International Journal of Surgery Case Reports 94 (2022) 107056

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Primary nasopharyngeal amyloidosis with nasal polyposis: Case report of a diagnostic challenge

Stacy A. Jamarun [a], Eugene Hung Chih Wong [b,*]

[a] Department of Otorhinolaryngology, Queen Elizabeth Hospital, Kota Kinabalu, Sabah, Ministry of Health, Malaysia
[b] Department of Otorhinolaryngology, Faculty of Medicine and Health Sciences, Universiti Malaysia Sabah, Kota Kinabalu, Sabah, Malaysia

ARTICLE INFO

Keywords:
Nasopharyngeal
Amyloidosis
Nasal
Polyposis

ABSTRACT

Background: Primary nasopharyngeal amyloidosis is a rare entity of localised amyloidosis. Patients usually present with symptoms that mimic other common nasal diseases. We report an unusual case of nasopharyngeal amyloidosis that *co*-exist with nasal polyposis at the same time.

Case presentation: We described a 72-year-old gentleman who presented with left-sided nasal obstruction, rhinorrhoea and hyposmia and right-sided hearing loss. Examination revealed bilateral intranasal polyposis with right lobular swelling at torus tubarius and right sided middle ear effusion. Biopsy revealed inflammatory nasal polyps with nasopharyngeal amyloidosis. Patient was treated successfully with functional endoscopic sinus surgery (FESS) for nasal polyposis and an en bloc wide local excision of the torus tubarius with no signs of recurrence at one year follow-up.

Conclusion: Clinicians should have raised index of suspicion of a possible primary nasopharyngeal amyloidosis in patients presenting with nasopharyngeal mass with *co*-existing nasal polyposis to avoid delay in diagnosis and treatment.

1. Introduction

Amyloid fibrils are protein polymers that play a variety of important physiological roles such as in memory formation and hormone storage. Amyloidosis is disease of abnormal intracellular or extracellular deposition of these amyloid fibrils that alters the normal function of tissues. Generally, amyloidosis can be categorised based on the number of organs involved. Involvement of a single organ is called localised amyloidosis while involvement of two or more organs is called systemic amyloidosis [1–3]. Majority of the head and neck amyloidosis are localised [4].

Primary nasopharyngeal amyloidosis is a rare benign tumour in the head and neck region where the usual clinical presentation includes nasal obstruction, rhinorrhoea, epistaxis and reduced hearing [5–7], all of which can mimic symptoms of other nasal diseases.

We report a rare case (in line with the SCARE criteria [8]) of primary nasopharyngeal amyloidosis that *co*-exist with bilateral nasal polyposis, which was successfully treated with surgical excision.

2. Case presentation

A 72-year-old gentleman presented with a five years history of left-sided nasal obstruction, associated with hyposmia and rhinorrhoea. There was no history of epistaxis, facial pain, post-nasal drip or mucopus discharge. He also complained of two years of reduced hearing on the right side with no other otological symptoms.

Nasendoscopy revealed bilateral intranasal polyposis (Grade 3 on the left and Grade 2 on the right) with a smooth, lobular swelling in the nasopharynx located at the right torus tubarius, partially obstructing the right Eustachian tube (Fig. 1). Otoscopy of the ears showed a right sided middle ear effusion. Other ENT examinations were unremarkable.

A biopsy of the right torus tubarius lesion was performed under local anaesthetic in clinic where the histopathological analysis demonstrated polypoidal tissue lined by respiratory epithelium containing deposition of amorphous eosinophilic material within the stroma (Fig. 2), which stained positive for amyloid with Congo red stain (Fig. 3) with apple green birefringence appearance under polarized microscopy (Fig. 4). Computed tomography (CT) scan of the paranasal sinus revealed soft tissue density in both nasal cavity and maxillary region with no

* Corresponding author.
E-mail address: eugenewong@ums.edu.my (E.H.C. Wong).

https://doi.org/10.1016/j.ijscr.2022.107056
Received 17 February 2022; Received in revised form 1 April 2022; Accepted 5 April 2022
Available online 7 April 2022

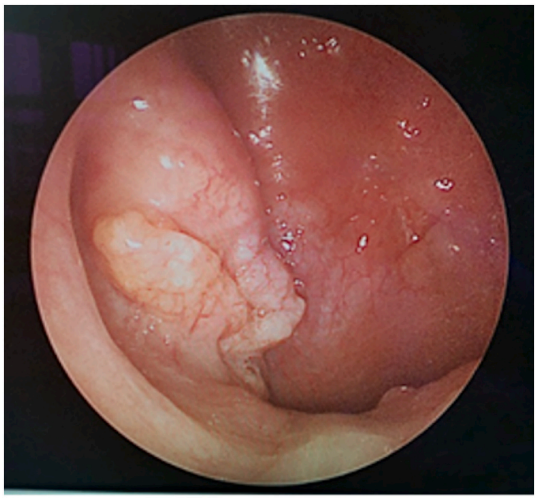

Fig. 1. Endoscopic view of the right nasopharynx showing a mass at the right torus tubarius.

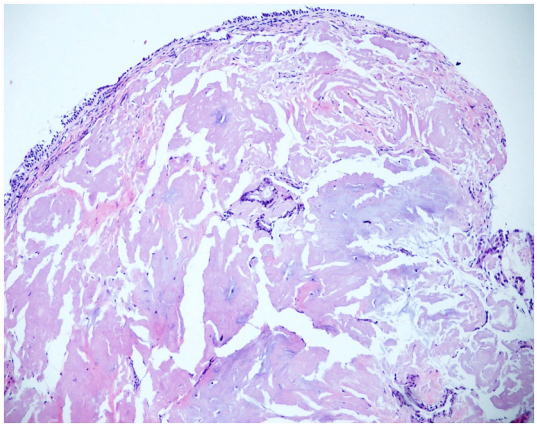

Fig. 2. Histopathological slide (with Haematoxylin and Eosin stain) showing polypoidal tissue lined by respiratory epithelium containing deposition of amorphous eosinophilic material within the stroma.

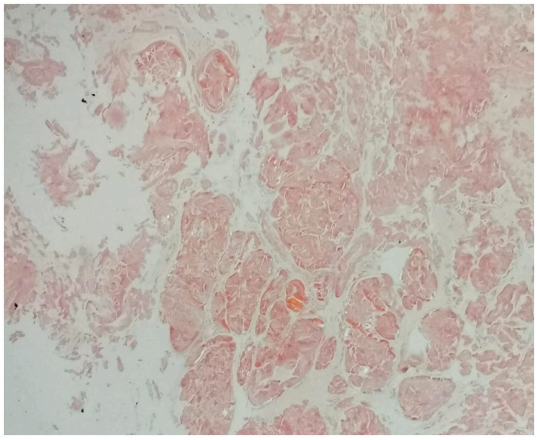

Fig. 3. Histopathological slide (with Congo Red stain) showing extensive amyloid deposition. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

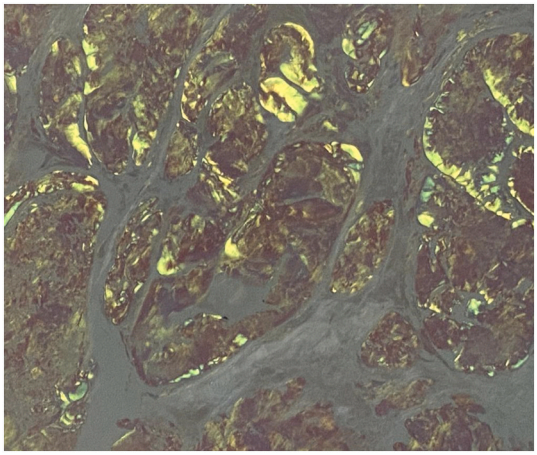

Fig. 4. Histopathological slide showing apple green birefringence appearance under polarized microscopy. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

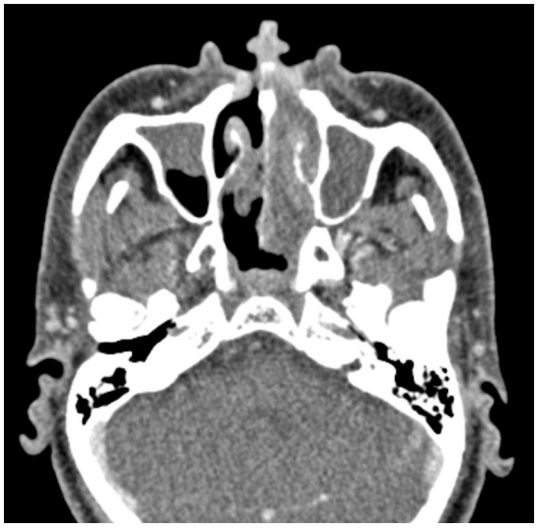

Fig. 5. Axial cut of CT scan of paranasal sinus showing soft tissue density at bilateral nasal cavity and torus tubarius with no obvious mass detected in the Eustachian tube.

detectable mass in the Eustachian tube (Fig. 5). Other investigations, including blood and urine profiles, chest X-ray and ECG, were all normal and systemic amyloidosis was ruled out. A diagnosis of primary nasopharyngeal amyloidosis with bilateral intranasal polyposis was made.

The patient was treated with tapering doses of Prednisolone prior to a functional endoscopic sinus surgery (FESS) to remove the nasal polyps. A wide local excision of the torus tubarius mass was also performed where a resection margin of 1 cm was outlined using monopolar diathermy tip and the lesion was dissected in a plane between the pharyngobasilar fascia and prevertebral muscles and was removed en bloc. Histopathology analysis confirmed the diagnosis of torus tubarius amyloidosis and inflammatory nasal polyps. Post-operatively, the symptoms of nasal obstruction and hearing loss resolved with no signs of disease recurrence at one year surveillance during follow-up in the clinic.

3. Discussion

Amyloidosis is a group of diseases characterised by accumulation of extracellular amyloid fibrils, which normally occur without any known

cause [2]. The build-up of these amyloid protein deposits disrupts the normal function of tissues and organs.

Approximately two third of patients with amyloidosis have several organs involved at presentation [3] (systemic amyloidosis) while localised amyloidosis is less common and accounts for about 10–20% of all cases of amyloidosis [6]. Common clinical manifestations of systemic amyloidosis include non-diabetic nephrotic range proteinuria, cardiac failure with left ventricular hypertrophy without evidence of aortic stenosis or hypertension, unknown cause of peripheral or autonomic neuropathy and hepatomegaly with and elevated ALP with normal imaging appearance [3].

On the other hand, localised amyloidosis usually presents with symptoms related to the affected organ and is usually not life-threatening. The typical sites for localised amyloid depositions are the skin, ocular or periorbital area, respiratory tract, gastrointestinal tract, urinary tract and head and neck region. Localised head and neck amyloidosis are most commonly seen in the larynx (61%), oropharynx (23%), trachea (9%) and obit (4%) while other rarer sites that can be involved include the tongue, nasal cavity, paranasal sinuses, salivary glands and cervical lymph nodes [6]. The local recurrence rate of localised amyloidosis is about 30% and they rarely progresses to systemic disease [9].

Primary nasopharyngeal amyloidosis is extremely rare with only 32 reported cases in the English and German literature so far [6]. Patients usually present with nasal obstruction, nasal discharge, post-nasal drip, epistaxis and conductive hearing loss due to otitis media with effusion [6]. Our patient is interesting because his initial presentation was for the nasal polyposis on the opposite side of the amyloidosis where he experienced nasal obstruction, rhinorrhoea and hyposmia. The only symptom he had secondary to the amyloidosis was two years history of reduced hearing on the side of lesion.

Imaging studies may be useful to help define the disease extent [10] where early enhancement on dynamic contrast-enhanced MRI may suggest the diagnosis [11]. Confirmation of localised amyloidosis is usually obtained through tissue biopsy with typical characteristics of congophilia with associated green birefringence appearance when viewed with polarized light [2,12]. Our patient's biopsy tissue had all the typical appearance described above.

Patients with confirmed nasopharyngeal amyloidosis also require further assessment to exclude systemic diseases since treatment for systemic amyloidosis can be challenging and require a multidisciplinary approach depending on the organs involved [6]. On the other hand, localised amyloidosis are mainly managed with surgical excision and surveillance for recurrence [6] although extensive amyloid deposits where excision would cause more significant morbidity can be managed with watchful waiting especially when disease progression has been found to be slow in general [6]. Some authors have also suggested adjuvant radiotherapy in surgically challenging cases [13]. In our patient, a FESS had to be performed first to remove the bilateral inflammatory nasal polyps to allow easier surgical access to the nasopharynx for en bloc surgical excision of the amyloidosis. Post-operatively, patient's symptoms resolved with no signs of disease recurrence during his one-year follow-up.

4. Conclusion

Primary nasopharyngeal amyloidosis is a rare clinical entity that can mimic other common nasal diseases. Clinicians should have raised index of suspicion for primary nasopharyngeal amyloidosis as a potential differential diagnosis in patients with nasopharyngeal mass, which can *co*-exist with other common nasal diseases such as nasal polyposis in our case, to ensure early diagnosis and disease treatment.

Source of funding

None.

Ethical approval

Ethical approval was not required for case report in the institution.

Consent

Written informed consent was obtained from the patient's relative for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

NA.

Guarantor

This is a retrospective case report. The primary author (Dr Eugene Wong) takes full responsibility for the work.

Provenance and peer review

Not commissioned, externally peer-reviewed.

CRediT authorship contribution statement

Stacy A Jamarun – Methodology, Data Curation, Writing - Original Draft.

Eugene Hung Chih Wong – Supervision, Data Curation, Writing – Review & Editing.

Declaration of competing interest

None.

References

[1] F. Perfetto, A. Moggi-Pignone, R. Livi, A. Tempestini, F. Bergesio, M. Matucci-Cerinic, Systemic amyloidosis: a challenge for the rheumatologist, Nat. Rev. Rheumatol. 6 (2010) 417–429.
[2] J.D. Sipe, M.D. Benson, J.N. Buxbaum, S.I. Ikeda, G. Merlini, M.J. Saraiva, et al., Amyloid fibril proteins and amyloidosis: chemical identification and clinical classification international society of amyloidosis 2016 nomenclature guidelines, Amyloid 23 (2016) 209–213.
[3] I. Vaxman, M. Gertz, When to suspect a diagnosis of amyloidosis, Acta Haematol. 143 (2020) 304–311.
[4] C.H. Chen, C.H. Sun, Multifocal head and neck amyloidosis as a diagnostic clue of systemic lupus erythematosus (sle): a case report, Medicine (Baltimore) 98 (2019), e16830.
[5] M. Durbec, A. Ambrun, R. Barnoud, M. Poupart, J.C. Pignat, O. Merrot, Localized nasopharyngeal amyloidosis, Eur. Ann. Otorhinolaryngol. Head Neck Dis. 129 (2012) 160–162.
[6] G. Sakagiannis, E. Giotakis, L.D.R. Thompson, Localized nasopharyngeal amyloidosis: a clinicopathologic series of 7 cases with a literature review, Head Neck Pathol. 12 (2018) 542–547.
[7] A. Patel, S. Pambuccian, R. Maisel, Nasopharyngeal amyloidosis, Am. J. Otolaryngol. 23 (2002) 308–311.
[8] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, Group S, The scare 2020 guideline: updating consensus surgical case report (scare) guidelines, Int. J. Surg. 84 (2020) 226–230.
[9] M.L. Biewend, D.M. Menke, K.T. Calamia, The spectrum of localized amyloidosis: a case series of 20 patients and review of the literature, Amyloid 13 (2006) 135–142.
[10] A.D. Gean-Marton, C.F. Kirsch, L.G. Vezina, A.L. Weber, Focal amyloidosis of the head and neck: evaluation with ct and mr imaging, Radiology 181 (1991) 521–525.
[11] U. Motosugi, T. Ichikawa, T. Araki, S. Endo, K. Masuyama, T. Nakazawa, Localized nasopharyngeal amyloidosis with remarkable early enhancement on dynamic contrast-enhanced mr imaging, Eur. Radiol. 17 (2007) 852–853.
[12] H.L. Nienhuis, J. Bijzet, B.P. Hazenberg, The prevalence and management of systemic amyloidosis in western countries, Kidney Dis. (Basel) 2 (2016) 10–19.
[13] M. Luo, G. Peng, L. Shi, X. Ming, Z. Li, S. Fei, et al., Intensity-modulated radiotherapy for localized nasopharyngeal amyloidosis: case report and literature review, Strahlenther. Onkol. 192 (2016) 944–950.