Journal of Experimental and Clinical Medicine
http://dergipark.ulakbim.gov.tr/omujecm

Case Report

J. Exp. Clin. Med., 2016; 33(3): 167-169
doi: 10.5835/jecm.omu.33.03.008

Paterson-Kelly syndrome in a patient with celiac disease

Piltan Buyukkaya[a*], Nevin Alayvaz Aslan[a], Sude Hatun Aktimur[a], Serdar Turan[b], Hilmi Atay[a], Seda Gün[c], Engin Kelkitli[a], Ahmet Bektas[d], Mehmet Turgut[a]

[a] *Department of Internal Medicine, Division of Hematology, Faculty of Medicine, Ondokuz Mayıs University, Samsun, Turkey*
[b] *Department of Internal Medicine, Faculty of Medicine, Ondokuz Mayıs University, Samsun, Turkey*
[c] *Department of Pathology, Faculty of Medicine, Ondokuz Mayıs University, Samsun, Turkey*
[d] *Department of Gastroenterology, Faculty of Medicine, Ondokuz Mayıs University, Samsun, Turkey*

ARTICLE INFO

Article History
Received 30 / 12 / 2014
Accepted 22 / 04 / 2015

*** Correspondence to:**
Piltan Buyukkaya
Department of Internal Medicine
Division of Hematology,
Faculty of Medicine,
Ondokuz Mayıs University,
Samsun, Turkey
e-mail: pitan79@hotmail.com

Keywords:
Bougie dilation
Celiac disease
Dysphagia
Iron-deficiency anemia

ABSTRACT

Paterson-Kelly syndrome is characterized with iron deficiency anemia and esophagial web. Association between Paterson-Kelly and Celiac disease is not well-known. Especially in our country, there is insufficient data about these two diseases. We report a case with a complaint of dysphagia and diagnosed as Paterson-Kelly syndrome with celiac disease. Dysphagia was resolved with bougie dilation, oral iron supplement and gluten free diet. We want to emphasize the importance of screening for celiac disease in patient with dysphagia and iron-deficieny anemia.

1. Introduction

Paterson Brown Kelly syndrome (Plummer Vinson syndrome) is described with triad of dysphagia, iron-deficiency anemia and esophagial webs. Loss of iron dependent enzyme in esophagial mucosa cause impaired peristaltism. So web formation occurs in esophagus. So there is a direct association with iron deficiency anemia and esophagial web. It has been known that treatment of iron deficiency resulted with improvement of dysphagia (Klifto et al., 1983). In celiac disease, iron deficiency anemia was devoloped due to impaired absorption of iron from duodenum in the 12-18% of patients (Ackerman et al., 1996). There are very few case reports suggesting association between celiac disease and Plummer Vilson syndrome (Dickey and McConnell, 1999; Malhotra et al., 2000, Sood et al., 2005; Sinha et al., 2008). We want to report a case with severe dysphagia and celiac disease who was succesfully treated with iron supplements.

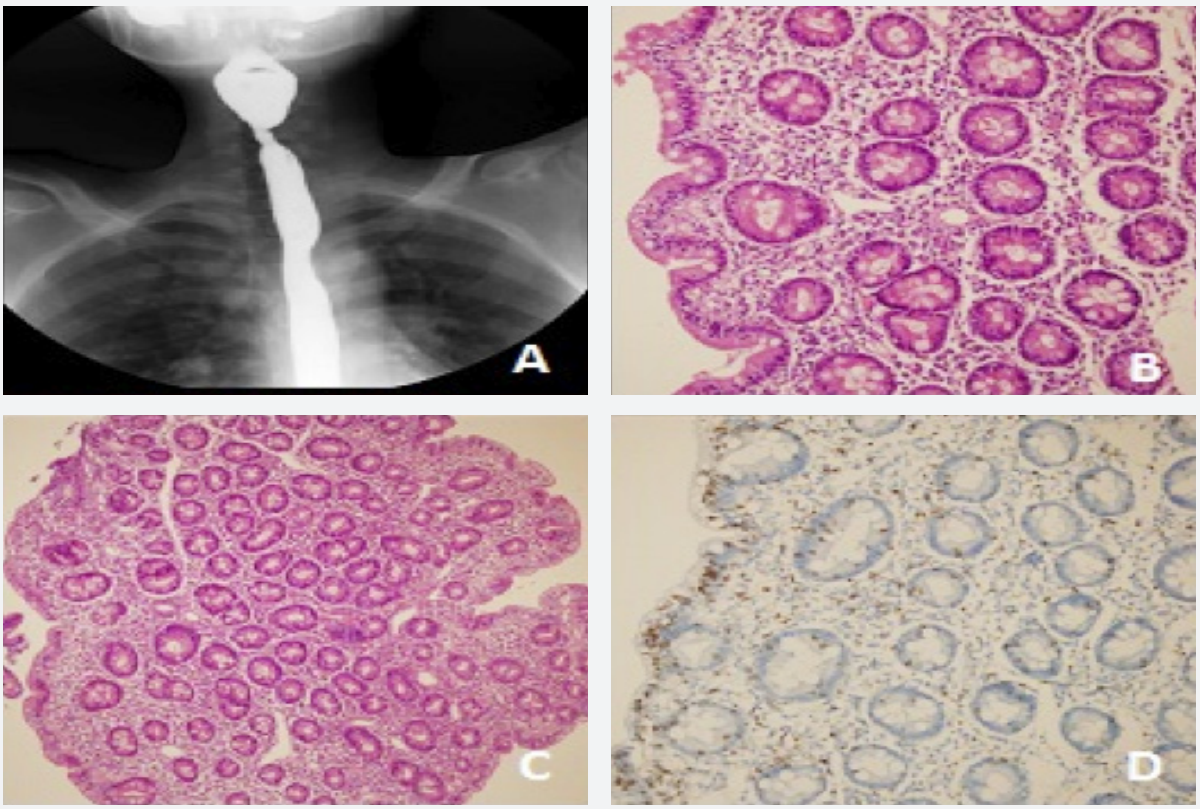

Fig. 1. A. Pharyngoesaphagography shows 1,5 cm lenght web on prowimal esaphagus. **B-C:** Pathologic examination of duodenal biopsy with high field **(B)** and low **(C)** power view shows villous atrophy and blunting; **D**: Lymphocytic infiltration with CD3 positive lymphocytes in surface epithelium of duodenum

2. Case report

A fourty-two year old woman came to hematology clinic with complaints of exhaustion, dizziness and palpitation. In her story, she had difficulties in swallowing especially with the solid foods for ten years. She have had nousea and vomiting for 15 days. Her difficulty in swallowing was increased recently. She lost 6 kg in the past six months. She has taken oral iron treatment discontinuously for ten years because of anemia. On physical examination she looked pale and angular stomatitis was present. Laboratory examination of the patient showed hemoglobin:7.9 g/dL, platelets (PLT): 1302 000/uL, iron saturation: 2,64%, serum iron level: 8 µg/dL (37-145), ferritin: 2.28 ng/mL (21.8-274), folic acid: 3,7 ng/mL (4.6-18.7) and vitamine B12:419 pg/dL (197-866) with normal liver and renal function. Examination of peripheral blood smear showed microcytic hypochromic anemia and excess thrombocytes with no atypical nucleated cells. For further investigation of iron deficiency anemia serologic test was done for celiac disease and antigliadin Ig A was 53.1 U/mL(<12), antigladin IgG was 62 U/mL(<12) and anti-tissue transglutaminase level was 69 U/L (0-10).

In gastroscopic examination there was narrow part that could not be passed with endoscope in the upper esophagus sphincter localization. In pharyngoesaphogography, after the pharyngoesophageal junction there was a web approximately 1.5 cm length on proximal esophagus (Fig. 1A). In another session, dilatation was done with 9-11-13 mm-bougies. On the second part of duodenum there was a ‘’cracked earth view ‘’and biopsy was taken from there. Villus atrophy and blunting were observed via microscopic examination (Fig. 1B,C). There was a lymphocytic infiltration with CD3 positive lymphocytes in surface epithelium (Fig. 1D).

Patient was diagnosed as gluten enteropathy and gluten free diet was started and intravenous ferric hydroxide was given with a dose of 1500mg. After one month of treatment her Hbg was raised to 11.2 gr/dL and her complaints were disappeared.

3. Discussion

There is a well-known relationship with iron deficiency anemia and celiac disease (Corazza et al. 1995; Ackerman et al., 1996). In celiac disease, 12-18% of patients have iron-deficieny anemia (Corazza et al., 1995) because of iron malabsorption.

Also iron deficienc anemia is one of the components of Paterson Kelly syndrome. Strong relationship with these two disease was shown in two case series with 72 and 63 patients (Chisholm et al., 1971; Bredenkamp et al., 1990). But there were few case reports and one prospective study regarding association of esophagial web and celiac disease. In a prospective study from India includes 21 patients with esophagial web and dysphagia. Eighteen of them had iron-deficiency anemia and five (23.8%) of them diagnosed as celiac disease with serology and endoscopic biopsy (Sinha et al., 2008). Duration of dysphagia before diagnosis of celiac disease ranged from six months to 15 years (Sinha et al., 2008). Sood et al. (2003) retrospectively investigated 96 cases with celiac disease and they found that three of them had esophagial web also. In most of case reports and study, none of the patients had chronic diarrhea at presentation (Sood et al., 2005; Sinha et al., 2008).

Celiac disease is misdiagnosed in case with esophagial web because serologic test and endoscopic biopsy is not routinely done. In our case, there was no symptom of diarrhea or skin lesions that indicate celiac disease. But patient had iron deficiency anemia and dysphagia since ten years and symptoms were resistant to iron replacement. Dickey and McConnell (1999) reported two cases with celiac disease and esophagial web and they were diagnosed as celiac disease after 13 and 9 years from dysphagia begun.

In many patients with Paterson-Kelly syndrome dysphagia is resolved after iron replacement (Hoffman and Jaffe, 1995). But in celiac disease in the absence of gluten free diet, iron deficiency anemia persist. In our patient bougie dilation was done for dysphagia and iron supplements were given orally. Also the importance of gluten free diet was emphasized to the patient. During a follow-up for two months dysphagia did not recur and iron deficiency anemia was improved. In other case reports (Dickey and McConnell, 1999; Malhotra et al., 2000; Sood et al., 2005) and prospective study (Sinha et al., 2008) dysphagia was resolved after bougie dilation and iron replacement with gluten free diet.

Patients with Paterson-Kelly syndrome have a risk of malignancy-carcinoma of the esophagus and postcricoid carcinoma (Novacek, 2006; Ben Gamra et al., 2007). So regular follow up these patients is important and necessary.

4. Conclusions

Celiac disease is usually misdiagnosed in patient with Paterson-Kelly syndrome. So patients with esophagial web and iron deficiency anemia should be investigated for celiac disease even if there is no diarrhea or skin manifestation of celiac disease.

REFERENCES

Ackerman, Z., Eliakim, R., Stalnikowicz, R., Rachmilewitz, D., 1996. Role of small bowel biopsy in the endoscopic evaluation of adults with iron deficiency anemia. Am. J. Gastroenterol. 91, 2099-2102.

Ben Gamra, O., Mbarek, C., Mouna, C., Zribi, S., Zainine, R., Hariga, I., El Khedim, A., 2007. Plummer vinson syndrome. Tunis Med. 85, 402-404.

Bredenkamp, J.K., Castro, D.J., Mickel, R. A., 1990. Importance of iron repletion in the management of Plummer-Vinson syndrome. Ann. Otol. Rhinol. Laryngol. 99, 51-54.

Chisholm, M., Ardran, G.M., Callender, S.T., Wright, R., 1971. A follow-up study of patients with post-cricoid webs. Q. J. Med. 40, 409-420.

Corazza, G.R., Valentini, R.A., Andreani, M.L., D'Anchino, M., Leva, M.T., Ginaldi, L., De Feudis, L., Quaglino, D., Gasbarrini, G., 1995. Subclinical coeliac disease is a frequent cause of iron-deficiency anaemia. Scand. J. Gastroenterol. 30, 153-156.

Dantas, R.O., 1999. Iron deficiency and dysphagia. Am. J. Gastroenterol. 94, 3072-3073.

Dickey, W., McConnell, B., 1999. Celiac disease presenting as the Paterson-Brown Kelly (Plummer-Vinson) syndrome. Am. J. Gastroenterol. 94, 527-529.

Hoffman, R.M., Jaffe, P.E., 1995. Plummer-Vinson syndrome. A case report and literature review. Arch. Intern. Med. 155, 2008-2011.

Klifto, E.J., Allen, S.K., Metzman, M., Chase P.J., Kupersmit, M., 1983. Plummer-Vinson syndrome: Report of a case and review of the literature. J. Am. Osteopath. Assoc. 83, 56-59.

Malhotra, P., Kochhar, R., Varma, N., Kumari, S., Jain, S., Varma, S., 2000. Paterson-Kelly syndrome and celiac disease a rare combination. Indian J. Gastroenterol 19, 191-192.

Novacek, G., 2006. Plummer-Vinson syndrome. Orphanet. J. Rare. Dis. 1, 36.

Sinha, S.K., Nain, C.K., Udawat, H.P., Prasad, K K., Das, R., Nagi, B., Singh, K., 2008. Cervical esophageal web and celiac disease. J. Gastroenterol. Hepatol. 23, 1149-1152.

Sood, A., Midha, V., Sood, N., Bansal, M., 2005. Paterson Kelly syndrome in celiac disease. J. Assoc. Physicians. India. 53, 991-992.

Sood, A., Midha, V., Sood, N., Malhotra, V., 2003. Adult celiac disease in northern India. Indian J. Gastroenterol. 22, 124-126.