Treatment of Plummer–Vinson syndrome with Savary–Gilliard dilatation

Mohamed I. Yasawy, MD, AFIM.

ABSTRACT

The association of postcricoid dysphagia, iron deficiency anemia and upper esophageal webs is known as the Plummer-Vinson syndrome (PVS). It is predominantly a disease of middle aged females. Four patients with cervical web of the esophagus were studied, dysphagia was mostly for solids, which were non-progressive and were present for years before they presented. Initial treatment in 3 patients consisted of esophageal dilatation with savary bougies after initial rupture of the web by endoscope. In one case, due to failure to visualize the esophageal lumen by fluoroscopy guidance, guide wire was introduced and breaking of the web was performed by the endoscope, followed by bougie dilatation. In 4 patients, webs were ruptured by the endoscope and had a single session of dilatation, with no recurrence during the 3-year period of follow up. Endoscopic esophageal dilatation is simple and the procedure of choice in the treatment of "PVS" and cervical web of the esophagus.

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H istorically Plummer-Vinson syndrome (PVS) also known as Paterson-Kelley syndrome and Sideropenic dysphagia has been largely described among middle aged white women. The presentation includes anemia, dysphagia, choking spells and aspiration due to the proximal location of the web. Esophageal web is a thin 2-3 mm membrane, consisting of mucosa and submucosa causing dysphagia. The postulated mechanical etiopathogenic mechanism includes iron deficiency, genetic predisposition and autoimmune factors.¹⁻⁴ This syndrome is rare nowadays and its decline has been attributed to better nutrition and health care.⁵ The majority of symptomatic webs are treated by dilatation with an excellent outcome.6,7 Surgery is rarely needed and in some cases laser interventions were used.^{8.9} Morbidity and mortality were attributed primarily to diet modification and repeated esophageal dilatation with a small risk of perforation in patients with recurrent dysphagia. Recent articles show its association with celiac

disease and high incidence of postcricoid carcinoma.¹⁰⁻¹³

Case Report. Four Saudi female patients between 30-40 years of age were suffering from dysphagia, anemia and its constitutional symptoms for a period of 3-10 years. All 4 patients had dysphagia to solids with choking spells. All of them had angular cheilitis, 3 had koilonychias and one had a palpable spleen. They were pale looking and were found to have iron deficiency anemia. It was proven by reduced serum iron, increased total iron binding capacity (TIBC) and reticulocyte count that were raised after iron therapy. Hemoglobin in all patients was ranging from 6-10 mg/dl. A gynecologist for interrupted menstruation cycle and secondary amenorrhea saw 2 of them. No gynecological cause was found and none of the patients had history of acute or chronic blood loss. Barium-swallow as well as the video esophagram

From the Department of Internal Medicine, King Faisal University, Dammam, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Mohamed I. Yasawy, Consultant Internist Gastroenterologist, King Fahad Hospital of the University, PO Box 40143, Al-Khobar 31952, Kingdom of Saudi Arabia. Tel/Fax. +966 (3) 8966741. E-mail: yasawy@hotmail.com

and esophagoscopy showed cervical web, a finding consistent with PVS. Oral iron therapy was given to 2 patients but due to difficulty in swallowing and complaint, the patients did not take the treatment regularly for a long period of time and did not show any obvious changes in the correction of anemia or improving the dysphagia. In one patient, again due to poor compliance in oral iron therapy, injectable iron was given for a total period of one month which showed some correction of iron deficiency anemia however, no obvious improvement in dysphagia (this patient was later found as having circumferential web). In all cases endoscopic dilatations were performed under topical xylocaine 20% and conscious sedation. In 3 patients, the endoscope was used as diagnostic, as well as an initial therapeutic tool such as the site of the web was documented and breaking and perforation of the web by passing the scope was achieved and followed by esophageal dilatation up to 15mm. In one patient, intubation under direct visualization failed. The whole lumen was almost obstructed by the membrane. In this case, a guide wire under fluoroscopy was inserted in to the stomach and esophageal dilatation using Savary-Gilliard polyvinyl bougies from 6 mm up to 15 mm was performed. Post dilatation upper gastrointestinal endoscopy was carried out to evaluate and to check for any possible complication resulting from the procedure. A week later, barium esophagogram was performed to evaluate the state of the web and the response of the dilatation. In all patients, dysphagia responded to the first session of endoscopy and dilatation. Rupture or break of the web by endoscopy was achieved in 3 patients with anterior

web. One patient had a circumferential web, which significantly occluded the lumen. Visualization of the lumen was endoscopically not possible. Under fluoroscopy, a guidewire was passed from the upper esophagus down to the stomach and the web was ruptured using Savary Gilliard dilators. A good response was achieved and no immediate or early complications were noted following the procedure (Figures 1a & 1b). In 2 of the patients, minimal hematemesis was noticed immediately after the procedure which did not cause any hemodynamic changes. In all 4 patients, early control contrast swallow study showed evidence of minimal mucosal irregularities. Follow up of 1-3 years showed complete correction of anemia and no recurrence of dysphagia in any of them. All patients were relieved of cheilitis, Iron chemistry and hemoglobin came to normal range. During the short period of follow up, no patient developed evidence of postericoid carcinoma.

Discussion. Cervical webs are one of the causes of dysphagia, seen almost in all age groups including children. However, previously it was reported that it mainly occurs in age groups of third to sixth decades in white females preponderance, although there are other reports from other parts of the world.^{5,6,14,15} The pathogenesis of PVS remains speculative. Postulated etiopathogenic mechanisms includes iron and nutritional deficiencies, genetic predisposition and autoimmune factors amongst others. Evidence for an association between iron deficiency and postcricoid dysphagia is the improvement of dysphagia after iron therapy, especially early and mild cases.16 in



Cineradiography is a sensitive method of investigating these patients but it is not routinely carried out. Conventional barium studies are not as specific sensitive as cineradiography. and Fiber-optic endoscopy is a safe and reliable tool for diagnosing upper esophageal webs.5,15 In addition to the establishment of the diagnosis, in majority of the cases, treatment can also be offered in the same endoscopic sitting. The early degree of dysphagia with its intermittent nature of symptoms response to medical management such as by treating iron deficiency. Advanced and long standing forms is unlikely to respond to iron replacement alone and requires interventional management, such as endoscopic tear of the web, balloon dilatation, Savary Gilliard dilators, endoscopic laser division and even surgical intervention has been reported to be successful in the treatment of the esophageal web.6-9,17 All our patients were symptomatic with dysphagia and the rupture of the web was achieved by flexible endoscope. In 3 patients with anterior web, rupture was incomplete. So the remaining parts of the web were cleared with Savary Gilliard dilator. patient with In one circumferential web. visualization of the esophageal lumen was not possible and required fluoroscopic guided Savary Gilliard dilator, which started by 6mm and continued up to 15mm. Forceful endoscopic dilatation for initial rupture of the web by an endoscope and direct use of a 15 mm dilator for further dilatation was found to be the approach of choice namely safe and effective with no morbidity or complications. In majority of the cases, a single session of dilatation is sufficient to achieve adequate dilatation and relief of dysphagia and patients can be discharged on the same day. During the 3 years of follow up period no recurrence or malignant changes were encountered.

We conclude that forceful endoscopic dilatation followed by the use of savary dilators is an effective, safe and economical mode for the management of dysphagia caused by cervical esophageal webs.

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