International Surgery Journal
Suseeharan M et al. Int Surg J. 2024 Jan;11(1):107-114
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20233843

Case Report

Localised sinonasal amyloidosis presenting with unilateral hearing loss: a case report and review of the literature

Melanie Suseeharan*, Keli Dusu, Dema Motter, Alistair Balfour

Department of Otolaryngology, East Kent NHS Hospital Trust, Kent, UK

Received: 06 November 2023
Accepted: 01 December 2023

***Correspondence:**
Melanie Suseeharan,
E-mail: melanie.suseeharan@gmail.com

ABSTRACT

Localised sinonasal amyloidosis is a rare occurrence with only twenty-nine documented cases in literature. This report follows the case of a 79-year-old gentleman with an atypical presentation of headaches and unilateral (right-sided) hearing loss. The patient initially underwent magnetic resonance imaging of his internal auditory meatus which was normal. Flexible nasoendoscopy was performed which identified a right middle meatus discharging possible polypoidal lesion. A computed tomography scan of his sinuses was performed which identified a large soft tissue lesion projecting into the upper nasal airway arising from the nasal recess, with the appearance suspicious for a polyp. Subsequently, the patient underwent functional endoscopic sinus surgery to manage right maxillary sinusitis and further examine the right-sided polypoidal mass lesion that was obstructing the maxillary antrum. Maxillary sinus biopsy revealed a diagnosis of sinonasal amyloidosis while tests for systemic amyloidosis were negative.

Keywords: Sinonasal, Amyloidosis, Nasal obstruction, Otolaryngology

INTRODUCTION

Amyloidosis is a rare systemic disease characterised by the extracellular deposition of protein in different organs of the body. It is a rare condition and can be separated into systemic and localised forms.[1] Systemic amyloidosis can present with non-specific symptoms and largely depend on the affected organ. Due to this, systemic amyloidosis is often diagnosed late and carries a higher morbidity and mortality rate, often requiring more aggressive treatment.[2] In localised amyloidosis, amyloid deposits form at the site of precursor protein synthesis, whereas in systemic amyloidosis, amyloid deposition occurs distant from the site of precursor protein secretion. Localised amyloidosis carries a better prognosis and often can be managed conservatively with a watchful waiting approach.[3]

CASE REPORT

This case reports a 79-year-old gentleman who was referred to the general otolaryngology clinic by his primary care doctor due to right-sided hearing loss, tinnitus and intermittent headaches. He has a past medical history of atrial fibrillation, hypertension and previous deep vein thrombosis and subsequently takes regular blood thinners. At presentation, the patient reported no sinonasal symptoms whatsoever. A magnetic resonance scan of his internal auditory meatus was performed to further evaluate his hearing loss and tinnitus and rule out an acoustic neuroma, which was not present. On clinical examination including flexible nasoendoscopy a right middle meatal presumed polypoidal lesion was identified. This lesion was noted to be actively discharging although the patients did not report symptoms of increased discharge from bilateral nostrils. To investigate this lesion further, a computed tomogrophay (CT) scan of his sinuses was performed. This identified a large soft tissue

lesion projecting into the upper nasal airway on the right arising from the frontal recess (Figure 1).

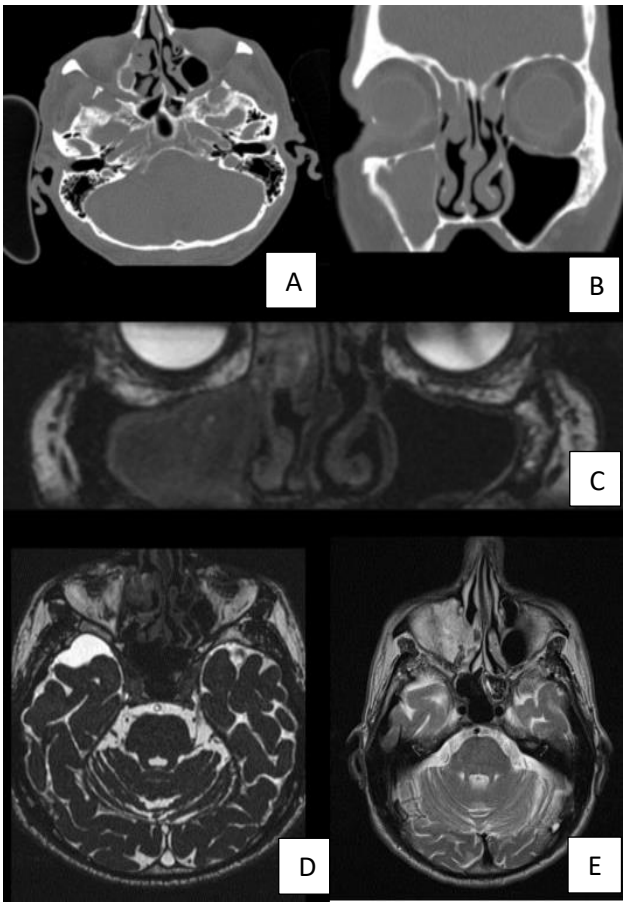

Figure 1: Complete opacification of the right maxillary sinus, right frontal sinus and obstruction of the recess. Large soft tissue lesion projecting into the upper nasal airway arising from the right sphenoethmoidal recess, A) Coronal CT opacified right maxillary sinus, B) Axial CT opacified right ethmoid air cells, C) MRI coronal T2, D) MRI T2 axial, E) Post contrast MRI T2 axial.

At this point, the main differentials were nasal polyp, hypertrophied turbinate, and suspected malignancy. The patient was started on nasal steroids and oral antibiotics for a week and booked in for an urgent functional endoscopic sinus surgery (FESS) and biopsy of the lesion. The patient underwent FESS surgery and biopsies of the lesion were taken with debulking also performed. Intraoperative puss and swollen mucosa of the right antrum was noted. All tissue samples were sent for histology. Histological analysis of the uncinate and anterior ethmoid showed evidence of chronic inflammation. Testing of tissue from the maxillary sinus, stained orange on congo red and apple green birefringence on polarisation - indicating the presence of amyloid tissue. After diagnosis of sinonasal amyloidosis, the patient was referred to a tertiary care centre to be tested for systemic amyloidosis. Extensive testing was done, and results were negative for systemic amyloidosis. He remains asymptomatic, takes regular nasal steroids, does regular nasal douching and is closely followed up in an outpatient otolaryngology clinic.

In our review of the literature available, we have identified twenty-nine documented cases of sinonasal amyloidosis, these have been published from the year 1990 to 2023, these cases have been succinctly presented in (Table 1).[4-31] The mean age at the point of presentation was found to be 46 years, with a range of 8 to 81 years old. Notably, four cases were identified in the paediatric population. It was further noted there was a female predominance with a 19 to 10, female-to-male ratio. Nasal obstruction, epistaxis, and nasal discharge emerged as the most frequently cite symptoms with frequencies occurrences documented as 17, 12 and 7 cases respectively. Less common symptoms included hypo/anosmia (3 cases), hearing loss (2 cases), headache (2 cases), epiphora (2 cases), nasal bridge broadening (2 cases), general malaise (1 case), facial pain (1 case), and facial swelling (1 case). Within the cohort of the 29 cases reviewed all patients underwent a computed tomography (CT) scan of their sinuses to investigate the lesion further and for preoperative planning. Additionally, 44% of patients underwent magnetic resonance imaging (MRI) as a supplemental diagnostic modality. Among the 25 patients that were subjected to testing for systemic amyloidosis, 92% (23 cases) yielded negative results whilst 8% (2 cases) tested positive, highlighting that the localised nature of sinonasal amyloidosis is the majority. Overall, 79% (22 cases) of patients underwent surgical excision of the lesions, surgical approaches varied from endoscopic excision, to combined septoplasty and turbinectomies. Of this cohort, five causes required subsequent management (including revision surgeries) due to documented recurrence or residual disease. Two patients had surgical management via the insertion of a ventilation tube in the eardrum to address aural symptoms. Conservative management, marked by a 'watchful waiting' approach was deemed appropriate for three cases. Additionally, three cases were managed with chemotherapy, two with systemic steroids and one case was managed with external beam radiotherapy due to an inoperable lesion. The average duration of follow-up across the cohort 15 months, with five cases noted to have no recurrence at follow-up appointments. One case, however, reports of a patient representing 8 years later with an inoperable recurrence necessitating palliative management.

DISCUSSION

The incidence of amyloidosis is 5-10 million per million per year, with approximately 20% of cases being in the head and neck region.[32] Within the head and neck region, 61% of cases occur in the larynx, 23% oropharynx, 9% trachea, 4% orbit and less than 3% identified in the nasopharynx.[33] Amyloidosis is diagnosed on histopathological testing of tissue samples, often the diagnosis can be a surprise to the clinical team as in this case.

Table 1: Review of previously reported cases of sinonasal amyloidosis; 1990-2023.

Source	Presenting complaint	Age	Sex	Past medical history	Invest-igation	Location of lesion	Histology results	Systemic amyloidosis testing	Management	Outcome
2023 Our case	Right sided hearing loss	78	M	Atrial fibrillation, hypertension and previous deep vein thrombosis	CT and MRI	Right nasal cavity	Apple-green birefringence under polarised light on Congo red staining.	Negative	Functional endoscopic sinus surgery	6 months follow up, no recurrence
2023 Wentland et al.[4]	Nasal obstruction	65	M	Hypertension, hypercholesterolemia and obstructive sleep apnoea	CT and MRI	Sphenoid sinus	Apple-green birefringence under polarised light on Congo red staining.	Negative	Endoscopic sinus surgery	n/a
2023 Gomes et al.[5]	Nasal obstruction and ear fullness	72	F	Hypertension and dyslipidemia	CT	Right nasal cavity	Apple-green birefringence under polarised light on Congo red staining.	Negative	Watchful waiting	n/a
2022 Lombo et al.[6]	Nasal bridge broadening	70s	M	Ischaemic and valvular heart disease and aortic aneurysm	CT	Right nasal cavity	Apple-green birefringence under polarised light on Congo red staining.	Negative	Watchful waiting	8 months follow up, no recurrence
2021 Takakura et al.[7]	Hyposmia and nasal discharge	41	M	Atopic dermatitis and allergic rhinitis	CT and MRI	Bilateral nasal cavities	Stained black by von Kossa staining, and Apple-green birefringence under polarised light on Congo red staining.	Negative	Septoplasty and bilateral inferior turbinectomy	8 months follow up, no recurrence
2020 Onishi et al.[8]	Unilateral blurred vision	75	F	Cervical cancer, cataract, hives, hyperlipidaemia, and hypertension	CT and MRI	Bilateral maxillary sinuses and right nasal septum	Hematoxylin-eosin staining of the resected tissues positively stained with both Congo red and DFS stain	Positive	Endoscopic sinus surgery	1 year follow up, no recurrence
2019 Singh et al.[9]	Nasal obstruction	14	M	Nil	CT	Bilateral nasal cavities	Hematoxylin and Eosin staining, homogenous pinkish material was seen which, on staining with Congo red, showed apple green birefringence	Negative	nil	n/a

Continued.

Source	Presenting complaint	Age	Sex	Past medical history	Invest-igation	Location of lesion	Histology results	Systemic amyloidosis testing	Management	Outcome
2019 Iliev et al.[10]	Nasal obstruction and nasal bridge broadening	81	F	Nil	CT	Frontal sinus, ethmoid sinus extending into the right orbit.	consistent with an amyloid structure; a periglandullar inflammation	Negative	Non-operapble, external beam radiotherapy	9 months follow up
2019 Nur Wahidah et al.[11]	Nasal obstruction and epistaxis	61	M	Hypertension, and elevated body mass index	CT	Left inferior turbinate, extending into the nasopharynx.	Deposits of amorphous eosinophilic material and the presence of amyloid on Congo-red staining with apple-green birefringence when viewed with polarized light	Negative	Surgical excision	12 months follow up
2016 Nishimura et al.[12]	Headache, proptosis and nasal obstruction	60	F	nil	CT	Across nasal cavity and extending into the left orbit	Eosinophilic amorphous deposits that had a greenish-yellow tone under polarized light	Negative	Chemotherapy followed by endsoscopic surgery	18 months follow up
2016 Kumar et al.[13]	Nasal obstruction, hearing loss and epistaxis	55	M	n/a	CT	Nasal cavity extending to skull base	Apple-green birefringence under polarised light on Congo red staining.	Negative	Surgical excision	n/a
2014 Forde et al.[14]	Epistaxis and nasal discharge	74	F	B12 deficiency, sigmoid amyloidosis	n/a	Left sphenoid sinus	Apple-green birefringence under polarised light on Congo red staining.	Positive	Surgical excision and SPA ligation, chemotherapy and steroids	8 months follow up, no recurrence
2014 Doshi et al.[15]	Nasal obstruction and epiphoria	70	F	Lymphoma	n/a	Right inferior turbinate extending to nasopharynx	n/a	n/a	n/a	n/a
2013 Rauba et al.[16]	Nasal stiffness, nasal discharge, anosmia and epistaxis	53	F	nil	CT and MRI	Left maxillary, frontal, and sphenoidal sinuses with deformation and erosions of the left orbital wall	Tissue stroma full of amorphous eosinophilic KR+ substance.	Negative	Ethmoidectomy	6 and 18 months follow up
2013 Cunningham	Nasal obstruction	67	F	n/a	CT	Left nasal cavity	Apple-green birefringence under	Negative	Functional endoscopic	Revision surgery 1

Continued.

Source	Presenting complaint	Age	Sex	Past medical history	Invest-igation	Location of lesion	Histology results	Systemic amyloidosis testing	Management	Outcome
et al.[17]	and epistaxis						polarised light on Congo red staining.		sinus surgery and debulking	year later
2013 Mirza et al.[18]	Hearing loss	31	F	Polycystic ovary disease and focal migraines	CT and MRI	Left nasopharynx lesion obstructing the eustachian tube opening	Apple-green birefringence under polarised light on Congo red staining.	Negative	Grommet, watchful waiting	f/u and 24 months. Recurrence at 8 years - unresectable and managed palliatively
2012 Naidoo et al.[19]	Nasal obstruction, nasal discharge and hyposmia	50	F	nil	CT	Roof of the nose.	n/a	Negative	Revision endoscopic surgery	6 weeks follow up
2012 Nakyama et al.[20]	Anosmia, facial pain and epistaxis	14	F	n/a	CT and MRI	Bilateral middle meatuses and olfactory clefts	acellular eosinophilic material which stained apple-green birefringence under polarised light on Congo red staining.	Negative	Endoscopic excision	Further surgery and 9 months, follow up for 4 years
2012 Nakyama et al.[20]	Epistaxis	27	F	n/a	CT	Bilateral inferior turbinates	Apple-green birefringence under polarised light on Congo red staining.	Negative	Grommet	n/a
2011 Sadeghipour et al.[21]	Nasal obstruction and epistaxis	9	F	n/a	CT	Bilateral nasal mass	Apple-green birefringence under polarised light on Congo red staining.	Negative	Surgically excised	n/a
2010 Pearlman et al.[22]	Nasal obstruction, malaise and diffuse arthralgia	55	F	Rheumatoid arthritis, and monoclonal gammopathy of undetermined significance	CT	Bilateral maxillary sinuses, nasal septum, and inferior turbinates	Apple-green birefringence under polarised light on Congo red staining.	Negative	Steroid treatment	n/a
2009 Sass et al.[23]	Nasal obstruction, nasal discharge and	46	M	n/a	CT	Left axillary sinus and nasal meatus, maxillary sinus medial wall, nasal	Squamous mucosa fragments with a stressed amyloid deposition at the core	Negative	Endoscopic excision	3 years follow up

Continued.

Source	Presenting complaint	Age	Sex	Past medical history	Invest-igation	Location of lesion	Histology results	Systemic amyloidosis testing	Management	Outcome
	epistaxis					infundibula and nasal septum	in the middle of fibroblast.			
2003 Teo et al.[24]	Nasal obstruction and epiphora.	18	F	n/a	CT and MRI	Right inferior turbinate	n/a	Negative	Right inferior turbinectomy	Recurrence at 3 years
2008 Prasad et al.[25]	Nasal obstruction and epistaxis	42	F	Diabetes	CT and MRI	Less nasal cavity	Apple-green birefringence under polarised light on Congo red staining.	Negative	Endoscopic excision	8 months follow up
2008 Ali et al.[26]	Cerebrospinal fluid leak	48	M	n/a	CT	Sphenoid body and sphenoid sinus extending posteriorly and to the roof of the nasopharynx	Apple-green birefringence under polarised light on Congo red staining.	n/a	Endoscopic excision	12 months follow up
Chin et al.[27]	Nasal stuffiness and dysphonia	21	M	nil	CT and MRI	Bilateral nasal cavities	Apple-green birefringence under polarised light on Congo red staining.	Negative	Surgical excision	n/a
2001 Tsikoudas et al.[28]	Nasal obstruction and nasal discharge	53	F	n/a	CT and MRI	n/a	Apple-green birefringence under polarised light on Congo red staining.	Negative	Biopsy and debulking	3 years follow up
2001 Pang et al.[29]	Nasal obstruction, nasal discharge, epiphora, and epistaxis	10	F	nil	CT and MRI	Right nasal cavity	n/a	Negative	Diagnostic surgery	3 years follow up
1997 Birchall et al.[30]	Facial swelling	38	F	nil	CT and MRI	Right maxilla with protrusion through the roof of the mouth	n/a	n/a	Hemi-maxillectomy and reconstruction	n/a
1990 Mufarrji et al.[31]	Nasal obstruction, epistaxis, and headache	8	M	n/a	CT	Roof of nasopharynx	n/a	n/a	Ethmoidectomy	15 months follow up

amyloid is recognised by its amorphous structure and affinity for the dye Congo red and its increased birefringence under polarised light after staining.[3] Once a histopathological diagnosis is made, further assessment is required to investigate systemic involvement of the disease. Investigations include kidney and liver function tests, echocardiography, serum and urine protein electrophoresis and abdominal or rectal biopsies.[27] As mentioned, the disease progression and management of amyloidosis is very different depending on location and systemic spread. Systemic amyloidosis is the more common form and carries a much higher morbidity and mortality rate. Amyloid deposition in vital organs such as the liver, kidneys, hear and gastrointestinal tract can lead to organ toxicity and subsequent dysfunction.[1] Treatment options of systemic amyloidosis include chemotherapy, immunotherapy, stem cell transplant and organ transplantation.[34] Treatment is often supportive rather than curative and prognosis is generally based on the type of amyloidosis and progression of the disease, with 5 year survival rates varying between 27 and 97%.[35] Medical treatment including chemotherapy and immunotherapy have little role in managing localised amyloidosis. The first line of treatment is surgical debridement to provide symptomatic benefit. 50% of cases recurrence occurs and revision surgery is often required.[19] If the patient is asymptomatic from the lesion a watchful waiting approach can be taken.[6] In our case, as with many others, as malignancy is of top differential, it is often inevitable the patient will undergo endoscopic sinus surgery to obtain biopsies of the lesion. In our particular case, as the patient was also suffering from sinus disease as identified on the CT scan, debridement of the sinonasal compartments was also performed alongside debulking of the lesion.

CONCLUSION

Given the rarity and complexity of sinonasal amyloidosis, it is important for clinicians to be aware of this condition and to consider it in the differential diagnosis of patients presenting with sinonasal symptoms and less commonly hearing loss. Endoscopic sinus surgery is the mainstay of treatment, with a high success rate in relieving symptoms and improving quality of life. Whilst, systemic amyloidosis can be treated with chemotherapy and anti-inflammatory medications, there is no definitive treatment for localised amyloidosis. Overall, localised sinonasal amyloidosis has good outcomes however recurrence of amyloid deposits is common, and long-term follow-up is necessary.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Zhuang Y-L, Tsai T-L, Lin C-Z. Localized amyloid deposition in the nasopharynx and neck, mimicking nasopharyngeal carcinoma with neck metastasis. J Chin Med Assoc. 2005;68(3):142-5.
2. Gertz MA, Dispenzieri A. Systemic amyloidosis recognition, prognosis, and therapy. JAMA. 2020; 324(1):79.
3. Blancas-Mejía LM, Ramirez-Alvarado M. Systemic amyloidoses. Ann Rev Biochem. 2013;82(1):745-74.
4. Wentland K, Shukairy MK, Picken MM, Patadia MO. Localized amyloidosis of the sphenoid sinus: A case report and a Descriptive Literature Review. Cureus. 2023.
5. Gomes S, Santos S, Silva M, Ferreira J, Ponte T. Amyloidosis localized to the sinonasal tract: From the diagnosis to management of disease. Cureus. 2023.
6. Lombo C, Matos C, Fonseca R. Sinonasal localised amyloidosis: An uncommon location. BMJ Case Rep. 2022;15(3):23-9.
7. Takakura H, Tachino H, Takii K, Imura J, Shojaku H. Localized amyloidosis of the nasal mucosa: A case report and review of the literature. Front Surg. 2021; 8:469-73.
8. Onishi T, Yasuda M, Koida A, Inui T, Okamoto S, Hirano S. A case of primary systemic amyloidosis involving the sinonasal tract. Ear Nose Amp Throat J. 2020;100(9):765-8.
9. Singh A, Handa KK, Kumar A. Idiopathic isolated nasal amyloidosis: report of a rare case with review of literature. Indian J Otolaryngol Head Neck Surg. 2019;71:2106-9.
10. Iliev G, Ivanova P, Kerimov K, Petya G, Kalchev K. Localised, isolated amyloidosis of the nose and paranasal sinuses. Otolaryngol Head Neck Surg. 2019;5:1-4.
11. Wahid NW, Abed T, Meghji S, Gilbertson J, Barnes M. Localized sinonasal amyloidosis. Allergy Rhinol. 2019;10:21.
12. Nishimura K, Tanaka S, Takahashi Y, Uchida Y, Tanigawa T, Ueda H, et al. Huge localized amyloidosis of the sinonasal cavity: a rare case report. J Clin Case Rep. 2016;6:797.
13. Kumar B, Pant B, Kumar V, Negi M. Sinonasal globular amyloidosis simulating malignancy: a rare presentation. Head Neck Pathol. 2016;10:379-83.
14. Forde R, Ashman H, Shah D, Williams E. Primary amyloidosis of the nose presenting with refractory epistaxis and systemic involvement a rare phenomenon. West Indian Med J. 2013;62:296-9.
15. Doshi PH, Roman B, Lim J, Shatzkes DR. A rare sinonasal entity. sinonasal amyloidosis. JAMA Otolaryngol Head Neck Surg. 2014;140:477-8.
16. Rauba D, Lesinskas E, Petrulionis M, Sukyte D, Valeviciene N, Palionis D, et al. Isolated nasal amyloidosis: a case report. Medicina. 2013;49:497-503.
17. Cunningham A, Kalwani S, Alsanjari N, Fayad G. Rare subtype of localised nasal amyloidosis. Otorhinolaryngol. 2013;6:60-3.
18. Mirza AH, El-Shunnar S, Sama A. Nasopharyngeal Amyloidosis: An unusual cause of unilateral hearing loss. J Surg Case Rep. 2013;2013(2):48.

19. Naidoo YS, Gupta R, Sacks R. A retrospective case review of isolated sinonasal amyloidosis. J Laryngol Otol. 2012;126:633-7.
20. Nakayama T, Otori N, Komori M, Takayanagi H, Moriyama H. Primary localized amyloidosis of the nose. Auris Nasus Larynx. 2012;39:107-9.
21. Sadeghipour A, Mirzaie AZ, Mohammadi Sh, Nilipour Y. Primary localized nasal amyloidosis in a child, a rare case report. Int J Pediatr Otorhinolaryngol. 2011;6(4):310-2.
22. Pearlman AN, Jeffe JS, Zynger DL, Yeldandi AV, Conley DB. Localized amyloidosis of the nasal and paranasal mucosa: a rare pathology. Am J Otolaryngol. 2010;31:130-1.
23. Saas SMG, Pinto MC, Campos DS, Maeda CAS, Mello PF. Localized nasopharyngeal amyloidosis. Intl Arch Otorhinolaryngol. 2009;13:207-10.
24. Teo D. Recurrent localized Sinonasal Amyloidosis: A case report. Otolaryngol Head Neck Surg. 2003; 129(2):23-9.
25. Prasad M, Abdulla M, Aroor R, Somayaji G. Primary nasal amyloidosis. J Otorhinolaryngol. 2009;9(2):43-7.
26. Ali E, Phillip R, Prepageran N, Peh S. Cerebrospinal fluid rhinorrhoea secondary to amyloidosis of the sphenoid sinus. Med J Malaysia. 2008;63(4):341-2.
27. Chin S, Fatterpeckar G, Kao C, Chen C. Amyloidosis concurrently involving the sinonasal cavities and larynx. AJNR Am J Neuroradiol. 2004;25(4):636-8.
28. Tsikoudas A, Martin-Hirsch DP, Woodhead CJ. Primary Sinonasal Amyloidosis. J Laryngol Amp Otology. 2001;115(1):55-6.
29. Pang KP, Chee LW, Busmanis I. Amyloidoma of the nose in a pediatric patient: A case report. Am J Otolaryngol. 2001;22(2):138-41.
30. Birchall D, Fields JM, Poon CL. Case report: Focal Amyloidosis of the maxillary antrum: Plain film, CT and mr appearances. Clin Radiol. 1997;52(5):392-4.
31. Mufarrij AA, Busaba NY, Zaytoun GM, Gallo GR, Feiner HD. Primary localized amyloidosis of the nose and paranasal sinuses. Am J Surg Pathol. 1990;14(4): 379-83.
32. Jacques T, Stearns M, Hawkins P, Giddings C. Head and neck manifestations of amyloidosis. Otolaryngologist. 2013;6(1):35-40
33. Panda NK, Saravanan K, Purushotaman GP, Gurunathan RK, Mahesha V. Localized amyloidosis masquerading as nasopharyngeal tumor: A Review. Am J Otolaryngol. 2007;28(3):208-11.
34. Girnius S. Overview of systemic and localized amyloidosis. Rev Health Care. 2013;4(4):231-47.
35. Joss N. Presentation, survival and prognostic markers in AA amyloidosis. QJM. 2000;93(8):535-42.

Cite this article as: Suseeharan M, Dusu K, Motter D, Balfour A. Localised sinonasal amyloidosis presenting with unilateral hearing loss: a case report and review of the literature. Int Surg J 2024;11:107-14.