

## Malignization of the Lichen Planus: Clinical Case and Review of the Literature

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### Abstract

The lichen planus is a chronic inflammatory disease usually located in skin and oral mucosa. On rare occasions, it can be located in the esophageal mucosa, with an unknown prevalence and with potential malignization to squamous esophageal carcinoma, a serious pathology with very few cases reported in the literature to date. The standard management and treatment of these patients is still unknown, given the limited clinical experience and the scarcity of the diagnosis. We present a case of a patient with oral lichen planus with years of evolution who developed dysphagia and weight loss, with endoscopic and radiological finding of esophageal stenosis. The biopsies taken were of high-grade dysplasia, thus indicating surgical treatment (3 field-esophagectomy). The final histological diagnosis was squamous carcinoma of the esophagus (pT3N0).

**Keywords:** Lichen planus; Esophagus; Squamous cell carcinoma; Malignant transformation

### Introduction

The lichen planus (LP) is a chronic idiopathic inflammatory disease that usually affects the skin and mucosa. It is a rare disease (up to 2% of the population), predominantly in middle-aged women [1]. Esophageal involvement of lichen planus is rare, and even more so its malignant transformation into squamous carcinoma of the esophagus, a pathology still not fully researched due to its low prevalence, posing a real diagnostic and therapeutic challenge for experts [1,2]. The malignancy of lichen planus in squamous cell carcinoma (SCC) is well known, despite its extremely low incidence [3]. In these patients, it is typical to observe lesions in the oral mucosa of years of evolution that end up developing symptoms of dysphagia and odynophagia, so it will require a clinical diagnostic suspicion despite its low incidence [1,3]. Currently there are no consensus guidelines on the management of esophageal lichen planus (ELP), and neither for esophageal cancer derived from it [1-3]. The role of chemotherapy or radiotherapy in the treatment of SCC of ELP is still unclear [3].

### Clinical Case

Female patient of 64 years old in monitoring since 2010 by oral leukoplakia (Figure 1), with biopsies without findings of dysplasia and for which received laser treatment of CO<sub>2</sub>. Progressively, she

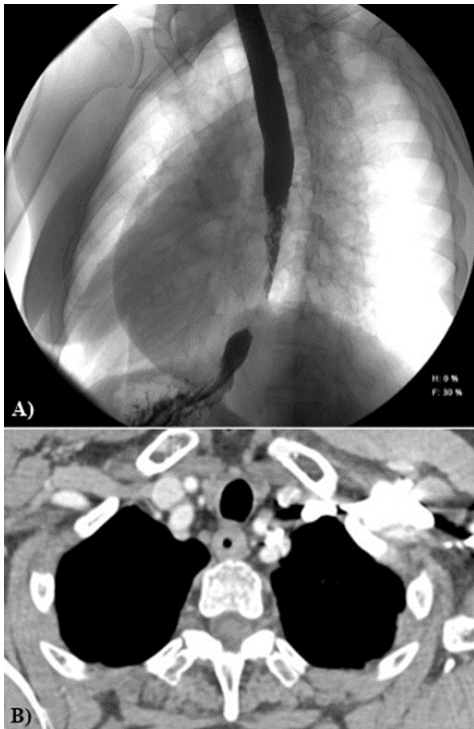
started having symptoms of dysphagia for solids and weight loss, for which a gastroscopy was performed, observing ELP of 11-12 cm in length, from which biopsies were taken, finding low-grade epithelial dysplasia. She was treated with oral corticoids and with Azathioprine without improvement, requiring two endoscopic dilatations. At 6 months, gastroscopy was repeated (Figure 2), with the beginning of lumen stenosis, and an esophagogram was performed (Figure 3A) showing esophageal involvement in the mid-distal third with marked mucosal irregularity and lack of distention. A thoracoabdominal CT scan was requested (Figure 3B) with findings of circumferential thickening of the esophageal wall and dilation of the lumen, at the level of the middle and distal esophagus. There were no lymphadenopathies or signs of the disease's extension.



**Figure 1:** Lichen planus on oral mucosa.



**Figure 2:** Gastroscopy with findings of mameloned esophageal mucosa suggestive of neoplasia.



**Figure 3:** A) Gastrografin esophagram with esophageal lumen stenosis. B) Thoracoabdominal CT scan with esophageal wall thickening.

The patient also began to develop fluid dysphagia. A new gastroscopy was performed for esophageal dilation, finding a mameloned, friable mucosa from 23 to 33 cm with even vegetative-looking areas, with a reduction in the caliber of the esophageal lumen. The biopsies taken showed high-grade dysplasia. Minimally invasive McKeown-type esophagectomy was indicated (Figure 4) with an anato-pathological result of pT3N0 squamous carcinoma of the esophagus.



**Figure 4:** Esophagectomy specimen showing mid-esophageal cancer (arrow).

## Discussion

The LP is a chronic inflammatory disease with predominant involvement of skin, mucosa and nails. It accounts for between 0.02 and 2% of dermatological diseases, affecting 2% of the population, with an especial predominance in middle-aged women (from 40 years of age) [1-4]. It has been associated with diseases such as alopecia, chronic hepatitis C and autoimmune disorders [4,5]. Autoimmune involvement is evident since this pathology responds to corticosteroid treatment in most cases [2,6].

Mucosal involvement occurs mainly in the oral cavity, pharynx, esophagus, genitalia and anus [1,4]. Carcinogenesis in oral lichen planus is associated with chronic irritation (chronic scratching of the skin lichen), tobacco use, alcohol abuse and human papillomavirus (HPV) infection. However, there is still little knowledge about the factors involved in the ELP malignization [6,7].

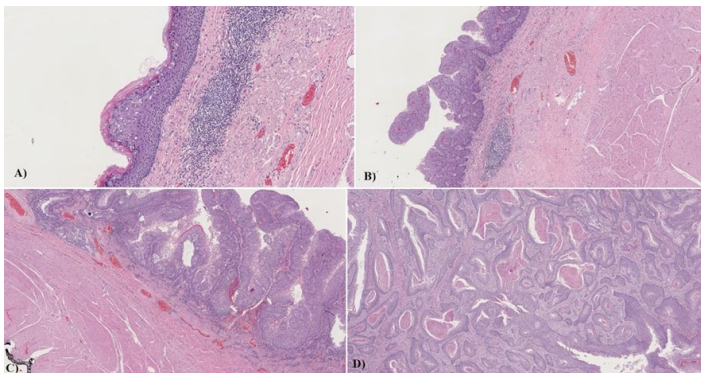
Esophageal involvement of lichen planus was first described by Al-Shihabi and Jackson in 1982 [2,4,5,7]. It is an exceedingly rare pathology, with a prevalence to this day still unknown and until 2019, with approximately 80 cases reported in the literature [2,5]. The apoptotic activity of TCD8 + lymphocytes on the keratinocytes of the basal layer is involved, generating a chronic inflammatory state with potential malignant transformation not yet well studied [8]. The usual symptoms are the appearance of dysphagia (80%), odynophagia (24%), esophageal impactions (33%) and/or weight loss (14%). It usually occurs in the upper third of the esophagus [1,2].

It has been proven that ELP is closely related to oral LP, a pathology sometimes confused with other diagnoses such as gastroesophageal reflux or oropharyngeal candidiasis [2],

therefore being treated with oral Fluconazole, something that also happened to the patient of this case. Usually, patients diagnosed with esophageal carcinoma have previous oral lesions of time of evolution [1,6]. An epidemiological study with 2,071 patients (based on the Swedish Cancer Registry) demonstrated that the single involvement of the oral cavity by LP is ultimately a risk factor for developing squamous cell carcinoma of the esophagus, with a risk between 0.5 - 2.6% [6].

The importance of the location of the ELP is its potential malignancy to squamous carcinoma, an extremely rare pathology, although underdiagnosed, requiring aggressive treatment subject to serious complications [1-6]. Currently there is no standardized diagnosis or treatment that ensures the correct management of this pathology, often incurable, and in which the role of radio or chemotherapy is yet uncertain [3]. The global prevalence of carcinoma development after diagnosis of esophageal lichen is unknown, but it should be suspected in patients with clinical symptoms and/or weight loss, with long-standing oral lesions and biopsies compatible with mucosal dysplasia.

The diagnosis of the ELP is endoscopic (whitish papules, pseudomembranes, friable mucosa) and confirmed by the presence of Civatte bodies on histopathologic study [1-6]. However, esophageal involvement may be underdiagnosed [3-6]. A retrospective analysis of the malignant transformation of oral lichen planus demonstrated a tendency for pathologists to underdiagnose it, confusing dysplasia with lichenoid infiltrates. In addition, the hyperkeratosis that covers the lesions makes it difficult to obtain biopsies with enough depth, making the diagnosis of ELP really complex [4,7]. Indigo staining has been suggested to facilitate evaluation of the esophageal mucosa during endoscopy. Lugol staining (commonly used to examine squamous epithelium) does not differentiate between dysplasia and tissue inflammation, making it difficult rather than easier to diagnose [6] (Figure 5).



**Figure 5:** A) Esophageal lichen planus; B) Esophageal lichen planus dysplasia; C) Esophageal squamous carcinoma on lichen planus; D) Esophageal infiltrating squamous carcinoma on lichen planus.

Regarding treatment, there are no established consensus guidelines for ELP [1-4]. For advanced and/or symptomatic cases, systemic corticosteroids can be used, with a response rate of 74% [1]. Other treatments such as topical intralesional corticosteroids, cyclosporine, retinoids or topical tacrolimus are showing good responses in certain patients [1-3]. If an esophageal stenosis occurs, the usual procedure used is endoscopic dilation with its repetition if stenosis recurs [1,2]. Treatment of malignant transformation to SCC of the esophagus is not standardized either and given its severity, it is often incurable. The esophagectomy is the treatment most used, being an aggressive, no free choice of further complications. The role of chemotherapy and radiotherapy in this type of neoplasm is still uncertain [3].

## Conclusion

A patient with oral LP and/or symptoms of dysphagia, odynophagia or impactions should lead us to suspect esophageal involvement, especially in middle-aged women and with abnormalities of the upper third esophageal mucosa on endoscopy. These patients should be closely followed with endoscopic controls and carefully analyzed biopsies, due to the existing risk of developing esophageal squamous carcinoma. It is a practically unknown and extremely rare pathology, on which there is very little literature at present and for which there is no standard treatment, so it must still be subject to study to ensure an adequate management of these patients.

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