Paterson-Brown-Kelly Syndrome


Introduction

This is a condition where iron deficiency is associated with a post-cricoid oesophageal web. A description of this syndrome was published by Henry Plummer in Chicago in 1911 and also by Porter Vinson in Philadelphia in 1919. Donald Ross Paterson and Adam Brown-Kelly published independently of each other in London in 1919. [1] Hence British doctors call it the Paterson-Brown-Kelly syndrome, although it is increasingly known as the Plummer-Vinson syndrome on both sides of the Atlantic.

Epidemiology

The syndrome most often affects middle-aged women (this may be related to a propensity for iron deficiency), although it can occasionally present in a child. [2] The web (containing mucosa and submucosa) occurs at the anterior post-cricoid area of the upper oesophagus. [3] It is an uncommon condition and much of the literature is case reports of one or a few individuals rather than large series. Figures for incidence and prevalence are not available but it is becoming rarer, probably because dietary inadequacy is becoming less common. [4] A high prevalence of Plummer-Vinson syndrome was reported in Sweden in the early 20th century. This reduced after the introduction of iron supplementation in food. [5].

Risk factors

- Iron deficiency - this is related to poor diet or blood loss. [2]

History [2] [3]

The presentation is usually with painless, intermittent dysphagia. It tends to be with solid foods but, if untreated, may progress to soft foods and even liquids. As with other causes of oesophageal strictures, webs and rings, the patient is able to give a good indication of the level of obstruction. In this condition the patient indicates the upper oesophagus. Dysphagia may lead to weight loss. This should be regarded as a sign of danger. [6]

There may also be features of iron-deficiency anaemia like lethargy, tiredness and shortness of breath on exertion.

Examination [2]

The features that appear on examination are those associated with iron deficiency. There may be pallor and even tachycardia if anaemia is marked. There may be koilonychia (spoon-shaped nails), angular cheilitis and glossitis.

Investigation

- FBC will show a microcytic, hypochromic anaemia. Ferritin is low.
- Barium swallow may show the web. [3] This may need to be enhanced with videofluoroscopy. [2]
- Biopsy may be required if malignancy is suspected clinically. [7]
Treatment

- Iron replacement can almost invariably be achieved by oral means. Adding vitamin C does not improve absorption significantly. There is rarely any need for parenteral iron. Supplements may be needed long-term because after correction it is important to maintain a normal iron status. Causes of blood loss like menorrhagia may require attention.
- Endoscopic dilatation or argon plasma coagulation therapy of the oesophageal web is occasionally required in cases of persistent dysphagia.

Complications

- Post-cricoid carcinoma or carcinoma of the oesophagus may develop.

Prognosis and prevention

- Correction of iron deficiency will correct the condition if malignancy has not yet occurred. Prevention of iron deficiency prevents the disease.

Historical notes

- Henry Plummer (1874-1937) was an American internist. He was a professor at the Medical School of Minnesota. He published: *Diffuse dilatation of the esophagus without anatomic stenosis (cardiospasm). A report of ninety-one cases.* Journal of the American Medical Association, Chicago, 1912, 58: 2013-2015.

Further reading & references

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