East African Medical Journal Vol 78 No. 6 June 2001
PLUMMER VINSON SYNDROME: CASE REPORT
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SUMMARY

Plummer Vinson syndrome is characterised by dysphagia, iron deficiency, anaemia and oesophageal web or webs. This is a case report of a 33 year old Asian female who presented with slowly progressive dysphagia and a long history of iron deficiency anaemia. The anaemia was confirmed on repeated haemograms and a barium swallow revealed an upper oesophageal web. Upper gastrointestinal endoscopy and forceful dilatation were necessary to effect relief of dysphagia.

INTRODUCTION

Plummer Vinson Syndrome encompasses dysphagia secondary to an upper oesophageal webs, thought to be related to chronic iron deficiency anaemia. It typically affects middle aged females who may have other features of chronic iron deficiency, namely papillary atrophy of the tongue, spoon shaped brittle nails, angular stomatitis and pica.

Since the dysphagia is associated with iron deficiency anaemia, the term sideropenic dysphagia has often been used to describe this condition. The oesophageal web is best diagnosed on barium swallow.

Dysphagia is graded as follows: grades 0 – no dysphagia; I – able to swallow some but not all solids; II – able to swallow liquids only; III – able to swallow semi-solids but not solids and IV – complete dysphagia.

The precise aetiology and pathophysiology is unknown but the consistent association with anaemia suggests the possibility of mucosal changes due to the chronic iron deficiency resulting into proximal oesophageal webs. A web is usually defined as a thin membrane like structure containing mucosa and sub-mucosa without the inclusion of muscle layers. A web is usually eccentric and presents with dysphagia if the diameter of the lumen through the web is less than 12 millimeters. The web occurs in the proximal 4 or 5 centimeters of the oesophagus and may be difficult to diagnose at endoscopy if located just beyond the cricopharyngeus muscle. Due to its location, the web may accidentally rupture when the endoscope is passed through the cricopharyngeus muscle.

Therapy of dysphagia in Plummer Vinson syndrome includes iron repletion, oesophageal dilatation and surgery(1,2). Some patients have relief of dyshagia with iron repletion but most require oesophageal dilatation and very few patients require surgical excision of the web. The patient had iron repletion therapy for almost four years but the dysphagia had persisted.

Plummer Vinson syndrome is thought to be a pre-malignant condition for post cricoid carcinoma(1). In view of its rarity no protocols are as yet established for surveillance endoscopy and if no symptoms recur, annual endoscopy is advisable.

CASE REPORT

A thirty-three year old female patient was referred in March 2000 with a five years history of slowly progressive dysphagia and weight loss. The dysphagia was of grade IV, associated with chest pains. The pain may have been due to the possible oedema secondary to the failed multiple attempts of intubation during her upper gastrointestinal (GI) endoscopy by a physician three days earlier. Her dysphagia before the endoscopy had been intermittent and varying between grade II to III, but slowly progressive. She had lost approximately fifteen kilograms in weight over the five years and attributed this primarily to her inability to swallow well.

She gave no history of epigastric pain, nausea, vomiting, heartburn or anorexia. Her bowels were regular and she had normal micturition habits. Her menstrual blood loss was excessive. Her general practitioner had diagnosed iron deficiency anaemia from the haemogram and peripheral blood film reports and attributed the anaemia to the menorrhagia. Her haemoglobin had remained around 9 grams/dl for many years despite being on oral iron supplements.

She had intermittent dysphagia which on a barium swallow was found to be secondary to an upper oesophageal web. She was then referred by her general practitioner to a physician for an upper gastrointestinal endoscopy which failed and was then re-referred.

In view of the dysphagia, iron deficiency anaemia and an oesophageal web, an impression of Plummer Vinson syndrome was formed and a repeat endoscopy planned. The endoscopy was performed under heavy sedation and upon direct vision entry into the oesophagus, an oesophageal web was seen just a few millimeters distal to the cricopharyngeup muscle. Keeping the luminal orifice in view, a spring tipped guide wire was introduced into the stomach and Eder-Peustow metal Olive dilatation performed. A repeat endoscopy confirmed the oesophageal web to be of moderately thick mucosa with minimal bleeding due to the forceful dilatation.

The patient was discharged on antibiotics and analgesics. She was able to swallow well upon review two weeks later. She was advised to have regular endoscopic surveillance of the upper oesophagus and a repeat endoscopy planned after one year.

DISCUSSION

Paterson and Kelly independently described a clinical state in 1919 with which the names of Plummer and Vinson later became associated in the United States.

The incidence of Plummer Vinson syndrome is decreasing in the developed world. This is most likely due to improved dietary status and the treatment of sideropenic anaemia with inorganic iron salts(2,4). In the author's personal experience of over 5000 upper gastrointestinal endoscopies, this was the first case of Plummer Vision syndrome. This was the first patient with an oesophageal web, out of the 100 patients with oesophageal narrowing so far seen, resulting in a local incidence of 1% of Plummer Vision syndrome which compares well with other studies. Of all the benign strictures seen locally, Plummer Vinson syndrome accounts for approximately 2%(6).

Although it commonly affects middle-aged females, Plummer Vinson syndrome has been described in children and adolescents(7,8).

Even though a barium swallow is the method of choice for diagnosis of Plummer Vinson syndrome, a careful upper GI endoscopy with introduction of the endoscope under vision past the cricopharyngeus, is better as it allows for therapy via endoscopic dilatation which is successful in most cases(8).

ACKNOWLEDGEMENTS

To Dr. J.P. Jani for her assistance in preparation of this manuscript.

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