



Case Report

Dysphagia in a patient with Plumer Vinson syndrome: An innovative surgical dilatation technique after failed endoscopy

Ikram ul Haq Chaudhry^{*}, Othman M. AlFraih, Meenal A AlAbdulhai, Yousif Al Qahtani, Hisham Al Maimon, Hasan Ali, Abdulhadi A Al Haddad, Abdullah M. Alghamdi

Division of Thoracic Surgery Damman Medical Complex, Saudi Arabia



ARTICLE INFO

Keywords:

Iron deficiency. esophagus
Dysphagia
Endoscopy
Surgery

ABSTRACT

For the last five years, a 43-year-old female presented with progressive dysphagia for solids and liquids. She was treated for iron deficiency anemia by systemic and oral iron therapy. Gastroenterologists failed to pass endoscopes through the upper esophagus. Gastrogriffin swallow revealed a critical narrowing of the esophagus. She was referred to a thoracic surgeon for further management. The cervical esophagus was exposed through the neck incision along the anterior border of the left sternocleidomastoid muscle, and esophageal dilatation was achieved using the silicon foleys catheter. It is a very safe and valuable surgical technique when dysphagia cannot be managed by endoscopically.

1. Background

The Plumer Vinson syndrome, a classical trait of iron deficiency anemia, dysphagia, and esophageal webs, is named after a mayo clinic physicians Henry Stanley Plummer and Porter Paisley Vinson (1874) [1, 2]. This condition is also described in medical literature by two British laryngologists, Donald Ross Paterson and Adam Brown-Kelly (1863) [3, 4]. The incidence of this disease is higher in middle-aged females (range 40–70 years). However, it has been reported to occur in children. In the 20th century, the incidence was high in the white population of Europe, North America, and Scandinavia. With economic growth and raised standard of living and better nutrition, the disease has almost disappeared in these areas. The incidence is higher in developing countries because of poor nutritional status and hygiene, high incidence of worm infestation, lack of iron supplementation, and fortification of dietary substances with iron [5–7]. This case is reported in line with scare criteria [8].

2. Case report

The is a 43 years old female known case of iron deficiency anemia following up with Internal medicine since 2013 in our institute. Hematological investigations revealed Hb 6 gm/dl, serum iron 27 mcg/dl, Total Iron Binding Capacity (TIBC) 397 mcg/dl, mean corpuscular volume (MCV)66.5fl. She was treated with Ferrous Sulfate 190 mg and

Folic acid 5mg orally once daily. Also, she required IV Ferrous Hydroxide 200mg IV or Iron Dextran 200mg IV once weekly to manage her severe anemia due to poor response to oral medications. She had multiple blood transfusions. She developed dysphagia progressively to solids and liquids. Upper GI Endoscopy showed complete occlusion of esophageal lumen distal to cricoid (15 cm from incisors) and failed to pass even the pediatric endoscope. Gastrogriffin swallow revealed narrowing in the upper esophagus Fig. 1 (A & B). In a multidisciplinary meeting, including a Gastroenterologist, Hematologist, Internist, and Thoracic surgeon, it was decided to proceed with surgical management. We approached the esophagus through a neck incision along the anterior border of the sternocleidomastoid muscle. And esophagus was mobilized and pulled into the operating field. We found an esophageal narrowing distal to the cricoid cartilage. A pediatric silicone foley catheter was passed orally up to the stricture level. It was further guided by digital manipulation through the stricture. Gradually, the balloon was inflated with saline. A similar procedure was performed with silicone adult foley's catheter balloon inflation with 2.5 ml of normal saline. The wound was closed in layers without any drain. On the second postoperative day, Gastrogriffin swallow showed normal esophagus and free contrast flow (Fig. 1 (C & D). Operative pictures Fig. 2(A&B), and the procedure is shown in the illustrative drawings in Fig. 3 (A, B, C & D). Postoperative recovery was uneventful, and the patient was discharged for follow-up in outpatient. Periodic endoscopic examination for three years was normal, and the patient remained asymptomatic.

^{*} Corresponding author.

E-mail address: drihc007@gmail.com (I.H. Chaudhry).

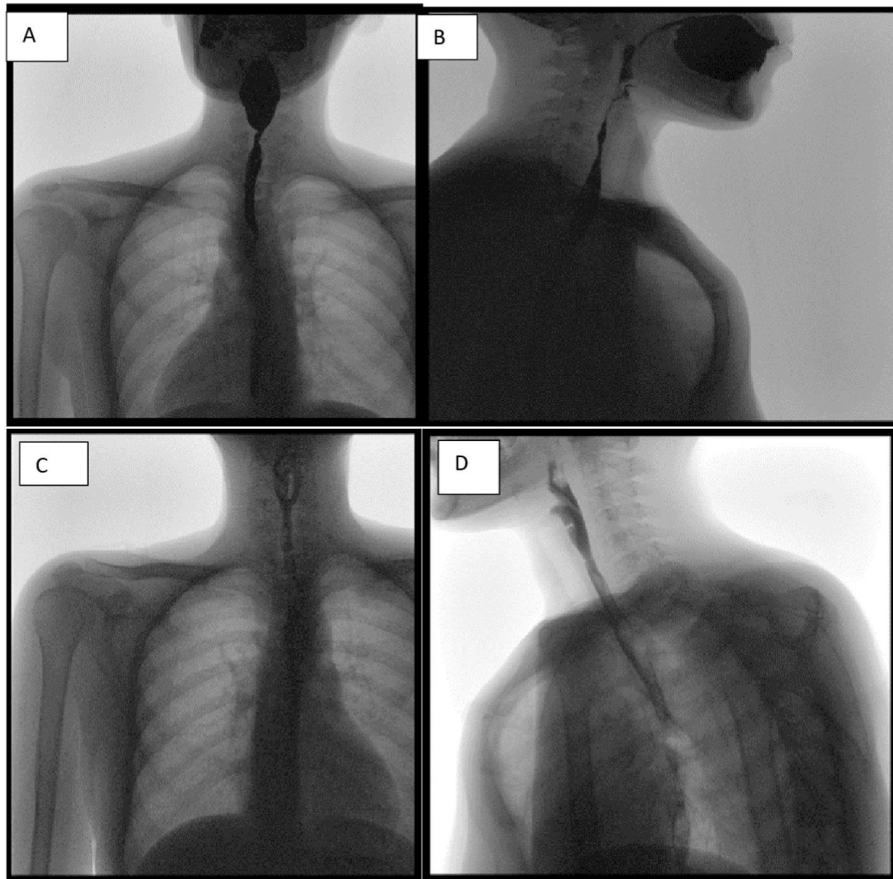


Fig. 1. (A&B) Gastrogriffin swallow preoperative showing contrast hold up and very narrow esophagus. (C&D) Post dilatation contrast swallow showing free flow of contrast.

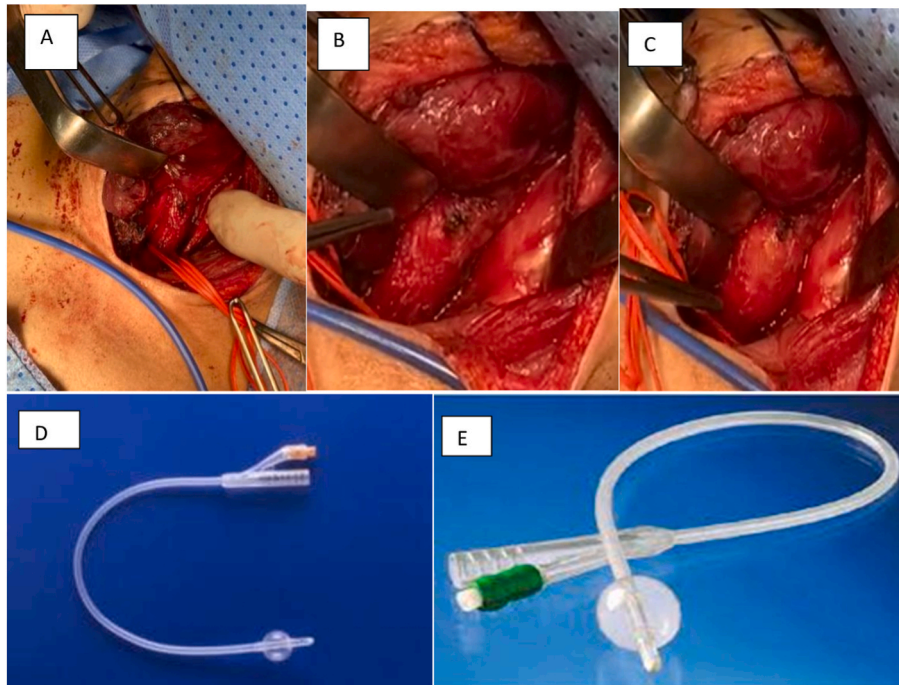


Fig. 2. (A) Esophagus exposed to demonstrate narrowing. (B) Digitally under vision esophagus dilated by passing pediatric foleys catheter. (C) Fully dilated esophagus (Using adult foleys catheter) (D) Pediatric silicon foleys catheter used. (E)Adult foleys catheter used. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

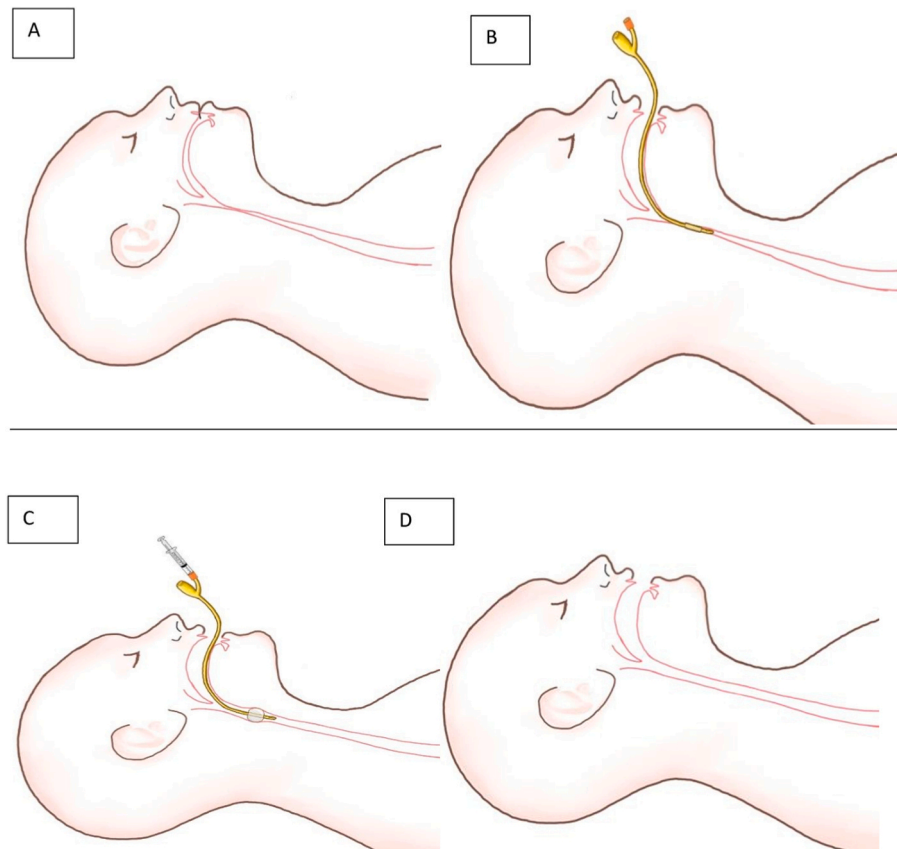


Fig. 3. Illustrative Drawings of the procedure. (A) Showing esophageal stricture. (B) Silicon Foley's catheter passed through the mouth and manually guided to stricture area. (C) Foley's catheter balloon gradually inflated to full capacity. (D) Esophageal lumen restored to normal. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

3. Discussion

Detailed data about the pathogenesis, prevalence, and Incidence of Plummer Vinson syndrome is not available in the medical literature. There are different proposed theories about the genesis of esophageal webs in this disease. There is a hypothesis that a reduction in the iron-dependent oxidative enzymes due to low iron levels, which in turn, can lead to myasthenic changes in muscles, mucosal degeneration, and eventually web formation [9,10]. Another theory is that there is an esophageal motility disorder due to iron deficiency. This is further supported by the evidence that iron therapy showed a reduction in the formations of webs; probably, the depletion of hemoglobin level mediates as a scavenger of nitric oxide, which improves esophageal tone motility [11]. Other factors implicated in the pathogenesis of this syndrome are iron deficiency due to malnutrition, worm infestations, low hygiene standards, genetic predisposition to autoimmune processes such as celiac disease, gastrointestinal bleeding, hiatus hernia, rheumatic arthritis, thyroid disease, and increased menstrual blood loss [12,13]. There data in medical literature epidemiology of this disease is limited to few case series and sporadic case reports. This is more common among middle-aged women (45–50years) female to male ratio is 4:1 [14]. Elwood et al. [15] in 1964 reported a single population-based study from south Wales. In this study, 2346 women and 1994 men were screened for post cricoid dysphagia by survey and performing barium swallow in symptomatic patients. The study revealed that the Incidence of post-cricoid webs and Dysphagia in women was 0.3%–1.15% and 8.4%–22.4%, respectively. There were no post-cricoid webs in men. This significant female preponderance may be due to the high prevalence of IDA in women [15]. Noshier et al., in 1975 reported a radiologic study of 1000 cineradiographs of the hypopharynx and cervical esophagus and

found esophageal webs in 5.5% of patients only [16]. In English medical case reports described from 1999 to 2005 revealed that 25 out of 28 patients (89%) were female, with a mean age of 47 years. All patients had an iron deficiency anemia with a mean hemoglobin level of 8.2g/dl [17]. The characteristic feature of Plummer Vinson syndrome is post cricoid dysphagia, iron deficiency anemia, and esophageal web. Most of the patients are female in the fourth to seventh decade of life, although this disease has been reported in children. The most common clinical manifestation is progressive, painless dysphagia to solids associated with fatigue and weight loss. These patients seek medical advice only when dysphagia worsens (Esophageal luminal diameter <12mm). Atkinson et al. described the severity of dysphagia in patients with Plummer Vinson syndrome in four grades. Grade I (Occasional Dysphagia for solids) Grade-II (can swallow semisolid only) Grade III (can swallow liquids only) Grade IV (cannot swallow liquids) [18–20].

Other symptoms such as fatigue, pallor, exertional dyspnea, and tachycardia are due to iron deficiency. The patient usually points out the site of obstruction at the neck or suprasternal notch. Other clinical findings in patients with iron deficiency anemia are dry mouth angular cheilitis, atrophic glossitis, dental issues, and dermatological problems such as seborrheic dermatitis, keratitis, and blepharitis. Rarely in women, genital mucosal changes due to iron deficiency can lead to itching and burning in vulval region [20–22].

Diagnosis is based on clinical, hematological, and radiological examinations. Blood investigation will show a decreased level of hemoglobin, serum ferritin MVC, and an increased level of iron-binding capacity. A blood smear study will show microcytic hypochromic anemia [23]. Radiologically barium swallow x-ray and videofluoroscopy is the best modality to demonstrate the webs. Another important diagnostic and therapeutic tool is Upper gastrointestinal fibro-optic

endoscopy [24,25]. Initial management of Plummer syndrome is to identify the cause of the iron deficiency. This can be due to menstrual abnormality in women and malignancy in men. In developing countries, it is due to poor hygiene, worm infestations, and dietary insufficiency [26]. Treat the underlying cause of iron deficiency anemia. Iron supplementation alone can improve the symptoms [27,28].

Various techniques are described for the mechanical dilatation of esophageal webs, endoscopic balloon dilatation, and Savary-Gilliard dilators. Endoscopic laser division and electro incision. Some patients may need more than one endoscopic procedure as some studies have reported a recurrence rate of 10% [29]. Bakari et al. reported in a retrospective study of 135 patients with esophageal webs that 97% were treated successfully with endoscopic dilatation, and the remaining 3% were treated spontaneously only by passing the endoscope through the webs [30]. Similarly, Goel et al. describe those 37 patients who were treated successfully with endoscopic dilatation and supplemental iron therapy [31]. A close follow-up and surveillance endoscopy are essential since the Plummer Vinson syndrome is associated with an increased risk of developing squamous cell carcinoma of the pharynx and the esophagus in 3%–15% of patients, mostly in women between 20 and 50 years of age. This is based on the hypothesis that due to iron deficiency and anemia lead to epithelial atrophy and c. impairs the mucosal repair capacity, which allows the carcinogen and cocarcinogens to act aggressively, predisposing the oral cavity hypopharynx, and esophagus to malignancy [32–35]. Non-operative management is the standard treatment for the esophageal webs or stricture in this disease. Recurrence dysphagia (10–20%) has been reported with non-operative management. Surgery has a very limited role. Three-year surveillance endoscopy revealed no abnormality, and the patient remained asymptomatic.

4. Conclusion

We report an innovative surgical technique for the dilatation of esophageal stricture in a patient with PVS. Endoscopists failed to pass even the pediatric endoscope through the web/stricture. We dilated the stricture under direct vision without esophagostomy by using the silicon foleys catheter balloon. The stricture was gradually dilated, and the patient had an excellent outcome. Postoperative follow-up, endoscopy showed good esophagus intraluminal diameter, dysphagia resolved completely, and the patient resumed her normal diet. The patient will continue iron supplementation and surveillance endoscopy in the future as such patients are at risk of developing squamous cell carcinoma of the hypopharynx and esophagus.

Sources of funding

No source of funding.

Ethical approval

IRB approval.

Author contribution

Ikram ul Haq Chaudhry Main author operating surgeon, Othman M Al Fraih Wrote Introduction Assisting surgeon, Hisham Al Maimon Highlights, Yousif A Alqahtani Structured abstract, Hasan Ali images, Abdulhadi A Alhaddad searched references, Meenal A Al Abdulhai Wrote Abstract, Abdullah M Al Ghamdi Wrote Part of discussion and Assisting surgeon.

Registration of research studies

Name of the registry: Research registry
Unique Identifying number or registration 7933

Hyperlink to your specific registration (must be publicly accessible and will be checked): <http://www.researchregistry.com/browse-the-registry#home/>.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request".

Guarantor

Ikram ul haq Chaudhry.

Provenance and peer review

Not commissioned externally peer-reviewed.

Declaration of competing interest

No conflict of interest, and there was no funding or financial assistance in this case.

References

- [1] S. Plummer, Diffuse dilatation of the esophagus without anatomic stenosis (cardiospasm) A report of ninety-one cases, *J. Am. Med. Assoc.* (58) (1912) 2013–2015.
- [2] P.P. Vinson, A case of cardiospasm with dilatation and angulation of the esophagus, *Med. Clin.* 3 (1919) 623–627.
- [3] A. Kelly, Spasm at the entrance of esophagus, *J. Laryngol. Rhinol. Otol.* 34 (1919) 285–289.
- [4] D. Paterson, Clinical type of dysphagia, *J. Laryngol. Rhinol. Otol.* 34 (1919) 289–291.
- [5] E.J. Klito, S.K. Allen, M. Metzman, P.J. Chase, Kupersmit. Plummer Vinson syndrome: report of a case and review of literature, *Am. Osteopath. Assoc.* 83 (1) (1983) 56–59.
- [6] M.D. Crawford, A. Jacob, B. Murphy, D.K. Peters, Peterson kelly syndrome in adolescence med, *J* (1965) 11189–11190.
- [7] S.S. Bakshi, Plummer Vinson syndrome, *Mayo Clin. Proc.* 91 (2016) 404.
- [8] Agha Ra, Franchi T, Sohrabi C, Mathew G, for the SCARE group. The SCARE 2020 Guidelines: updating Consensus Surgical case report (SCARE) Guidel. *Int. J. Surg.* 2020; 84:226–2304.
- [9] R.O. Dantas, Esophageal motility impairment in Plummer Vinson syndrome. Correction by iron treatment, *Dig. Dis. Sci.* 38 (1993) 968–971.
- [10] K. Atmatzidis, B. Papaziogas, T. Pavlidis, C. Mirelis, Papaziogas plummer Vinson syndrome, *Dis. Esophagus* 16 (2003) 154–157.
- [11] Y. Sugiura, M. Nakagawa, T. Hashizume, E. Nemoto, S. Kaseda, Iron supplementation improved dysphagia related to Plummer Vinson syndrome, *Keio J. Med.* 64 (3) (2015) 48–50.
- [12] J. Estadella, L. Villamarin, A. Felio, J. Pellrolo, J. Calf, Characterization of the population with severe iron deficiency anemia at risk of requiring iron supplementation, *Eur. J. Obstet. Gynecol. Reprod. Biol.* 224 (2018) 41–44.
- [13] S. Hasan, N.I. Khan, A. Siddiqui, Plummer- Vinson syndrome. A premalignant condition and over the review of the literature, *J. Med. Dental Sci.* 1 (1) (2013) 28–30.
- [14] Priyadarshini K, Nalini A, and Ramesh K Plumer. Plummer Vinson Syndrome in Males with a Review of the Literature.
- [15] P.C. Elwood, A. Jacobs, R.G. Pitman, Entwistle epidemiology of the Paterson- kelly syndrome, *Lancet* 2 (7362) (1964) 716–720.
- [16] J.L. Noshier, W.L. Campbell, W.B. Seaman, The clinical significance of cervical esophageal and hypopharyngeal webs, *Radiology* 117 (1) (1975) 45–47, 13.
- [17] Gottfried novacek. Plummer -Vinson syndrome, *Orphanet J. Rare Dis.* 1 (2006) 1–4, 36.
- [18] Kevin Bryan Lo, Jeri Albano, Naemat Sandhu, Nellowe Candelario, Plummer Vinson syndrome: improving outcome with a multidisciplinary approach, *J. Multidiscip. Healthc.* (9) (2019) 471–476.
- [19] A. Goel, C.P. Lakshmi, S.S. Bakshi, N. Soni, Koshy single-center prospective study of plummer Vinson syndrome, *Dis. Esophagus* 29 (7) (2016) 837–841.
- [20] Goel Amit, Satvinder sing Bakshi, Neetu Soni, Chhavi Nanda, Iron deficiency anemia and Plummer Vinson syndrome: current insights, *Hematol. Res. Rev.* (8) (2017) 175–178.
- [21] Y.C. Wu, P. Wang, Y.F. Chang, S. Cheng, H.M. Chen, A. Sun, Oral manifestations and blood profile in patients with iron deficiency anemia, *J. Formos. Med. Assoc.* 113 (2) (2014) 83–87, vol.
- [22] R.M. Hoffmann, P.E. Jaffee, Plummer Vinson syndrome and case Report and review of the literature, *Archit. Inter. Med* 155 (1995) 2008–2111.

- [23] A.C. Massey, Microcytic anemia Differential diagnosis and management of iron deficiency anemia, *Med. Clin. North A* 76 (3) (1992) 549–566.
- [24] O. Ekberg, Cervical esophageal webs in inpatient with dysphagia, *Clin. Radiol.* 32 (6) (1981) 633–641.
- [25] K. Vittal, S.S. Pandian, T. Malarkodi, Plumer Vinson syndrome: a case report and medical management's, *Case Rep. Rev.* 6 (2015) 10–13.
- [26] T.S. Chen, P.S. Chen, Rise and fall of the plummer Vinson syndrome, *J. Gastroenterol. Hepatol.* 9 (6) (1994) 654–658.
- [27] A. Lopez, P. Cacoub, I.C. Macdougall, L. Peyrin-Biroulet, Iron deficiency anemia, *Lancet* 387 (10021) (2016) 907–916.
- [28] J.D. Cook, Diagnosis and management of iron deficiency anemia, *Best Pract. Res. Clin. Haematol.* 18 (2) (2015) 319–332.
- [29] Krevsky B, Pusateri JP, Laser lysis of esophageal web. *Gastrointest. Endosc.* 19889; 35(5)451-453.
- [30] G. Bakari, I. Benelbarhadadi, I. Bahije, El feydi Essaaid, A endoscopic treatment of 135 cases of Plummer Vinson web: a pilot experience, *Gastrointest. Endosc.* 80 (4) (2014) 738–741.
- [31] A. Goel, C.P. Lakshmi, S.S. Bakshi, N. Soni, Koshy single-center prospective study of plummer Vinson syndrome, *Dis. Esophagus* 29 (7) (2016) 837–8411.
- [32] M. Chisholm, The association between webs, iron and post cricoid carcinoma, *Postgrad. Med.* 50 (528) (1974) 215–219.
- [33] L.G. Larson, A. Sandstrom, Westling, Relationship of Plummer Vinson disease to cancer of the upper alimentary tract in Sweden, *Cancer Res.* 35 (11 part2) (1975) 3308–3316.
- [34] G. Lanke, P. Koduru, M.S. Bhutani, Plummer Vinson syndrome presenting as squamous cell carcinoma of the esophagus, *J. Dig. Endosc.* 7 (2) (2016) 71–73.
- [35] S. Hasan, N.I. Khan, A. Siddiqui, Plummer Vinson syndrome -a premalignant condition -an over a review of literature, *Unique J. Med. Dental Sci.* 1 (1) (2013) 28–30.