Primary Amyloidosis of the Nose Presenting with Refractory Epistaxis and Systemic Involvement – A Rare Phenomenon

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INTRODUCTION

Amyloid, a fibrillar protein deposited in the extracellular space, may be localized to a specific organ or it may be systemic, involving various organs (1, 2). Localized amyloidosis is uncommon in the head and neck (1). Systemic amyloidosis involving the nasal cavity presenting as refractory epistaxis to our knowledge has not been previously reported. The purpose of this case report is to present a rare presentation of refractory epistaxis which was secondary to systemic nasal amyloidosis and to draw attention to the recognition of this form of disease in order to initiate systemic therapy and to be vigilant for surgical intervention.

Keywords: Nasal amyloidosis, refractory epistaxis

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CASE REPORT

A 74-year old male, known to have vitamin B12 deficiency and no other chronic illnesses, presented with a one-day history of left-sided epistaxis progressing to both sides and a bloody postnasal discharge. There was a one-year history of intermittent bloody postnasal discharge not requiring admission. He denied any other sinonasal or otological symptoms, trauma or history of blood dyscrasias. There was a past history of sigmoid amyloidosis diagnosed on colonoscopy and biopsy. The rest of his history was non-contributory.

On examination, he was haemodynamically stable. Nasoendoscopy revealed moderate bilateral epistaxis with the left greater than the right. Oropharyngeal examination revealed postnasal bleeding. No identifiable bleeding point was seen. The rest of his examination was normal. He was assessed as moderate posterior epistaxis. He underwent posterior nasal packing with a 16 French catheter and bilateral anterior packing with antibiotic impregnated ribbon gauze. His initial haemoglobin level was found to be 9.8 g/dL. He was admitted to the ward and placed on prophylactic amoxicillin and clavulanic acid. On day two post admission, his vital signs were normal and his haemoglobin level did not fall. The anterior and posterior packs were removed followed by moderate epistaxis which warranted repacking. On day four, while the packs were *in situ*, mild epistaxis persisted and his haemoglobin level fell to 7.5 g/dL. On day five, he became haemodynamically unstable. He was taken to the operating theatre for an examination under anaesthesia and cautery. The surgical findings were severe left posterior epistaxis originating from the sphenoethmoidal recess. The brisk bleeding obscured endoscopic visualization. Because of the lack of the appropriate blood type and his instability, he was repacked. He was admitted to the Intensive Care Unit (ICU) post procedure because of soft palate oedema. On day seven, blood products were available. The patient was returned to the operating theatre to obtain haemostasis *via* ligation of the left external carotid artery followed by endoscopic ligation of the left sphenopalatine artery. The epistaxis was markedly reduced. The surgical findings were a bleeding friable mass on the anterior wall of the left sphenoidal sinus which was removed prior to cauterization. This resulted in haemostasis. Light anterior packs were placed intra-operatively. He was discharged from ICU on day nine to the ward and the nasal packs were discontinued. His progress was complicated by an aspiration pneumonia and urinary tract infection which were both treated. He was discharged home on day 27. The histopathology revealed amyloidosis (Figure). Further haematological work-up including a bone marrow aspirate did not reveal a cause. He was assessed as primary systemic amyloidosis, amyloid light chain (AL) type and treated with melphalan and prednisone. On follow-up eight months later, he denied any epistaxis. His last haemoglobin was 9.8 g/dL.

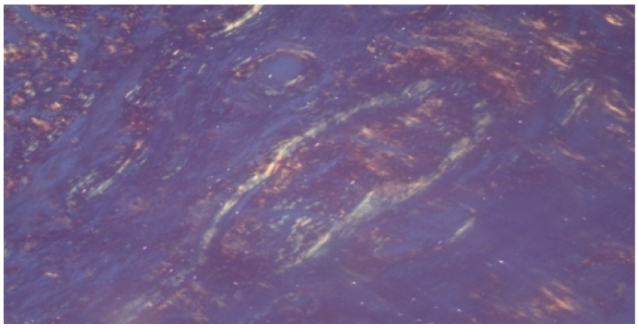

Figure: Histology of amyloidosis from tissue section of mass from sphenoethmoidal recess demonstrating birefringence on polarizing microscopy after staining with Congo red.

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DISCUSSION

Amyloidosis may be classified either by the type of amyloid protein or by the clinical presentation. The type depends on the fibrillar protein precursor. In this case, it was the AL type. It may be a primary disorder which may be localized in a specific organ, or systemic, compromising various organs or systems with a more serious prognosis (3). It may also be secondary to processes such as multiple myeloma or rheumatoid arthritis.

Head and neck amyloidosis is rare and usually localized (3). The tongue may be associated with systemic disease (3). Utilizing a PubMed and Medline search, there has only been one reported case of nasal cavity involvement in systemic disease (4). The patient in this case report presented with primary systemic amyloidosis AL type involving the nasal cavity and sigmoid colon. From the literature available to us, this is the first reported case of systemic amyloidosis involving the nasal cavity presenting as refractory epistaxis requiring sphenopalatine and external carotid artery ligation. Amyloidal deposits cause vascular fragility (5) which may lead to epistaxis. Removal of the localized form has a good prognosis (3) and low recurrence rate. The treatment of primary systemic amyloidosis includes systemic therapy. In studies of different regimes of intermittent oral melphalan and prednisone, the response rates were low with a median survival of seven to nine months in patients not receiving chemotherapy *versus* 12–18 months in those who did (6). The patient in this case report has been treated with melphalan and prednisone with no further recurrences of epistaxis and stabilization of his haemoglobin eight months after.

CONCLUSION

This case illustrates that although nasal amyloidosis is a rare cause of refractory epistaxis, it should be considered in the differential diagnosis. Where amyloidosis is detected as a cause, the patient should be evaluated for systemic involvement since this type has a worst prognosis and would require more aggressive treatment with systemic therapy. Vigilance is necessary for surgical interventions.

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