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A Case Report On Plummer Vinson Syndrome

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A B S T R A C T

Plummer Vinson syndrome is a rare association of post cricoid dysphagia, upper oesophageal webs and iron deficiency anemia. There is hypothesis that iron deficiency anemia plays a major etiological role in causing PVS. It typically affects a middle aged female, who has other conditions like chronic iron deficiency anemia. Plummer Vinson syndrome can be treated successfully with iron supplements. This condition is gradually decreased with better nutrition supply. The exact ethiopathogensis of Plummer Vinson syndrome is idiopathic. This paper presents a case of 36 year old woman with Plummer Vinson syndrome who was successfully treated.

Keywords: Esophageal Web, Iron Deficiency Anemia, Iron Therapy

Abbreviations: PVS- Plummer Vinson Syndrome, IDA- Iron Deficiency Anemia, SG- Savary Gilliard Dilators, BG- Bougies Dilators

1. Introduction

Plummer Vinson syndrome is also known as Paterson- Kelly syndrome/ Patterson brown Kelly syndrome/ sideropenic dysphagia is defined as the classical traid of dysphagia, iron deficiency anemia, oesophageal webs. It is commonly seen in middle- aged females. PVS is characterized by dysphagia due to upper oesophageal webs, anemia, and other features like weakness, pallor, tachycardia, fatigue. Dysphagia is usually painless and intermittent or progressive over the years which is limited to solids, sometimes PVS is also associated with weight loss. The exact etiology of PVS is unknown but it was hypothesized that PVS is due to iron deficiency anemia and some other etiological factors include genetic predisposition, malnutrition, autoimmune response. The diagnosis is mostly depended upon the identification of oesophageal webs by using barium swallow x rays. It's primarily diagnosed with the evidence of iron deficiency anemia. The other ways of identifying the PVS are gastrointestinal endoscopy, videochloroscopy is the best way for detecting the webs. The exact pathogensis is unknown.

2. Pathogenesis

Plummer Vinson syndrome is due to iron deficiency leading to rapid loss of iron dependent enymes

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Like tryptophan dioxygenase, ferrodoxin and 2- oxoglutarate dioxygenase

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Hence the loss of the following enymes causes mucosal degeneration with atropic changes of mucosa

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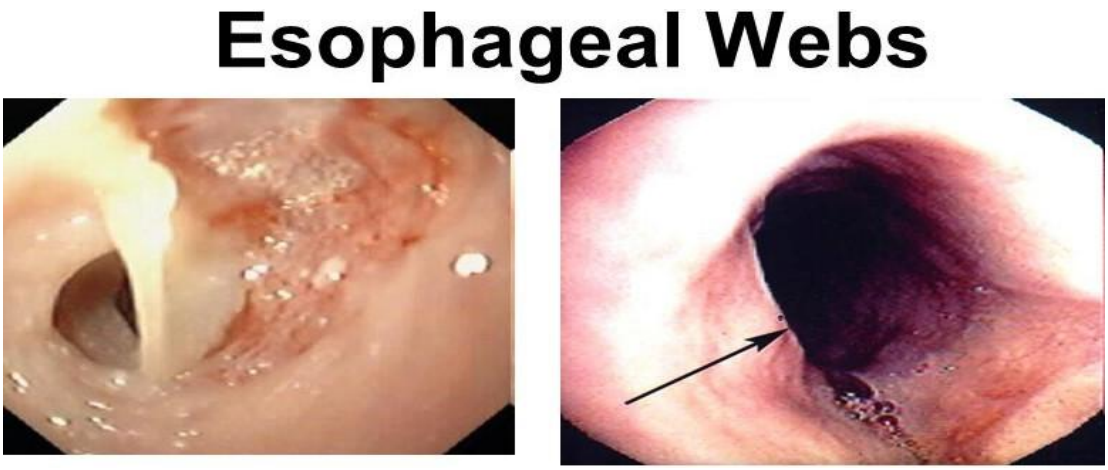
Thereby results in formation of oesophageal webs which are associated with dysphagia.



Iron deficiency is relieved by iron supplements. PVS can be managed by giving iron supplements and in case, when there is no cure with iron supplementations then a web dilation technique is done to allow a normal swallowing for the passage of food. A case study of PLUMMERS VINSON SYNDROME which was collected from a tertiary care hospital was attached below.

3. Case Presentation

A 36 years old female was admitted to the hospital with the chief complaints of dysphagia for both solids and liquids since two months, loss of weight , vomiting, anemia. She was diagnosed as severe iron deficiency anemia and was given iron supplements to manage the iron deficiency condition. The patient medical history was notable for Plummer Vinson syndrome, post cricoid web dilation up to 14mm (2013) which was now improved.

Physical examination:

Parameters	Observed range
Blood Pressure	110/70 mmhg
Temperature	98.6F
Pulse rate	66 beats/minute
Spo2	100

Complete blood picture:

Parameters	Observed Range	Normal Range
WBC	5400 Cells/mm^3	4000-10000
RBC	4.6mill/mm^3	3.8-4.8
HB	9.5gm/dl	12-15
HCT	35%	36-46
MCV	74fl	83-101
MCH	20pg	27-32
MCHC	27g/dl	32-33
RDW	29%	11.6-14

Liver function test:

Parameters	Observed Range	Normal Range
Total Bilirubin	0.5mg/dl	0.3-1.2
Direct Bilirubin	0.1mg/dl	Upto0.2mg/dl
Indirect Bilirubin	0.4mg/dl	Upto0.2mg/dl
SGPT	19U/L	0-40U/L
SGOT	27U/L	Upto40U/L

Video Endoscopy:

It is clinically diagnosed as a known case of anemia. There is a narrowing of post cricoid area in oesophagus scope not negotiable beyond.

USG of abdomen:

It has revealed that all the findings are within normal limits.

Dilation report:

Web at post cricoid area dilated upto14mm in oesophagus. Endoscopy dilation was done with SG dilators upto14mm.

Disease name and synonyms:

Plummer Vinson syndrome (PVS)

Paterson-Kelly syndrome (History)

Paterson-Brown Kelly syndrome

Sideropenic dysphagia (Since dysphagia is associated IDA it is termed as siderophemic dysphagia)

History:

The most frequently used name is Plummer Vinson syndrome, which was named after Henry Stanley Plummer(1874-1936) who reported a series of patients with longer duration of iron deficiency anemia ,dysphagia and spasm of esophagus.Paterson- Kelly syndrome which was named after Donald Ross Paterson(1863-1939) and Adam Brown- Kelly (1865-1941) who were first to describe the characteristic clinical features of the syndrome.

4. Discussion

Anemia is disordered process in which the rate of red cells production fails to match the rate of destruction ,which results in reduction of Hemoglobin concentration. Iron is important in the body for maturation and to maintain the integrity of the epithelium. PVS is a manifestation of severe, long term, IDA causing dysphagia because of esophageal webs. Esophageal webs are defined as extensions of normal esophageal tissues and may develop at anypoint along the oesophagus, patient may be asymptomatic and may develop dysphagia. They are described as a thin membranous extension of the oesophagus mucous and sub mucous.

Dysphagia from the webs of PVS is commonly painless and intermittent or progressive. Other features include glossitis(inflammation of tongue) glossopyrosis(burning sensation of tongue), glossodynia(multifunctional disorders characterized by painful sensations in mouth and throat), angularcheilitis(inflammation and small cracks in one or both corners of the mouth), koilonychias(spoon shaped nails), fragility, thinning of nails, brittle hairs. Symptoms secondary to anemia such as pallor, fatigue, weakness.

Plummer Vinson syndrome is most common in white females of fourth to seventh decade, but same cases are also reported in the children and adolescents. Most commonly patients first have dysphagia to solids but over time symptoms can progress to dysphagia to liquids as observed in our case. In this case usually dysphagia is painless and associated with weight loss. Dysphagia was the main symptom, which made the patient to seek medical help and dilation. The patient in this case is not associated with any systemic disorder.

The approach begins with lab evaluation of CBC count, iron and ferritin studies. Barium swallow studies and fluoroscopic evaluation suggest the diagnosis and degree of stenosis, esophageal gastroduodenoscopy confirms the diagnosis and helps therapeutically in the dilation of webs.

Mercury/tungsten filled bougies (maloney/hurst) bougienage dilators(bougie passed over guide wire) and through the scope(ballon dilation)are esophageal dilators commonly available. Complications of esophageal dilation may range from odynophagea(painful swallowing) hematemessis to severe sepsis, bleeding. In most cases, one session of such dilation is usually enough for long term relief but, rarely, multiple sessions may also be warranted. In this case, the patient is treated with sevary gilliard dilator and iron supplementation.

A variety of dilatord have been associated with PVS such as celiac disease, inflammatory bowel disease, Pernicious anemia, thyroid disease and Rheumatoid arthritis. Iron replacement is recommended atleast until normalization of the hematocrit and ferritinn levels. The benefit from dilation may be short-lived and satisfactory regulation of dysphagia can be concluded only upon longer follow-up periods.

5. Conclusions

Iron deficiency is a common cause of anemia. Severe anemia with koilonychias and dysphagia are the features of PVS. PVS is considered to be a premalignant condition .Early diagnosis is of utmost importance for a better prognosis. A case of PVS with barium swallow performed has been described here with the literature review

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