive tract needs no treatment; but when symptoms are present, total extirpation is essential (Krammer and Som, 1935).

References

Cohen, A. S. and Rubinow, A. (1983) Amyloidosis. In: *Internal Medicine* (Stein, J. H. ed), pp 1118–1121, Little Brown and Company, Boston.

Holinger, P. H., Johnston, K. C., and Delgado, A. (1959) Amyloid tumour of the larynx and trachea. *Archives of Otolaryngology,* **70:** 555–561.

https://doi.org/10.1017/S0022215100107571 Published online by Cambridge University Press

Krammar, R. and Som, M. L. (1935) Local tumour like deposits of amyloid. *Archives of Otolaryngology,* **21:** 324–334.
Rokitansky, C. L. (1842) Handbuch der Speciellen Anatomie. Vol 2, pp 311, Brawmuller and Seider, Vienna.
Simpson II, G. T., Strong, S. M., Skinner, M. and Cohen, A. S. (1984) Localized amyloidosis of the head and neck, and aerodigestive and lower respiratory tract. *Annals of Otology, Rhinology, and Laryngology,* **93:** 374–379.
Symmers, W. (1956) Primary amyloidosis: A review. *Journal of Clinical Pathology,* **9:** 187–211.
Virchow, R. L. K. (1860) Cellular pathology based upon physiological and pathological history. pp 409–437, Robert De Witt, New York.

Address for correspondence
Prof. Y. N. Mehra,
Dept. of Otolaryngology,
Postgraduate Institute of Medical Education & Research,
Chandigarh—160012,
India.

https://doi.org/10.1017/S0022215100107571 Published online by Cambridge University Press