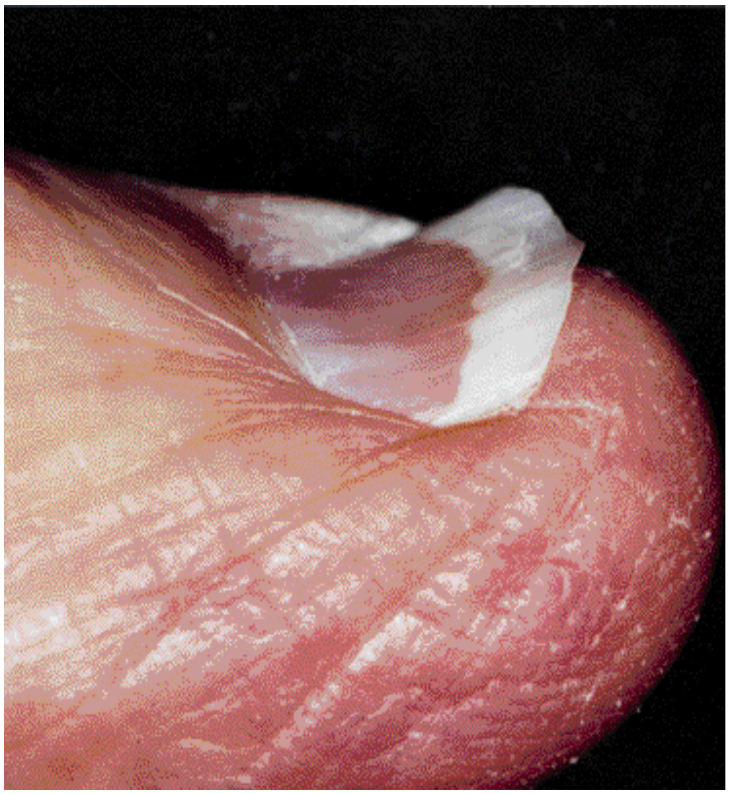
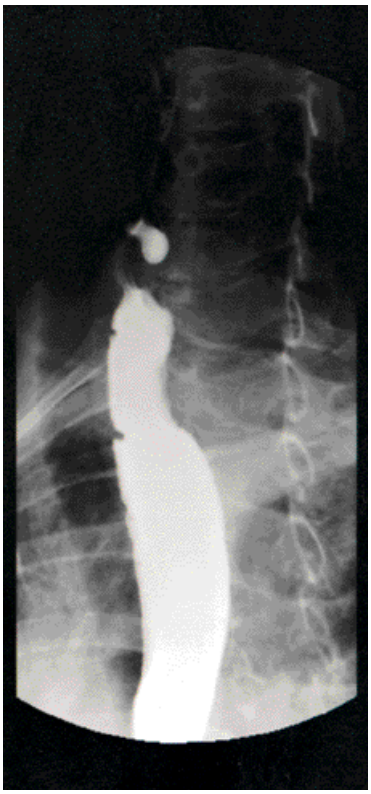
What Is Your Diagnosis?





A 74-year-old woman presented with nail changes. After further questioning and physical examination, it was discovered that she had iron deficiency anemia and a history of long-standing dysphagia. An x-ray study was performed.

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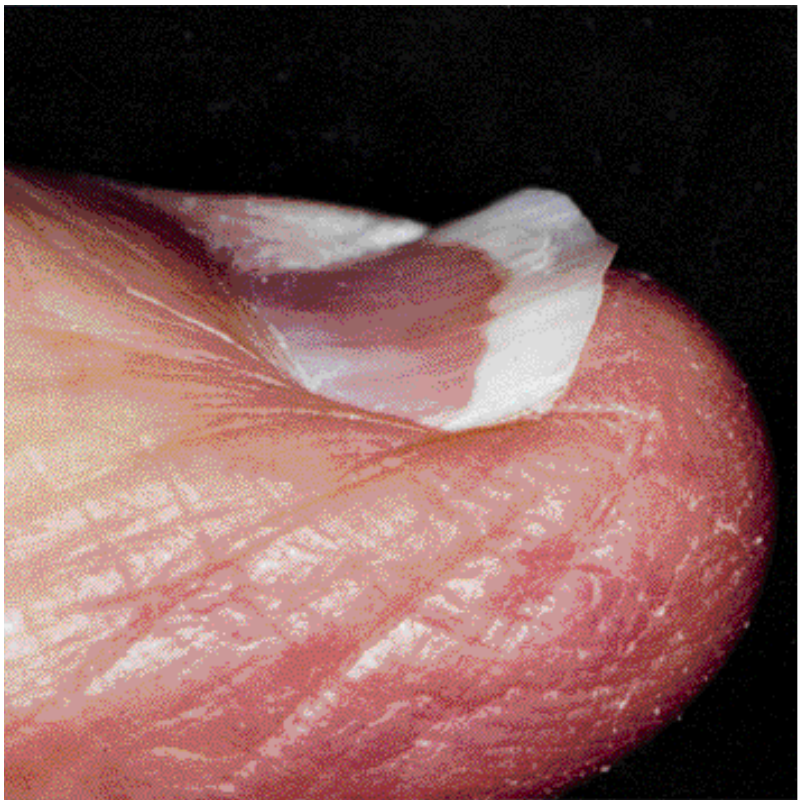
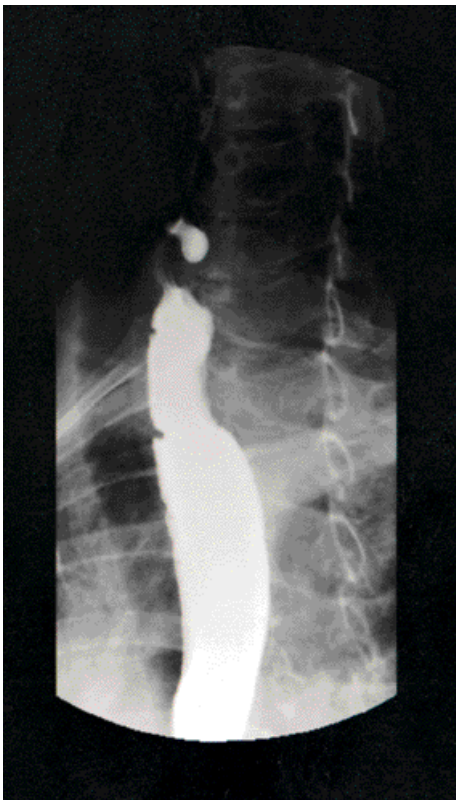
The Diagnosis: Plummer-Vinson Syndrome





The patient presented with koilonychia. Laboratory results revealed iron deficiency anemia. She had a long history of intermittent dysphagia, especially to solid foods. Barium swallow results revealed 2 anterior filling defects compatible with esophageal webs, and esophagogastroduodenoscopy results confirmed the presence of anterior webs. An incidental Zenker diverticulum also was noted.

The classic triad of Plummer-Vinson syndrome (PVS) includes iron deficiency anemia, esophageal webs, and dysphagia. However, all 3 of these manifestations are not present in all cases. The iron deficiency can present as anemia, koilonychia, angular cheilitis, glossitis, and/or pallor. Esophageal webs do not occur in all cases, but dysphagia is a symptom in most cases.[1,2]

PVS is also known as *Paterson-Kelly syndrome*, *Brown Kelly-Paterson syndrome*, *Paterson-Brown Kelly syndrome*, *sideropenic dysphagia*, and *Waldenstrom and Kjellberg syndrome*. *Paterson-Kelly syndrome* is the preferred eponym in Great Britain, whereas *sideropenic dysphagia* is preferred in Scandinavia. PVS is commonly used in the United States.[1-4]

In 1926, British gastroenterologist Arthur Hurst gave this condition its first eponym by coining the term *Plummer-Vinson syndrome*. Yet controversy exists as to who published the first work describing the features of this syndrome. In 1919, Scottish-born otolaryngologists Donald Rose Paterson and Adam Brown Kelly independently reported this syndrome; American physician Porter Paisley Vinson published his work in 1921. Vinson's mentor, fellow American Henry Stanley Plummer, reported cases of esophageal spasms in 1908 and 1912, but some feel these cases were unrelated to this

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syndrome. The term sideropenic dysphagia was created from Waldenstrom and Kjellberg's work in 1939 describing iron deficient cases without anemia. Early descriptions of a similar syndrome date from the eighteenth century.[1,3-6]

PVS is most commonly seen in middle-aged white women.[1,2,4-6] The pathophysiology is uncertain.[1,2,4,5] It is rare in the United States, and the incidence is declining. The decline may be secondary to better nutrition and healthcare. Internationally, PVS was common in the early to mid twentieth century, especially among Scandinavian women, but has since declined rapidly. This rapid decline has paralleled improvement in nutritional status.[2,4]

Evaluation should include a complete blood count and an iron panel to confirm the presence of anemia and its origins. In addition, a barium esophagram and an esophagogastroduodenoscopy can be performed to evaluate the dysphagia and esophageal webs. Periodic follow-up esophageal examinations are needed to monitor the potential development of hypopharyngeal and esophageal cancers.[1,2,5]

Management is determined by laboratory results and symptoms. Iron replacement is needed when iron deficiency is present. Trapped food should be mechanically removed. Symptomatic esophageal webs can be treated by mechanical dilation, surgery, or laser.[1,2,6]

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