

Primary nasal amyloidosis

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Citation

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Abstract

Introduction: Amyloidosis in the head and neck is a rare but benign disease. Primary amyloidosis localized to the sinonasal tract is extremely rare. This is a report of one such case with a brief review of the literature. **Case report:** A 42 year old lady who presented with epistaxis and left sided nasal obstruction was found to have a mass in the left nasal cavity, which was proved to be amyloidosis after histopathological examination. **Conclusion:** Localized amyloidosis, though rare, can be considered as a differential diagnosis for a nasal mass.

INTRODUCTION

Amyloidosis is characterized by extra cellular deposition of amorphous proteinaceous material in the form of fibrils.

Amyloidosis can be systemic or localized. Localized amyloidosis is rare, frequently involves the head and neck region without systemic manifestations. Larynx is the most commonly affected region (61%) followed by the oropharynx, trachea, orbit and nasopharynx. Sinonasal amyloidosis is extremely rare. We report one such case managed by us along with a brief review of the literature.

CASE REPORT

A 42 year old, known diabetic lady presented with recurrent left sided nasal obstruction since four years and recurrent epistaxis since one month. On examination, the nasal airway was significantly reduced on the left side by a smooth, solitary, soft to firm, non tender, pale pink mass between the septum and the lateral wall above the inferior turbinate. The inferior turbinate was bulky. Post nasal examination showed a similar mass in the left choana obscuring the view of inferior turbinate. The diagnostic nasal endoscopy confirmed the findings. But, the scope could not be passed beyond the mass. There was minimal bleeding on touching the mass. A biopsy taken from the mass was reported as amyloidosis. (Figure 1 & 2)

Figure 1

Figure 1: – HPE – Amyloid deposit beneath the nasal mucosa

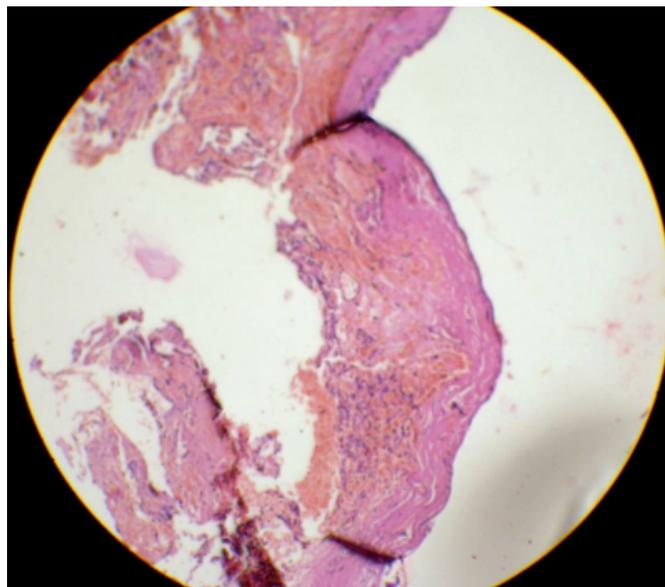
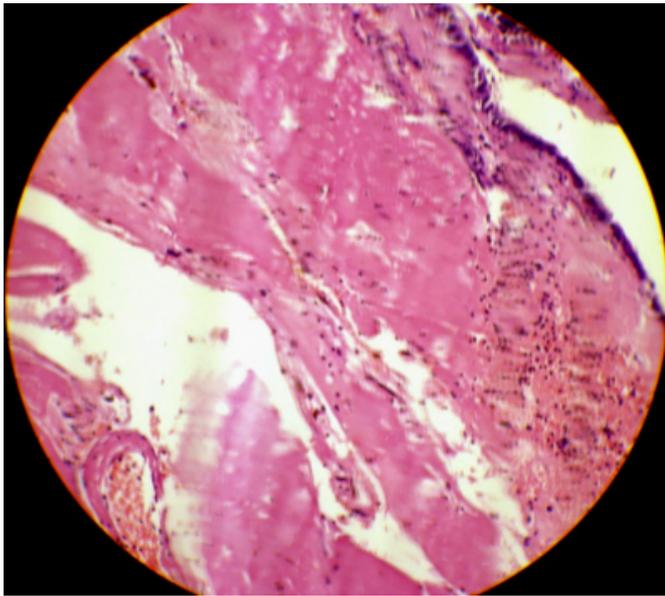


Figure 2

Figure 2: Amyloid deposits beneath the nasal mucosa 20 X



Confirmation was done by Congo red staining. MRI of the nose and PNS showed a soft tissue mass in the left nasal cavity involving left inferior turbinate and retained secretions in the left maxillary sinus without bone erosion. (Figure 3 &4)

Figure 3

Figure 3: Coronal MR Image showing the mass in the left nasal cavity

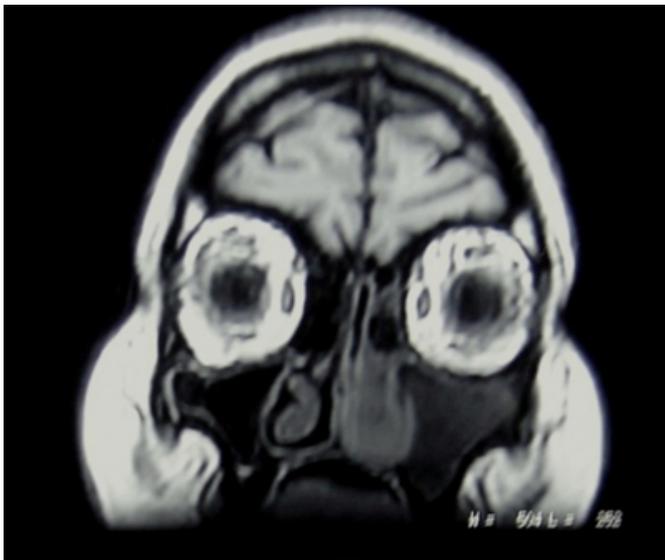


Figure 4

Figure 4: Axial MR image showing the mass



Patient underwent investigations like liver function tests, renal function tests, serum alkaline phosphatase, urine analysis, chest X-ray, abdominal ultrasound, ECG, echocardiogram, abdominal fat aspiration and rectal biopsy to rule out foci of systemic amyloidosis, but all the tests were negative. Endoscopic removal of the mass was done under general anesthesia. (Figure 5)

Figure 5

Figure 5: Photograph of the excised specimen



The post operative period was uneventful. Presently, patient

is asymptomatic and recurrence free eight months after the surgery.

DISCUSSION

Amyloidosis is the accumulation of extra cellular deposits of proteinaceous substance in one or more body parts.

Clinically, amyloidosis can be systemic or localized.¹ The systemic amyloidosis can be either primary, wherein there is no evidence of coexisting disease or secondary, which may be associated with chronic inflammatory diseases like tuberculosis, rheumatoid arthritis etc.² Localised amyloidosis is extremely rare,¹ but, can be seen in the head and neck region. Hereditary or familial amyloidosis is even rarer and is due to an autosomal recessive disorder.

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.² Larynx is the most frequently involved site in the head and neck region (61%), followed by oropharynx (23%), trachea (9%), orbit (4%), sinonasal tract and nasopharynx (3%).¹

About twenty three cases of isolated sinonasal amyloidosis have been found to be reported on reviewing the literature. Tsikoudas A has reviewed twenty cases² and added one further case. Upto 1935, there were only seven reported cases of sinonasal amyloidosis.³ Muffarrij et al⁴ found seven subsequent cases from 1935 to 1990 and added one further case. A Medline search up to 2001 revealed five other reported cases.^{5,6,7,8,9} We have found 3 more case reports of sinonasal amyloidosis in English literature.^{10,11,12} 13 cases of nasopharyngeal amyloidosis are reported till date.¹ Isolated nasal amyloidosis is extremely rare.²

Amyloidosis most commonly affects individuals between 50-70 years with a male to female predominance of 3:1 to 3:2. The Aetiology of amyloidosis is still unclear. It is currently believed to be a derangement in immunoregulation after protracted antigenic challenge.¹ Symptoms are related to the sites of involvement. Sinonasal amyloidosis presents with nasal obstruction, nasal discharge, epistaxis (due to vessel wall invasion), glue ear and post nasal drip.¹ On examination, the mass is seen as yellowish polypoidal lesion. The extension is better assessed by CT or MR imaging. Amyloidosis is confirmed by tissue biopsy.¹ Histologically, amyloid is an eosinophilic amorphous extra cellular protein deposit which can be confirmed by Congo red staining wherein a characteristic apple green

birefringence is seen under polarizing microscope. Systemic amyloidosis should be ruled out by a detailed clinical examination and investigations like peripheral blood smear, ESR, liver function tests, serum electrophoresis, radiographic imaging and bone marrow biopsy.¹³ Abdominal fat aspiration is risk free and as sensitive as rectal mucosal biopsy (70-80%) to rule out systemic amyloidosis.¹³

There is no satisfactory treatment for systemic amyloidosis.¹ For localized amyloidosis, symptomatic removal is required.¹ Bleeding can be a complication in such cases. Close follow up is essential as recurrence rates are as high as 50%.¹ Medical treatment, including corticosteroids and chemotherapy have not been successful.² Radiotherapy is also not advisable.²

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References

1. Panda NK, Saravanan K, Purushotaman GP et al. Localised amyloidosis masquerading as nasopharyngeal tumor: A review. *Am J of Otolaryngology, Head Neck Medicine and Surgery* 2007; 28:208-11
2. Tsikoudas A, Martin-Hirsch DP and Woodhead CJ. Primary sinonasal amyloidosis. *The Journal of Laryngology & Otology* Jan 2001; 115: 55-56
3. 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
4. Muffarrij AA, Busaba NY, Zaytoun GM et al. 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
5. 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
6. Lim JS, Lebowitz RA, Jacobs JB. Primary amyloidosis presenting as a nasopharyngeal mass. *Am J Rhinol* 1999; 13:209-12
7. Kobayashi T, Taguchi O, Ysui H et al. A case of amyloidosis of the tracheobronchial tree and inferior nasal concha. *Jap J Thorac Dis* 1971; 35:1378-82
8. Dominguez S, Wienberg P, Claros P et al. Primary localized nasopharyngeal amyloidosis. A case report. *Int J Pediatr Otorhinolaryngol* 1996; 36:61-7
9. Clevers RA, Wiatrac BJ, Myers MW. Multifocal amyloidosis of the pediatric airway. *Arch Otolaryngol Head Neck Surg* 1995; 121:229-32
10. Kenny PP, Lincoln WJ Chee, and Inny Busmanis. Amyloidoma of the nose in a pediatric patient: A case report. *Am J of Otolaryngology* 2001; 22 (2): 138-41
11. Teo DT, David PL and Sethi DS. Recurrent localized sinonasal amyloidosis: A case report. *Otolaryngology- Head and Neck Surgery*. Aug 2003; 270

12. Chin SC, Girish F, Kao CH et al. Amyloidosis concurrently involving the sinonasal cavities and larynx. Am J Neuroradiol April 2004;25:636-638

13. Carla R Penner and Susan Muller. Head and Neck Amyloidosis: A clinicopathological study of 15 cases. J of Oral Oncology Apr 2006;42(4): 421-429

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