Paterson Kelly Syndrome in Celiac Disease

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Abstract
The association of Paterson Brown Kelly Syndrome and celiac disease is not widely reported. Both the entities have iron deficiency anemia. This case report emphasizes the need of screening for celiac disease in patients with Paterson Kelly Syndrome.

INTRODUCTION

Paterson Brown Kelly Syndrome (Plummer Vinson Syndrome, sideropenic dysphagia or post cricoid dysphagia) comprises of a triad of troubling findings: iron deficiency anemia, dysphagia and esophageal webs. The pathogenesis of the syndrome is not well understood although underlying iron deficiency of long duration is implicated as the possible explanation. However, its occurrence in association with celiac disease which almost always has iron deficiency anemia, has rarely been reported.

CASE REPORT

Patient 1

A 40 year female was aware of having anemia since 6-7 years & had been on iron supplements. The cause of anemia was possibly considered to be nutritional but not investigated. She presented to us with dysphagia to solids of 2 years duration, loss of appetite and loss of 2 kg weight over 2 years. She had no history of vomiting, altered bowel habits, GI bleed, pain abdomen or any other significant history. Menstrual history was normal. She had moderate pallor & koilonychia; rest of the general physical and systemic examinations being normal. Her investigations revealed iron deficiency anemia (hemoglobin 6.4 g/dl(12-18), serum iron 12 µg/dl(45-158), microcytic hypochromic RBCs). Her serum ferritin was 10 ng/mL(30-140) and bone marrow aspiration revealed iron deficiency anemia with absent iron stones. Serum calcium was 7.8 g/dl, serum albumin 3.2 g/dL, blood sugar, renal and liver function tests were normal. Serial tests for occult blood in the stool were twice negative. She underwent an upper GI endoscopy which revealed a post cricoid web. The web was disrupted endoscopically and dilated. Duodenal mucosa revealed scalloping of the folds and duodenal biopsy revealed total villous atrophy. Serum anti-tissue transglutaminase level was 74 u/ml(0-10). She was put on gluten free diet and her hemoglobin had improved to >10 g/dl after 18 months follow-up. Her weight has increased and she has no recurrence of dysphagia.

Patient 2

A 36 year female presented with a two year history of progressively increasing difficulty in swallowing and easy fatigability. She had lost 3 kgs of weight over past 2 years. There was no other positive history. Physical examination revealed a fairly well built lady with pallor and koilonychia and angular stomatitis; systemic examination was normal. Her investigations were as follows: hemoglobin 7.8 g/dl(12-18), MCV 56 fl(77-93), MCH 20 pg(27-32), MCHC 28%(31-35), total leucocyte count 5.9x10⁹/l(4-11) and platelets 220x10⁹/l(150-400). Her serum iron was 18 µg/dl (45-158), total iron binding capacity 481 µg/dl (250-400) and serum ferritin 8.5 ng/ml(30-140); peripheral blood smear was suggestive of iron deficiency anemia. Serum albumin was 3 g/dL (3.5-5.0); rest of the blood biochemistries being normal. Upper gastrointestinal endoscopy revealed esophageal web in the post cricoid region, which yielded to Savary Gilliard dilators. The distal duodenal mucosa revealed effacement of folds. Duodenal biopsy revealed total villous atrophy. Her anti-tissue transglutaminase levels were 56. She was started on gluten-free diet. Her hemoglobin improved gradually and she was asymptomatic during one year follow-up.

DISCUSSION

In the last decade, iron deficiency anemia alone, has been recognized as the presenting feature of underlying celiac disease. The prevalence of celiac disease is on the rise in India. We came across two patients of Plummer Vinson syndrome in last 2 years and duodenal biopsies in both these patients showed total villous atrophy. Both patients have responded to gluten-free diet and iron supplements. For iron deficiency to progress to web formation it needs to be chronic and insidious, a
situation not uncommon in India. However, esophageal webs are not reported in proportions to the prevalence of iron deficiency anemia in our country. This suggests that although iron deficiency plays an important role in the pathogenesis, factors other than iron are likely to be involved.

Both our patients of Plummer Vinson Syndrome turning out to be actually celiac disease raises suspicion whether iron deficiency state in the background of malabsorption is the cause rather than just nutritional iron deficiency. As all patients of Plummer Vinson syndrome fail to respond to iron supplements, it further lends support to this contention. We feel that all patients with Plummer Vinson syndrome should undergo endoscopic work up for celiac disease.

REFERENCES


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