A Rare Cause of Nasopharyngeal Stenosis

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The case of a Saudi female from south west Saudi Arabia with nasopharyngeal stenosis due to late bejel is presented. The clinical picture and management were described. It is concluded that bejel can present as nasal obstruction and cause nasopharyngeal stenosis.

Nasopharyngeal stenosis can result from different pathologies, namely surgical or non-surgical traumas, acute or chronic nasopharyngitis, tumours (mainly of malignant type) and can be congenital. Bejel disease is one of the chronic, non-venerereal forms of syphilis affecting the nasopharynx in arid, subtropical areas such as the Arabian peninsula.

Case Report

A 50-year-old Saudi female from Bisha (a rural area in south west Saudi Arabia) had attended the otorhinolaryngeal (ORL) clinic with 3 years’ history of headache on and off, bilateral nasal obstruction, nasal tone speech and epigastric pain with decreased appetite and loss of weight. There was no history of trauma or previous surgery. There was no history of extramarital sexual contacts revealed on questioning both the patient and her husband.

On examination, the patient looked generally underweight, her vital signs were stable. The nose showed a pale mucosa and multiple nasal polypi on the left side. The airway was decreased on both sides. The throat showed a thick post-nasal drip and an adhesion between soft palate and posterior pharyngeal wall with a small slit on right side of the nasopharynx through which secretions could be seen. There was scarring of the soft palate and its movements were restricted.

The larynx showed normal anatomy. The neck showed no palpable mass. The patient’s laboratory and radiological investigations, apart from hazy maxillary sinuses, were normal.

The patient was taken to theatre where it was found that the oropharynx showed an adhesion between the soft palate and the posterior pharyngeal wall with a small slit-like opening on the right side. A biopsy was taken from the adhesion area of the soft palate and sent for histopathological studies. The nose showed multiple polypi on the left middle meatus; they were removed and sent for histopathological study.

The oropharynx histopathology showed a non-specific chronic inflammatory reaction with moderate lymphocytic infiltrate and a few plasma cells. No vasculitis or endarteritis obliterans and no gumm formation were seen. The nasal polypi were allergic polypi.

Postoperatively, blood was sent for Venereal Disease Research Laboratory (VDRL) and fluorescent treponemal antibody (FTA) tests which were strongly positive for the patient and negative for her husband.

The diagnosis of bejel was made and the patient was given injections of procaine penicillin 600 000 I/U intramuscularly for 21 days.

She was admitted again for excision of the adhesions and insertion of two Portex tubes—size 7. The tubes were removed 6 weeks later and the nasal airways were quite patent.

Discussion

Nasal obstruction can result from nasopharyngeal stenosis. There are many factors which can contribute to this stenosis such as trauma, acute and chronic nasopharyngitis, tumours and congenital anomalies. Bejel is one of the rare causes of nasopharyngeal stenosis. Bejel is a form
of chronic endemic contagious non-venereal syphilis caused by *Treponema pallidum* which is morphologically and serologically identical to the organism causing pinta and yaws. It seems that racial and environmental factors modify the pathology caused by this organism. Bejel occurs in arid, subtropical areas and shows proximal cutaneous and mucosal lesions while pinta and yaws are present in the tropics and have distal cutaneous lesions.\(^1\) Bejel is characterized by self-limited primary and secondary lesions with a silent course undetected clinically and late lesions with a destructive course. The diagnosis is based on positive serological tests, the clinical manifestations and the geographical distribution. Bejel can be transmitted from one person to another by kissing in the early primary and secondary stages and also by the common use of drinking and eating utensils.

The most common initial lesions in early bejel are buccal mucous patches, skin rashes, condylomata lata in moist skin folds and ostalgias.\(^2\) The early disease is followed by a latent period of undetermined duration. Late manifestations include nasopharyngeal ulcerations which range from palatal perforation to rhinopharyngitis mutilans, gummatous ulceration of skin, palate and nasal septum. Bone lesions are the most frequent manifestations of late bejel affecting the clavicle, the frontal bone, the tibias and other long bones\(^3\) giving rise to swelling, tenderness and pain. There have been no confirmed cases of congenital bejel or of systemic manifestations like those seen in syphilis such as cardiovascular and neurological lesions.\(^4\) Bejel is treated with injectable penicillin, either with one injection of 2.4 mega units of long-acting penicillin or with 600 000 IU procaine penicillin intra-muscularly daily for 3 weeks (which is mandatory in tertiary manifestations of the disease). The long-acting benzathine penicillin is preferred in contact treatment among nomadic tribes.\(^5\) The incidence in Saudi Arabia in nomadic and semi-nomadic Bedouin peoples is 16.75%, whereas in urban people it is 0.32%\(^6\).

The clinical picture described above with the strongly positive serological tests for syphilis without history of extramarital sexual contact and the geographical area from which the patients came confirmed the diagnosis of bejel. Adhesion and scarring of the nasopharynx may have resulted from mucous ulcerations due to bejel. These ulcerations started to heal by fibrosis causing the nasopharyngeal adhesions. The nose and oral cavity were spared as well as other areas. Rhinoscleroma in the cicatrizing stage was excluded as a cause of nasopharyngeal stenosis on the basis of the histopathological report which failed to reveal Mikulicz cells and Russell bodies and the strongly positive VDRL and FTA which were in favour of bejel.

Bejel should be suspected in any case of nasopharyngeal ulceration or stenosis especially in Saudi Bedouins coming from endemic areas. The clinical manifestations, serological test results and epidemiological pattern of this disease make the diagnosis easy.

### References