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Primary sinonasal amyloidosis

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Abstract
Primary amyloidosis localized to the sinonasal tract is extremely rare with only 20 reported cases in the English literature. We describe a further case and review the literature.

Key words: Amyloidosis; Nose; Paranasal Sinuses

Case report

A 53-year-old woman was referred to the ENT Department with a two-year history of chronic discomfort over the maxillary sinuses, nasal blockage and serosanguinous nasal discharge.

Examination revealed a large lesion affecting the nasal septum and right lateral nasal wall that was friable and bled with mild trauma. A computed tomography (CT) scan and magnetic resonance image (MRI) demonstrated the lesion to involve the right nasal cavity eroding the lateral wall with extension to the upper half of the antrum and involving the anterior ethmoid cells (Figure 1). A debulking biopsy was taken and antral lavage was clear.

Histology revealed deposits of hyaline eosinophilic material at the periphery of which foreign body-type giant cells were present. The hyaline material stained with Congo red dye and showed ''apple green'' birefringence typical of amyloid. The deposits were of the amyloid light chain (AL) type, demonstrated by immunohistochemistry. Full blood count blood, urea and creatinine, liver function tests, urinanalysis, erythrocyte sedimentation rate, Rh testing, antinuclear antibody testing, serum/urine immunoelectrophoresis, tuberculin skin test, bone marrow aspiration, ECG and chest radiography were all normal. Subsequent endoscopy of the aerodigestive tract excluded multifocality. Serum albumin was low at 28. A isotope Serum Amyloid Precursor (SAP) scan confirmed no evidence of systemic amyloid deposition.

The patient's symptoms following initial debulking ameloriated and further surgical excision was not warranted. The patient has remained under review for nearly three years without further treatment.

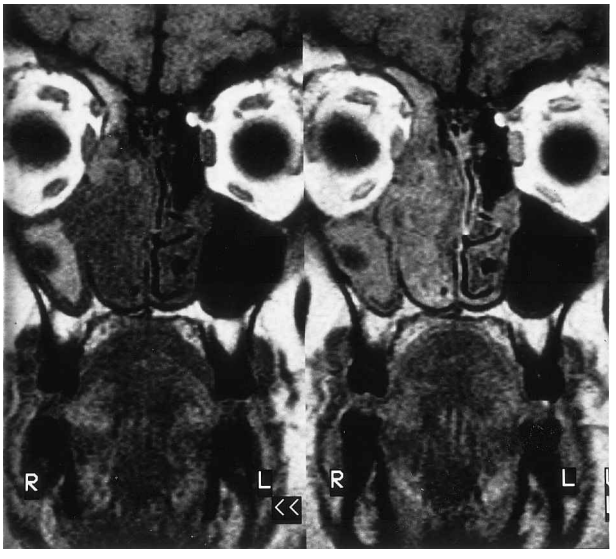

Fig. 1
Coronal T1 MRI image showing lesion eroding right antral wall extending to frontoethmoid junction and into anterior ethmoid cells.

Discussion

Amyloid is a pathologic proteinaceous substance deposited between cells in various tissues and organs of the body in a variety of clinical settings.[1] Its diagnosis depends on identification in biopsy specimens.

Although amyloid deposits have a uniform appearance, it is clear that amyloid is not a single entity. There are three major and several minor biochemical forms. Of the 15 biochemically distinct forms of amyloid proteins that have been identified, three are the most common.[2,3]

(1) Amyloid light chain (AL) is derived from plasma cells and contains kappa or lambda immunoglobulin light chains,
(2) Amyloid-associated (AA) is a unique non-immunoglobulin protein synthesized by the liver.
(3) Aβ amyloid is found in the cerebral lesion of Alzheimer disease.

Clinically amyloidosis may be broadly classified as systemic (generalized), involving scleral organ systems, or it may be localized, when deposits are limited to a single organ. Hereditary or familial amyloidosis constitutes a separate group, with several distinctive patterns of organ involvement.

The systemic or generalized pattern is subclassified into primary amyloidosis, when associated with some plasma cell dyscrasia, or secondary amyloidosis, when it occurs as a complication of an underlying chronic inflammatory process (tuberculosis, rheumatoid arthritis, inflammatory bowel disease etc.).

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There is no consistent pattern or organ involvement in systemic amyloidosis. Nevertheless, the secondary subclass tends to be the most severe, and usually involves kidneys, liver, spleen, lymph nodes, adrenals and thyroid.

The primary subclass although it cannot be reliably distinguished from the secondary by the organ distribution, usually affects heart, kidney, gastrointestinal tract and tongue.

Plasma cell dyscrasias predispose to AL amyloid,[4] whereas, systemic amyloidosis secondary to chronic inflammatory diseases predispose to deposition of AA amyloid.

Amyloid deposition in the head and neck region can occur as an isolated pathology, or be part of a systemic amyloidosis with or without plasma cell dyscrasias or malignant lymphoma.[3] The most frequent site of involvement in the upper respiratory tract is the larynx followed by the base of tongue, trachea, pharynx and nasal cavity.[5]

Isolated nasal amyloidosis is extremely rare and is a localized, idiopathic, primary process composed of AL, kappa or lambda light chain amyloid. Up to 1935 there were only seven reported cases of sinonasal amyloidosis.[6] Mufarrij *et al*.[7] found seven subsequent cases from 1935 to 1990 and added one further case. A Medline search up to the present revealed five extra[8–12] reported cases. The disease can present at any age and has no sex predilection.[7]

The initiating factors remain obscure and the clinical manifestations of nasal amyloid are often insidious, with symptoms that are slowly progressive over months to years prior to diagnosis.[13]

The extent of the localized disease is best evaluated by CT and/or MRI scan and systemic causes of amyloidosis such as multiple myeloma, tuberculosis, and rheumatic diseases must be ruled-out. A thorough examination of the entire respiratory tract is indicated including nasopharynx, oropharynx, larynx and tracheobronchial tree given the multifocality of the disease.

Primary sinonasal amyloidosis is a slowly progressive disease that does not respond to non-surgical treatments. Medical treatment, including local and systemic corticosteroids and chemotherapy have been unsuccessful.[14] Radiotherapy is contra-indicated because it is ineffective and has significant side-effects.[15]

Therapy in this site has been dictated by experience in other more common sites e.g. larynx. Recurrences following surgery are common and further surgical therapy is dictated by symptomatology.[16]

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References

1 Cohen AS, Rubinow A. Amyloidosis. In: Stein J. H., ed. *Internal Medicine*, Boston: Little Brown and Company, 1983;1118–21

2 Husby G, Araki S, Benditt EP. The 1990 guidelines for nomenclature and classification of amyloid and amyloidosis. In: Natvig J. B., ed. *Amyloid and Amyloidosis*. Dordrecht, the Netherlands: Kluwer Academic Publishers, 1990;7–11

3 Cotran R, Kumar V, Collin T. Amyloidosis. In: *Pathologic Basis of Disease*. Philadelphia: WB Saunders, 1999

4 Dubey SP, Mehra YN, Banergee AK. Nasal involvement in systemic amyloidosis. *J Laryngol Otol* 1988;**102**:1153–5

5 Holinger PH, Johnston KC, Degado A. Amyloid tumour of the larynx and trachea. *Arch Otolaryngol* 1959;**70**:555–61

6 Kramer R, Som ML. Local tumour-like deposits of amyloid in the larynx. Report of a case and review of the literature. *Arch Otolaryngol* 1935;**21**:324–34

7 Mufarrij AA, Busaba NY, Zaytoun GM, Gallo GR, Feiner HD. Primary localized amyloidosis of the nose and paranasal sinuses. A case report with immunohistochemical observations and a review of the literature. *Am J Surg Pathol* 1990;**14**:379–83

8 Birchall D, Fields JM, Poon CL. Case report: focal amyloidosis of the maxillary antrum: plain film, CT and MR appearances. *Clin Radiol* 1997;**52**:392–4

9 Lim JS, Lebowitz RA, Jacobs JB. Primary amyloidosis presenting as a nasopharyngeal mass. *Am J Rhinol* 1999;**13**:209–12

10 Kobayashi T, Taguchi O, Yasui H, Hataji O. A case of amyloidosis of the tracheobronchial tree and inferior nasal concha. *Jap J Thorac Dis* 1979;**35**:1378–82

11 Dominguez S, Wienberg P, Claros P, Claros A, Vila J. Primary localised nasopharyngeal amyloidosis. A case report. *Int J Pediatr Otorhinolaryngol* 1996;**36**:61–7

12 Clevens RA, Wiatrac BJ, Myers MW. Multifocal amyloidosis of the paediatric airway. *Arch Otolaryngol Head Neck Surg* 1995;**121**:229–32

13 Garret JA. Amyloid deposits in the nose and maxillary sinuses. *Arch Otolaryngol* 1968;**87**:103–4

14 McAlpine GC, Fuller AD. Localised laryngeal amyloidosis. A report of a case with a rreview of the literature. *J Laryngol Otol* 1964;**78**:296–314

15 Mitriani M, Biller HS. Laryngeal amyloidosis. *Laryngoscope* 1985;**95**:1346–7

16 Hardingham M. Diffuse amyloidosis of the larynx. *Ear Nose Throat J* 1987;**66**:61–3

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