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Case Report

Dysphagia in a Middle Aged Woman. Was it a Plummer Vinson Syndrome with Squamous Cell Carcinoma of Hypopharynx?

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Introduction

The incidence of hypo-pharyngeal cancers in Indian women is 1:100,000. Plummer-Vinson syndrome is a rare disease defined by the classic triad of dysphagia, iron-deficiency anemia and esophageal webs. It is considered a pre-malignant condition of squamous cell carcinoma of hypo pharynx and esophagus. The exact incidence of Plummer Vinson/Patterson Kelly syndrome is unknown and its prevalence appears to be declining worldwide, due to improvements in nutrition and standard of living over time. It was first described by Patterson and Kelly in 1919.

Case Report

A 35-year-old lady presented to the outpatient clinic with complaints of gradually progressive difficulty in swallowing of solids more than liquid. Presently she was dependent on semisolids and liquids for a duration of one year. This was associated with weight loss and generalized weakness. No complaints of hoarseness of voice/cough/difficulty in breathing/neck swelling/aspiration. No history of medical or surgical co-morbidities. Her menstrual and obstetric history is uneventful. No habit of tobacco/alcohol consumption. Patient physically looked emaciated, severely dehydrated, pale and lethargic. She weighed 45kg with a height of 1.56m (BMI-18.73kg/m^2). Physical examination revealed Pallor, atrophic glossitis (Figure1), angular stomatitis (Figure 2), koilonychia, dry skin, large volume pulse and tachycardia. Oral cavity and cervical lymph node examination was normal. Laboratory evaluation revealed anemia, with a hemoglobin level of 6.8gm/dl, hematocrit of 29.1%, mean corpuscular volume of 62.7fl, mean corpuscular hemoglobin 14.7pg and mean corpuscular hemoglobin concentration 23.4g/dl. Thyroid profile was within normal limits. Peripheral smear showed Microcytic hypochromic anemia with anisopoikilocytosis (Figure 3). No evidence of hepato-spleenomegaly on palpation. Her initial evaluation with indirect laryngoscopy revealed ulcero-proliferative growth in pyriform fossa and posterior pharyngeal wall (Figure 4). Biopsy from the ulcero-proliferative lesion showed keratin pearls with pleomorphic squamous cells showing well differentiated Squamous cell carcinoma (Figure 5). USG neck showed Hypo echoic mass lesion of 5cm length involving the pharyngoesophageal junction. CT scan of Neck and Chest showed circumferential irregular thickening with heterogeneous enhancement noted in the hypo-pharyngeal-esophageal junction and enlarged bilateral level two lymph nodes (Figure 6). Well differentiated squamous cell carcinoma of hypo pharynx, T3N2cM0 with (Group Staging IV A). She was planned to be treated with concurrent radio-chemotherapy with 70Gy/35# with weekly Cisplatin.

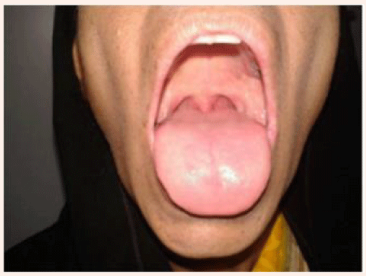

Figure 1: Physical examination revealed Pallor, Atrophic Glossitis.

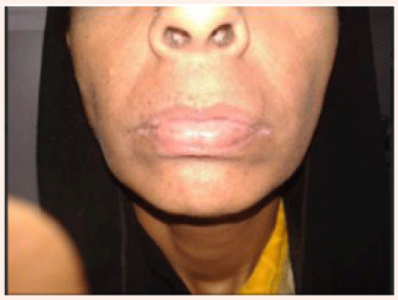

Figure 2: Angular stomatitis.

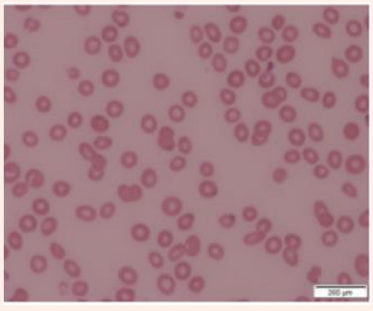

Figure 3: Peripheral smear showed Microcytic hypochromic Anaemia with Anisopoikilocytosis.

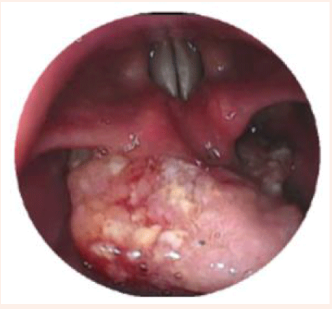

Figure 4: Her initial evaluation indirect laryngoscopy revealed ulcero-proliferative growth in pyriform fossa and posterior pharyngeal wall.

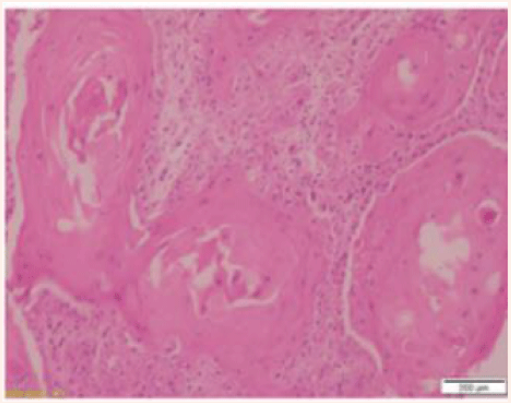

Figure 5: Biopsy from the Ulcero-Proliferative lesion showed Keratin pearls with pleomorphic squamous cells showing well differentiated Squamous cell carcinoma.

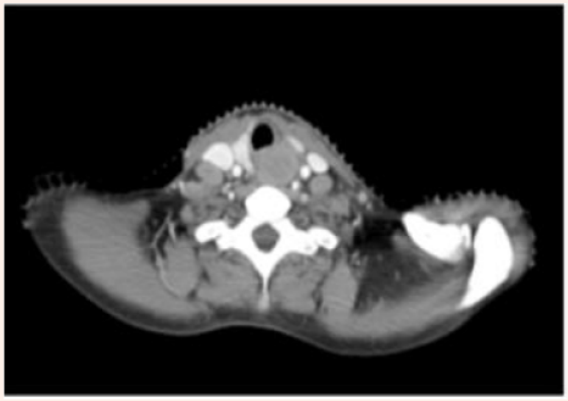

Figure 6: CT scan of Neck and Chest showed Circumferential Irregular thickening with Heterogenous enhancement noted in the Hypo-Pharyngeal-Oesophageal junction and enlarged bilateral level two Lymph nodes.

Discussion

Plummer-Vinson syndrome is a rare disease defined by the classic triad of dysphagia, iron-deficiency anemia and esophageal webs. Most of the patients are middle aged women, in the fourth to seventh decade of life. The dysphagia is painless and intermittent or progressive over years. The esophageal webs in PVS are thin mucosal folds, which are best seen either in lateral views at barium swallow or at esophagoscopy. These are usually semilunar or crescentic, being located most often along the anterior esophageal wall, but can be concentric. Proposed etiopathogenesis for Plummer Vinson syndrome includes iron, micronutrient deficiencies, genetic predisposition and autoimmunity. Iron deficiency anemia is the primary cause of dysphagia and not esophageal webs. The depletion of iron-dependent oxidative enzymes produces myasthenia changes in muscles involved in the swallowing mechanism, atrophy of the esophageal mucosa and formation of webs as epithelial complications. Hence a complete panel of Iron Deficiency Anemia which include Serum Iron, Serum Ferritin, Total iron binding capacity and Reticulocyte count. A test for Helicobacter pylori should be included. Celiac disease is commonly seen associated with this syndrome; hence investigations should include anti-endomysial IgA and anti-tTG. PVS has also been viewed as an autoimmune phenomenon. Treatment of Patterson Kelly syndrome is with iron supplementation and mechanical endoscopic dilation which usually produces complete resolution of the dysphagia. In of diagnosed cases of carcinoma hypo pharynx, radio chemotherapy and surgery are the treatment options. Multiple studies report that hemoglobin during radiation therapy is a predictor for loco regional disease control and survival [1].

In diagnosed cases of hypo pharyngeal cancer, it is of importance to diagnose the cause of anemia [2].Firstly packed red blood cell transfusion is considered. Transfusion goal is to maintain Hb 8-10g/dl as needed for prevention of symptoms. Iron sucrose injection can be used to decrease the requirement of blood transfusions. Post treatment use of oral iron supplements in absence of mal-absorption is recommended. Around 10-20% of them develop second primary malignancy. It may be due to "field cancerization" like effect caused by iron deficiency anemia [3].

Conclusion

i. Even though tobacco use is the most common modifiable risk factor for head and neck malignancies, micronutrient deficiencies are also a preventable risk factor for potentially malignant conditions like Plummer Vinson syndrome.

ii. Awareness should be created regarding the importance of nutritional deficiencies in etiology of carcinoma

iii. Pre-treatment, treatment and post treatment hemoglobin values were found to be an independent prognostic factor for loco-regional tumor control in head and neck cancers.

iv. A radiation oncologist plays an important role in the diagnosis and treatment of the anemia and cancers associated with Plummer Vinson syndrome.

References

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