Sinonasal localised amyloidosis: an uncommon location

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DESCRIPTION

A male patient in his seventh decade of life presented to the otorhinolaryngology department due to a 3 month history of progressive nasal pyramid broadening (figure 1). He denied nasal obstruction, epistaxis, rhinorrhea or post nasal drip. There were no systemic symptoms such as fever, weight loss or night sweats. The patient also denied facial pain, numbness or visual symptoms. His medical history included ischaemic and valvular heart disease and aortic aneurysm. He had non-smoking habits and moderate alcohol intake.

Rhinoscopy revealed right enlargement of nasal pyramid at the level of nasal bones and a rightsided painless submucosal hard mass occupying the superior meatus and superior aspect of nasal vestibule, as shown in figure 2. Remaining ENT examination was otherwise unremarkable. CT-scan findings demonstrated a heterogeneous mass with inside calcifications (figure 3). Incisional biopsy was undertaken in an outpatient setting and histopathological analysis demonstrated the presence of amorphous deposits, suggesting the presence of amyloid protein after apple-green birefringence under polarised light in Congo-red stained sections. Patient was then referred for observation by internal medicine. Systemic amyloidosis was excluded after unremarkable blood counts, biochemistry, erythrocyte sedimentation rate, serum protein electrophoresis and absence of Bence Jones protein in urine analysis. Rectal biopsy specimens, bone marrow aspirates, pulmonary function tests, echocardiography and abdominal ultrasound were also unremarkable. So, the final diagnosis of nasal localised amyloidosis was achieved. Due to cardiovascular disease and absence of symptoms, it was decided to keep the patient under clinical surveillance. After 8 months of follow-up, the lesion remains stable.

Amyloidosis is a rare group of diseases characterised by idiopathic extracellular deposition of insoluble amorphous protein fibrils (amyloid) in multiple organs and tissues. It is usually systemic, but it can be localised to a specific organ or tissue in 10%–20% of the cases. Localised amyloidosis most commonly occurs at head and neck and it primarily affects larynx and oropharynx. The involvement of nasal cavities is exceedingly rare, with less than 40 cases described in literature to date. The pathophysiology of localised amyloidosis is poorly understood, but it has been proposed that light chains are locally produced and deposited by plasma cells. Imaging findings usually consist in well-defined...

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Figure 1 Right-sided nasal pyramid broadening at presentation.

Figure 2 Inferior view of right nasal cavity through nasofibroscopy demonstrating a smooth submucosal mass, occluding superior meatus (arrows). NS, nasal septum; NP, nasopharynx; MT, middle turbinate; IT, inferior turbinate.

Figure 3 Sagittal (A) and axial (B) views of paranasal CT scan showing a heterogeneous mass with inside calcifications between nasal septum and nasal bones (arrows).
homogeneous soft tissue mass, although it has been rarely reported to appear as heterogeneous and calcified masses, such as the present case. Bony erosion is only reported at advanced stages. Unlike the systemic form, localised amyloidosis generally has good prognosis, rarely progressing to systemic involvement. Evidence regarding localised amyloidosis management is sparse, but it usually involves surgical excision, radiotherapy or clinical surveillance.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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