

Photoclinic Diagnosis:

Progressive mechanical dysphagia in a patient with longstanding intermittent solid dysphagia

Rings or webs may occur in any part of the esophagus. They are muscular or membranous and may be asymptomatic or present as dysphagia. Webs, unlike rings, often encircle the esophageal lumen incompletely. Esophageal web and rings can occur as the consequence of a developmental defect or an inflammatory disorder; however, the exact etiopathogenesis, especially in upper esophageal webs, is not known. Esophageal webs may be present as an isolated lesion or a component of the Plummer Vinson syndrome (esophageal web in association with iron deficiency). There is evidence that the Plummer Vinson syndrome is linked with squamous cell carcinoma (SCC) of the esophagus. The mechanism of developing esophageal SCC in this setting is not clear.

Asymptomatic cervical esophageal webs are demonstrated in about 10% of people and typically originate along the anterior aspect of the esophagus. When circumferential, they can cause dysphagia. The combination of proximal esophageal webs, dysphagia and iron-deficiency anemia in middle-aged women constitutes the Plummer Vinson syndrome or the Paterson-Kelly syndrome¹; angular stomatitis and glossitis may be another finding in this syndrome. However as in our patient, iron deficiency may be absent in this disorder.

The primary symptom in our patient was mild intermittent solid dysphagia, especially with extraordinary large pieces of solid food particularly at the beginning of eating since teenage; this history is consistent with mucosal ring or web of the esophagus. Nevertheless, progressive dysphagia over the recent months that was associated with significant weight loss in our patient was suggestive of a new serious pathology such as esophageal cancer. Radiologic images and endoscopy of esophagus in our patient revealed a membranous web incompletely encircling the lumen in the upper part and a circumferential infiltrative ulcerated lesion in distal esophagus. Histopathologic assessment of endoscopic biopsies from lower esophageal lesion confirmed SCC in our patient. Contrast computerized tomography scan revealed few mediasti-

nal lymphadenopathies; after replacing a covered self-expandable metal stent in tumor site, the patient was scheduled for chemo-radiation.

The Plummer Vinson syndrome raises the risk of oral and esophageal squamous cell carcinoma. This syndrome, although uncommon, occurs primarily in women. It is an important syndrome because it identifies a group of patients at increased risk of squamous carcinoma of the pharynx and esophagus.¹ Also, recent reports have shown an association between the Plummer Vinson syndrome and celiac disease.² Correction of iron deficiency in the Plummer Vinson syndrome may result in resolution of the associated dysphagia as well as disappearance of the web.¹

The main risk factors of esophageal SCC are excess alcohol consumption, cigarette smoking, and ingestion of nitrites, smoked opiates, and fungal toxins in pickled vegetables, physical insults including long-term exposure to extremely hot tea, the ingestion of lye, radiation, and longstanding achalasia. None of these risk factors were found in our patient; therefore, esophageal SCC in our case may be linked to esophageal web.

The exact prevalence of esophageal SCC in the Plummer Vinson syndrome and the importance of surveillance endoscopy are challenges in patients with esophageal web. Large prospective studies are needed for these purposes.

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References

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2. Jessner W, Vogelsang H, Püspök A, Ferenci P, Gangl A, Novacek G, et al. Plummer- Vinson syndrome associated with celiac disease and complicated by poststeroid carcinoma and carcinoma of the tongue. *Am J Gastroenterol*. 2003; **98**: 1208 – 1209.