Plummer-Vinson Syndrome with Simultaneous Mid-esophageal Growth

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Abstract

Patterson Brown Kelly or Plummer-Vinson syndrome is characterized by dysphagia, iron deficiency anemia and post-cricoidal esophageal web. Waldenstrom introduced the term 'sideropenic dysphagia' because of absence of stainable iron in the bone marrow. There is increased incidence of upper aerodigestive tract carcinoma in patients with Plummer-Vinson syndrome has been well established. The reported rates range from 4% to 16%, with almost all cases occurring at the postcricoid location. We have reported here a case of a 48-year-old woman with dysphagia, upper esophageal web and iron deficiency anemia. Dilatation of esophageal web with subsequent endoscopy showed mid-esophageal growth which on biopsy showed squamous cell carcinoma.

Introduction

Patterson Brown Kelly or Plummer-Vinson syndrome is characterized by dysphagia, iron deficiency anemia and post-cricoidal esophageal web. Waldenstrom introduced the term 'sideropenic dysphagia' because of absence of stainable iron in the bone marrow. There is increased predisposition to upper aerodigestive tract malignancies in these patients. In 1937 Ahlbom found out that the risk of carcinoma was not confined to the postcricoid area but also to the buccal mucosa, tongue, and all levels of the esophagus. Paterson commented on the association with "malignant disease at the mouth of the gullet" in his 1919 report.¹ Since that time, the increased incidence of upper aerodigestive tract carcinoma in patients with Plummer-Vinson syndrome has been well established. The reported rates range from 4% to 16%, with almost all cases occurring at the postcricoid location. We have reported here a case of a 48-year-old woman with dysphagia, upper esophageal web and iron deficiency anemia. Dilatation of esophageal web with subsequent endoscopy showed mid-esophageal growth which on biopsy showed squamous cell carcinoma.

Case

A 48 year-old woman presented with nausea, progressively increasing dysphagia to solids and generalized weakness since 4 months.

Her past history and family history were not specific. She had no history of tobacco or alcohol use. Her examination revealed a BMI of 20.2 kg/m². On examination, she had pallor, koilonychia with glossy tongue. There was no palpable lymphadenopathy.

Her blood investigations were normal except hemoglobin 7.7 gm/dl, MCV 74.8 F Lt, serum ferritin 10.2 ng/ml. Barium swallow that was done at other centre did not show any abnormality.

She underwent an Upper GI endoscopy for evaluation of dysphagia, it revealed an upper esophageal web in the subcricoid region and the endoscope could not be negotiated across the web. The web was typically attached to the anterior wall of esophagus and form just inferior to the cricopharyngeus (Figure 1).

Esophageal web was dilated with through-the-scope controlled radial expansion balloon (13 mm diameter) (Figure 2). Post dilatation, the scope could be negotiated further into the esophagus. It revealed a ulceroproliferative growth at 26 cm from the incisors, extending upto 29 cm (Figure 3). The scope could be negotiated across the growth. Multiple biopsies were taken and sent for histopathological examination. The stomach, duodenum did not show any abnormality. The biopsy revealed poorly differentiated squamous cell carcinoma. A whole body PET-CT showed hypermetabolic diffuse circumferential mural thickening in mid esophagus with enlarged gastrohepatic, superior mediastinum and para aortic lymphnodes. A EUS guided FNA was done from the para aortic nodes which was suggestive of a metastatic carcinoma. She underwent palliative chemotheraphy with Docetxel, Cisplatin and 5-flurouracil. She was advised concurrent radiotherapy but she refused. A repeat PET CT was
fluoroscopy. The radiologic method is way for demonstration is the video barium swallow X-ray but the best Esophageal webs can be detected by tortuous esophagus instead of include clubbing instead of koilonychia, however some unusual manifestations are the hallmark of this syndrome; anaemia, dysphagia and koilonychia esophageal webs, iron deficiency


Discussion

In the first half of 20th century Plummer-Vinson syndrome was more common in Caucasians in northern countries, particularly amongst middle aged women. The incidence of the Plummer-Vinson syndrome has recently decreased because of improved nutrition and health care especially for pregnant women. The recognition of this malady is important because it identifies a group of patients at increased risk of squamous cell carcinoma of the pharynx and the esophagus. The presence of postcricoid esophageal webs, iron deficiency anaemia, dysphagia and koilonychia are the hallmark of this syndrome; however some unusual manifestations include clubbing instead of koilonychia, tortuous esophagus instead of esophageal webs and celiac disease. Esophageal webs can be detected by barium swallow X-ray but the best way for demonstration is the video fluoroscopy. The radiologic method is more suitable than endoscopy because endoscopy can sometimes miss the point of benign stricture, and does not verify most of the motility disorders. The web is usually detected at the upper esophagus below the cricoid by showing a thin membrane across esophagus. Pathologically it shows hypertrophy or atrophy of squamous cells and it is sometimes combined with chronic inflammation.

This syndrome can be effectively treated with iron supplementation and mechanical dilation. Iron supplementation can alone relieve the dysphagia. However, a few data suggests that for many patients iron deficiency is neither a necessary nor a sufficient cause of web formation. The esophageal web can be managed with endoscopic dilatation, balloon dilatation, endotracheal dilatation, and incision of web. On occasion, more than one sitting may be required.

Plummer-Vinson syndrome is known to be associated with an increased risk of upper alimentary tract cancers, and the incidence rate of upper esophageal cancer in this syndrome is 3-15% so a close monitoring of these patients is advisable. Repeat endoscopy on an annual basis may be done in the initial period of follow-up.

A decreasing trend in the overall incidence of hypopharyngeal cancer in women is reported, probably due to decreased prevalence of Plummer-Vinson syndrome. A history of smoking, alcohol consumption, and diets low in fruits and vegetables accounted for almost 90 percent of esophageal SCC. Head and neck cancers are found in approximately 10 to 15 percent of patients diagnosed with esophageal cancer. In addition, patients with one aerodigestive malignancy have an increased risk of synchronous and second primary tumors of the aerodigestive tract. SCC invades the submucosa at an early stage, and extends along the wall of the esophagus usually in a cephalad direction, as seen in our case. Local lymph node invasion occurs early and quickly because the lymphatics in the esophagus are located in the lamina propria, in contrast to the rest of the gastrointestinal tract, in which they are located beneath the muscularis mucosa. The tumor spreads to regional lymph nodes along the esophagus, the celiac area, and adjacent to the aorta. Invasion of local structures may result in fistula formation (such as to the trachea).

In our case the presence of an esophageal web was accompanied by a mid esophageal SCC. This case provided the rare opportunity to diagnose and effectively treat an esophageal carcinoma as well as its accompanying Plummer-Vinson syndrome. In addition, this patient’s evaluation confirms earlier findings that conventional barium swallow studies are suboptimal for esophageal web diagnosis in Plummer-Vinson syndrome. Cineradiography of swallowing is recommended for evaluating webbing, which is best visualized when the hypopharynx and cervical esophagus are maximally dilated.

References


