Section: Medicine

Case Report

PLUMMER-VINSON SYNDROME IN A YOUNG FEMALE WITH COEXISTING RHEUMATOID ARTHRITIS

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ABSTRACT

Background: Plummer-Vinson Syndrome (PVS) is a rare cause of dysphagia associated with iron deficiency anemia and esophageal webs. Rheumatoid arthritis (RA), an autoimmune systemic condition, may share overlapping features and complicate diagnosis. **Case Presentation:** A woman in her 30s presented with progressive dysphagia, iron deficiency anemia, and joint symptoms. Endoscopy revealed a pharyngeal web. Serology confirmed coexisting RA. She was managed with iron supplementation and immunomodulators. **Conclusion:** Coexistence of PVS and RA is uncommon but clinically significant. Early recognition and a multidisciplinary approach are essential for optimal outcomes.

INTRODUCTION

Plummer-Vinson Syndrome (PVS), also known as Paterson-Brown-Kelly syndrome, is a rare disorder primarily characterized by the triad of iron deficiency anemia (IDA), dysphagia, and esophageal webs. It typically affects middle-aged women, though recent reports suggest that it may also present in younger age groups, especially in settings of chronic nutritional deficiencies. The prevalence of PVS has significantly declined in developed countries due to improved nutrition and early detection of iron deficiency. However, it remains underrecognized in developing countries, where iron deficiency is common. The exact prevalence is unknown due to underreporting, but studies suggest it is more frequently encountered in females aged 40–70 years, with some estimates indicating a female-to-male ratio of 3:1 to 10:1.[1,4]

Rheumatoid Arthritis (RA), in contrast, is a chronic systemic autoimmune disease affecting approximately 0.5–1% of the global population, with a clear female predominance (F:M ≈ 3:1). It is characterized by symmetric polyarthritis and systemic inflammation, leading to joint destruction and extra-articular complications. Anemia of chronic disease (ACD) is common in RA due to chronic inflammation, but coexisting iron deficiency anemia may also occur, especially in female patients with poor dietary intake, chronic NSAID use, or heavy menstrual losses.[3,7]

The coexistence of RA and PVS, although rarely reported, is pathophysiologically plausible. Both disorders predominantly affect women and involve immune dysregulation and chronic inflammatory pathways. Chronic inflammation in RA can contribute to iron deficiency via hepcidin-mediated suppression of iron absorption and utilization. Moreover, systemic autoimmune diseases like RA can produce overlapping symptoms such as fatigue, weight loss, mucosal changes, and anemia, potentially masking the diagnosis of PVS.[8]

This case report presents a rare but insightful occurrence of PVS in a young woman with newly diagnosed RA, highlighting the need for clinicians to maintain a high index of suspicion for overlapping syndromes, especially when constitutional symptoms are disproportionate or refractory to standard treatment.

CASE REPORT

A female in her early 30s presented to the outpatient department with complaints of progressive dysphagia for solids for the past 5–8 months, associated with generalized weakness, fatigue, and significant weight loss. She also reported multiple joint pains, primarily affecting small joints of the hands and wrists, associated with morning stiffness lasting more than 30 minutes. There was no history of odynophagia, regurgitation, or hematemesis. Her menstrual history

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was unremarkable, and she denied any significant past medical or surgical history.

On general examination, she had severe pallor, angular stomatitis, glossitis, and spoon-shaped nails (koilonychia), suggestive of chronic iron deficiency. No lymphadenopathy was noted. Systemic examination revealed tenderness and swelling over the metacarpophalangeal and proximal interphalangeal joints bilaterally. There were no signs of organomegaly or lymphadenopathy. Her vital signs were stable.

INVESTIGATIONS

- Hemoglobin: 6.4 g/dL
- Mean Corpuscular Volume (MCV): 68 fL
- Serum Iron: 7 µg/dL (normal: 50–170 µg/dL)
- Serum Ferritin: 12 ng/mL (low)
- Total Iron Binding Capacity (TIBC): 460 µg/dL (elevated)
- Peripheral smear: microcytic hypochromic anemia
- Vitamin D: 8.0 ng/mL (deficient)
- Rheumatoid Factor (RF): 25 IU/mL (positive)
- Anti-Cyclic Citrullinated Peptide (Anti-CCP): 81 IU/mL (strongly positive)
- C-reactive protein (CRP): 20.86 mg/L (elevated)
- Erythrocyte Sedimentation Rate (ESR): 65 mm/hr (elevated)
- Barium Swallow: showed pharyngeal constriction suggestive of a cervical web
- Upper GI Endoscopy: revealed a thin esophageal web located at approximately 15 cm from the incisors.

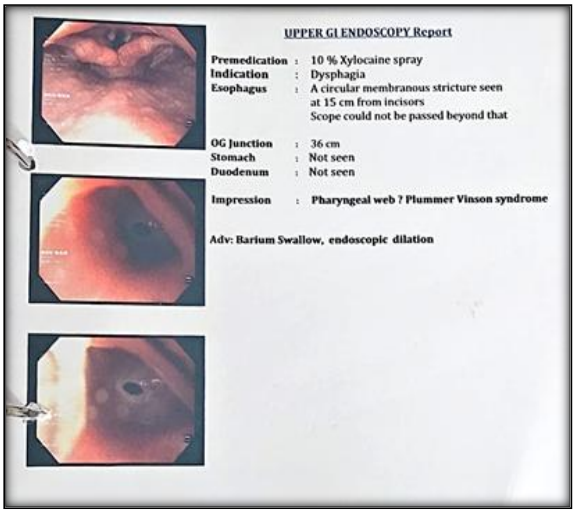

Figure 1: Endoscopic findings confirming a pharyngeal web at 15 cm from the incisors, suggestive of Plummer-Vinson Syndrome

Differential Diagnosis

The initial clinical picture of progressive dysphagia raised concerns for structural or neoplastic causes, such as:

- Esophageal carcinoma
- Esophagitis (infectious or reflux-related)
- Achalasia or other motility disorders
- Post-inflammatory strictures

However, the presence of iron deficiency anemia, mucocutaneous findings, and the identification of an esophageal web on imaging supported the diagnosis of Plummer-Vinson Syndrome. The associated joint pain and positive serology (RF and anti-CCP) pointed toward an autoimmune etiology—specifically, Rheumatoid Arthritis as a coexisting diagnosis.

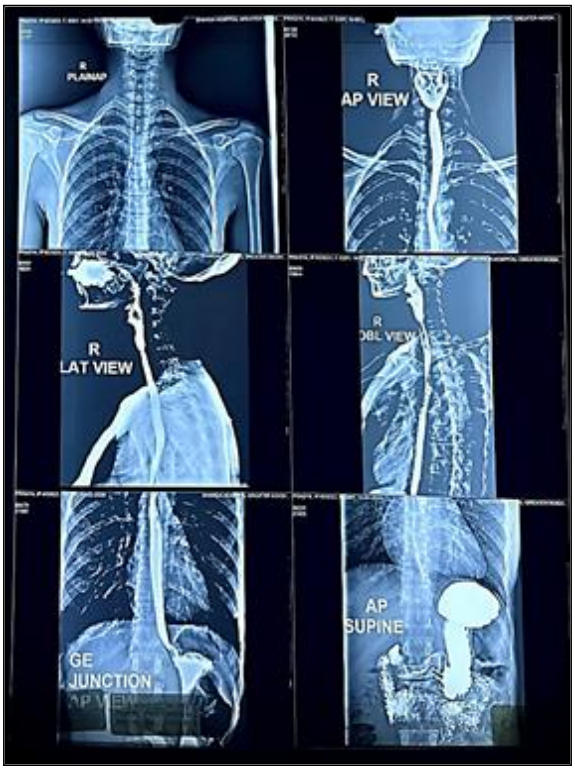

Figure 2: Barium swallow image showing pharyngeal constriction

Treatment

The patient was managed with a multidisciplinary approach:

- Iron Supplementation: Intravenous ferric carboxymaltose (Orofer FCM) was administered for rapid replenishment of iron stores.
- Deworming: A single dose of Albendazole (400 mg) and Ivermectin (12 mg) was given to eliminate any parasitic causes of chronic anemia.
- Autoimmune Management: She was initiated on hydroxychloroquine 200 mg twice daily and short-term NSAIDs (naproxen) for joint symptoms under rheumatology guidance.
- Nutritional Support: High-iron diet with oral iron later transitioned from IV, along with vitamin D (60,000 IU weekly) and calcium supplementation.
- Lifestyle Advice: Adequate hydration, oral hygiene, and physiotherapy for joint stiffness.

Outcome and Follow-Up

The patient experienced significant symptomatic relief in dysphagia within two weeks of intravenous iron therapy. Her hemoglobin improved to 9.8 g/dL by the fourth week.

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Nutritional status and energy levels improved, and she tolerated semisolid foods without difficulty.

Joint pains became less severe with hydroxychloroquine and NSAID therapy, and she was referred for long-term rheumatology follow-up. Vitamin D repletion was ongoing. She was counseled about long-term surveillance for esophageal carcinoma and advised to follow up every 3 months for anemia monitoring and autoimmune disease management.

DISCUSSION

Plummer-Vinson Syndrome (PVS) is an uncommon condition in modern clinical practice, typically presenting with the triad of iron deficiency anemia, dysphagia, and esophageal webs. The pathogenesis is thought to be multifactorial, involving mucosal atrophy due to chronic iron deficiency, which impairs epithelial regeneration and contributes to web formation.[1,2] These esophageal webs are typically located in the post-cricoid area and are best visualized via barium swallow studies or endoscopy. Though rare, PVS carries clinical significance due to its potential reversibility with iron therapy and its association with upper gastrointestinal malignancies, particularly squamous cell carcinoma of the hypopharynx and upper esophagus. Early recognition and treatment can reverse the dysphagia and prevent malignant transformation.[5,6]

Rheumatoid arthritis (RA), a systemic autoimmune disease, commonly presents with symmetrical joint pain, systemic fatigue, and chronic inflammation. Anemia in RA is typically normocytic and related to chronic inflammation; however, nutritional deficiencies and gastrointestinal blood loss (especially from NSAID use) may contribute to iron deficiency in this population. The combination of RA with PVS is extremely rare and not well-documented in medical literature, though both conditions predominantly affect women and share some overlapping systemic features.[3,7,8]

In this case, the coexistence of RA and PVS posed a diagnostic challenge, with the patient's fatigue and constitutional symptoms potentially attributable to either disorder. The diagnosis of PVS was established based on endoscopic evidence of a pharyngeal web and correction of dysphagia following iron repletion. Serological evidence of RA, along with joint involvement and elevated inflammatory markers, necessitated concurrent autoimmune management.

This case underscores the need for comprehensive assessment in patients presenting with multiple systemic complaints, especially in young females with iron deficiency anemia and signs of autoimmune pathology. It also highlights the importance of interdisciplinary management, involving gastroenterologists, rheumatologists, and nutritionists for optimal outcomes.

CONCLUSION

This case illustrates the classical presentation of Plummer-Vinson Syndrome in a young female, complicated by coexisting Rheumatoid Arthritis. The presence of progressive dysphagia and mucocutaneous signs of iron deficiency prompted endoscopic evaluation, leading to diagnosis of an esophageal web. Serological markers confirmed the concurrent diagnosis of RA, explaining the patient's joint complaints and chronic inflammation.

Timely diagnosis and management with intravenous iron therapy, disease-modifying anti-rheumatic drugs (DMARDs), and nutritional support led to a favorable outcome. Clinicians must consider overlapping etiologies in patients presenting with systemic symptoms and anemia, particularly in young females. Early identification of PVS is crucial to prevent complications, including upper GI malignancies. Multidisciplinary care is essential for addressing the complexities associated with such overlapping syndromes.

LEARNING POINTS

- Suspect PVS in middle-aged women with dysphagia and iron deficiency
- Iron therapy can reverse esophageal webs
- Rule out coexisting autoimmune conditions
- Monitor for potential upper GI malignancies
- Multidisciplinary approach improves outcomes

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