Lichen Planus With Esophageal Involvement

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Abstract. Lichen planus is an inflammatory disease of unknown etiology which may affect the esophagus. Patients with esophageal lichen planus are usually elderly women. The most frequent symptoms are dysphagia and odynophagia. There is usually concomitant oral involvement in cases of esophageal lichen planus. Endoscopic findings include erosions, exudates and stenosis. Systemic steroids are the most effective therapy for these patients. We report a 68-year-old woman with oral, genital and submammary lesions of lichen planus of nine months duration, that referred dysphagia and odynophagia. Biopsy of oral and cutaneous lesions showed typical features of lichen planus. Treatment with systemic steroids significantly improved the symptoms.

Key words: esophageal diseases, esophagus, lichen planus, oral lichen planus.

LIQUEN PLANO CON AFECTACIÓN ESOFÁGICA

Resumen. El liquen plano es una enfermedad inflamatoria de etiología desconocida que puede afectar al esófago. Los pacientes con liquen plano esofágico suelen ser mujeres de mediana o avanzada edad. Los síntomas más frecuentes son la disfagia y la odinofagia. Suele haber afectación oral concomitante en los casos de liquen plano esofágico. Los hallazgos endoscópicos incluyen la presencia de erosiones, exudados y estenosis. Los corticoides sistémicos son el tratamiento más efectivo en estos pacientes. Presentamos el caso de una mujer de 68 años de edad con lesiones orales, genitales y submamarias de liquen plano de nueve meses de evolución y que aquejaba disfagia y odinofagia. El estudio endoscópico reveló la presencia de placas blanquecinas y zonas de mucosa friable. No había datos de estenosis. Los hallazgos histológicos no fueron concluyentes, pero excluyeron los cambios debidos a esofagitis fúngica o por reflujo. La biopsia de las lesiones orales y cutáneas demostró datos típicos de liquen plano. El tratamiento con corticoides sistémicos produjo una considerable mejoría de los síntomas.

Palabras clave: esofagopatías, esófago, liquen plano, liquen plano oral.

Introduction

Lichen planus is a very common condition in daily clinical practice. Although erosive oral forms of the disease are less common, they are highly debilitating due to the discomfort they cause patients. Oral lesions rarely extend to involve the esophageal mucosa, but when they do, they cause dysphagia and odynophagia.

Case Report

We describe the case of a 68-year-old woman who attended the emergency department in May 2005 with retrosternal pain that was diagnosed as angina pectoris. The patient was resident in Madrid, Spain and had previously undergone cholecystectomy for treatment of gallstones. She was prescribed acetylsalicylic acid on discharge and readmitted in June 2005 with retrosternal pain and dysphagia. Two endoscopic examinations of the digestive tract revealed superficial ulcers with fibrinous membranes and slough (Figure 1). The pathological diagnosis was esophagitis with ulceration but no specific etiology was identifiable. The condition was attributed to the use of acetylsalicylic acid and treated with gastroprotective drugs. However, the symptoms did not improve.
In July 2005, the patient moved to the north of Lugo, Spain, and attended the emergency department of our hospital with intense pain, dysphagia, and ulcerated oral lesions that she reported had been present since May 2004. Physical examination revealed several erosive lesions with well-defined borders on the sides of the tongue and on the jugal and gingival mucosae (Figure 2). The lesions were very painful to the touch. The rest of the skin examination revealed violaceous papules and brownish macules in the dorsolumbar region, the genital area, and under the breasts. The patient said that these lesions had been present since May 2004 but that she had not considered them to be serious or thought to have them checked (Figure 3).

Lichen planus with esophageal involvement was suspected, and an oral and lumbar skin biopsy were performed. Both biopsies revealed a dense lymphocytic infiltrate in the upper dermis, vacuolar degeneration, and disruption at the dermoeipidermal junction. In addition, the lumbar biopsy revealed hyperorthokeratosis, sawtooth acanthosis, and Civatte bodies (Figures 4 and 5). Direct and indirect immunofluorescence studies were negative. The results of all the other tests ordered (which included a complete blood count and laboratory workup, a protein profile, a chest radiograph, antinuclear antibodies, and serological tests for hepatitis B and C) were normal. The patient was treated with oral corticosteroids (30 mg of prednisone a day) and the lesions responded favorably. Both the oral and esophageal symptoms had improved by the time the patient was discharged to home, 1 month later.

Discussion

Lichen planus is an inflammatory disease that affects under 1% of the population. Although it can occur at any age, onset is typical in middle-aged patients, and it is particularly common in women in their 50s. The disease is of unknown etiology, although cases exist in which it has been associated with infectious processes and the use of drug treatments. Around 25% of patients report recent situational stress. The disease presents with characteristic shiny, polygonal, violaceous papules and fine, whitish striae in the lumbar region and on the wrists and lower limbs. Sixty percent of patients develop oral lesions. Although these lesions can occur in any area of the buccal and lingual mucosae, they are more common in the posterior region and are often bilateral and symmetrical. Oral lichen planus typically presents as asymptomatic, whitish, reticulated plaques, although other clinical forms such as atrophic and erosive lesions and blisters also exist. Nail and genital involvement occurs in 6% and 11% of patients, respectively. The most debilitating form of the disease is erosive lichen planus, which can vary greatly in terms of ulcer number, size, and location. It affects 6% of patients with oral lesions. Causes include dental material such as copper, mercury, cobalt, gold, and resins, nonsteroidal antiinflammatory drugs, angiotensin-converting enzyme inhibitors, diuretics, antibiotics, viral hepatitis A, B, and C, cytomegalovirus,
Epstein-Barr virus, psychological factors such as anxiety or depression, food allergies, and Koebner phenomenon due to ill-fitting dental prostheses, dental treatments, smoking/heat, friction, or bites. Oral lichen planus lesions undergo malignant transformation in 0.8% to 10% of cases. This presumed transformation of mucosal lesions, however, has been called into question as many of the cases published have been isolated and poorly documented, and the potential role played by topical and systemic immunosuppressant drugs, tobacco consumption, or repeated trauma due to dental devices cannot be ruled out.

Esophageal involvement is rare, with only 36 documented cases in the literature as of 2004. All of the cases described involve women aged between 50 and 60 years. The only exception is 1 documented case involving a man with esophageal involvement associated with cranial nerve paralysis. The incidence of the disease varies enormously from one study to the next. Esophageal involvement can be missed due to the difficulty of performing an endoscopic examination or the lack of a detailed clinical history focusing on the digestive symptoms associated with the disease. In 1 study involving 19 patients with active lichen planus, 5 patients had esophageal involvement. They presented subtle papular lesions in which the superficial layers often lifted away in the process of biopsy, highlighting a difference between these lesions and the lesions observed in reflux esophagitis. Only 1 of the 19 patients had dysphagia, and 4 of those with esophageal disease had concomitant oral disease. According to the authors of the study, symptomatic esophageal lichen planus is very rare, with symptoms appearing in fewer than 10% of patients. In another review of 584 patients with oral lichen planus, only 8 patients reported dysphagia, and of those, just 4 had endoscopic esophageal changes consistent with lichen planus.

As with classical lichen planus, the cause of esophageal lichen planus is unknown. There is, however, 1 case report of a patient undergoing alcohol detoxification treatment who developed a lichen-planus–like eruption with esophageal involvement following the administration of cyanamide. Most patients with esophageal lichen planus normally have concomitant oral disease that tends to develop first and generally presents as erosive lesions. The oral cavity is involved in almost 100% of cases, the genitals in 58%, and the skin in 29%. Esophageal lichen planus without cutaneous involvement is rare. Over 20 years can pass between disease onset and the development of dysphagia. There are also reports of intervals of up to 10 years between the onset of dysphagia and the appearance of oral lesions. It is estimated that the average diagnosis delay is 10 years.

The main symptoms are dysphagia and odynophagia, which present in two thirds of cases. Stenosis is another complication of esophageal lichen planus. It occurs in over 60% of cases and generally affects the upper third of the esophagus. Diagnostic clues to esophageal stenosis due to lichen planus include the patient being a middle-aged woman, associated mucosal lesions that are often disseminated and erosive, stenosis in the upper third of the esophagus, histologic changes consistent with lichen planus (although these are often nonspecific with a dense dermal lymphohistiocytic infiltrate and localized disruption in the basal cell layer), outbreak of buccal lesions coinciding with therapeutic dilatation, and considerable improvement following the administration of systemic corticosteroids.
Diagnosis is also possible when a patient without esophageal reflux has mucocutaneous lesions with concomitant inflammation, ulceration, and stenosis in the upper third of the esophagus.7

Macroscopic examination of the esophagus reveals erosions or lesions in the form of whitish plaques and a hemorrhagic and friable mucosa.12 Histologic findings for esophageal lichen planus can be similar to those for lichen planus13 but, in the majority of cases,6,12 they also reveal a nonspecific lymphocytic infiltrate.12,13,17 Direct immunofluorescence staining shows the presence of immunoglobulin (Ig)M, and less frequently, IgA, IgG, and complement C3, in subepidermal colloid bodies.12 Many documented cases, however, do not indicate whether or not this test was performed,7,11,14-17 while others report a negative result.10,16

The differential diagnosis of esophageal lichen planus with stenosis should include peptic esophagitis and esophageal adenocarcinoma.17 It is very important to rule out peptic esophagitis before treatment with corticosteroids as they are contraindicated for this condition.6

The majority of patients described by the literature were treated with systemic corticosteroids.7,11,12,14-17 In isolated cases, patients were treated with etretinate,16 triamcinolone acetonide injected intraesophageally,1 oral tacrolimus,1 and cyclosporine. Response to treatment was quick and effective in those treated with corticosteroids and more irregular in those treated with etretinate. Parotein pump inhibitors, H2-receptor antagonists, and sucralfate are generally ineffective.11,12 Although many studies mention treating stenosis with esophageal dilatation, this is not recommendable due to the risk of Koebner phenomenon.12 Systemic corticosteroids are again the best treatment option, and dilatation should only be used in extreme, life-threatening cases.12

Although there are no reports of malignant transformation in patients with esophageal lichen planus, regular follow-up should nevertheless be undertaken in these patients given that there is a theoretical risk.7,16 Some authors recommend performing repeated endoscopic examinations to rule out malignant degeneration.16

Finally, we believe that it is important for all women with oral lichen planus lesions to be questioned about esophageal symptoms, and when symptoms are present, to perform a digestive endoscopy.

Conflicts of Interest
The authors declare no conflicts of interest.

References