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PLUMMER VINSON SYNDROME – A CASE REPORT ON ESOPHAGEAL STRICTURE MANAGED BY ESOPHAGEAL DILATATION AND THEIR TREATMENT PATTERNS

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ABSTRACT :

Background: Plummer Vinson syndrome is found to be other synonym as sideropenic dysphagia is a disease characterized by chronic iron- deficiency anemia, dysphagia and esophageal web. Mostly affects in women as compared to men. Most of dysphagia and iron- deficiency can be treated by iron supplementation and procedure using by web dilatation.

Setting: Department of laryngology, Head and Neck surgery at PRS hospital PVT.LTD, Killipalam, Karamana, Trivandrum, Kerala.

Case report: A 55-year old female patient with complaints of foreign body sensation of lump seen in throat since 1 month and upper esophageal stricture was found by using esophagoscopy.

Intervention: Conservative line of management with blood transfusion and dilatation with esophagoscopy.

Results: A good symptomatic line of treatment and improvement was seen after blood transfusion and managed by dilatation with esophagoscopy.

Conclusion: Dilatation with esophagoscopy is a better way of managing upper esophageal stricture with minimal complications under general anesthesia.

KEYWORDS: Plummer- vinson syndrome, dysphagia**,** Iron- deficiency anemia, post cricoid region, esophageal stricture and management by using dilatation with esophagoscopy.

INTRODUCTION :

Plummer- Vinson syndrome, otherwise known as Paterson- Brown Kelly syndrome and sideropenic dyshagia this name is reportedly named in United Kingdom is a very rare condition which comprises a classic triad of symptoms like dysphagia, iron- deficiency anemia and upper oesophageal webs. Even though the syndrome is very rare nowadays, its recognition is important because it identifies a group of patients at increased risk of squamous cell carcinoma of the pharynx and the esophagus. It is more common in middle-aged women at an increased risk. This syndrome is associated with an increased incidence of post- cricoid carcinoma and for surveillance, an endoscopy is recommended under general anesthesia. Although it was first described as early as 1912, there is still limited knowledge about it due to the low and progressively decreasing incidence of syndrome. It is more common among middle-aged females. It is also important to recognize the plummer- vinson syndrome with esophageal and hypopharyngeal cancer.

The term sideropenic dysphagia has also been used, because the syndrome can occur with iron deficiency (sideropenia), but it is not associated with anemia. The exact cause and pathogenesis of PVS remains unclear, although iron and other nutritional deficiencies, genetic predisposition and autoimmunity have all been implicated in formation of webs. Treatment includes correction of iron deficiency and endoscopic dilation of esophageal webs to relieve dysphagia. PVS is associated with an increased risk of hypo pharyngeal and esophageal malignancies. Correction of iron deficiency may arrest and reverse the mucosal changes and possibly reduces this risk [(1, 2)].

EPIDEMIOLOGY

United States

Reliable prevalence data on PVS are lacking. The syndrome is now a rare, and its decline has been attributed to better nutrition and health care. Webs may be found in 5-15% of patients presenting with dysphagia, but most of these patients do not have PVS.

International data

Plummer- Vinson syndrome is rare but clinical data gathered come from case reports and case series. It is more common in females, accounting for 90% of the cases. It is also more common at ages 40-70years old, although it has been reported to occur in children as young as six years old.
In the first half of the 20th century, PVS was a relatively common finding, particularly in middle-aged Scandinavian women. The rapid fall in prevalence of the syndrome in the latter part of the 20th century has paralleled an improvement in the nutritional status.

Race-, sex-, and age-related demographics

PVS has mainly been described in whites, and it is more frequently observed in women. In earlier studies from Scandinavia, up to 90% of patients were women. The typical age range at diagnosis is 40-70 years. A handful of cases have been reported in children [(3)].

ETIOLOGY

The cause of Plummer-Vinson syndrome (PVS) is unclear. Proposed etiopathogenic mechanisms include iron and nutritional deficiencies, genetic predisposition, and autoimmunity. The underlying cause of iron deficiency anemia (eg, gastrointestinal blood loss, celiac sprue) [(4)].

PATHOPHYSIOLOGY

The pathogenesis of Plummer-Vinson syndrome (PVS) remains speculative, and the existence of the syndrome has been challenged. Postulated etiopathogenic mechanisms include iron and nutritional deficiencies, genetic predisposition, and autoimmune factors, amongst others.
The prevalent iron deficiency theory remains controversial. Older reports have implicated iron deficiency in the pathogenesis of esophageal webs and dysphagia in predisposed individuals. The depletion of iron-dependent oxidative enzymes may produce myasthenic changes in muscles involved in the swallowing mechanism, atrophy of the esophageal mucosa, and formation of webs as epithelial complications.
The improvement in dysphagia after iron therapy provides evidence for an association between iron deficiency and postcricoid dysphagia. Reports have also been made of patients with PVS exhibiting impaired esophageal motility (with dysphagia) that recovers following iron therapy. Moreover, the decline in PVS seems to parallel a universal improvement in the nutritional status, including iron supplementation.
However, population-based studies have shown no relationship between post cricoid dysphagia and anemia or sideropenia. Other studies have demonstrated that patients with webs are as likely to be iron deficient and webs are often found in patients without iron deficiency or dysphagia. Lastly, the iron deficiency theory does not explain the predilection of webs for the upper esophagus and the rarity of the syndrome in populations in which chronic iron deficiency is endemic. PVS has also been viewed as an autoimmune phenomenon. The syndrome has been associated with autoimmune conditions, such as rheumatoid arthritis, pernicious anemia, celiac disease, and thyroiditis. In one study, a significantly higher proportion of patients with PVS had thyroid cytoplasmic autoimmune antibodies compared to controls with iron deficiency. The autoimmune theory, however, has gained little acceptance to date [(5)].
A complicated inlet patch (heterotopic gastric mucosa) has also been implicated in the pathogenesis of PVS. An ulcerated inlet patch in the upper esophagus can cause structuring (web like formation) and bleeding (with subsequent iron deficiency). However, most studies with biopsy or autopsy specimens have not demonstrated the presence of gastric metaplasia in the samples.
There are multiple theories occurs in PVS is the exact cause. Low iron levels lead to reduction in the activity of iron- dependent oxidative enzymes that may leads to myasthenia changes in muscles, atrophy of the mucosa and development of webs. The evidence includes the improvement of dysphagia after iron supplementation. However not everyone improves with only taking supplementation and they will undergo and managed by procedure- dilation with esophagoscopy [(6)].

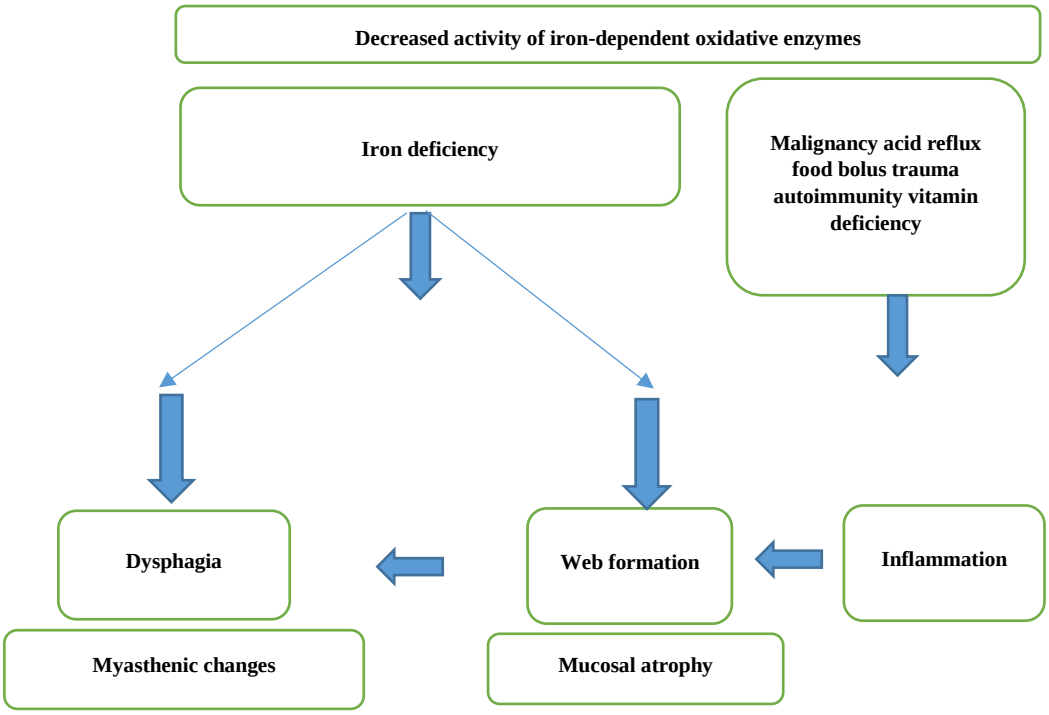

Diagram 1: Illustration which shows formation of Plummer- Vinson Syndrome (PVS)

CASE REPORT

A 55- year old female patient with MRD NO- 958539 consulted by Dr. Jaya kumar- complaints of sensation of lump was seen in throat since 1 month admitted at PRS hospital and diagnosed with plummer- vinson syndrome in 2/6/24 and procedure was done by using dilatation with esophagocopy and discharged on 4/6/24. There is no history of dysphagia and dyspnea. She had a past history on known case of dilatation and post- cricoid region appears on 3times from 2007-2013 diagnosed at KIMS hospital and history of hypothyroidism for $1^{1/2}$ years was on medication Tab. Thyronorm 88mcg p/o 1-0-0 and post menopause for 6years. On 23/5/24 was diagnosed with this case shows symptomatically no improvement. After diagnosing from KIMS hospital she took medicines on Tab. Razo (Rabeprazole 20mg p/o 1-0-1*15days) and Tab. Gavison liquid (Sodium alginate+ sodium bicarbonate+ calcium carbonate) and plan for esophagoscopy+ dilatation on 3/6/24 at PRS hospital and review after 2weeks if symptoms persist. Her vitals chart shows BP- 140/80mmHg, pulse rate- 65bt/min, respiratory rate- 18/min, spo2- 99%. Her ECG shows sinus rhythm and echo shows good LV function. Her clinical examination B/L cords and movement found to be normal, no pooling of saliva. Initially she was treated with stat medicines on 3/6/24 IVF. DNS-33/mins and IVF. RL- 500ml 33/mins. She was treated INJ. AUGMENTIN (AMOXYCILLIN+ CLAVULANIC ACID 1.2gm IV 100ml NS 1-0-1), INJ. PANTOP (PANTOPRAZOLE 40mg IV 1-0-0), TAB. ALPRAX (ALPRAZOLAM 0.5mg P/O HS), TAB. PANTOP. D (PANTOPRAZOLE P/O HS), NEB. DUOLIN (LEVOSALBUTAMOL+ IPRATROPIUM P/N HS), NEB. BUDECORT (BUDESONIDE P/N HS), TAB. THYRONORM (LEVOTHYROXINE 88mcg P/O 1-0-0), INJ. PCM (PARACETAMOL 1gm IV 1-1-1).

According to investigation reports, chloride level was found to be elevated and her urine routine examination shows colour- yellow, PH-7, appearance- clear, specific gravity- 1.005, urine albumin, urine sugar, bile pigments, salts and urobilinogen was found to be normal and her urine microscopic examination shows pus cells and epithelial cells was found to be elevated- crystals, cast and bacteria found to negative [(7)].

Table 1: Blood and Urine Examination Reports

PARAMETERS	**TEST VALUE WITH DATES**
	2/6/24
Hemoglobin	13.7
PCV/ Hematocrit	41.4
MCV	88.8

MCH	29.4
MCHC	33.1
WBC	6180
Polymorph	52.2
Neutrophils	55.3
Lymphocytes	35.8
Eosinophils	3.9
Monocytes	5.2
Basophils	0.5
ESR	18
PC	2.04
RBS	99
Urea	14
Creatinine	0.77
ALT	29.5
AST	54.4
Sodium	142
Potassium	3.89
Chloride	107.5
TSH	2.6
Total. Bilirubin	0.393
Direct bilirubin	0.17
Indirect bilirubin	0.223
Total protein	7.37

Albumin	4.64
Globulin	2.72
A/G ratio	1.71
ALP	74
HIV	NR
HCV	NR
BT	2mins
CT	5mins
Prothrombin time	13.2
Activated partial thromboplastin	31.5
INR	1.05
Colour	Yellow
PH	7
Appearance	clear
Specific gravity	1.005
Pus cells	2-4
Epithelial cells	4-6
RBC	0-1

Her pre- operative notes were diagnosed as plummer-vinson syndrome and was managed with dilatation+ esophagoscopy on 3/6/24.

Operation notes: Under general anaethesia, patient positioned. Direct larynogoscopy introduced. Granuation noted over posterior surface of body of right arytenoid, biopsied web seen in cricopharynx R>L esophageal web and dilatation done. Hemostasis achieved.

Post- Anesthesia orders on medication: INJ. TRAMADOL (TRAMADOL) 50mg IV SOS
INJ. EMESET (ONDRANSETRON) 4mg IV SOS
INJ. PARACETAMOL(PARACETAMOL) 1gm IV Q8H

Advise on discharge: TAB. CEFTUM (CEFUROXIME) 500mg p/o 1-0-1* 5days
TAB. PAN (PANTOPRAZOLE) 40mg p/o 1-0-1* 1 week
TAB. DOLO (PARACETAMOL) 650mg p/o SOS

Review in laryngology OPD after 1 week with HPE report.

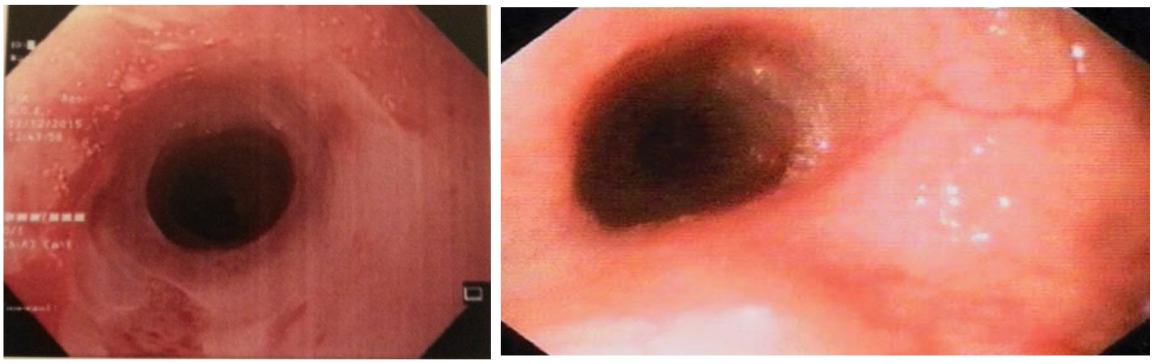

Figure 1: Endoscopic findings shows web in esophagus in partial view.

TREATMENT PATTERNS :

Various reports have shown that treatment of iron deficiency anemia alone may resolve the symptoms of dysphagia. The mechanism underlying the efficacy of iron supplementation in the resolution of symptoms in PVS is unclear, iron deficiency has been implicated to reduce iron dependent oxidative enzymes leading to degradation of pharyngeal muscles. The repletion of hemoglobin levels which act as scavengers of nitric oxide which in turn influences esophageal tone and peristalsis leads to stabilization of the appropriate nitric oxide levels and symptomatic improvement. The treatment of dysphagia can be done by esophageal dilators. In studies which shows ten patients only three needed multiple sessions of dilatations while all were treated successfully with endoscopic dilatations [8, 9].

Patients with PVS need treatment for iron-deficiency anemia for dysphagia:

IRON DEFICIENCY ANEMIA

First step in the management of IDA is to exclude the blood loss such as menstrual bleeding, gastrointestinal tract malignancy or iron malabsorption, celiac disease. Majority of people affects with PVS, the iron deficiency is nutritional and can be treated easily with iron supplementation, either oral or parenteral. Iron supplementation alone resolves dysphagia in many patients. This treatment alone could be considered for those with mild dysphagia if endoscopy facilities are not available. Prolonged dysphagia is unlikely to respond to iron replacement alone and requires dilatation of the web. Iron therapy should be considered in all patients with web, regardless of hemoglobin status, to replenish the iron stores [10, 11].

POST-CRICOID WEB

Esophageal webs have been dilated using various endoscopic techniques, in use of endoscopic balloon dilatation. The procedure using endoscopic balloon used after anesthetizing the throat mucosa with xylocaine spray. Light sedation helps improves patient comfort and does not appear to increase the risk of complication. Dilatation is performed under fluoroscopic guidance after passing a guidewire into the stomach at endoscopy. The rupture of web is recognized at endoscopy by the presence of a small amount of fresh blood at the location of web.

In some studies, shows that a single session of balloon dilatation provided complete response in 94% of patients, with only some patients need a repeat session. The recurrence of dysphagia is very uncommon. The rate of recurrence of dysphagia appears to be lower after endoscopic balloon dilatation. Treatment of recurrence is easy and usually needs only a single session of dilatation [12].

DISCUSSION :

PVS was described as a symptom complex which includes dysphagia, iron deficiency and presence of superior esophageal web. The syndrome mainly affects white women, in the 4th to 7th decade of life, but some pediatric and adolescent cases are also reported. The disease is rarer nowadays and the incidence is decreasing even in African nations where iron deficiency and malnutrition are common. The incidence and prevalence are not reliable as hematological parameters are not included. The pathogenesis of the syndrome is unclear but iron deficiency anemia, malnutrition, genetic predisposition and autoimmune etiologies are postulated. Iron-deficiency anemia is the widely accepted etiology as dysphagia and esophageal webs improve with iron supplementation. Upper esophageal webs are seen in only 10% of patients with iron deficiency which denotes a multiple etiology in the syndrome. Iron deficiency causes reduction of iron-dependent oxidative enzymes which results in gradual degradation of the muscles of the pharynx leading onto mucosal atrophy and development of webs [13, 14].

Reduction of these enzymes may cause mucosal degenerations, atrophic changes and web formation which causes neoplastic changes in the lower pharynx and upper esophagus. Also iron deficiency decreases the rate esophageal muscle contraction amplitude leading to motility impairment. The webs limited to the proximal and middle part of the esophagus can be explained on the finding that the transit time is slower in the upper esophagus in iron deficient individuals compared with normal volunteers. The patients usually complain of dysphagia to solids, but the symptoms can progress to dysphagia to liquids as well. The dysphagia is usually non-painful, leading onto progressive weight loss. As the progressive dysphagia is painless, patients tolerate it for long before presenting to the clinician. Symptoms of iron-deficiency anemia like pallor, fatigue and weakness dominate the clinical picture along with dysphagia [15].

The syndrome has a risk of squamous cell carcinoma of the post-cricoid pharynx and upper esophagus. Around 3 to 15% of the patients mostly women between 15 and 50 years of age, have been reported to develop esophageal or pharyngeal cancer. The syndrome is proven to be precancerous as 10% of the neoplastic changes in the hypopharynx, oral cavity and upper esophagus are malignant squamous cell carcinomas. Management includes diagnosing the cause of anemia like active hemorrhage, malignancy or celiac disease. The syndrome can easily be treated by iron therapy and the webs by mechanical dilatation. Iron supplementation can resolve dysphagia in many patients without mechanical dilatation. Iron therapy is advised even if the hemoglobin percentage is normal in the presence of web formation. Patients with choking and aspirations need dilatation therapy along with iron supplementation [16, 17].

The prognosis of this condition is good as anemia and dysphagia can be effectively treated by iron therapy and the webs by dilatation. The prognosis worsens dramatically if the syndrome is associated with complications like squamous cell carcinomas of hypopharynx and upper esophagus. Endoscopic dilatations are the procedure of choice for treating esophageal webs. In our case, dilatation of the esophageal web was done using esophageal dilatation under general anesthesia. This is the first time reported in literature where esophageal web dilatation using esophagoscopy has been done although balloon dilatation has already been reported. The patient had good symptomatic and radiological improvement after therapy [18].

CONCLUSION :

Plummer Vinson syndrome is a rare disease usually affecting middle aged females. It forms an important differential diagnosis of dysphagia. Since the dysphagia is slowly progressive, patients often present with iron deficiency anemia and treatment of anemia usually regresses signs and symptoms of PVS. Endoscopic dilatation or incision of esophageal web leads to marked symptomatic improvement in short span of time [19]. Here, we have described a unique way of dilatation of the upper esophageal web using the esophageal dilatation after iron supplementation. The prognosis is good but due to the possibility of malignant transformation, regular follow-up is necessary [20, 21].

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