International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Plummer - Vinson Syndrome in a 65 Year Old Male in Sokoto, North - Western Nigeria

Joshua Chukwuemeka Okoro[1], Sufiyanu Yabo Umar[2], Abdulmajid Usamatu[3]

Department of Otorhinolaryngology, Usmanu Danfodiyo University Teaching Hospital Sokoto
Corresponding Author Email: *joshalmighty5096[at]gmail.com*

Abstract: ***Plummer - Vinson syndrome is a rare entity characterized by dysphagia, oesophageal web formation and iron deficiency anaemia. It is uncommon among black Africans and very rare in males. We present a case of Plummer - Vinson syndrome in a 65year old man with a 6 - year history of dysphagia to solids, progressive weight loss and body weakness. He was found to be cachectic, severely pale with presence of angular stomatitis, glossitis and koilonychias. Contrast oesophagogram (barium swallow) revealed multiple oesophageal webs which were confirmed by flexible fibre - optic upper gastrointestinal endoscopy. Severe anaemia was corrected with multiple blood transfusions. He was subsequently treated with iron replacements and vitamin supplementations resulting in total resolution of symptoms. He remained in good health four months after treatment***

Keywords: Plummer - Vinson syndrome, Iron - deficiency anaemia, males, North Western Nigeria

1. Introduction

Plummer - Vinson syndrome, also known as Paterson - Kelly syndrome or sideropaenic dysphagia is a rare entity characterized by dysphagia, oesophageal web formation and iron deficiency anaemia. Most of the patients are middle aged women in the fourth to seventh decade of life. Very few cases of Plummer - Vinson syndrome affecting males have been reported. Dysphagia is usually intermittent or progressive over the years, and limited to solids.[1 - 3] This uncommon syndrome is most often seen in middle aged women from the northern latitudes and is associated with hypothyroidism, autoantibodies and hypopharyngeal carcinoma.[4] Iron repletion often improves the dysphagia, although some patients require oesophageal dilatation or bougienage. The syndrome is associated with an increased incidence of post - cricoid carcinoma and surveillance endoscopy is recommended.[5]

2. Case Report

A 65 - year - old man presented to the otorhinolaryngology clinic of Usmanu Danfodiyo University Teaching Hospital Sokoto on 10th August, 2020 with a six - year history of persistent dysphagia to solids, progressive weight loss, generalized body weakness and easy fatigability. He was found to be severely pale, grossly cachectic with bitemporal muscle wasting; having angular stomatitis, smooth beefy tongue (glossitis) and koilonychias. His weight at presentation was 40kg, blood pressure 110/60mmHg, pulse rate 78bpm, respiratory rate 18cpm while his axillary temperature was 36.7^{0}C.

The laboratory evaluation of the patient revealed iron deficiency anaemia with a haemoglobin level of 6.7g/dl, packed cell volume was 20%, TLC - 3.1X 10^{9}/L, and platelet count of 545 X 10^{9}/L. The peripheral blood film revealed severe hypochromic microcytosis of the red cells with reactive thrombocytosis. Both the total protein and albumin serum levels were reduced at 5.4g/dl and 3.2g/dl respectively. Renal function test showed slightly decreased serum sodium, potassium and chloride while urea level was slightly increased at 7.9mmol/L. The serum creatinine however, was normal. Fasting blood sugar was also noticed to be slightly reduced. It is noteworthy that the serum iron assay was not done for the index patient due to paucity of facilities in our hospital.

Barium swallow oesophagography showed two indentations at the laryngopharynx and proximal oesophagus (cervical oesophagus). Flexible, fibre - optic oesophagogastroscopy confirmed the presence of multiple webs in the laryngo - pharynx and cervical oesophagus. A small hiatus hernia was also noted. The distal mucosa appeared normal up to the duodenum. Moreover, multiple oesophageal and gastric biopsies were taken which excluded presence of malignancy. The patient also had an abdomino - pelvic ultrasound scan which revealed normal findings. A diagnosis of Plummer - Vinson syndrome was made.

The patient was transfused with three pints of packed cells raising his packed cell volume to 31%. He also received oral iron repletion 300mg/day, vitamin C 1000mg tds, vitamin E 500IU per day and vitamin B complex supplements. Surprisingly, it was amazingly observed that dysphagia to solids dramatically resolved as early as mere 5th day of commencement of therapy. Patient could tolerate all solid and liquid diets. At one month following initiation of therapy, follow - up haemaoglobin was 11g/dl and packed cell volume was 33%. He remains in good general condition without dysphagic complaints several months after treatment.

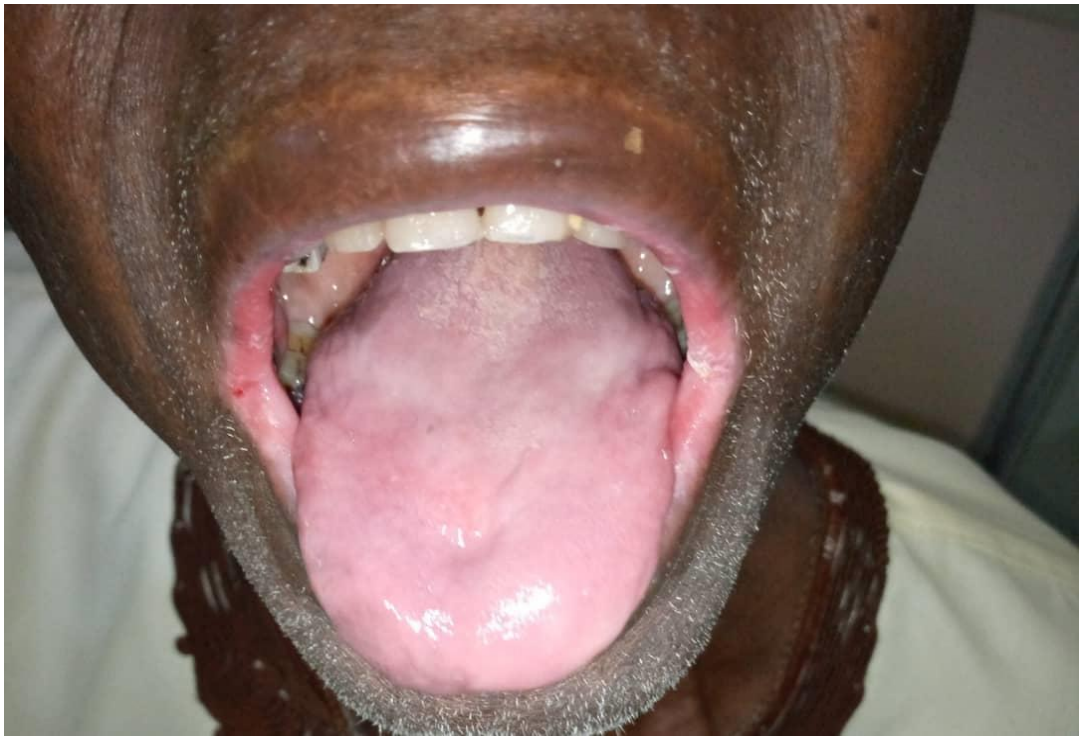

Figure 1: A picture showing angular stomalitis and smooth tongue

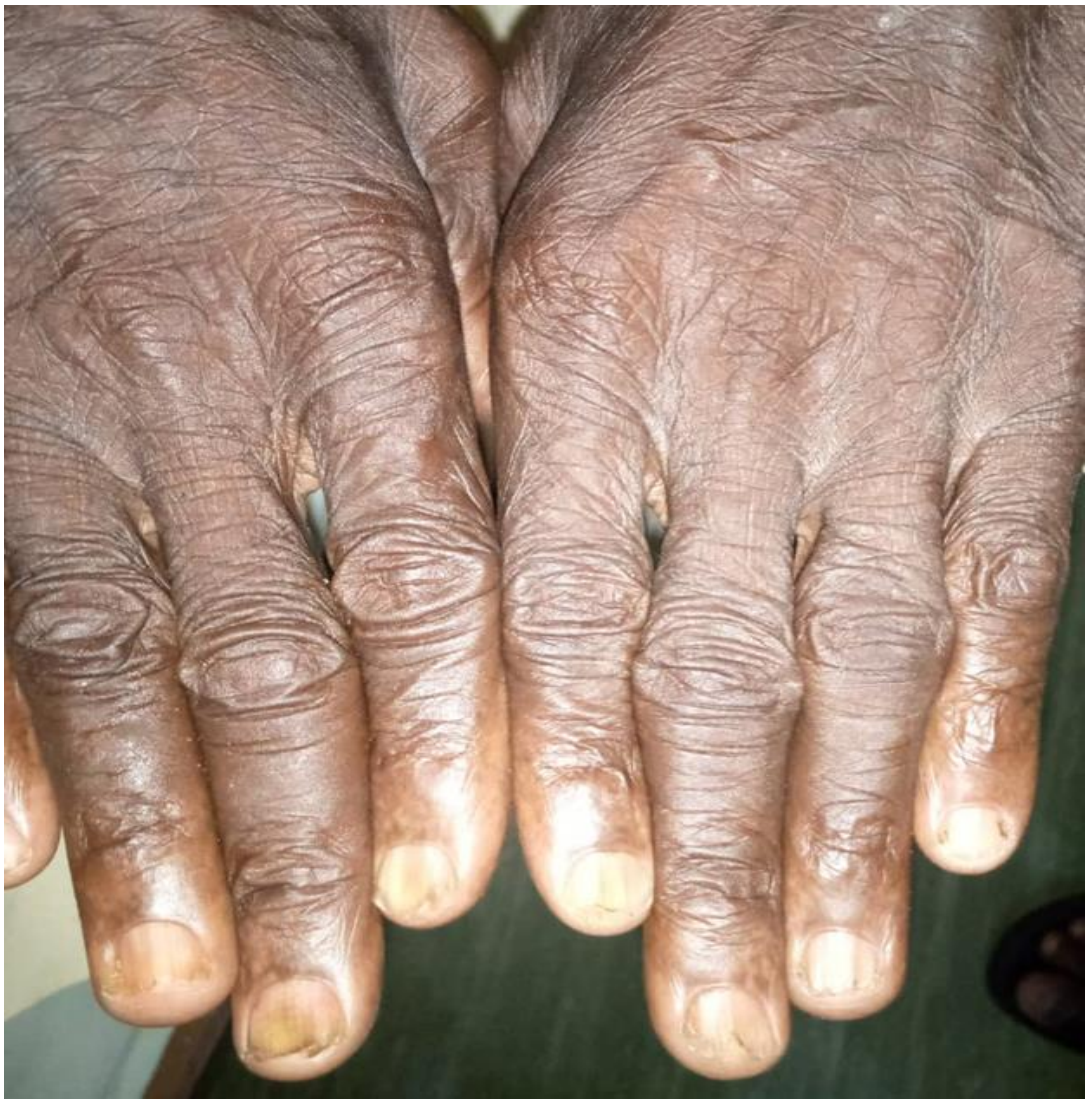

Figure 2: A picture showing koilonychias

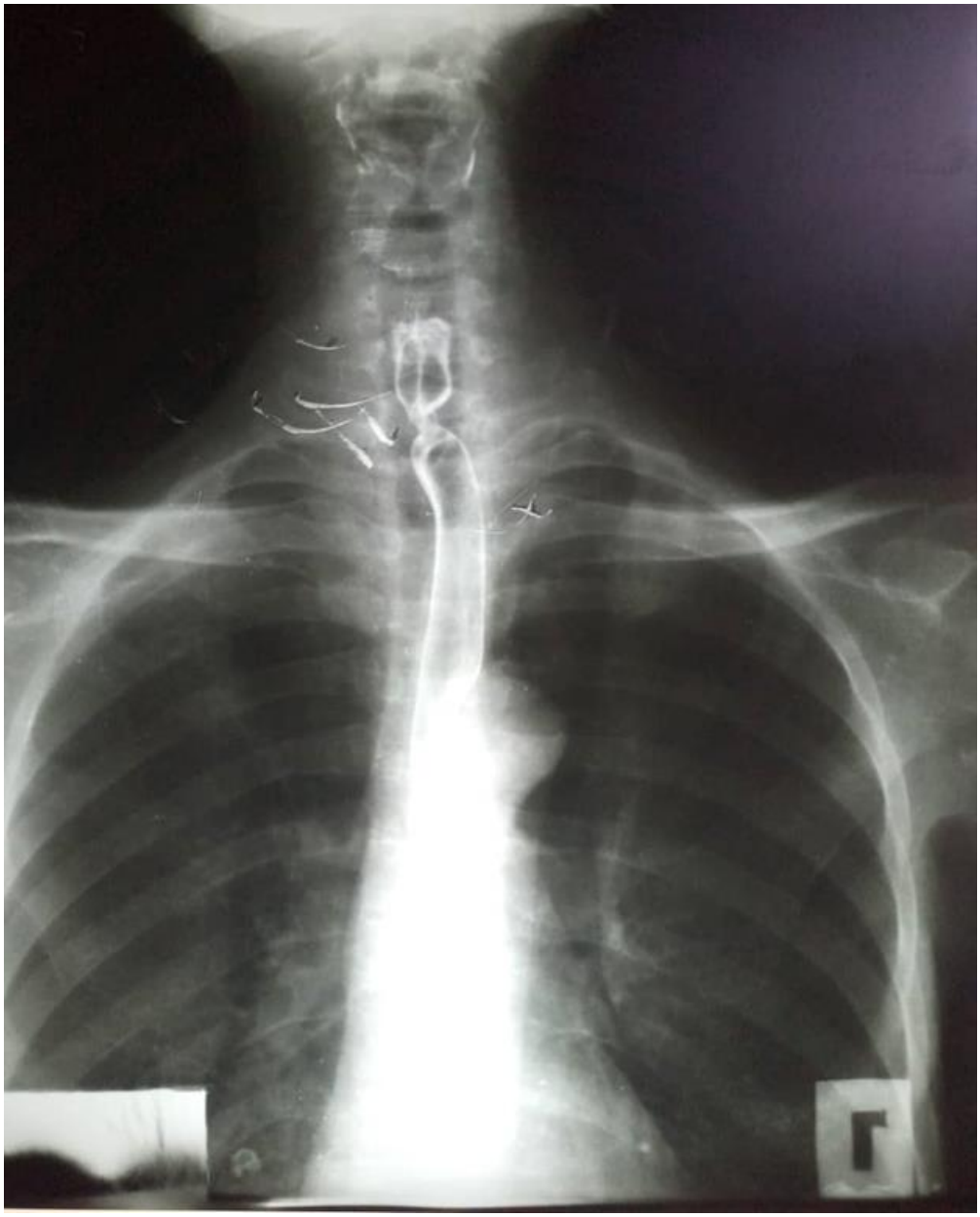

Figure 3: Barium swallow X - ray showing strictures

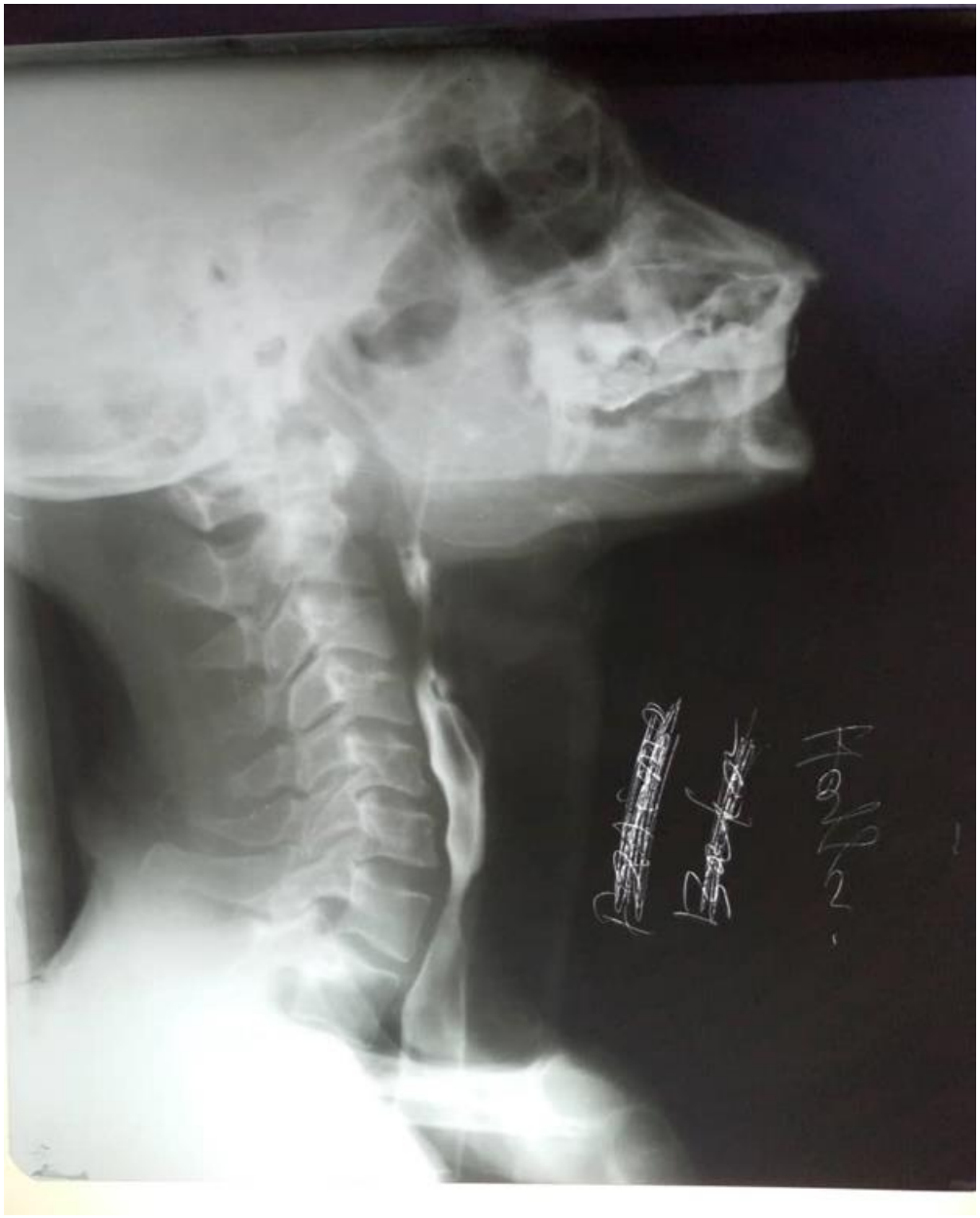

Figure 4: Barium swallow X - ray showing strictures

3. Discussion

Plummer - Vinson or Patterson - Kelly syndrome was first reported by two American Physicians: Henry Stanley Plummer (1874 - 1936) and Porter Paisley Vinson (1890 - 1959) and two British laryngologists: Donald Ross Paterson (1863 - 1939) and Adam Brown - Kelly (1865 - 1941). In 1912, Plummer reported a series of patients with long standing iron deficiency anaemia, dysphagia and spasms of the upper oesophagus without anatomic stenosis, which was described as hysterical. In 1919, Vinson reported another case of 'angulation' of the oesophagus and attributed the first description of this entity to the earlier report of Plummer. He subsequently published a series of patients with dysphagia who were successfully treated with the passage of bougies; most of the patients were women. Paterson and Kelly, who published their findings independently in 1919, were first to describe the characteristic clinical features of the syndrome. Paterson gave the fullest description but without reference to anaemia.[6, 7]

The index case is a 65 - year - old African male who had persistent dysphagia to solids, progressive weight loss, generalized body weakness and easy fatigability, pallor, angular cheilosis, glossitis and koilonychias. These findings are comparable with observations in other studies that attributed these features to iron deficiency.[8, 9] In addition to the three main symptoms, namely hypochromic anaemia, dysphagia and atrophic glossitis, a diversity of other symptoms including cheilosis, early loss of teeth, oesophagism, cardiospasm, web formation, achlorhydria, nail deformation, conjunctivitis, dermatitis seborrhica, hyperkeratosis, blepharitis and visual disturbances have been reported in patients with Plummer - Vinson syndrome.[10] Dysphagia mostly is an indirect marker of presence of oesophageal web; however, its absence does not prevent the diagnosis of Paterson Kelly syndrome.[11]

Although the racial predilection that this syndrome occurs most frequently in Caucasians from Northern countries has been advocated, there are papers reporting several cases in Japan including males. The condition is also known to be relatively common in the Indian subcontinent. However, it is very rare in Africa.[2, 10, 11] The index case is the first reported case in North - Western Nigeria and there are very scanty reported cases in other regions in Nigeria and Africa.

Our patient is a 65 - year - old male. This is at variance with most of the earlier reported cases as only very few cases of Plummer - Vinson syndrome affecting males have been reported. Most patients with Plummer - Vinson syndrome are middle aged women and it is thought to occur via blood losses from menstruations and pregnancies; and it is also very rare in children.[2, 3, 10] Novacek[6] did analysis of English language case reports of Plummer - Vinson syndrome published in the literature during a 7 - year period (1999 - 2005). He observed that 25 out of the 28 adult patients with Plummer - Vinson syndrome were women (89%). He also noted that the mean age at presentation was 47 years (range28 - 80 years). However, Bakshi[2] reported five (5) cases of males (age range 40 - 51years) over a mere 5 month period between April and August 2012 in a single health facility [in] India. Review of English literature by Hamza et al[12] in 2018 revealed a total number of nine cases of Plummer - Vinson syndrome, mainly adult females but very rare in children and adolescents.

Although Plummer - Vinson has decreased in the west due to improvement in nutrition, healthcare and decreased pregnancy, it is still relatively common in the Indian subcontinent.[2] Cases of Patterson - Kelly syndrome were more during the twentieth century than now in developed countries; improved nutritional status which has reduced iron deficiency was given to explain this. Paradoxically, Patterson - Kelly syndrome is still very rare in developing regions such as Africa, despite the preponderance of iron deficiency anaemia and malnutrition. The real mechanism accounting for Patterson Kelly syndrome is still not fully understood but some investigators have hypothesized iron deficiency, genetic predisposition, malnutrition and autoimmunity.[11] It has been proposed that prolonged sideropaenia predisposes the digestive tract to mucosal atrophy, and ulcerous changes or cracks may occur at the entrance of the oesophagus as trauma from the intake of solid food. This process induces a web which results in organic stricture, finally causing dysphagia. However, many investigators have doubted the causal relationship between the anaemia and dysphagia, the landmark study among which was performed by Elwood et al[13] who concluded that the prevalence of anaemia did not increase in patients with dysphagia, nor iron deficiency appear more often in women with webs. Wynder et al[14] found that the serum iron values were normal in 70% of female patients with Plummer - Vinson syndrome, but that their diets were deficient in fresh fruits, vegetables and meats. Whatever the source of iron deficiency, the theory is based on the rapid losses of iron - dependent enzymes due to its high cell turnover. Reduction of these enzymes may cause mucosal degeneration, atrophic changes and web formation and may even lead to cancer development in the upper gastrointestinal tract.[3, 10, 15]

Iron deficiency severely affects skeletal muscle and tissues with rapid cell turnover, such as the mucosal lining of the oesophagus. These changes are due to local myoglobin and oxidative enzyme depletion, which can develop earlier and move profoundly than circulating haemoglobin deficiency. The oesophageal webs are not the primary cause of dysphagia in this syndrome. This has been evidenced by the resolution of dysphagia before the web decreases in size following iron replacement therapy.[16]

The diagnosis is based on the evidence of iron - deficiency anemia and one or more oesophageal webs in a patient with post - cricoid dysphagia. Oesophageal webs can be detected by barium swallow X - ray but the best way for demonstration is the video - fluoroscopy. Webs are also detectable by upper gastrointestinal endoscopy. They appear smooth, thin, and gray with eccentric or central lumen. The webs typically occur in the proximal part of the oesophagus and may be missed and accidentally ruptured unless the endoscope is introduced under direct visualization. Laboratory examinations typically reveal iron deficiency anemia with decreased values of haemoglobin, haematocrit, mean corpuscular volume, serum iron and ferritin, and increased total iron binding capacity.6 Supplementary investigations like video - fluoroscopy, biopsy for histopathological examination might be helpful in some patients. Differential diagnosis of PVS has to be performed keeping in view of all the other related causes of dysphagia such as malignant tumors esophagus, benign strictures, spastic motility disorders, scleroderma, diverticula, and gastroesophageal reflux disorders all such conditions have to be evaluated.7 The index case had evidence of iron - deficiency anaemia, multiple oesophageal webs and post - cricoid dysphagia. These features were confirmed clinically, radiologically and endoscopically. Biopsy was also done which ruled out malignancy.

Correction of aetiology is the mainstay of treatment. Iron supplementation in the form of ferrous sulfate orally can be given or parenteral doses of iron in severe cases. Iron supplementation alone can resolve dysphagia in many patients. Mechanical dilation of webs and strictures are done by endoscopic dilatation in single or multiple sessions depending on the severity of the web, in which wire - guided bougies or balloons are used to disrupt and rupture the fibrous tissue of the stricture. For tight or highly constricted webs which prevent the passage of the endoscope a fluoroscopic guidance is usually required. Endoscopic balloon dilation becomes difficult when the webs are located proximally, but the same can be performed under fluoroscopic guidance. Other successful means of disrupting an esophageal web are by ND: YAG laser therapy or needle - knife electro incision which is seldom used. Patient should be kept on a high vitamin diet to maintain integrity and maturative potential of the oral epithelium since PVS is a potentially malignant disorder. Regular follow - up is mandatory. It is wise to perform an upper gastrointestinal endoscopy annually.[1 6, 17] Our patient had multiple blood transfusions with packed red cells to correct anaemia. He was also placed on iron and vitamin supplementation with complete resolution of symptoms.

4. Conclusion

Plummer - Vinson is rare in black Africans, generally uncommon in males and has been on a downward trend globally. However, a strong index of suspicion should be maintained in males with features of Plummer - Vinson syndrome in order not to miss the diagnosis. Iron replacement therapy sometimes suffices but some patients will require oesophageal dilatation. Periodic upper gastrointestinal endoscopy and biopsy is necessary in all patients during follow - up as they are at risk of squamous cell carcinoma.

Consent
As per international standard, patient's consent has been collected and preserved by the authors.

Ethical Approval
As per international standard, written ethical approval has been collected and preserved by the authors.

Competing Interests
The authors have declared that no competing interests exist.

Funding
The authors have declared no source of funding.

Acknowledgement
All teams involved in the management of this patient are fully acknowledged.

Authors' Contributions
This work was carried out in collaboration among all authors. Author JCO designed the study, wrote the protocol and the first draft of the manuscript. Authors SYU, AU managed the literature searches. All authors read and approved the final manuscript.

References

[1] Takashi H, et al. Plummer - Vinson syndrome with oropharyngeal web formation in which detailed endoscopic images were obtained. *Intern Med 2019; 58: 785 - 789*
[2] Bakshi SS. Plummer - Vinson syndrome – is it common in males? *Arg Gastroenterol 2015; 53: 250 - 2*
[3] Dinler G, Tander B, Kalaci AG, Rizalar R. Plummer - Vinson syndrome in a 15 year old boy. *The Turkish Journal of Paediatrics 2009; 51: 384 - 386.*
[4] Brendenkamp JK, Castro DJ, Mickel RA. Importance of iron repletion in management of Plummer - Vinson syndrome. *AnnotolRhinorLaryngol 1990; 99: 51 - 54*
[5] Hoffman RM, Jaffe PE. Plummer - Vinson syndrome: A case report and Literature Review. *Arch Intern Med 1995; 155: 2008 - 11*
[6] Novacek G. Plummer - Vinson syndrome. *Orphanet Journal of Rare Diseases 2006; 1: 36*
[7] Vanam C, Thammisetty DP. Case Report on Plummer - Vinson syndrome. *Int J Basic Clin Pharmacol 2019; 8: 2334 - 7*
[8] Patterson DR. A clinical type of dysphagia. J Laryngol Otol 1919; 34: 289 - 91
[9] Kelly AB. Spasm at the entrance of the oesophagus. *J Laryngol Otol 1919; 34: 285 - 89*
[10] Kitabayasi K, et al. Gastric cancer occurring in a patient with Plummer - Vinson syndrome: Report of a case. *Jpn surg 1998; 28: 1051 - 55*
[11] Ahmadu BU, Abew KA, Claphton DH, Pwavimbo AJ. Diagnostic challenge in an 11 year old boy with clinical features of Patterson - Kelly syndrome in a health resource poor setting: A case report from Yola, North - Eastern Nigeria. *International Journal of Medicine and Applied Sciences 2013; 2: 2320 - 3137.*
[12] Hamza RE, Sabu NM, Ali B, Kodithodi F, Villyoth MP. Plummer - Vinson syndrome in a 6 year old boy. *Gastroenterol Hepatol open access 2018; 9: 00283.*
[13] Elwood PC, Jacobs A, Pitman RG, Entwistle CC. Epidemiology of the Patterson - Kelly syndrome. *Lancet 1964; 2: 716 - 720*
[14] Wynder EL, Fryer JH. Etiologic considerations of Plummer - Vinson (Paterson - Kelly) syndrome. *Ann Intern Med 1958; 49: 1106 - 1128*
[15] Chen TSN, Chen PSY. Rise and fall of Plummer - Vinson syndrome. *Journal of Gastroenterology and Hepatology 1994; 9: 654 - 658*
[16] Anderson SR, Sinacori JT. Plummer - Vinson syndrome heralded by postcricoid carcinoma. *American Journal of otolaryngology - Head and Neck Medicine and surgery 2007; 8: 22 - 24.*
[17] Samad A, Mohan N, Balaji RVS, Augustine D, Patil SG. Oral manifestation of Plummer - Vinson syndrome: a classic Report with Literature Review. *Journal of International oral Health 2015; 7: 68 - 71.*