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CLINICAL RECORD

Acquired nasopharyngeal stenosis in a patient with sarcoidosis

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Abstract

Introduction: Acquired nasopharyngeal stenosis typically occurs as a result of surgery or irradiation of the nasopharynx. Sarcoidosis has numerous manifestations in the head and neck region, although an association with nasopharyngeal stenosis has not previously been reported.

Case report: A 40-year-old man with sarcoidosis developed severe acquired nasopharyngeal stenosis. This was successfully managed with balloon dilatation, followed by pharyngoplasty with local pharyngeal flap reconstruction.

Conclusion: This report is intended to prompt consideration of nasopharyngeal stenosis as a potential cause of nasal obstruction in patients with sarcoidosis, and to draw attention to the need to consider sarcoidosis in the differential diagnosis of patients with acquired nasopharyngeal stenosis. We also demonstrate the viability of pharyngoplasty in the management of nasopharyngeal stenosis in the setting of sarcoidosis.

Key words: Sarcoidosis; Nasopharynx; Stenosis

Introduction

Sarcoidosis has many common head and neck manifestations, including chronic rhinosinusitis, chronic otitis media and laryngeal stenosis. Histopathological analysis shows non-caseating granulomas. Treatment includes steroids, immunosuppressants and occasionally surgery.

Acquired nasopharyngeal stenosis is typically associated with radiotherapy[1] or nasopharyngeal procedures such as adenoidectomy[2] and uvulopalatopharyngoplasty.[3] Assorted surgical interventions have previously been reported, with varied success.[1–5]

We report a case of a patient with sarcoidosis who developed severe acquired nasopharyngeal stenosis, an association not previously reported. We describe successful management with endoscopic balloon dilatation followed by transoral pharyngoplasty.

Case report

A 40-year-old man with a previous diagnosis of sarcoidosis had been treated for chronic rhinosinusitis and chronic otitis media for six years. He had undergone three functional endoscopic sinus surgery procedures, two tympanostomy tube placements and two glottic dilatations. Some of these had been performed elsewhere prior to the diagnosis of his sarcoidosis, which was established by the presence of non-caseating granulomas on sinonasal and glottic biopsy specimens. He had been commenced on methotrexate and prednisone by a rheumatologist.

In December 2008, he developed nasal obstruction, snoring and restless sleep. Flexible nasopharyngolaryngoscopy revealed severe stenosis of the nasopharyngeal port, with an estimated diameter of 4 mm, which had not been present on previous endoscopies. He underwent surgery, with transnasal visualisation of the stenotic nasopharyngeal port via a 70° rigid endoscope (Figure 1), serial dilatation to 15 mm with a balloon catheter (Boston Scientific, Natick, Massachusetts, USA) (Figure 2), and injection with 20 mg of topical triamcinalone (Kenalog; Bristol-Myers-Squibb, Princeton, New Jersey, USA). Improvement was seen on follow up, and the patient reported partial resolution of his symptoms; however, some nasal obstruction and sleep difficulty persisted.

In August 2009, the patient again underwent surgery, during which uvular bifidity and evidence of a submucous cleft palate were observed. A pharyngoplasty was performed by bilateral suturing of the superiorly based posterior tonsillar pillar flaps to the lateral pharyngeal walls, allowing the nasopharyngeal port to be further widened while preserving velopharyngeal competency.

Following this procedure, the patient's symptoms dramatically improved, and he maintained this improvement one year post-operatively. At that time, his only remaining problem was persistent eustachian tube dysfunction.

During subsequent surgery for a tympanostomy tube change and rigid nasal endoscopy, we were able to pass a lighted balloon sinuplasty guidewire (Acclarent, Menlo Park, California, USA) through the patient's myringotomies

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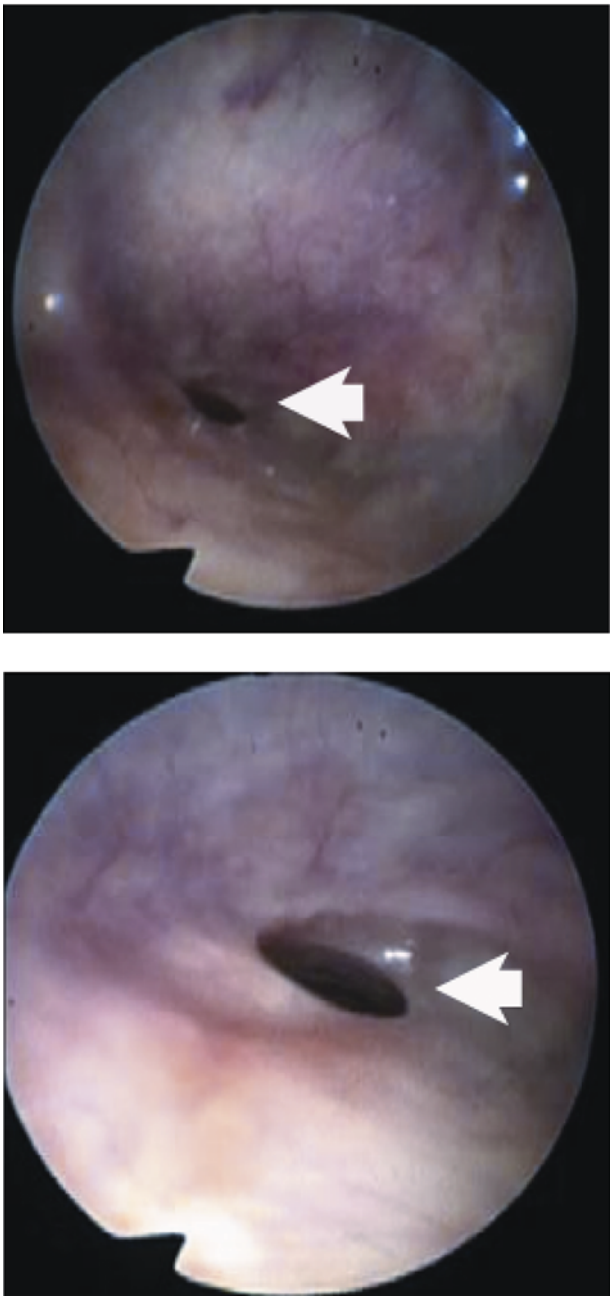

FIG. 1
Pre-operative endoscopic views of the patient's acquired nasopharyngeal port stenosis (arrows), obtained via transnasal flexible fibre-optic endoscopy.

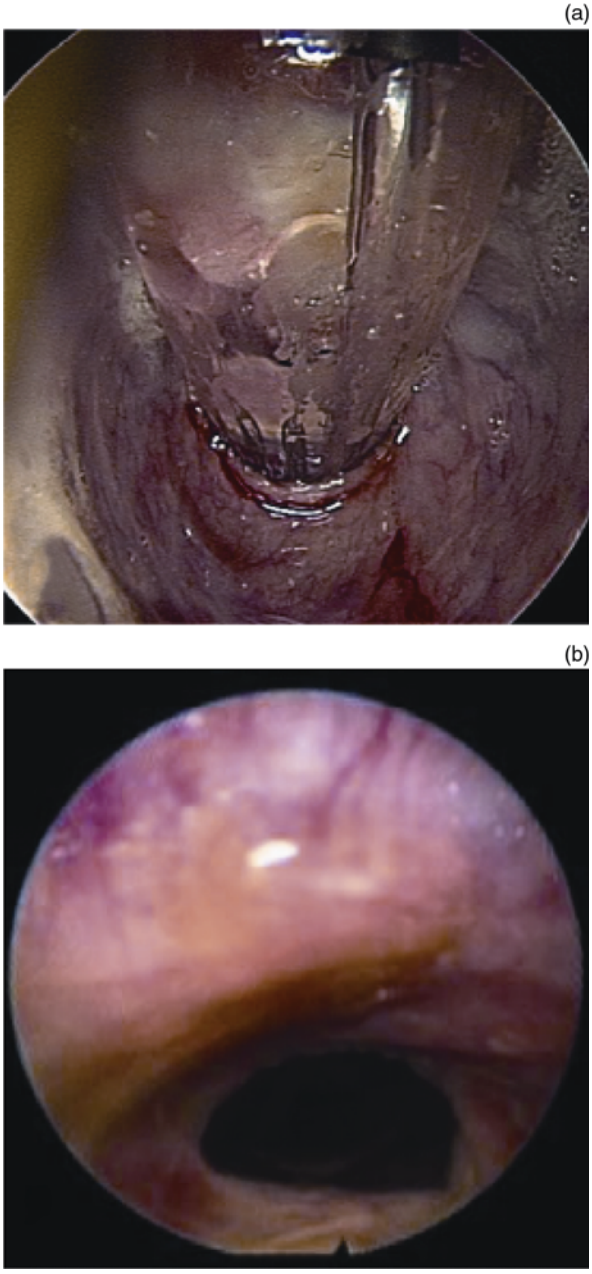

FIG. 2
Endoscopic views showing (a) the balloon catheter expanding the nasopharyngeal port, and (b) the nasopharynx one week after balloon dilatation.

and into the eustachian tubes via the middle-ear cavities, allowing transnasal endoscopic visualisation of a layer of tissue blocking both eustachian tube orifices (Figure 3).

Discussion

Sarcoidosis is a multisystem disease primarily affecting adults younger than 50 years of age, with a peak incidence at 20 to 59 years of age.[6]

The aetiology of sarcoidosis is poorly understood. The most accepted theory is that a non-degradable foreign agent, the identity of which is unclear, initiates an inflammatory reaction mediated by macrophages and cluster of differentiation 4 protein positive ($CD4^+$) helper T cells, resulting in the release of cytokines which further propagate the inflammatory response, eventually leading to the formation of non-caseating epithelioid granulomas.[6,7]

The lungs and intrathoracic lymph nodes are primarily affected, although other areas can also be involved. Approximately 9 per cent of affected patients have head and neck involvement, which can include the middle and inner ears, sinonasal cavities, oral cavity, hypopharynx, and larynx.[8] Nasopharyngeal involvement has been described but is rare.[8]

The diagnosis of sarcoidosis is made from the combination of a consistent clinical picture, typical radiological findings and a positive biopsy. Biopsies are usually obtained from the most easily accessible affected organ, such as the skin, lacrimal glands, nasal mucosa or oral mucosa. Affected tissues tend to have a nodular appearance

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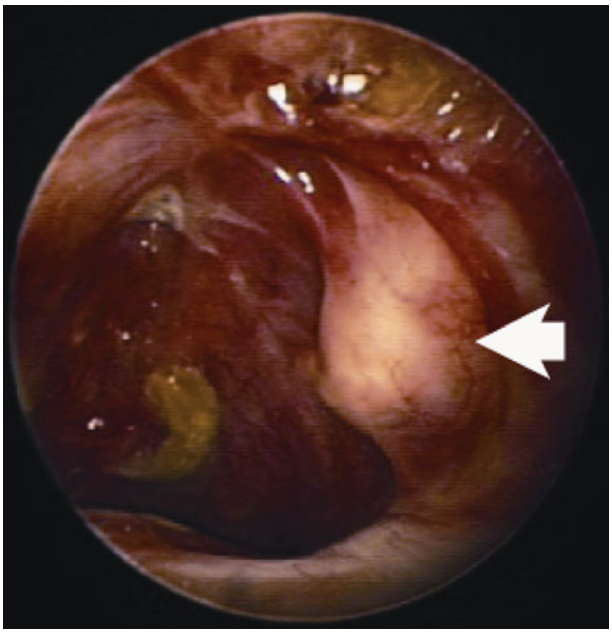

FIG. 3

Transnasal, endoscopic view of the obstructed left eustachian tube orifice (arrow) in the nasopharynx, obtained using a 0° rigid endoscope with a lighted guidewire in the eustachian tube (passed through the middle ear via a previously created myringotomy).

with white or yellow discoloration. Transbronchial biopsy may be appropriate in patients without more easily accessible lesions. The serum concentration of angiotensin-converting enzyme is elevated in 30–80 per cent of patients with sarcoidosis, but it is considered a poor diagnostic test due to its highly variable sensitivity and low specificity.[6,7] An evaluation for sarcoidosis, with a chest X-ray and biopsy, should be considered in any patient presenting with acquired nasopharyngeal stenosis of unknown aetiology.

- **Acquired nasopharyngeal stenosis typically occurs due to surgery or irradiation**
- **In spontaneous cases without a clear aetiology, sarcoidosis should be considered**
- **In sarcoidosis with nasal obstruction, nasopharyngeal stenosis should be considered**
- **A patient with sarcoidosis and acquired nasopharyngeal stenosis is reported**
- **Management with endoscopic balloon dilatation and transoral pharyngoplasty was successful**

We searched the PubMed database using the open-ended key words 'nasopharyn*' and 'sarcoid*' along with the closed-ended key words 'stenotic' and 'stenosis'. We did not find any previous reports of acquired nasopharyngeal stenosis occurring in conjunction with sarcoidosis.

There have been many treatments reported in the literature for the management of acquired nasopharyngeal stenosis, including laser excision,[3,5] 'cold knife' excision[4] and balloon dilatation.[1] Modalities for preventing recurrence have included the placement of stents and obturators,[3,5] the application of mitomycin C,[5,6] and local flap reconstruction.[4] Our case illustrates the successful initial management of acquired nasopharyngeal stenosis in the setting of sarcoidosis, using balloon dilatation. Local steroid injection was also performed in an attempt to prevent recurrence. Unfortunately, our patient showed evidence of symptomatic, residual, low-grade stenosis post-operatively (Figure 2b). Chheda and Postma[1] recently reported the successful use of a similar balloon dilatation technique on three patients with radiation-induced acquired nasopharyngeal stenosis, although one of their patients did require a second dilatation. It is unclear whether a persistent inflammatory process such as sarcoidosis would affect the incidence of long-term recurrence following balloon dilatation, compared with radiation-induced stenosis.

Our patient subsequently underwent transoral pharyngoplasty, which further improved his symptoms to his pre-morbid baseline. The only adverse consequence of this transoral pharyngoplasty was the development of permanent eustachian tube obstruction. It is difficult to determine whether the patient's eustachian tube dysfunction had been worsened by the pharyngoplasty or had simply persisted as a result of the continued inflammatory disease process.

Conclusion

Acquired nasopharyngeal stenosis typically occurs as a result of surgery or irradiation of the nasopharynx. Investigation for sarcoidosis should be considered in patients with spontaneous acquired nasopharyngeal stenosis of unclear aetiology.

Sarcoidosis can present with numerous sinonasal manifestations. Acquired nasopharyngeal stenosis should be considered as a potential cause of nasal obstruction in patients with sarcoidosis.

The presented case demonstrates successful management of acquired nasopharyngeal stenosis in a patient with sarcoidosis, using a combination of endoscopic balloon dilatation and transoral pharyngoplasty. Both these procedures may be considered as first-line treatments for acquired nasopharyngeal stenosis, particularly in patients with rheumatological diseases such as sarcoidosis.

References

1 Chheda NN, Postma GN. Balloon dilation of an acquired nasopharyngeal stenosis. *Otolaryngol Head Neck Surg* 2009;**140**:939–41
2 Cheung VW, Manoukian JJ. Two cases of nasopharyngeal stenosis after multiple adenoidectomies. *J Otolaryngol Head Neck Surg* 2009;**38**:E98–9
3 Krespi YP, Kacker A. Management of nasopharyngeal stenosis after uvulopalatoplasty. *Otolaryngol Head Neck Surg* 2000;**123**:692–5
4 Toh E, Pearl AW, Genden EM, Lawson W, Urken ML. Bivalved palatal transposition flaps for the correction of acquired nasopharyngeal stenosis. *Am J Rhinol* 2000;**14**:199–204
5 Jones LM, Guillory VL, Mair EA. Total nasopharyngeal stenosis: treatment with laser excision, nasopharyngeal obturators, and topical mitomycin-C. *Otolaryngol Head Neck Surg* 2005;**133**:795–8
6 Iannuzzi MC, Rybicki BA, Teirstein AS. Sarcoidosis. *N Engl J Med* 2007;**357**:2153–65
7 Lazarus A. Sarcoidosis: epidemiology, etiology, pathogenesis, and genetics. *Dis Mon* 2007;**55**:649–60
8 Braun JJ, Gentine A, Pauli G. Sinonasal sarcoidosis: review and report of fifteen cases. *Laryngoscope* 2004;**114**:1960–3

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