A case report of Plummer–Vinson Syndrome in a young adult with poor eating habits

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SUMMARY

Iron deficiency anaemia (IDA) is a very common disease, especially in poor and developing countries. It has a high prevalence in children and young adults. In young patients with no other confounding factors other than nutritional deficiency, the treatment is straightforward with either an oral or intravenous iron supplementation. Nonetheless, IDA in older patients requires attention to red-flag symptoms such as dysphagia, loss of appetite, early satiety and loss of weight. Further investigations including an oesophagogastroduodenoscopy (OGDS) are required to identify any sinister causes in such cases. We report an IDA case of a young woman who presented with dysphagia, in which further investigations with OGDS and a barium swallow test revealed a diagnosis of Plummer–Vinson syndrome.

INTRODUCTION

Oesophageal web is a thin, semi-circular membranous structure in cervical oesophagus consisting of mucosa and scanty fibrous tissue. It is a rare cause of dysphagia. Plummer–Vinson syndrome (PVS) is characterised by the triad of oesophageal web, dysphagia and IDA. PVS is a predominantly disease of middle age females and rarely happen in young adult or childhood.[1] The overall incidence of PVS has declined over the years due to improved nutritional status.[2] However, profound transition in lifestyle and eating habits among young adult has placed them in nutritionally vulnerable population that fail to achieve dietary requirements, predisposing them to develop PVS. We have reported a case of PVS in a young woman who presented with dysphagia and history of bad eating habits.

CASE PRESENTATION

A previously fit 18-year-old Malay lady presented to a gastroenterology clinic with a complaint of dysphagia for a two-month duration. She described the dysphagia as difficulty to swallow solid food which was slightly relieved with a small amount of fluid. Otherwise, there was no significant loss of weight, loss of appetite, gastroesophageal reflux symptoms or change in bowel habit. She also denied any haematochezia or melaena and autoimmune disease symptoms. She had an uneventful birth history with normal vaginal delivery without developmental delay. There was no family history of malignancy.

Her clinical examination revealed a pale underweight woman of body mass index 18 kg/m^2 with angular cheilitis. Other systemic physical examinations were otherwise unremarkable. There was IDA from her blood investigations with a hypochromic microcytic anaemia with haemoglobin level of 7.7 g/dL, mean cell volume (MCV) of 54 fL, and mean cell haemoglobin (MCH) of 15.6 pg, and low iron, ferritin and transferrin saturation levels of 2.2 µmol/L, 7.9 µg/L and 2.7% respectively. Other blood tests including renal profile, liver function test and thyroid function test were unremarkable.

An OGDS demonstrated a web-like narrowing of upper oesophagus immediately post-cricoid at 17 cm from incisor (Figure 1). A normal calibre gastroscope was not able to advance beyond the stricture. Therefore, a slim calibre gastroscope with a diameter of 5.5 mm was subsequently used to pass through the oesophageal web, which spontaneously tore the web and dilated the stricture. No biopsy was done due to the mild bleeding of the tear. The remaining parts of the oesophagus were unremarkable without white exudates, concentric rings, or longitudinal furrows to suggest an eosinophilic oesophagitis. Stomach mucosa was normal, while the second and third part of duodenum did not show any endoscopic features of celiac disease, such as mucosal or villous atrophy or scalloping of the mucosa. Subsequently, a barium swallow test revealed a suspicious filling defect in the oesophagus (Figure 2). She was eventually diagnosed with Plummer–Vinson syndrome taking into consideration overall clinical findings. On further questioning of the patient's dietary habit, she admitted having poor eating habits since childhood. She usually skipped breakfast with only one heavy meal per day, which consists mainly of carbohydrates. Besides, she preferred to eat snacks and sweet beverages which contained high sugar with low fibre, protein and micro- or macronutrients. She denied any incidence of body shaming or fear of gaining weight. She had no issues at school or with peers.

She was given iron infusion with 1 g of ferric carboxymaltose followed by oral iron supplementation for 6 weeks. Oral proton pump inhibitors were also given to help in mucosal healing. A follow-up OGDS 6 weeks later revealed a wider oesophageal lumen at the previous oesophageal web, passable by a normal calibre gastroscope with gentle manoeuvre (Figure 3). Her symptoms resolved completely 6 weeks after OGDS and iron supplement. Her repeated blood parameters after 6 weeks showed haemoglobin of 11.8 g/dL with MCV of 72 fL and MCH of 26.6 pg, and iron, ferritin and

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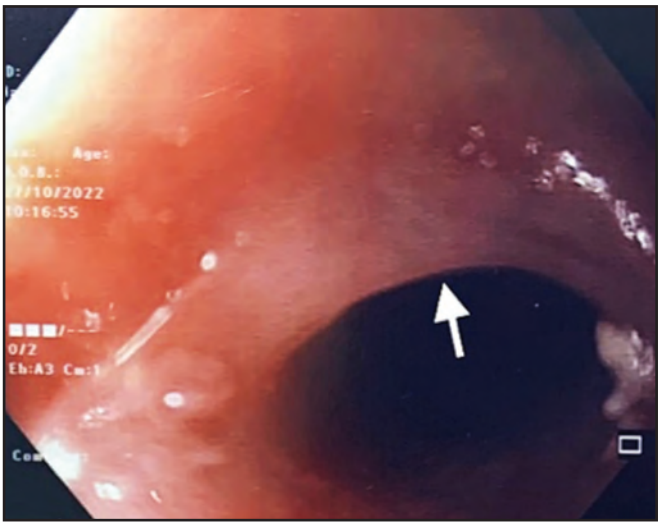

Fig. 1: White arrow showing the circular thin membrane (web) at upper oesophagus.

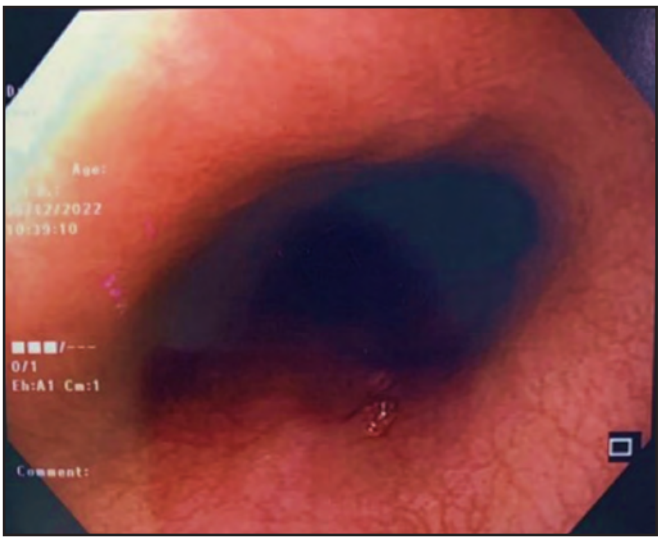

Fig. 3: Follow-up oesophagogastroduodenoscopy 6 weeks later showed resolution of oesophageal web and wider oesophageal lumen.

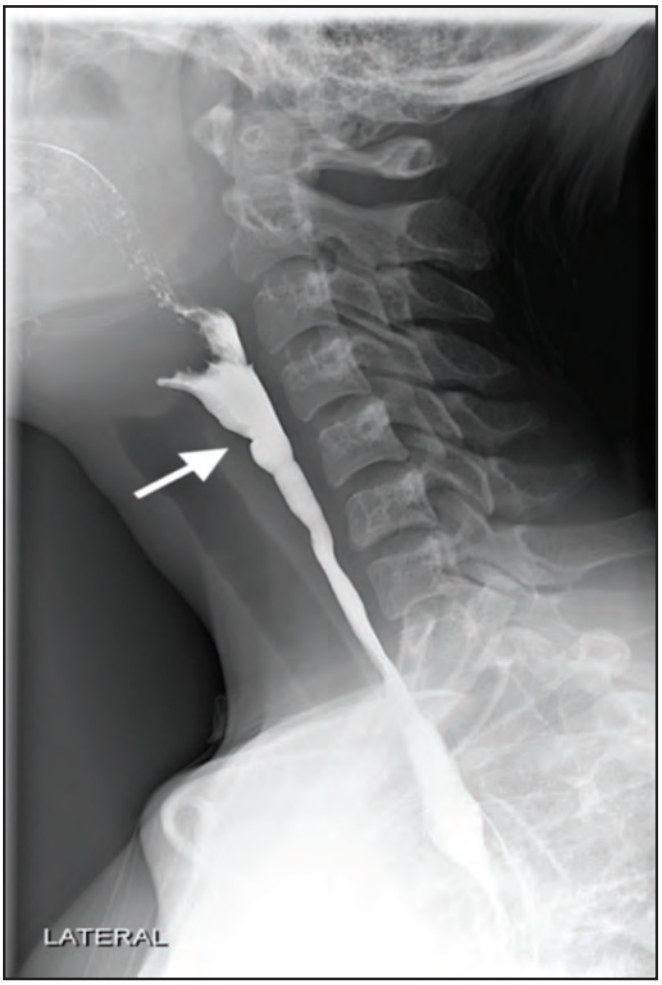

Fig. 2: White arrow showing suspicious filling defect in oesophagus.

transferrin saturation levels of 12.9 µmol/L, 114 µg/L and 24.5% respectively. She remained asymptomatic during a follow-up 4 months later with normalisation of her blood parameters with haemoglobin, iron, ferritin and transferrin saturation levels of 12.5 g/dL, 21.2 µmol/L, 31.8 µg/L and 34% respectively.

DISCUSSION

Plummer–Vinson syndrome is also known as the Paterson Brown–Kelly syndrome, which is a syndrome of a classical triad of dysphagia, oesophageal web and hypochromic microcytic anaemia. It was first reported by Henry Stanley Plummer in 1912 and is commonly found in middle-aged women. Nowadays, the syndrome is very rare due to improvements in nutritional status in developed countries. There is no recent data on the incidence and prevalence of this syndrome due to its rarity.[3] PVS often presents with symptoms of painless dysphagia, which is intermittent or progressive over years. It is usually confined to solid food and is occasionally associated with weight loss. Patients with PVS may manifest the signs and symptoms of anaemia, such as lethargy, palpitation, glossitis, angular cheilitis and koilonychias.

The exact aetiology of PVS is still unknown; numerous potential causes were proposed, including iron deficiency, autoimmune-related disorders, thyroid disease and malnutrition. IDA is postulated to be the aetiology as the symptoms usually improve with iron supplementation. This can be explained by the myasthenic changes in the alimentary tract muscles due to rapid depletion of iron-dependent oxidative enzymes in IDA, leading to mucosal degeneration, muscle atrophy and oesophageal web formation.[4] Inadequate iron intake is the predominant cause of IDA in developing countries. In our case, poor eating habits and a dietary deficiency of iron were the culprits in developing PVS.

Oesophageal webs can be detected by barium swallow radiography and endoscopy. The webs appear smooth, thin,

eccentric, and commonly located at the anterior wall of the oesophagus. OGDS has the advantage of direct visualisation of the webs, taking tissue biopsy and permitting treatment in the same setting. However, the webs may be missed by OGDS as its location is very close to the upper oesophageal sphincter.[5]

The management of Plummer–Vinson syndrome is generally straightforward. Iron supplementation is the standard first-line therapy which can effectively alleviate the symptoms. Intravenous iron therapy is more efficacious and can rapidly increase iron and ferritin levels compared to oral iron supplements.[6] An observational study by Das et al.[7] comparing the efficacy of oral and intravenous iron in raising the haemoglobin and ferritin levels after 28 days of treatment showed a statistically significant result ($p < 0.001$) in the intravenous iron group. Other than giving iron supplements, a healthy diet is important as these patients usually have concomitant vitamins and electrolytes deficiencies. However, in those patients with dysphagia who do not improve with iron supplementation, a mechanical dilation of the oesophageal web is needed. A single or serial dilatation might be needed to relieve the oesophageal stricture by using endoscopic bougies or balloon dilatation.[8] A study by Huynh et al.[9] showed that a single session of balloon dilation is able to dilate the web with low risk of perforation and recurrence. Although the treatment is simple and symptoms can completely resolve after the correction of IDA, most patients need a scheduled follow-up as this syndrome is considered a precancerous state with a risk of squamous cell carcinoma of hypopharynx and upper oesophagus. About 10% of patients with Plummer–Vinson syndrome have a high risk of developing squamous cell carcinoma thus a surveillance endoscopy three-yearly is recommended.[10] In addition, for patients who have lack of symptoms and biochemical resolution despite medical and endoscopic interventions, relevant investigations to look for autoimmune and rheumatological disorders need to be performed, especially if the clinical suspicion index is high.

CONCLUSION

High index of suspicion for Plummer–Vinson syndrome in patients with dysphagia and IDA is crucial to prompt diagnosis and initiate treatment in order to prevent the development of complications such as oesophageal stricture and malignancy.

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