Missed Plummer-Vinson Syndrome Presenting as an Acute Airway Obstruction

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ABSTRACT
Plummer-Vinson syndrome (PVS) presents as a triad of dysphagia, iron-deficiency anemia, and esophageal webs. Nowadays, it is an extremely rare lesion in our community. Most of the patients are middle-aged women, in the fourth to seventh decade of life but the syndrome has also been described in children and also adolescents. The common presentations of PVS are dysphagia, upper esophageal webs, and iron deficiency anemia. We reported a case of a 32-year-old woman who was suffering from anemia and dysphagia for fifteen years. The anemia of PVS was masked by the gynaecological symptoms. Later she presented with worsening dysphagia, hoarseness and stridor which necessitate an emergency tracheostomy. The biopsy taken from hypopharyngeal area mass turned out to be squamous cell carcinoma.

Keywords: Dysphagia, symptom, anemia, stridor

1. INTRODUCTION
Plummer-Vinson syndrome (PVS) is one of the names given to the constellation of dysphagia, esophageal webbing, and iron-deficiency anemia. The syndrome is associated with an increased risk of hypopharyngeal carcinoma, which usually presents at an advanced stage. Dysphagia is the commonly encountered symptom in a primary care setting. A positive history of concurrent gynecological symptom causing anemia should lead to the diagnosis of PVS. Thus, a patient presenting with a prolonged history of dysphagia, hoarseness and symptomatic anemia should be referred for further evaluation of the pathology such as hypopharyngeal squamous cell carcinoma (SCC). Although it is uncommon, the condition carries the worst prognosis amongst all upper aerodigestive tract malignancy (1).
2. CASE SUMMARY

A 32-year-old Indian lady presented to Emergency Department with a history of shortness of breath associated with sudden onset of stridor. Upon presentation, she deteriorated and laryngoscopy prior to intubation showed the presence of a mass in the laryngeal inlet. She was intubated with difficulty with tube sized 6.0 mm.

On a further question from her husband, there was a history of dysphagia since 16 years prior to presentation. She had difficulty in swallowing even the oral tablets. She had presented to gynecologist six years prior to this presentation with the symptom of per vaginal bleeding and prolonged menstruation. She was treated as anemia secondary to prolonged menses and primary infertility. She was a nonsmoker and no family history of cancer.

She was planned for direct laryngoscopy and tracheostomy. A very friable mass was noted at right arytenoid region covering the laryngeal inlet. The mass extended to involve the right aryepiglottic fold, posterior cricoid region, and posterior pharyngeal wall. It appeared circumferential at the esophageal inlet, bled with contact, right cricoarytenoid joint was fixed, oedematous both false cords and subglottic region. Other structures were normal. Biopsies were taken from right arytenoid and posterior pharyngeal wall.

White blood count during admission was 18.8 x10^9/L, hemoglobin was 9.4g/dL, Peripheral blood film taken noted moderate hypochromic microcytic anemia secondary to iron deficiency anemia (IDA) and leucocytosis with neutrophilia.

Chest radiograph showed bilateral perihilar haziness, with opacities at the right lower zone. Otherwise, there was no cardiomegaly and no pleural effusion.

Computed tomography (CT scan) done noted laryngeal mass about 1.7 x 1.2cm causing significant airway narrowing with bilateral level II cervical lymphadenopathy (Figure 1). Patchy ground glass opacity in bilateral lungs likely due to the infective cause. Fullness at para-aortic and aorto-caval region with soft tissue density lesion were most likely due to lymph nodes enlargement. OGDS was deferred due to the risk of perforation, as the mass obstructing the opening of the esophagus. A barium swallow was done via feeding through Ryle`s tube, and there was no filling defect seen in mid and lower 3rd esophagus. We were unable to proceed with the upper third due to the risk of aspiration.

The biopsy of mass from right arytenoid and posterior pharyngeal wall came back as squamous cell carcinoma, moderate-to-well differentiated. A pharyngeal biopsy is consistent with SCC, at least moderately differentiated carcinoma.

Currently, the patient was well under room air and gastrostomy tube feeding, clinically improving with the ongoing fourth cycle of neo-adjuvant chemotherapy awaiting for concurrent chemoradiotherapy (CCRT).

![Figure 1: Computed tomography (CT scan) done noted laryngeal mass about 1.7 x 1.2cm causing significant airway narrowing with bilateral level II cervical lymphadenopathy.](image-url)
3. DISCUSSION

Squamous cell carcinoma of the hypopharynx is relatively uncommon and is a challenge to diagnose and treat. It accounts for approximately 95% of all primary tumors of the hypopharynx and is typically encountered in patients with a long history of smoking tobacco and alcohol consumption(2). It is more common in males, with perhaps the exception of posterior cricoid tumors which may be more common in women with PVS(3). Common presentations include dysphagia, hoarseness of voice, iron deficiency anemia and weight loss which were present in this patient. PVS complicate with hypopharyngeal lesion presenting as an acute upper airway emergency is an extremely rare situation.

There are few differential diagnosis of hypopharyngeal cancer which can be non-squamous cell malignancy, such as accessory salivary gland tumors, lymphoma or radiation change in the setting of irradiation for malignancy elsewhere in the head and neck.

Any patient who presented with symptomatic anemia and menorrhagia should be examined and investigated. Another detailed history from the patient should be included, such as her menstrual cycle, irregularity of her cycle, and amount of each menstruation. History of prolonged dysphagia in this patient should alert the medical personnel to refer for further assessment.

Although the co-existence of gynecological symptoms happen in this patient, the history of prolonged dysphagia with symptomatic anemia should be regarded as the strong diagnosis of PVS. The symptoms and history that strongly indicate the diagnosis of PVS include the prolonged history of dysphagia, hoarseness, and iron deficiency anemia(4). If the lesion is expanding, stridor can be the presenting symptoms. Sometimes a concurrent gynecological symptom may present, masking the PVS as the cause of anemia leading to delay in establishing the right diagnosis.

The incidence of post-cricoid cancer, which is a type of hypopharyngeal cancer, is related to PVS(5). It appears that the PVS is associated with a 30 percent risk of future development of post-cricoid cancer. The epidemiology of post-cricoid cancer has been considered historically to be different to that of cancer of the pyriform fossa. The former is more common in women(7).

Hypopharyngeal cancer, in general, is a rare disease representing about 0.5% of all human malignancies and constituting only 3–5% of all head and neck cancer(7), with a tumor-specific five-year survival only 13 percent(8). Owing to its rarity, post-cricoid carcinoma can be easily overlooked and confused with other similar symptoms and related presentations(9). Plus the presentation also quite late compared to the oral cavity or laryngeal carcinoma because if relatively wider space in the pharynx for the lesion to exert its mass effect. This scenario was the most likely reason for delayed diagnosis of post-cricoid carcinoma in this patient.

Post-cricoid carcinoma is uncommon, but its treatment continues to excite interest, since as many as 30% of these patients may be unsuitable for treatment(10). Criteria of incurability include bilateral neck glands; fixation of the vocal cord; the well known, but very rare, fixation to the prevertebral fascia; distant metastases which are also very uncommon; and a tumor with a vertical length of more than 5 cm. Radiotherapy if used for carefully selected patient, gives a five-year survival rate of approximately 30% in favorable cases. These include those without palpable lymph node metastases in the neck and with a small tumor less than 3 cm in vertical height. Involvement of the vocal cord and also fixation to the prevertebral fascia (cT4bN2cM0) in this patient clearly indicate that a tumor is inoperable for her.

Post-cricoid carcinoma is uncommon and extremely rare under the age of 30 years. The youngest case of post-cricoid carcinoma reported occurred at the age 22 years old man which is not associated with Plummer Vinson syndrome. In our case, it occurred in a young otherwise healthy non-smoker and non-alcoholic female.

4. CONCLUSION

Any patient who presents with a history of dysphagia and iron deficiency anemia has to be treated with suspicion. These patients need an early referral. Persistent of symptoms would require a complete examination and investigation to avoid a delayed true diagnosis.
REFERENCES
