(*Refer to page 208*)

Answer: Plummer-Vinson or Patterson-Kelly-Brown Syndrome

Plummer-Vinson syndrome (PVS) is defined as a triad of upper oesophageal web (or ring), dysphagia and iron deficiency anaemia (IDA). It was first reported in the early twentieth century, and is also widely known as the Patterson-Kelly-Brown syndrome (PKBS) or sideropenic anaemia with epithelial lesions.

Between 1908 and 1912, Plummer [1, 2] reported a series of 21 patients who presented with a combination of chronic IDA and dysphagia with suspected upper oesophageal spasm. However, there were no obvious anatomical obstructions demonstrated and this led Plummer to attribute the dysphagia to hysteria. In 1919, Vinson [1] reported his finding of "angulation of oesophagus" in another patient with similar presentation, and based on the report from Plummer, Vinson attributed the dysphagia to neurosis and hypothesised that IDA in these patients was secondary to poor nutritional status as a consequence of dysphagia. In that same year, Patterson and Kelly, [1] both laryngologists from the United Kingdom associated the dysphagia to what they believed was oesophageal spasm or web and disregarded Plummer-Vinson's theories of hysteria. They were also the first to describe the clinical features of the classic triad of the syndrome.

PVS or PKBS is very rare nowadays, although the incidence of benign dysphagia and oesophageal webs in isolation are still relatively common. It is more commonly found in Caucasian females, [1] between the ages of 40 and 70, with occasional reports in children and adolescents. The exact pathogenesis is still unclear. A number of mechanisms have been proposed and these include genetic predisposition, auto-immunity and secondary to acid related injuries from heterotopic gastric mucosal patch or cervical inlet patch.

The clinical features include intermittent dysphagia for solid foods (rarely or never for liquid). Symptoms and signs of anaemia (weakness, pallor, fatigue, tachycardia, glossitis, koilonychia and cheilitis) may predominate. Diagnosis requires the presence of IDA, oesophageal webs/rings and dysphagia. These webs/rings can be detected by barium swallow study or video-fluoroscopy and appear as a shelf in the proximal oesophagus, usually on the anterior wall. The proximal locations differentiate webs/rings in association with PVS from other types of webs/rings of the oesophagus (*Refer to supplementary text for types of rings*). Due to its proximal location, it can be missed during endoscopy.

Management includes iron replacement therapy with or without dilatation of the webs/rings. Prognosis is usually excellent, but an increased risk of squamous cell carcinoma of the oesophagus has been noted and hence the need for long term follow up may be indicated.

REFERENCES

1: Novacek G. Plummer-Vinson Syndrome: A review. Orphanet J Rare Dis. 2006; 1:36.

2: Ganesh R, Janakiraman L, Sathiyasekaran. Plummer-Vinson Syndrome: An unusual cause of dysphagia. Ann Trop Paediatr. 2008; 28:143–7.

3: Dantas R O. Iron deficiency and dysphagia. Am J Gastroenterol. 1999; 94:3072–3.