BREAKING THE WEB:

A CASE OF PLUMMER VINSON SYNDROME IN THE PHILIPPINES

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Abstract

Significance: Plummer Vinson syndrome (PVS) is an extremely rare disease characterized by the triad of iron deficiency anemia, dysphagia and cervical esophageal web. The most accepted possible etiology of this syndrome is iron deficiency anemia. The prevalence of anemia in third world countries is high and yet there is no published case to date of Plummer Vinson syndrome in the Philippines.

Clinical Presentation: We report a case of a 44 year old female, Filipino with two decades of iron deficiency anemia, a decade of dysphagia with a cervical esophageal web on esophagogastroduodenoscopy (EGD).

Management: After EGD with mechanical dilatation, dysphagia was resolved and patient now is able to eat a full regular meal. She was given iron supplementation and advised annual esophagogastroduodenoscopy.

Recommendation: The triad of iron deficiency anemia, dysphagia and cervical esophageal web is Plummer Vinson syndrome which can be readily managed by iron supplementation, and esophageal dilatation. Patients need to be on annual surveillance EGD due to risk for esophageal or pharyngeal malignancy.

Keywords: Case report, anemia, dysphagia, esophageal web, Plummer Vinson syndrome, esophageal dilatation

Introduction

Iron deficiency anemia (IDA), dysphagia and cervical esophageal web are the triad of Plummer Vinson syndrome (PVS). This is an extremely rare disease with no reported prevalence or incidence since it was first described in 1912.[1] Patients are mostly white middle-aged women, in the fourth to seventh decade. Exact cause is not yet established but iron deficiency is the most important possible etiologic factor. Correction of iron deficiency anemia contributed to the paucity of cases.

In the Philippines, the prevalence of anemia in women in the reproductive age group (15-49 years old) ranged from 25.4 to 40.0%.[2] Despite the high prevalence of anemia in the Philippines, there is no published case of PVS in a Filipino to date. Hence the possibility of anemia as a presentation rather than a cause of PVS. Here, we present a Filipino with Plummer Vinson syndrome managed with esophageal dilatation and iron supplementation.

Case

We report a case of a 44 year old female, single, Filipino, from Davao City, who consulted due to dysphagia for more than 10 years. Since she was in her teens, she was noted to be pale and with low hemoglobin ranging from 7 to 9 g/dL. No blood transfusion was given. She was still ambulatory without easy fatigability and dizziness. Work-up was done in the province and apparently showed iron deficiency anemia. She was advised bone marrow aspiration however patient refused. She was initially given ferrous sulfate tablet but didn't tolerate the medication and was lost to follow-up. She was able to do activities of daily living, finished her education and even joined sports activities. For the past 10 years, she experienced progressive difficulty swallowing solid foods with feeling of food getting stuck in the throat. She sought consult with a surgeon at another institution who advised surgery. She and her family refused any invasive intervention. She adapted by only eating soft rice and soft viands cut into small pieces, chewing thoroughly and very slowly. She has no previous surgery, no exposure to radiation or chemicals and no family history of hematologic disease, celiac disease or cancer. She has no maintenance medications and denied NSAID use. She has no overt bleeding and reported regular menstruation consuming 3 to 4 pads per day, 4 to 5 days every month.

She decided to seek second opinion because she wanted to be able to eat normally again. She is ambulatory with stable vital signs. Her weight is 48.18 kg and her height is 5' 4" with a body mass index (BMI) of 18.23 kg/m2. She was pale with pale conjunctivae and anicteric sclerae. No cheilosis or koilonychias noted. The rest of the physical examination was unremarkable.

Her blood count showed a hemoglobin of 6.4 g/dl, hematocrit of 25%, white blood cell count of 5,310 mm^3, platelet count of 358,000/mm3 with MCV of 61 fl, MCH 16 pg and MCHC 26%. An esophagogram showed pooling of contrast material seen at the proximal esophagus at the level of C4-C5 that may relate to an esophageal stricture.

On esophagogastroduodenoscopy (EGD), a neonatal gastroscope (6.3 mm diameter) was inserted to 15 cm level showing a narrowed opening which did not allow passage of the scope. A 0.025 inch Zebra guidewire was inserted through the opening which allowed visualization of the normal esophageal mucosa beyond the narrowed area. Subsequent dilatations using Savary-Gilliard dilators from French 21 to 45 were done with easy passage of dilators to 40 cm level with minimal bleeding. Repeat EGD using an adult gastroscope (10.7 mm diameter) showed the mucosal break at 15 cm level without active bleeding. The rest of the esophageal mucosa is intact all the way down to the Z line which coincides with the diaphragmatic indentation at 35 cm level. The stomach and the duodenum down to the descending portion were unremarkable. The patient tolerated the procedure which lasted for an hour and 15 minutes. Four hours after the procedure, the patient was able to eat solid foods without any difficulty. She returned to her work as an overseas Filipino worker with no complaints to date. She was given ferrous gluconate tablet as maintenance medication.

Discussion

Plummer Vinson syndrome (PVS) was named from American physicians Henry Stanley Plummer and Porter Paisley Vinson. [3,4] The syndrome was also termed Paterson-Brown-Kelly syndrome based from independent reports by two British laryngologists, Donald Ross Paterson and Adam Brown-Kelly.[1] In 1939, Waldenstrom and Kjellberg introduced the term sideropenic dysphagia based on the constant finding of dysphagia and iron deficiency anemia. Since its discovery, there is no figure of its incidence because of its rarity. Its etiology is still a controversy; possible causes suggested were streptococcal infection, tertiary syphilis, vitamin deficiency, hysteria and gastric inadequacy. Difficulty in swallowing was once thought to be the primary cause of PVS leading to the secondary deficiencies. To date, improvement of symptoms with iron supplementation points to iron deficiency as the possible etiology. It is rarely seen in males and almost always in females, which may be due to increased iron requirements. In addition to being a component of hemoglobin and myoglobin, iron is also an essential component of the intracellular enzymes catalase, peroxidase and cytochromes. Its deficiency produces a generalized disturbance affecting the whole body. Since the fortification of food with iron, the syndrome has become progressively rarer.[5] The most common reason for consult is dysphagia which is painless and intermittent or progressive and is limited to solids as seen in our case.[6] This is caused by a characteristic thin fold arising anteriorly and stretching a variable distance across the esophageal lumen. These webs were composed of two mucus membrane layers with minimal intervening fibrous tissue on biopsy. PVS may also be associated with koilonychias, glossitis, cheilitis and splenomegaly. These are not appreciated in the case. A variety of disorders have been known to associate PVS such as celiac disease, inflammatory bowel disease, pernicious anemia, thyroid disease, Sjogren's disease, and rheumatoid arthritis.[7] Symptoms usually reverse with iron administration even without endoscopic intervention. However it was observed that only

in the early stages of the disease can iron therapy prevent occurrence of malignant changes.[6] In 1957 Weisberger stated that treatment of anemia in 18 cases with PVS had no effect on eventual development of carcinoma. Out of eight patients with PVS given iron therapy, seven developed carcinoma at some later stage of the disease.[9,10] Esophageal or pharyngeal cancer can develop in 3 to 15% of PVS patients hence annual EGD is advised.[11]

Esophagogastroduodenoscopy is essential in all patients with PVS to exclude other causes of dysphagia, exclude malignancy, for therapy to improve dysphagia and regular follow-up for possible malignant change. If dysphagia is persistent, esophageal dilatation is warranted and results in immediate relief.[1,7,] Our patient improved immediately after one session of esophageal dilatation. Once fully awake after the procedure, she was able to eat regular meals. However this benefit may be short-lived hence regular follow-up is recommended. She was discharged on iron supplementation. Argon plasma coagulation therapy of esophageal webs may also be utilized. The prognosis is mostly good at short-term follow ups.

Conclusion

Despite a high prevalence of anemia in the country, this is the first case to our knowledge of a Filipino with Plummer Vinson syndrome. She presented with more than two decades of anemia, a decade of dysphagia, cervical esophageal web, with instant relief after one session of esophageal dilatation. She was given iron supplementation and advised regular follow-up to check for recurrence of dysphagia, improvement of anemia and surveillance for possible development of malignancy in the upper alimentary tract.

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IMAGES

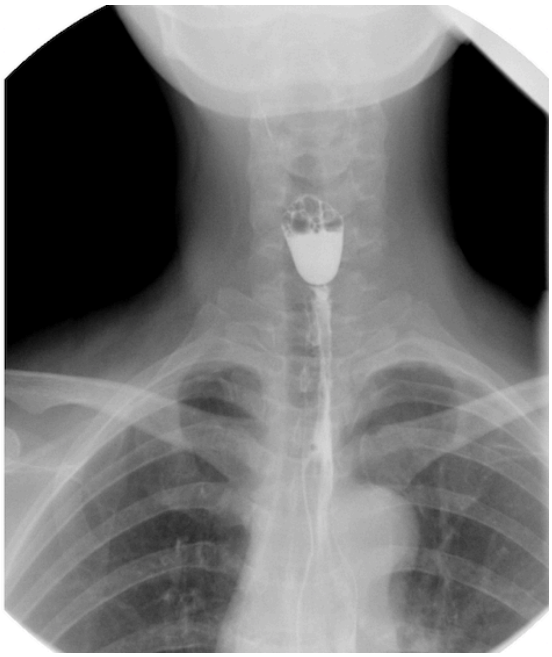

Image 1: Esophagogram: pooling of contrast material seen at the proximal esophagus at the level of C4-C5.

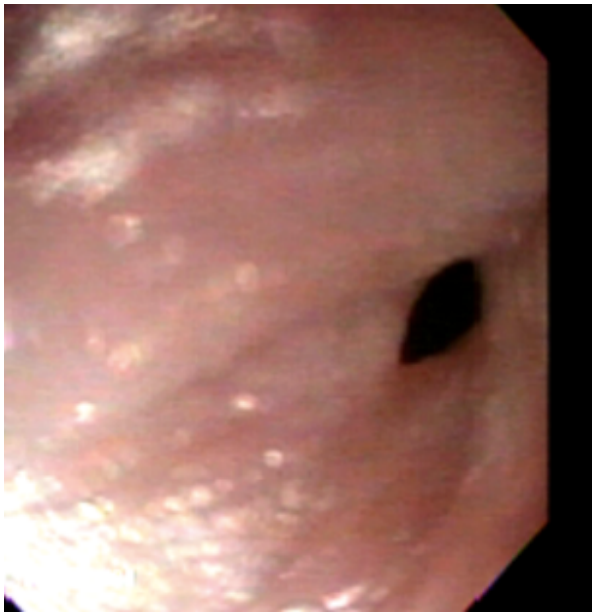

Image 2: Esophageal web at 15 cm level from the incisors

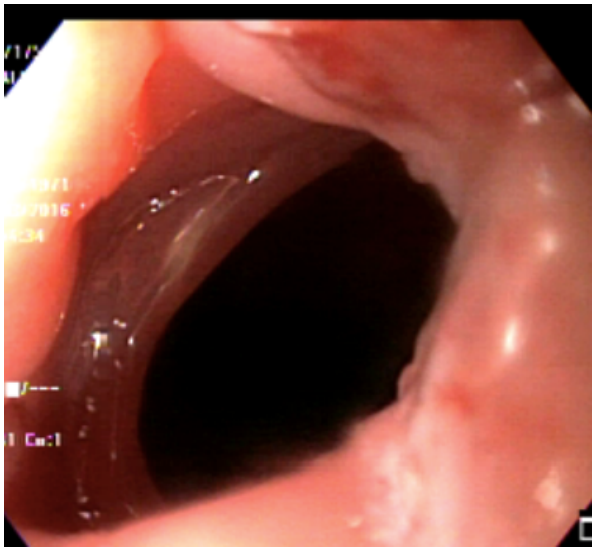

Image 3: Narrowed area post dilatation, proximal esophagus