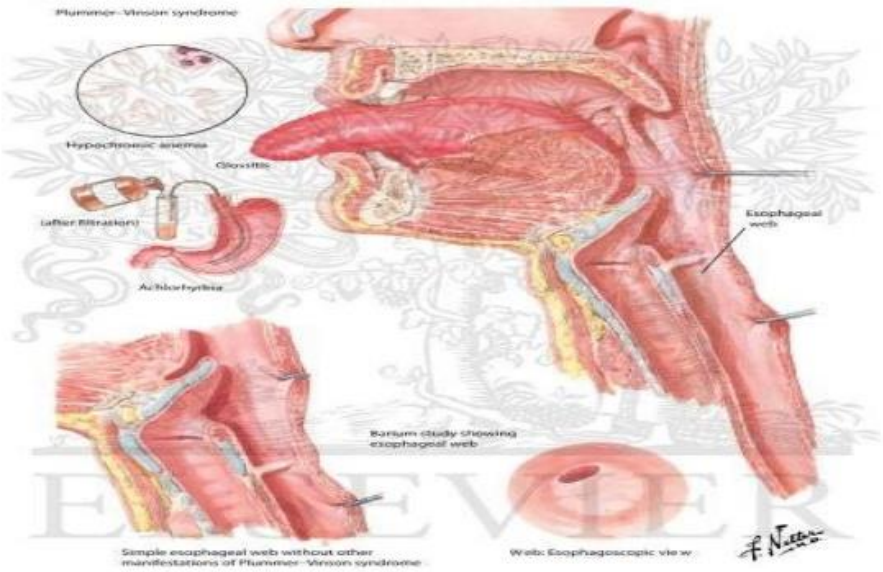
Plummer Vinson Syndrome



- Also called the Paterson–Kelly syndrome or sideropenic dysphagia
- Postcricoid dysphagia
- Upper esophageal webs
- Iron deficiency anemia
- 90% of patients were women
- Typical age range at diagnosis is 40-70 years

Pathophysiology

- Postulated etiopathogenic mechanisms include iron and nutritional deficiencies
- Genetic predisposition
- Autoimmune factors

Iron and nutritional deficiencies

Iron-dependent oxidative enzymes may produce myasthenic changes in muscles

Involved in

1. Swallowing mechanism
2. Atrophy of the esophageal mucosa
3. Formation of webs as epithelial complications

Autoimmune theory

- Associated with autoimmune conditions such as
  - Rheumatoid Arthritis
  - Pernicious Anemia
  - Celiac Disease
  - Thyroiditis
- Significantly higher proportion of patients with Plummer Vinson syndrome (PVS) had thyroid cytoplasmic autoimmune antibodies
- Little acceptance to date

Clinical features

Histroy

- Dysphagia - Typically intermittent and limited to solids, It is usually felt in the throat
- Choking spells
- Aspiration
- Weakness, fatigue, and dyspnea
- Weight loss is uncommon

Physical examination

- Angular cheilitis
- Glossitis
- Koilonychia (spoon nails)
- Pallor
- Splenomegaly, edentia (loss of teeth), and enlarged nodular thyroid glands in a few patients with PVS
- Some patients may have oropharyngeal leukoplakia

Investigations

Laboratory Studies

- Full blood count
- Peripheral blood smears
- Iron studies (eg, serum iron, total iron-binding capacity [TIBC], ferritin, saturation percentage)
- Other specific tests as necessary for the evaluation of the etiology of iron deficiency

Imaging Studies

- Barium esophagram
- Videofluoroscopy

The most sensitive methods and diagnostic tests of choice to detect esophageal webs

- Esophagogastroduodenoscopy

Histology

- Web is composed of a thin layer of normal squamous mucosa and submucosa

- Sometimes, chronic inflammatory cells may be observed in the submucosa

Treatment

- Usually managed on an outpatient basis

Management of dysphagia

- Diet modification may be sufficient in mildly symptomatic patients
- Advise patients to eat slowly and chew thoroughly
- Solid foods should be prepared and cut in small pieces, especially meats

Mechanical dilatation

- Significant and long-standing dysphagia usually require mechanical dilation
- Single large dilator is adequate and is thought to be more effective than serial progressive dilations

- Treat iron deficiency and its underlying cause
- Iron replacement - Ferrous sulfate
- Dysphagia may improve with iron replacement alone
- Address the cause of the iron deficiency (eg, celiac sprue, bleeding angiectasias)

Surgical Care

- Surgery is rarely needed
- Reserved for patients whose webs are recalcitrant to dilation or associated with Zenker diverticulum

Complications

Increased risk for,

- Hypopharyngeal cancer
- Oesophageal squamous cell cancers

Prognosis

- Generally good
- Unless PVS is complicated by hypopharyngeal or esophageal carcinoma
- Usually respond well to iron therapy, diet modification, and, if necessary, esophageal dilation
- Mortality is low