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CASE REPORT

Squamous cell carcinoma in the tongue and Plummer-Vinson syndrome. A case report

Carcinoma de células escamosas en lengua en un paciente con síndrome de Plummer-Vinson. Presentación de un caso

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ABSTRACT

The Plummer-Vinson syndrome, called sideropenic dysphagia, is characterized by dysphagia, iron deficiency, anemia and the presence of esophageal webs, and it has been identified as a risk factor for developing squamous cell carcinoma of the upper gastrointestinal tract. The cases of patients suffering Plummer-Vinson syndrome that develops intraoral carcinomas are very rare. We present one case of 45 years-female with diagnosis of sideropenic dysphagia who develops a squamous cell carcinoma of the tongue. The patient present at Oral Medicine and Oral pathology clinic at Dental School of the Autonomous University of Ciudad Juárez, México by glossodynia, glossopyrosis and dysphagia of 8 months of evolution. The medical file revealed antecedents of non-specified chronic anemia, with blood transfusions at medical institutions. The intraoral examination showed depapilation of two anterior thirds of the tongue. An ulcerated swallow with indurate borders site in the left lateral border of the tongue of unknown evolution was observes. An incisional biopsy was done and a histopathological diagnosis of squamous cell carcinoma of the tongue was emitted. In our knowledge this is the third case reported in the scientific literature of a lingual squamous cell carcinoma develops in a patient suffering Plummer-Vinson syndrome. The 3 reported cases are coincident in age, gender and oral features. The pertinence of to continue including to sideropenic dysphagia like a risk factor to develop intra-oral carcinomas is discuses.

RESUMEN

El síndrome de Plummer-Vinson, también llamado disfagia sideropénica, se caracteriza por disfagia, presencia de membranas esofágicas y anemia ferropénica crónica, así como un aumento en el riesgo de desarrollar carcinomas del tracto gastrointestinal alto. Los casos reportados de pacientes con síndrome de Plummer-Vinson con carcinomas de cavidad oral son muy raros. Se presenta el caso de una paciente de 45 años con diagnóstico de síndrome de Plummer-Vinson que desarrolló carcinoma de células escamosas de lengua. La paciente se presentó en la Clínica de Patología y Medicina Bucal de la Facultad de Odontología de la Universidad Autónoma de Ciudad Juárez por presentar disfagia de 8 meses de evolución, glosodinia y glosopirosis. Su archivo médico reveló antecedente de anemia crónica no especificada, transfundida en varias ocasiones de manera intrahospitalaria. La exploración intraoral mostró depapilación de los dos tercios anteriores del dorso lingual, así como un aumento de volumen, ulcerado, de bordes indurados, de evolución desconocida en el borde lateral izquierdo de lengua. Se realiza biopsia incisional emitiéndose un diagnóstico histopatológico de carcinoma de células escamosas. En nuestro conocimiento este es el tercer caso reportado en la literatura científica de carcinoma de células escamosas lingual en una paciente que padece síndrome de Plummer-Vinson. Los tres casos son coincidentes en género, edad y depapilación del dorso de la lengua. Se discute la pertinencia de seguir incluyendo a la disfagia sideropénica como una condición que favorece la presencia de carcinomas intraorales.

Key words: Plummer-Vinson syndrome, sideropenic dysphagia, squamous cell carcinoma, tongue.
Palabras clave: Síndrome de Plummer-Vinson, disfagia sideropénica, carcinoma de células escamosas, lengua.

INTRODUCTION

Plummer-Vinson syndrome is a rare syndrome characterized by dysphagia, esophageal webs and chronic iron deficiency anemia. Oral characteristics include glossitis, glossopyrosis, glossodynia, and angular cheilitis.[1] Its etiology is unknown although autoimmune, genetic, infectious and nutritional factors have been proposed as a cause.[2] Approximately 10% of patients suffering Plummer-Vinson syndrome

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develop squamous cell carcinoma principally in the hypopharynx and esophagus.[3] Reported cases of patients suffering Plummer-Vinson syndrome who develop oral squamous cell carcinomas are scarce. Cases of squamous cell carcinoma in the tongue are even rarer.[2] In order to increase the knowledge of the association between oral squamous cell carcinoma and Plummer Vinson syndrome, a case of a patient with diagnosis of Plummer Vinson syndrome who developed squamous cell carcinoma of the tongue is reported.

CASE REPORT

In May 2004, a 45 years female came to the Oral Medicine and Oral Pathology clinic at Dental School of the Autonomous University of Ciudad Juárez (México). She presented dysphagia, glossodynia and glossopyrosis. The dysphagia had approximately eight months of evolution, having increased in the past six months. The glossodynia and glossopyrosis were constant and exacerbated with hot or spicy foods. A history of unspecified chronic anemia treated with several hematologic transfusions was mentioned in the anamnesis. A weight loss greater than 10% of the total body weight of unexplained origin was also referred. The esophageal endoscopy, performed by a private physician, identified esophageal webs. The blood cells analysis showed low hematocrit and hemoglobin values as well as anisocytosis and hypochromic microcytosis. The iron's kinetic test corroborated iron deficiency. For all the aforementioned reasons, a diagnosis of Plummer-Vinson syndrome was emitted, and consequently treatment with iron supplement was initiated.

The intraoral physical examination revealed atrophy of the dorsum of the two anterior thirds of the tongue. In the left lateral border of the tongue a size increase of unknown evolution was observed. The tumor mass extended towards the dorsum and ventral region of the tongue. Clinically, this lesion was composed by an erythematous-whitish ulcer of irregular borders *(Figure 1)*. To palpation the tumor was firm and painful. The patient disacknowledged alcoholic and smoking habits. With a presumptive diagnosis of squamous cell carcinoma, the patient was referred to the maxillofacial surgery clinic of the same institution for incision biopsy. The surgical sample was fixed at 10% buffered formalin, embedded in paraffin and cut at 5 micrometers, to be stained with the Hematoxylin and Eosin stain technique.

Microscopically the lesion was characterized by neoplastic cells of epithelial origin, with individual or group keratinizations; cellular and/or nuclear pleomorphism; hyperchromatic nuclei; increased mitosis and abnormal mitosis *(Figure 2)*. The basal membrane was lost with infiltration of neoplasic epithelial cells into muscle and connective tissue. A diagnosis of squamous cell carcinoma of the tongue was emitted. The patient was referred to an Oral Cancer Center to receive surgical and radiotherapy treatment. To date, after 5 years, the patient is in remission and under close follow up.

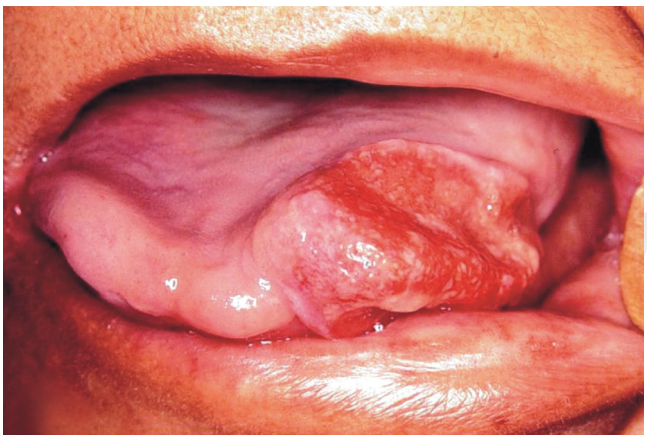

Figure 1. Clinic aspect. This pictures shows the depapilation of the two anterior thirds of the lingual dorsum and the increase of size of the left border of the tongue.

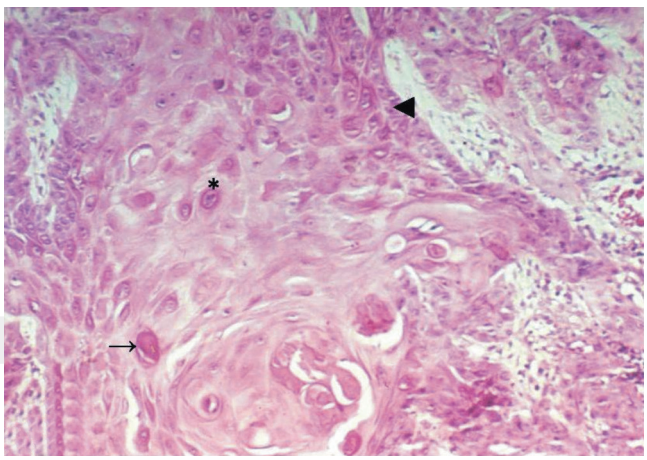

Figure 2. Histopathological characteristics. Microscopically, it can observe nuclear and cellular pleomorphism and conspicuous nucleolus (*). The arrow head (◀) point out basilar hyperplasia and arrow (→) point out individual keratinizations, all of this microscopic figures are characteristics of squamous cell carcinoma.

DISCUSSION

It has been proposed that patients suffering Plummer-Vinson syndrome show increased sensitivity to develop squamous cell carcinoma. This leads to consider Plummer-Vinson syndrome as a pre-cancerous condition to esophagus and stomach.[4] An increased incidence of post-cricoid squamous cell carcinoma is associated with Plummer-Vinson syndrome. The association between Plummer-Vinson syndrome and pseudopapilar carcinoma of the pancreas[5] has also been suggested. Although the etiology of Plummer-Vinson syndrome is unknown, iron deficiency has been suggested as the principal factor in its development. The epithelium of the upper digestive tract is susceptible to iron deficiency because its fast cellular exchange rate produces a decrease in the iron-dependent enzymes.[1] Therefore, epithelial changes observed in patients with Plummer-Vinson syndrome may be due to a depletion of the oxidative enzymes of epithelial cells. This change produces atrophic glossitis, angular cheilitis, coiloniquia, fragility and thinning of nails, brittle hair, and of particular relevance to this report, promotes the development of squamous cell carcinomas.[6] However in some patients, iron therapy is insufficient to reverse the changes and improve patients afflicted with sideropenic dysphagia. For the aforementioned reasons, it has been proposed that factors other than iron deficiency exist in the pathogenesis of the syndrome.[1]

In scientific literature, to our knowledge, only two cases of squamous cell carcinoma developed in the tongue of Plummer-Vinson syndrome patients have been reported.[7,8] In 1967, a 52-year female with iron deficiency and squamous cell carcinoma of the dorsal surface of the tongue was reported.[7] In 1970, Santoro et al[8] reported a case of a 42-year old woman afflicted with dysphagia, anemia, and asthenia who clinically showed loss of lingual papilla and presence of a firm borders lingual ulcer. Histopathologically, the diagnosis of squamous cell carcinoma was also confirmed. The clinical description of only two reported cases of lingual squamous cell carcinoma in patients with Plummer-Vinson syndrome is coincident with the case presented here, e.g. women in their fifth decade showing a bordered firm ulcer located in the posterior area of the lateral borders of the tongue.

The etiology of the squamous cell carcinoma of the tongue is associated with tobacco and alcohol consumption.[9-11] The medical file of the present case didn't show any habit (alcohol, tobacco), suggested as in association with Plummer-Vinson syndrome. However, the fact that in the last decade the presence of squamous cell carcinoma of the oral cavity,[10] especially in the tongue,[11] has been reported in young subjects who had not been exposed to risk factors raises the possibility that in the present case, both entities have developed simultaneously.

It is important to point out that the Plummer-Vinson syndrome has been definite as a risk condition for the development of oral cavity squamous cell carcinoma. Review of the literature shows few reported cases of squamous cell carcinoma of the oral cavity in patients with Plummer-Vinson syndrome. The latest one reported a patient with celiac disease and Plummer-Vinson syndrome also affected with lingual squamous cell carcinoma.[2] The only two reports of patients with carcinoma of the oral cavity also suffering of Plummer-Vinson syndrome have been published over 30 years ago. A possible reason for this fact could be that the Plummer-Vinson syndrome is associated with vulnerable social conditions, specifically poverty and malnutrition; therefore, improvements in socio-economic changes in different societies will bring as a consequence a decrease in incidence and prevalence of this syndrome. This supposition was established by the mid-70's[3] and subsequently confirmed in Swedish population.[12] However, in developing or underdeveloped countries where malnutrition could be a common event, the general dentist, and in particular the specialist in oral medicine must be alert to the presence of probably malignant lesions in women afflicted malnutrition (anemia) and lingual depapilation.

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