Plummer-Vinson syndrome in a 15-year-old boy

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Plummer-Vinson syndrome presents as a classical triad of dysphagia, iron deficiency anemia and upper esophageal web(s). The syndrome usually occurs in adults, and is rare in childhood. We report a case of this syndrome occurring in a 15-year-old boy. He presented with dysphagia and anemia. Radiological examination showed the presence of webs at the cervical esophagus. The boy was treated with endoscopic balloon dilation and iron supplementation and remains in good general condition six months after the treatment.

Key words: Plummer-Vinson syndrome, adolescent, iron-deficiency anemia, esophageal web, balloon dilatation.

Plummer-Vinson syndrome (PVS) is characterized by dysphagia, iron deficiency anemia and upper esophageal web(s). This syndrome is also known as Paterson-Kelly syndrome or sideropenic dysphagia[1]. Dysphagia is usually intermittent or progressive over years, and limited to solids. It is predominantly a disease of middle-aged females and very rare in childhood[2,3]. To our knowledge, to date only nine cases of PVS have been reported in the English-language literature in children and adolescents[4-8]. We report a case of this uncommon syndrome in an adolescent boy.

Case Report

A 15-year-old boy presented with symptoms of fatigue, pallor and progressive dysphagia over a one-year period. He had dysphagia with solid foods and had poor nutritional status. He never ate meat or meat products and had no pica. Physical examination was normal except for pallor. The weight and height were at the 10th percentile for age.

The laboratory evaluation revealed an iron deficiency anemia with a hemoglobin level of 6.2 g/dl, mean cell volume 52 fl (80.7-95.5), mean cell hemoglobin 14.9 pg (27.2-33.5), serum iron 7.8 μg/dl (59-158), total iron binding capacity 422.8 μg/dl (245-450), transferrin saturation of 1.8%, and ferritin level 1.2 ng/ml (30-400). Stool test for occult blood and the antigliadin and antiendomysial tests were negative.

A barium swallow esophagography revealed two indentations on the proximal part of the cervical esophagus and 1 cm long stenotic area between them (Fig. 1). The esophagogastroscopy confirmed the presence of a web in the cervical esophagus (Fig. 2). The instrument did not pass through at the level of the web. Balloon dilation was made under scopic control by the pediatric surgeon; afterwards, the endoscope passed through the esophagus easily and the distal mucosa was normal up to the duodenum.

The patient received oral iron, 6 mg/kg daily, and at the end of one month of iron therapy, fatigue and dysphagia disappeared and the patient began to gain weight. At one month after the initiation of therapy, follow-up hemoglobin and mean cell volume level showed an improvement to 11.6 g/dl and 68.2 fl, respectively. He remains in good general condition without any dysphagic complaints six months after the treatment.

Discussion

The main clinical features of PVS are upper esophageal web(s), dysphagia and iron deficiency anemia[1]. Most of the patients are middle-aged women and it is thought to

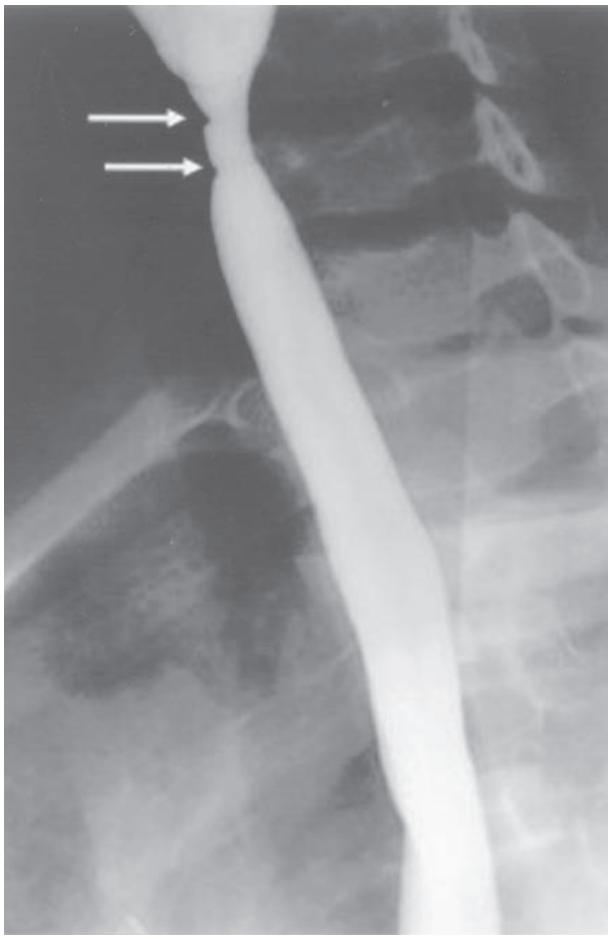

Fig. 1. Radiographic study showed the indentations (arrows) of the proximal esophagus indicating webs (oblique view).

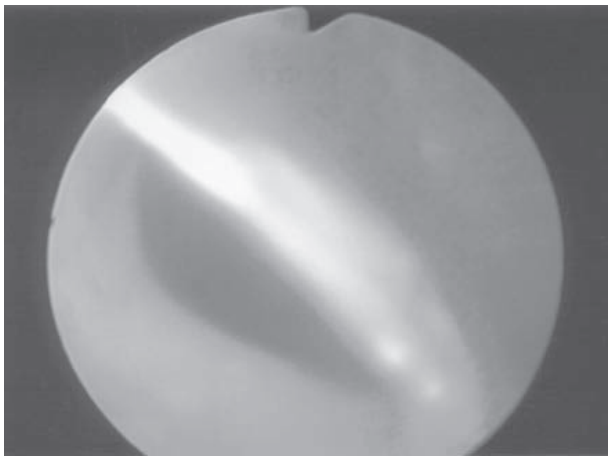

Fig. 2. Endoscopic view of the proximal esophageal web.

occur via blood losses from menstruations and pregnancies[2,3]. No exact data about the incidence and prevalence of the syndrome exist; only case reports have been published in the literature, and it is also very rare in childhood[4-8].

Symptoms resulting from anemia such as pallor, fatigue and weakness may dominate the clinical picture, and in our case as well, pallor and fatigue were the main symptoms and the patient had suffered from dysphagia for one year. The pathogenesis of the syndrome is not exactly understood but the most probable mechanism is iron deficiency[9]. The other factors, such as genetic predisposition, malnutrition and autoimmunity have not been proven to play an exact role in the pathogenesis of the syndrome, although celiac disease, thyroid disease and rheumatoid arthritis have been reported with PVS[1,10,11]. Whatever the source of the iron deficiency, the theory is based on the rapid losses of iron-dependent enzymes due to its high cell turnover. Reduction of these enzymes may cause mucosal degenerations, atrophic changes and web formation and may even lead to cancer development of the upper gastrointestinal tract[8,12]. The previous studies showed that iron deficiency decreased the contraction amplitude of the esophageal muscle resulting in motility impairment[13,14]. Miranda and Dantas[15] reported that patients with iron deficiency anemia had slower transit time at the proximal and middle parts of the esophagus than normal volunteers, but no significant difference at the distal esophagus was detected. This theory may explain why these webs occur at the proximal part of the esophagus.

The development of anemia in adolescence is thought to result from increased growth and erythropoietic activity and loss of blood with menstruation in females. In our case, the anemia was thought to be secondary to dietary deficiency of iron and increased growth. Although iron deficiency anemia is not uncommon in our country, we did not find any other case in childhood reported from Turkey, in contrast to adults[2,16].

Plummer-Vinson syndrome is rare, especially in childhood. We think that any child presenting with dysphagia and iron deficiency symptoms should be investigated for PVS. The prognosis is good but due to the possibility of malignant transformation, regular follow-up is necessary.

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