Pharyngeal webs in a patient with dysphagia and Behcet's disease



Fig. 1 Views during upper gastrointestinal endoscopy in a 64-yearold woman with a previous history of Behcet's disease showing fibrotic rings (black arrows) in: a the hypopharynx; **b, c** the pharynx. **d** A small diverticulum (white arrow) was al so present just above the larynx.

A 64-year-old woman presented with longstanding dysphagia to solids and a recent onset of nocturnal cough and change in her voice. The patient described restriction to the passage of solids at the level of the neck, but denied odynophagia or weight loss. She had a background of Behcet's disease with previous colonic and oropharyngeal involvement. She had also been treated for many years with esophageal dilations for an upper esophageal web. Her symptoms of dysphagia improved temporarily after each dilation; however, typically there was recurrence within 2 weeks of treatment. Video fluoroscopy was performed, which demonstrated a small web in the region of the pharyngo-esophageal junction, above which there was pharyngeal dilatation. At least one episode of aspiration occurred during this procedure.

After informed consent had been obtained from the patient, repeat esophagogastroduodenoscopy was undertaken using a 5-mm pediatric endoscope (GIF-180: Olympus, Ontario, Canada). Multiple fibrotic-looking rings were noted throughout the hypopharynx (> Fig.1a) and pharynx (> Fig. 1b, c). There was no ulceration or erythema present. A small diverticulum was seen just above the larynx (> Fig. 1 d). The pediatric endoscope was passed into the esophagus, stomach, and duodenum with no other abnormalities being detected.

The patient was referred to an ear, nose, and throat (ENT) surgeon for further management. ENT examination confirmed the presence of several fibrotic rings affecting the pharynx and hypopharynx with considerable narrowing toward the glottic opening. It was felt that the degree of restriction at the oropharynx was sufficient to account for the patient's symptoms. The etiology of the rings remained unclear. Because of her persistent symptoms, it was recommended to the patient that she undergo excision of the rings during direct laryngoscopy using a laser.

Histopathology of the specimens confirmed squamous mucosa with submucosal fibrosis and mild chronic inflammatory changes. At a follow-up visit after 2 months, the patient reported considerable improvement in her symptoms.

Behcet's disease is a type of systemic vasculitis that may affect small, medium, and large vessels (arteries or veins). The diagnosis of Behcet's disease is a clinical one and requires the presence of recurrent mouth aphthous ulcers (at least three

times per year), and two of the following: recurrent genital aphthous ulcers, eye lesions, skin lesions, or a positive pathergy test [1].

In the oropharynx, pharyngeal stenosis has been reported in addition to ulceration and it has been postulated that this is secondary to myositis [2]. Although there are reported cases of cicatricial pharyngeal stenosis in patients with Behcet's syndrome and previous oropharyngeal surgery, this is the first reported case of pharyngeal webs associated with this syndrome. Established associations with pharyngeal webs include iron deficiency anemia, pernicious anemia, rheumatoid arthritis, carcinoma, epidermolysis bullosa, and pemphigoid [3].

Webs in the pharynx are a rare cause of dysphagia that should be considered in patients with persistent dysphagia once esophageal lesions and motility disorders have been excluded. Behcet's disease may be associated with pharyngeal webs.

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