Plummer Vinson Syndrome: A Case Report And Literature Review

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Citation

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Abstract

Plummer-Vinson also known as sideropenic dysphagia is a disease that is characterized by chronic iron deficiency anemia, dysphagia and esophageal web. It is known to affect white female mainly but cases have been reported from other ethnic group in the literature. Treatment management is very promising when carcinoma is not involved.

INTRODUCTION

Plummer-Vinson Syndrome (PVS) was first described by Patterson and Kelly in 1919.[1] The syndrome consist of dysphagia, atrophic oral mucosa, glossitis and anemia, and most of the patient affected are post meno-pausal women. Other presenting symptoms may include cracks or fissure at the corners of the mouth along with painful tongue. Koilonychia (spoon shaped finger nails) or nails that are brittle and break easily.[2,3] In approximately 10% of cases , this uncommon syndrome is associated with hypopharyngeal, esophageal or oral cavity carcinoma which emanates from the degenerative changes in the mucosae of the oral cavity, pharynx, and esophagus. [4,5,6]

The aim of this report is to highlight the unique presentation of almost all the clinical features attributable to this syndrome along with a concurrent tongue carcinoma in one patient, and to share our experience in the diagnosis and management of this case.

CASE REPORT

A 27 year old Saudi female presented to the Emergency Department of the King Fahad Hospital at Al-Baha , Saudi Arabia on the 12th of June 1995 with a six weeks history of gradual difficulty in swallowing liquids and a tongue ulcer. The patient had an associated history of mental retardation, speech impairment and easy fatiquability. Prior to the onset of the illness, the patient was reported to be ambulant.

CLINICAL EXAMINATION

Patient physically looked emaciated, severely dehydrated, pale and lethargic. She weighed 29 kg and her temperature (oral) was 39.8°C. Blood pressure was 90/60 mmHg; Pulse 120/min and respiration was 22/minute. There was bilateral angular stomatitis with epithelial crust on the lips.The skin appeared generally dry and the finger nails were spoon shaped (figs 1 &2). There was a small discrete jugulodigastric node palpable on the right side of the neck. Chest auscultation revealed a normal heart sounds and bilateral basal crepitation in the lungs. The abdomen was soft, non tender and the liver, spleen and kidneys were not palpably enlarged. Intra-orally the oral mucosa was dry with thick patches of saliva. The tongue demonstrated a crater-like ulcer on the right side extending from the anterior third to the base.(fig 3)

LABORATORY AND X-RAY

Admitting laboratory values revealed a WBC of 10.5 x 10^9/L., RBC 4.34; Hemoglobin 7.6 gm/dL., hematocrit 23.8%, MCV 54.8; MCH 17.5 and serum ferritin of 38mg/dL. Blood chemistry revealed an elevated urea at 1.7 mmol/L., and creatinine of 117 mmoL/L. Chest x-ray showed scattered bronchopneumonic changes in the mid-and lower zones.

The patient was admitted to the Oral & Maxillofacial service with a diagnosis of severe dehydration, dysphagia, and queried advance carcinoma of the tongue

HOSPITAL COURSE

On the first day of admission a rehydration measure was commenced with close monitoring of fluid imput and output. The on-call internist reviewed the patient and place her on combination of parentheral Benzyl Penicillin 2 gm qid,

ceftazidime 1 gm q6h, and ferrous sulphate supension 300mg twice daily. Twenty four hour fluid intake was 240°Cc and the output stood at 50°Cc.Patient was transferred to the internal medicine service after 48 hour and was followed by the oral surgery unit. Culture swab from the tongue was taken and sent for AFB, mycology and routine culture and sensitivity. All were negative.

Attempt to pass a NG tube to facilitate feeding and instillation of contrast medium for esophagograph failed (fig 3) On the fourth day of admission 2 units of packed cell was transfused to boost the hemoglobin. General condition seemed to improve as patient was becoming more responsive to verbal dialogue and was also able to sip some fluids. On the eighth day of admission, patient underwent esophagogastroscopy, tongue biopsy, and insertion of NGT under general anesthesia. EGD did not reveal any esophageal web, but a mild cricopharyngeal edema was noted. Subsequent histopathology report of the tongue specimen confirmed a poorly differentiated squamous cell carcinoma of the tongue.

On the eighteenth day of admission patient developed a high temperature with worsening of pneumonia. A central line was inserted to facilitate TPN (total parentheral nutrition) Patient condition went on a decline and she went into cardiopulmonary arrest on the twenty third day of admission and expired.

Figure 1

Figure 1: Showing angular cheilitis, and dry skin

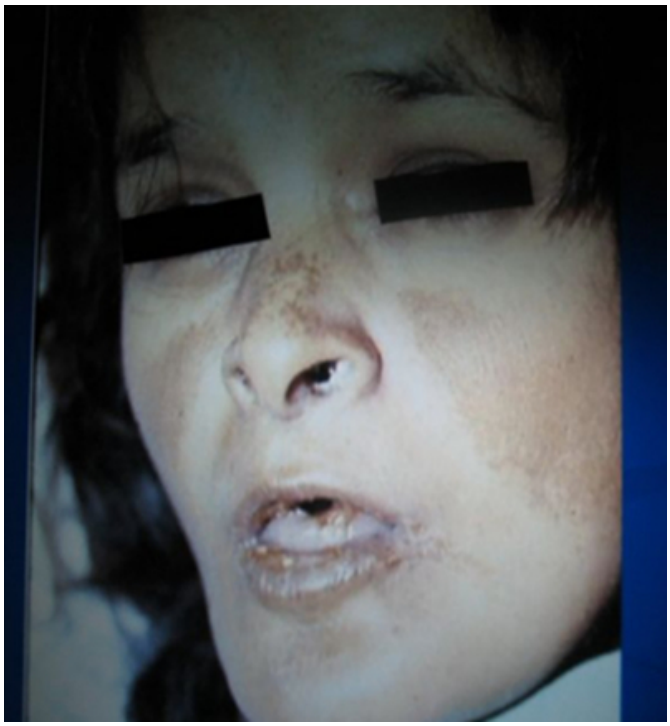


Figure 2

Figure 2: Spoon shaped finger nails

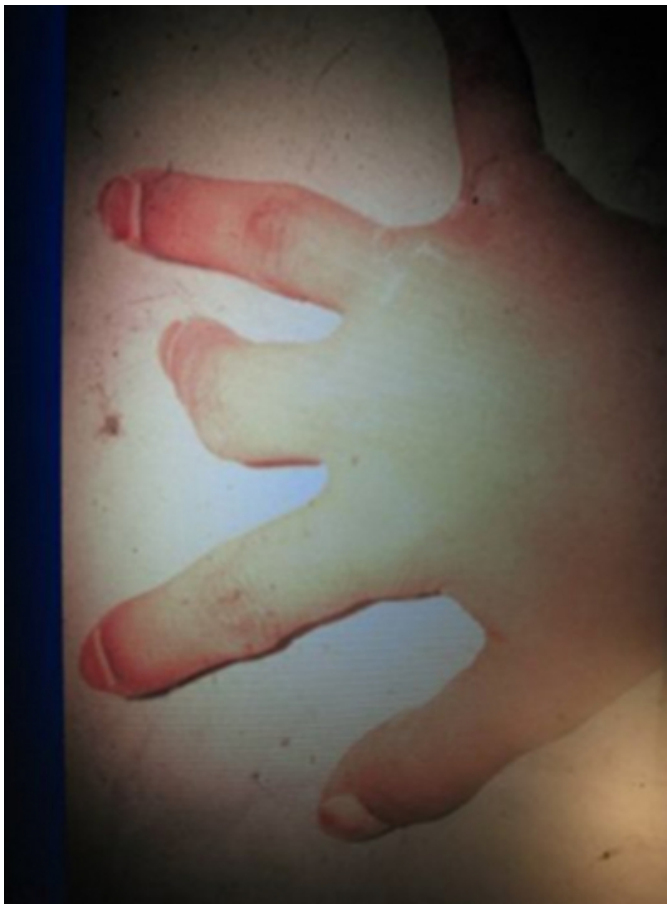


Figure 3

Figure 3: Showing a failed attempt to pass NGT

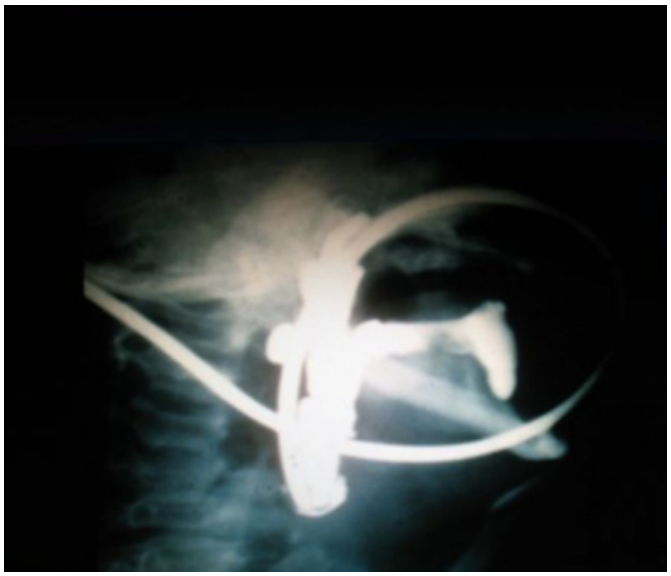
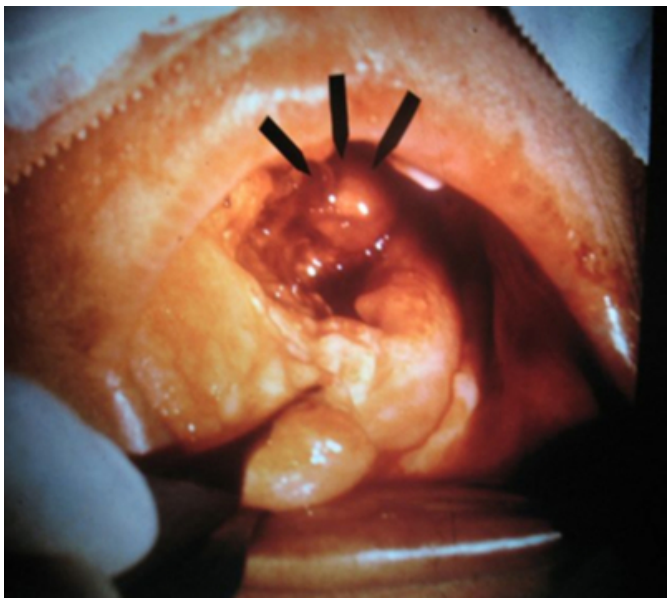


Figure 4

Figure 4: Arrows showing a crater-like lesion on the left posterior aspect of the tongue



DISCUSSION

In 1919, Patterson and Kelly independently described a clinical state (the Patterson-Kelly Syndrome) with which the names of Plummer and Vinson were later associated in the United States. The syndrome which is simply the association of dysphagia with anemia was earlier recognized by Plummer around 1914. Vinson described two additional cases later under the term “hysterical dysphagia”.[7] None of the earlier authors presented a full picture of the syndrome as it is known today.[8] PVS is a rare disease which affects mainly white women and it is characterized by dysphagia, iron deficiency and presence of superior esophageal web.[9,10] The age range is between 40-70years, however PVS has also been described in children and adolescents.[11,12]

The pathogenesis of PVS is unknown. The most important possible etiological factor is iron deficiency. This theory is primarily based on the finding that iron deficiency is a part of the classic triad of PVS together with dysphagia and esophageal webs and that dysphagia can be improved by iron supplementation.[10] Another opinion believes that it is unlikely that iron deficiency anemia is the sole explanation for the development of the webs, because webs are found in only approximately 10% of patients with iron deficiency anemia.[12]Other etiologic factors including malnutrition, genetic predisposition, or even auto-immune process have been proposed.[10]

Most commonly, patient first have dysphagia to solids, but

over time, symptoms can progress to involve dysphagia to liquids as well.[11] According to Novacek 10 the dysphagia is usually non painful, and it's progression can eventually lead to weight loss. [13,14] Logan[15] reported that patients often tolerate progressive dysphagia for considerable period of time without seeking medical attention, thus leading to late presentation. This probably explain why our patient did not seek medical attention for a long time.

PVS has been identified as a risk factor for developing squamous cell carcinoma of the upper gastrointestinal tract. Three to fifteen percent of the patients with PVS, mostly women between 15-50 years of age , have been reported to develop esophageal or pharyngeal cancer.[16,17] According to Kim et al [18] PVS is thought to be precancerous because squamous cell carcinoma of hypopharynx, oral cavity or esophagus takes place in 10% of those patient suffering from this malady. The incidence of cancer of the tongue while comprising 25-50% of all intra-oral cancers is relatively uncommon among women except in certain geographical localities chiefly Scandinavian countries, where it has been been associated with pre-existing PVS.[3,19] The prevalence among Saudi women are extremely rare despite the fact that iron deficiency is relatively high among Saudi population.[21]

The complete triad of PVS include cervical dysphagia, iron deficiency anemia and post cricoid web. The patient we are reporting presented with the first two, as no esophageal web was detected. However, according to Uygur-Bayramicli et al [20] ; some patient with all of the classical features of PVS will present with esophageal stricture instead of web. A barium swallow study is advised for the detection of a web, and other method will be an upper gastrointestinal endoscopy.[10,12]Due to the extensive malignant ulcer of the tongue demonstrated by our patient barium swallow was not successful. The treatment of PVS is hinged on the clarification of the cause of iron deficiency if a patient demonstrate one, and the rupture of and dilation of the web if it causes obstruction.10,[21,22]. The prognosis for the PVS is generally good from the standpoint that two of the triads (anemia,and dysphagia) can be effectively treated. In case of an associated squamous cell carcinoma of the hypopharynx or upper esophagus the prognosis worsens dramatically.[10]The patient that we present must have succumbed from the combination of an advance oral carcinoma and the pre-existing PVS.

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