Plummer–Vinson syndrome in celiac disease

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Received - 01 March 2020 Initial Review - 19 March 2020 Accepted - 06 April 2020

ABSTRACT

The association of Plummer–Vinson syndrome (PVS) and celiac disease is not widely reported. Both entities have associated iron deficiency anemia. The following case report signifies that the diagnosis of celiac enteropathy must be kept in mind in every iron deficiency case that we encounter, principally when associated with Plummer–Vinson. We report two cases which emphasize the need for definitive screening for celiac disease in outdoor patients with PVS.

Key words: *Celiac disease, Dysphagia, Iron deficiency anemia, Plummer–Vinson syndrome*

Plummer–Vinson syndrome (PVS) (Paterson Brown Kelly syndrome or post-cricoid dysphagia) comprises a triad of disconcerting findings of iron deficiency anemia, dysphagia, and cervical esophageal webs. The pathogenesis of the syndrome is not very well understood although the underlying iron deficiency of long duration is implicated as a possible explanation. The most apparent mechanism of PVS is iron deficiency, leading to a quick loss of iron-dependent enzymes due to its high cell turnover. Loss of these enzymes causes mucosal degenerations, atrophic changes, and web formation, which are associated with dysphagia [1]. However, its incidence in association with celiac disease which almost always has iron deficiency anemia has rarely been reported [2].

CASE REPORT

Case 1

A 28-year-old male presented to our center with complaints of dysphagia to solids for 2.5 years, loss of appetite and loss of weight (12 kg) over 2 years. The patient was aware of having anemia for 10 years and had been on and off iron supplements. The cause of anemia was possibly considered to be nutritional deficiency but was never investigated. There was no history of nausea, vomiting, altered bowel habits, fever, gastrointestinal bleed, pain abdomen, or any other noticeable history.

On examination, he had pallor and koilonychia. Rest of the general physical and systemic examinations was normal. The blood pressure was 90/68 mmHg, pulse rate was 102/min, and afebrile. Investigations revealed iron deficiency anemia with hemoglobin of 5.8 g/dl (11–15) and serum iron of 12 g/dl (45– 158). Peripheral blood picture showed microcytic hypochromic anemia. His serum ferritin was 10 ng/mL (30–140) and bone marrow aspiration demonstrated iron deficiency anemia with absent ironstones. Serum calcium was 7.6 g/dl and serum albumin 3.1 g/dL. Blood sugar, liver, and renal function tests were all normal. The stool examination for occult blood was negative.

He underwent an upper gastrointestinal endoscopy which showed a post-cricoid web. The web was endoscopically dilated without any complication and duodenal mucosa showed scalloping of the folds. A duodenal biopsy was taken which showed total villous atrophy. Serum anti-tissue transglutaminase (TTG IgA) level was 114 u/ml (0–10). He was put on a strict gluten-free diet and later his hemoglobin gradually improved to >10 g/dl after 8 months of outpatient department follow-up. His weight has increased and did not complain of any repeat episode of dysphagia.

Case 2

A 49-year-old female presented with 1 year of progressively increasing difficulty in swallowing history. She had lost 8 kg of weight over the past 3 years. There was no other significant history documented.

Her physical examination showed an average built lady with pallor, koilonychia, and angular stomatitis. The systemic examination was normal. Hemodynamically, the patient had a blood pressure of 100/72 mmHg, pulse 98/min, and afebrile. Investigations revealed iron deficiency anemia with hemoglobin of 6.2 g/dl (11–15), serum iron was 18 g/dl (45–158), and serum ferritin 8.2 ng/ml (30–140). Peripheral blood picture was suggestive of iron deficiency anemia. Serum albumin was 3 g/dl (3.5–5.0), serum calcium was 7.8 g/dl. Other blood biochemistries were normal.

Upper GI endoscopy revealed the esophageal web in the post-cricoid region, which required endotherapy (dilatation). The

duodenal mucosa revealed scalloping of folds. Duodenal biopsy showed total villous atrophy. Her anti-TTG levels (TTG IgA) were 200 u/ml. She was started on a strict gluten-free diet. Her hemoglobin improved gradually and she was not complaining of any fresh episodes of dysphagia on follow-up. Her follow-up hemoglobin was 9.8 g/dl.

DISCUSSION

In the past few decades, iron deficiency anemia alone has been identified as the presenting feature of underlying celiac disease. The prevalence of the celiac disease is increasingly rising in India. We came across two patients of PVS in the past 3 years and the duodenal biopsies in both of our patients showed classical total villous atrophy. Both patients have responded extremely well to a gluten-free diet and iron and folic acid supplements. For iron deficiency to progressing to esophageal web formation, it needs to be chronic and insidious, a situation not uncommon in our country.

Although, esophageal webs are not reported in proportions to the prevalence of iron deficiency anemia in India [3]. This suggests that although iron deficiency plays a key role in the pathogenesis, factors other than iron are likely to be involved [4,5]. Both our patients of chronic iron deficiency anemia with dysphagia came out to be of celiac disease, raising suspicion whether iron deficiency anemic state in the setting of malabsorption is the cause rather than just a nutritional iron deficiency. There was a similar case reported by Dubey *et al*. which showed that the patient presenting with dysphagia has atypical celiac disease [6]. Another case report was described by Hefaiedh *et al*. showed an association of PVS in celiac disease [7].

CONCLUSION

As all patients of PVS failed to respond to iron supplements, it further lends support to this debate. We experience that all patients with PVS should undergo an endoscopic evaluation to diagnose celiac enteropathy.

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Funding: None; Conflict of Interest: None Stated.

How to cite this article: Sharma ZP, Sharma D, Sharma R, Yadav SK. Plummer–Vinson syndrome in celiac disease. Indian J Case Reports. 2020;6(4):197-198.

Doi: 10.32677/IJCR.2020.v06.i04.015