

Refractory Esophageal Strictures with Wegener's Granulomatosis: A Treatment Experience.

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Citation

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Abstract

Wegener's granulomatosis (WG) is a pauci-immune vasculitis mainly involving pulmonary and renal vasculature. Here we report an extremely rare case of esophageal involvement with Wegener's granulomatosis resulting in stricture formation. The case report also describes the use of endoscopic through-the-scope (TTS) balloon dilations in treatment of the stricture

CASE

A 34-year-old white female with a history of chronic sinusitis and bloody nasal discharge presented with acute renal failure. She also developed painful oral ulcerations, target like lesions with central necrosis and digital necrosis. Laboratory testing revealed antineutrophil cytoplasmic antibody with a cytoplasmic staining pattern (C-ANCA) titer of 1:160, anti proteinase-3 antibody (anti PR-3) of > 100 U/mL (< 3.5 U/mL) and myeloperoxidase (MPO) antibody was negative. Punch biopsy of the skin lesions was consistent with necrotizing vasculitis involving small vessels. Based on the constellation of clinical manifestations and laboratory findings diagnosis of WG was made. High-dose steroids and cyclophosphamide were initiated.

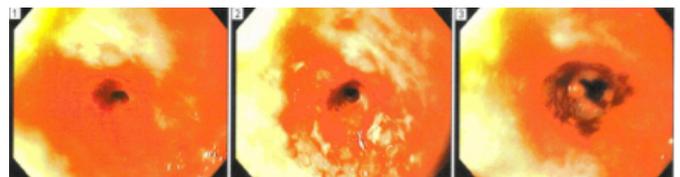
Patient complained of dysphagia and odynophagia on the fifth day of hospitalization leading to Esophagogastroduodenoscopy (EGD). EGD revealed multiple oral ulcers, severe circumferential ulceration with punched out areas from 21 cm down to the gastroesophageal (GE) junction at 35 cm from the incisors. Esophageal biopsy showed acute ulcerative esophagitis with no evidence of atypia, dysplasia or malignancy and gastroduodenitis with multiple ulcers. Symptomatic treatment with proton pump inhibitors was initiated. Patient was discharged home on oral steroids and cyclophosphamide.

Within three months after initial presentation, patient had a 40-pound weight loss due to poor oral intake and continued dysphagia. Repeat EGD showed a circumferential esophageal stricture between 30-34 cm from the incisors. The stricture was dilated up to 15 mm using a through-the-

scope (TTS) balloon. The weight loss continued necessitating percutaneous endoscopic gastrostomy (PEG) tube placement and was continued on tube feeds for approximately six months after which PEG tube was removed.

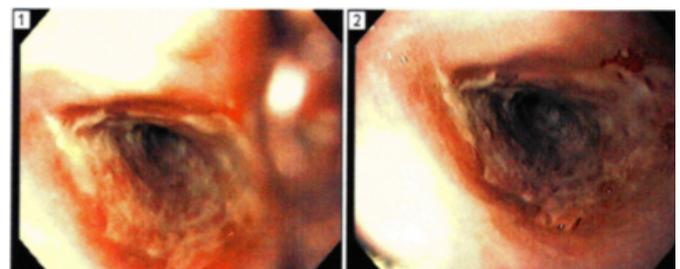
Eighteen months after the initial EGD and nine months after PEG tube removal, patient again complained of dysphagia. Endoscopy this time revealed a 3-4 mm pinhole esophageal stricture at 24 cm. The stricture was dilated to 11 mm using TTS balloon.

Figure 1



Dysphagia symptoms did not improve. EGD done ten days later showed a 5-6 mm esophageal stricture at 31 cm from the incisors.

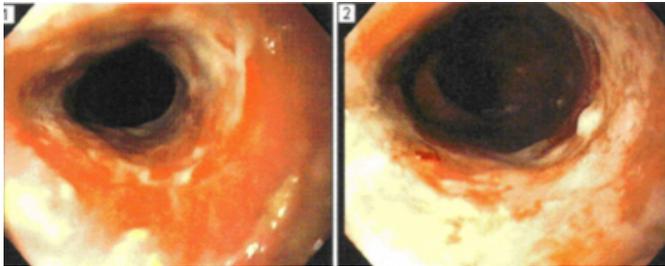
Figure 2



Patient subsequently underwent six endoscopic TTS balloon

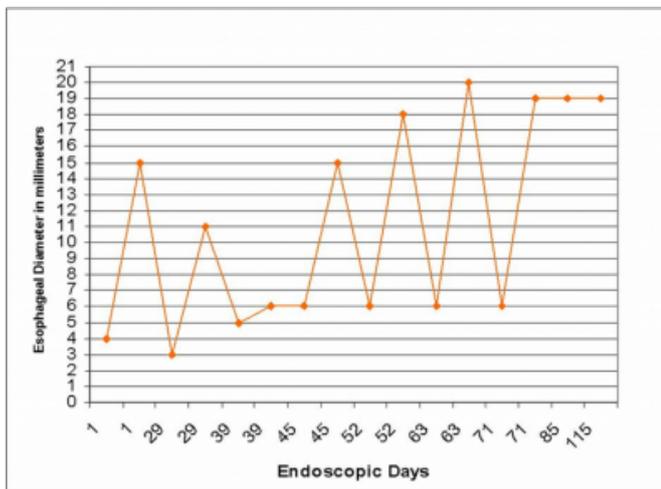
dilations ranging from 12-20 mm and one controlled radial expansion (CRE) balloon dilation within 8 weeks time. Currently the stricture remains open at 18-20 mm.

Figure 3



Endoscopy days with dimensions of dilation are shown in the graph. Patient was continued on immunosuppressants with WG in remission.

Figure 4



DISCUSSION

WG is a multi-systemic disease of unknown etiology.

Gastrointestinal (GI) involvement with WG is very rare. Endoscopic literature pertaining to the GI manifestations in WG is very limited (1-3). WG with esophageal ulcerative lesions have been previously reported (1-4). However, stricture formation requiring multiple dilations has not been described before (1).

The present case of esophageal Wegener's with circumferential ulceration and stricture formation needed multiple endoscopic dilations. In total, the patient had fifteen endoscopic TTS balloon dilations in eighteen months. Seven progressive TTS balloon dilations within a period of 8 weeks were required before the esophagus remained open at 18-20 mm.

Refractory benign esophageal strictures or recurrent strictures can also be treated using Savory dilation over a guide wire preferably under fluoroscopy, Eder-Peustow metal olives over a guide wire under fluoroscopy or endoscopic placement of an esophageal prosthesis. In our patient, the esophageal lumen has remained open for five months. Patient is currently asymptomatic with a steady weight. WG is in remission with Imuran and prednisone.

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