

Esophageal Lichen Planus: a Narrative Review of the Literature

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Abstract

Background: Lichen planus is a mucocutaneous chronic inflammatory disorder that can also affect the esophagus in a minority of patients. Esophageal lichen planus (ELP) is likely an underdiagnosed and underreported disease entity, although it can lead to significant morbidity with esophageal stenosis or malignant transformation.

Methods: A non-systematic search of Pubmed was performed using search terms ‘esophageal lichen planus’ and variants thereof combined with terms related to prevalence, diagnosis, and management.

Results: The cutaneous and oral forms of lichen planus are relatively common, but certain mucosal forms such as genital lichen planus are rare and esophageal planus (also a mucosal form) is very rare. ELP occurs predominantly in middle-aged women. Current guidelines regarding lichen planus do not mention a screening strategy for ELP. Diagnosis of ELP is based on endoscopic and histopathological criteria. The foremost complication of ELP is formation of esophageal strictures. Malignant transformation of ELP to squamous cell carcinoma, although rare, has also been reported. We summarize the current evidence regarding optimal management of ELP.

Conclusion: Even if esophageal lichen planus is a rare cause of esophageal strictures, it is important to raise awareness of this condition among gastroenterologists and endoscopists. The cornerstone of ELP treatment is an anti-inflammatory agent in conjunction with endoscopic dilatation of strictures. Further research is needed to devise an evidence-based treatment algorithm that describes the optimal anti-inflammatory drug in different stages/severities of ELP. (*Acta gastroenterol belg.*, 2025, 88, 129-140).

Keywords: lichen planus, esophageal lichen planus; esophageal stricture; esophageal dilatation.

Introduction

Lichen planus is a chronic mucocutaneous inflammatory disorder that can affect the skin, hair, nails, and the oral, genital, and conjunctival mucosal tissue (1, 2).

The cutaneous form typically presents as confluent pruritic polygonal and flattened papules. Commonly involved cutaneous surfaces are the inner forearms, wrists and ankles (3). A specific clinical finding of lichen planus is the presence of thin white stripes superimposed on affected papules, termed “Wickham’s striae”.

Oral lichen planus is a heterogeneous disease. The severity of oral lesions, as well as the level of discomfort they cause, differ along a few established subtypes. Oral manifestations of lichen planus range from white lacy lines that are commonly observed on routine dental examination and are usually asymptomatic (categorized as reticular oral lichen

planus) to more severe forms, such as the plaque-like; erythematous; or erosive lesions, which may cause significant pain, burning or swelling (4).

Esophageal lichen planus is considered one of the rarest manifestations of lichen planus. There is often a long delay between onset of esophageal symptoms - mainly dysphagia and odynophagia - and formal diagnosis (5). At the time of diagnosis, many patients will have undergone endoscopic dilatations for esophageal strictures already.

The aim of this narrative literature review, is to provide a clear and concise summary of the current academic understanding of esophageal lichen planus. The main purpose of this text is to inform clinicians, pathologists and endoscopists about this rare but underdiagnosed disease that, in severe untreated cases, can lead to complications such as esophageal stenosis and development of esophageal squamous cell carcinoma (SCC). Besides raising awareness of this condition among gastroenterologists and endoscopists, we also strove to provide a guide to its diagnosis and treatment.

Materials and methods

We searched Pubmed for reports in English containing the MeSH terms “esophageal lichen planus” and “esophagus AND lichen planus”, as well as “lichen planus” in combination with all of the following MeSH terms: “prevalence”, “epidemiology”, “pathophysiology”, “diagnosis”, “cancer”, “management”, “treatment”; from inception until July 2024.

We also checked the reference list of all retrieved publications to identify any additional relevant articles.

Results

Epidemiology

The prevalence of lichen planus is unknown (6). Because its many manifestations fall within the scope

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of multiple branches of medicine (dermatology; gynaecology; maxillofacial surgery and dentistry; otorhinolaryngology; gastroenterology) that do not often intersect, there is a lack of overarching studies using data extracted from patient cohorts across all these medical specialties. Due to the fragmented nature of lichen planus research, prevalence studies tend to focus on the subtypes of lichen planus that align with their corresponding disciplines. For example, the respective prevalences of oral lichen planus, cutaneous lichen planus, and genital lichen planus have been the focus of research much more often than the prevalence of lichen planus in general. Research using population-based data gathered in Sweden in 1970 suggests that the prevalence of cutaneous lichen planus is 0.3% in men and 0.1% in women (7).

The prevalence of oral lichen planus has been more thoroughly investigated. Two systematic reviews and meta-analyses studying the global prevalence of oral lichen planus have found similar results and estimate this prevalence around 1% (8, 9). In the context of a European population, the most reliable prevalence estimate resulted from a series of studies conducted on a large Swedish population. A 2008 critical review of these population studies estimate the prevalence of oral lichen planus at 1.27% (0.96% in men; 1.57% in women) (10).

In a study of 274 patients with known lichen planus, the cutaneous and oral forms of lichen planus were shown to be quite prevalent, with a smaller percentage of patients presenting with genital lichen planus (11). Significant overlap between these 3 forms was observed (Figure 1). The finding that, in this case series, despite

the low number of patients with genital lichen planus, a disproportionately high number of patients were observed to have both oral and genital lesions, seems to suggest the existence of a disease subtype with a predilection for mucosa rather than keratinized tissue (skin, scalp, nails).

Esophageal involvement of lichen planus is not as common as oral involvement, as the prevalence of esophageal lichen planus (ELP) has been estimated to be 0.19%(3). However, diagnostic issues may lead to underdiagnosis and underreporting since only a minority of patients with lichen planus will undergo an endoscopy. Quispel et al. studied 24 patients with known cutaneous and/or oral lichen planus who reported no more than mild upper GI tract symptoms (12). In this group, 50% (n=12) of patients were found to fulfill the endoscopic and histopathologic criteria for ELP, indicating that esophageal involvement of lichen planus can occur in patients with no or limited symptoms, underscoring its potential underdiagnosis. When symptomatic, the most common complaints are dysphagia and odynophagia (13).

This esophageal manifestation of lichen planus seems to present predominantly in middle-aged female patients (3). Furthermore, isolated ELP, i.e. without any other manifestation of lichen planus, is rare. In a study of 72 cases, 71 out of 72 patients (99%) were found to also exhibit extraesophageal disease activity (13).

In many cases, a specific diagnosis of ELP is preceded by years of symptomatic disease and repeated dilatation therapy for a presumed GERD-induced esophageal stenosis/stricture (5).

ELP can be the presenting manifestation of lichen planus. In the previously mentioned case series of 72

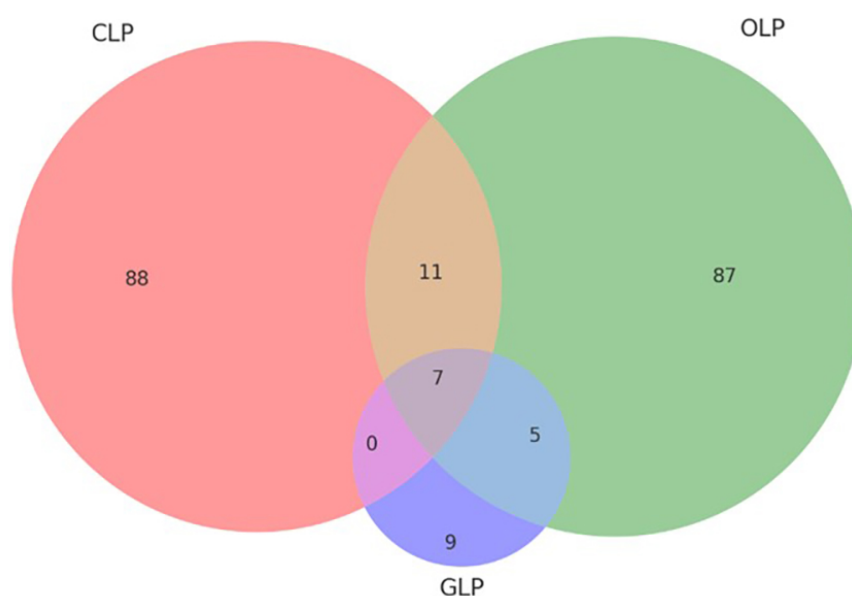


Figure 1. — A visual representation of the significant overlap between different types of lichen planus, as described by Cassol-Spanemberg et al. Abbr.: CLP: cutaneous lichen planus. GLP: genital lichen planus. OLP: oral lichen planus.

ELP patients, ELP was the first manifestation of lichen planus in 13 patients (and the sole manifestation of lichen planus in 1 patient) (13). With this information in mind, endoscopists should strongly consider a diagnosis of ELP when faced with proximal esophageal strictures, especially if these occur in middle-aged female patients.

Pathophysiology

Endoscopic findings such as white discoloration and sloughed-off appearance of esophageal mucosa with pseudo-membrane formation indicate damage at a superficial mucosal level. Commonly used diagnostic criteria for ELP, such as epithelial detachment, lymphocytic infiltrate, dyskeratosis and intraepithelial apoptosis also give insight into the type of cellular damage underlying ELP (1).

In oral lichen planus, studies have found involvement of myeloid and plasmacytoid dendritic cells, CD4+ and CD8+ T-cells, natural killer (NK) cells, and mast cells, whereas B-lymphocytes, plasma cells or antibodies were shown not to be present in oral lichen planus (14). This pattern indicates a type 1 immune response, which is also compatible with clinical evidence that corticosteroids and calcineurin inhibitors can improve inflammation in lichen planus.

The inflammation in lichen planus targets the basal epithelium, thereby explaining the predilection of lichen planus for squamous cutaneous and mucosal tissue (penile, vulvar, oral, ...). Lichen planus is not known to occur in the columnar mucosae distal to the gastro-esophageal junction.

Diagnosis

Screening

The 2019 European Dermatology Forum guidelines on lichen planus do not include guidance for endoscopic screening in patients with lichen planus to exclude ELP (15). It is mentioned, however, that “Some authors suggest that screening endoscopy to rule out esophageal involvement may be warranted in all patients with mucocutaneous lichen planus who complain of dysphagia or odynophagia.” However, Quispel et al. found that ELP is often present even in patients with no or only mild suggestive symptoms, indicating that this screening strategy might not be sufficiently sensitive (12). However, when using the results of this study to formulate a screening strategy, two study weaknesses are noteworthy: (i) the small sample size of 24 patients should be taken into account, and (ii) patients included were described as being diagnosed with cutaneous and/or oral lichen planus, without distinguishing these two forms later on. Indeed, it is of relevance to

know if ELP was observed more frequently in patients with oral lichen planus than in those with cutaneous lichen planus. Observations from case studies (cf. Epidemiology paragraph) and from clinical experience seem to suggest that patients with mucosal lichen planus are more likely to form other mucosal manifestations (such as ELP) than patients with cutaneous lichen planus. It is important to keep this distinction in mind when trying to formulate guidelines about screening for ELP in patients with lichen planus.

In the absence of consensus recommendations regarding screening for ELP, we propose to take 3 factors into account when considering a screening endoscopy for ELP: (i) the presence of symptoms that could point to undiagnosed ELP, in casu dysphagia, or odynophagia (not explained by oral lichen planus lesions), (ii) the predominance of mucosal rather than cutaneous disease manifestation, and (iii) the severity of mucosal lichen planus (if present): for example, we would advise a higher threshold for screening in patients with asymptomatic reticular oral lichen planus vs. in patients with plaque-like or erosive oral lichen planus).

There are no guidelines on the biopsy protocol, but it is our practice to take multiple biopsy samples from at least two regions of the esophagus, similar to the recommended method for diagnosing eosinophilic esophagitis (EoE), a condition that can present with similar endoscopic findings (16).

Endoscopic findings

As mentioned above, there are currently no consensus guidelines regarding the diagnostic criteria for ELP. Kern et al. proposed a set of endoscopic and histological diagnostic criteria, which were further refined in 2019 by Schauer et al. (17) (1).

One possible manifestation of ELP is a white and rough mucosal surface caused by hyperkeratosis (Figure 2A) (5). Additionally, the mucosa can display varying degrees of denudation, which results from the underlying cellular damage at the subepithelial/junctional level, evidenced histologically as the so-called lichenoid infiltrate (Figure 3A). As a result of this process, superficial layers of mucosa may separate from underlying tissue. Mucosal denudation can range from iatrogenic mucosal sloughing, i.e. only after endoscope passage or biopsy sampling, to extensive areas of mucosa peeling off (Figure 2B) (1). In extreme cases, this can form impressive tubular casts, warranting the description “esophagitis dissecans superficialis”, as discussed below under Differential Diagnosis” (Figure 2C) (18). Trachealization, an endoscopic finding often associated with eosinophilic esophagitis, can also be observed. Lastly, chronic inflammation due to ELP may lead to the formation of focal or multifocal strictures of the esophagus (Figure 2D).

Endoscopists should be aware that ELP has a predilection for the proximal esophagus for two reasons

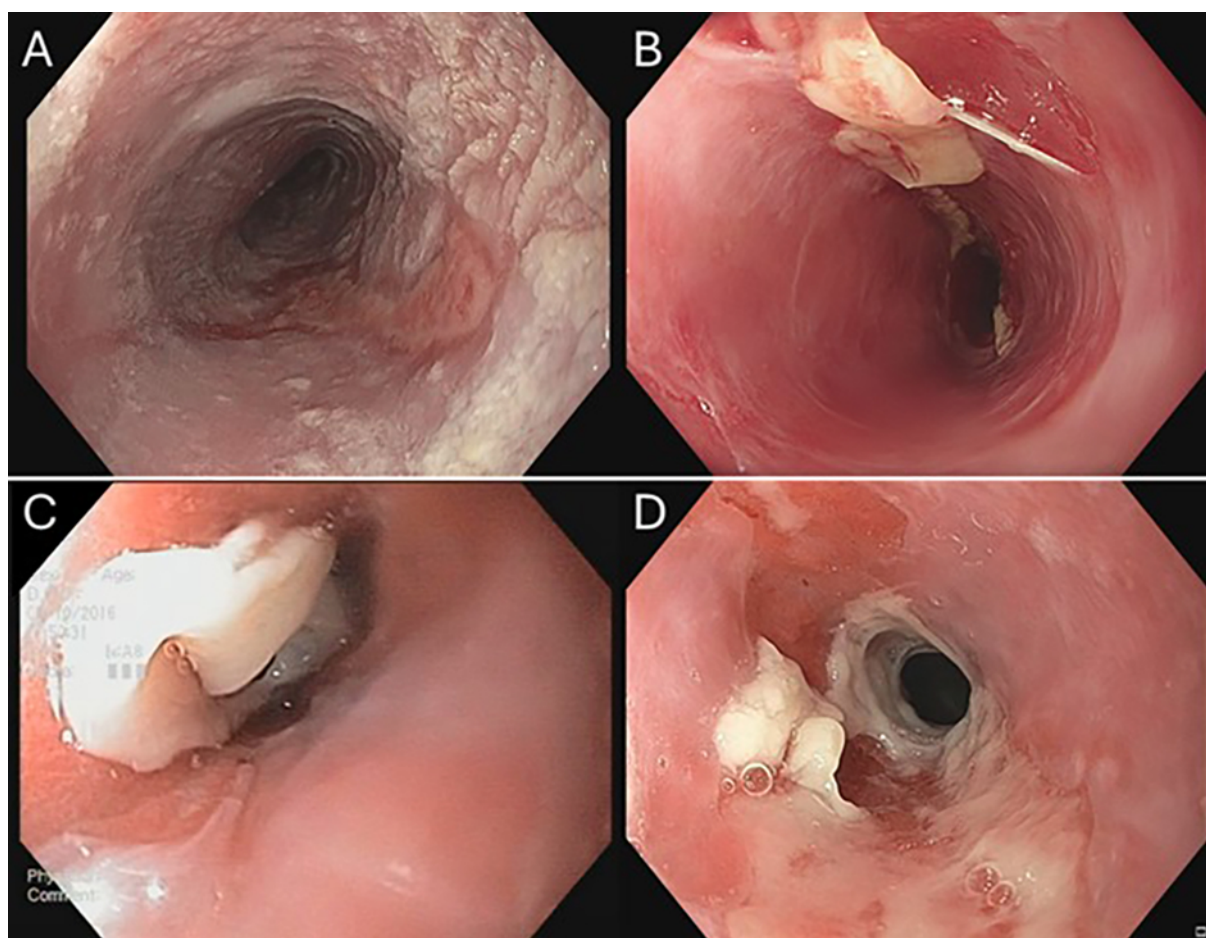


Figure 2. — Endoscopic findings of esophageal lichen planus. A: rough mucosal surface: hyperkeratosis. B: denudation of mucosal patch. C: intraluminal cast consisting of sloughed-off mucosa. D: stricture caused by chronic inflammation.

(13). This area can be (i) easy to overlook, as clinical experience tends to steer our attention towards the distal esophagus where we expect evidence of reflux disease, and (ii) more difficult to evaluate, as patient's tolerance to the position of the endoscopy near the upper esophageal sphincter is low.

In an attempt to quantify the severity of ELP using only endoscopic metrics, Kukreja et. al have devised the so-called ELP severity score (ELPSS; Table 1) (19).

Histopathological findings

Lichen planus, whether cutaneous or mucosal, is histologically characterized by the so-called lichenoid infiltrate: a dense band-like T-cell lymphocytic infiltrate at the subepithelial/subepidermal or junctional level caused by basal cell damage (Figure 3A) (19). Damage at this level may lead to epithelial detachment. In this context, it is crucial to emphasize the importance of proper biopsy sampling: at least some samples must include areas of seemingly “normal” mucosa where no macroscopic mucosal detachment is visible, as these biopsies are more likely to survive processing/staining and show the characteristic lichenoid infiltrate.

A specific histological finding in ELP is the presence of so-called Civatte bodies (Figure 3B) (20). These are eosinophilic deposits caused by intraepithelial squamous cell apoptosis. Additionally, any type of dyskeratosis (hyperkeratosis, parakeratosis) is suggestive of ELP.

On direct immunofluorescence, a clear fibrinogen deposition along the basement membrane is indicative of ELP. IgM deposition in the degenerating epithelial cells has also been described as a finding specific to ELP(3). In case of doubt, direct immunofluorescence on non-fixed tissue is recommended to confirm or reject the diagnosis.

Differential diagnosis

● Gastroesophageal Reflux Disease (GERD)

Elements of a patient's history that should make the endoscopist consider alternative diagnoses to GERD are (i) the absence of typical reflux symptoms, i.e., heartburn and regurgitation, and (ii) lack of response to previous anti-reflux therapy, including proton-pump inhibitors (PPI) (21).

When differentiating between GERD and ELP, the location of affected mucosa should be considered as well. Generally, endoscopic abnormalities limited to

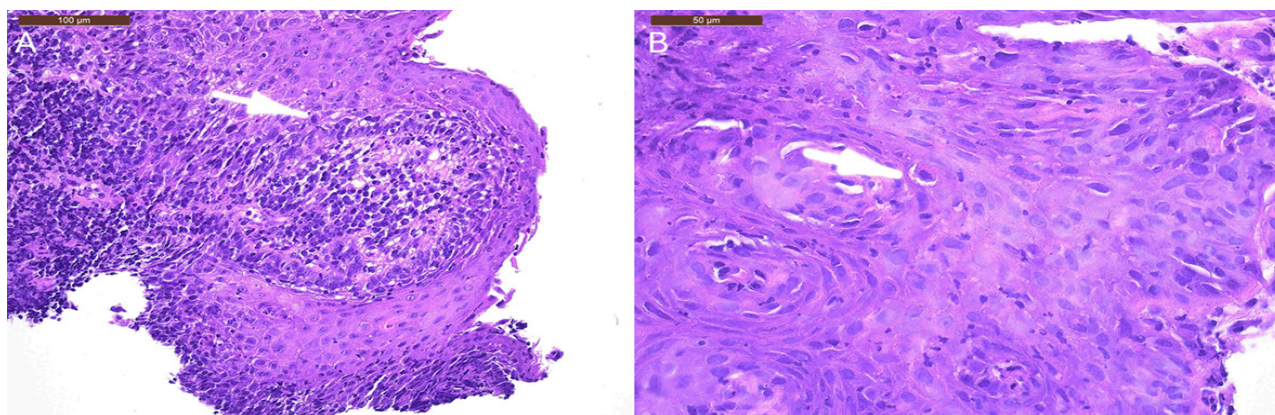


Figure 3. — Histological findings in esophageal lichen planus. A: Lichenoid infiltrate: a dense band-like T-cell lymphocytic infiltrate at the junctional level (interphase pathology); B: Civatte body.

Table 1. — The ELP severity score (ELPSS).

Inflammation (Max 6)	Fibrosis (Max 4)
Pale, edematous, or lacy appearance (1)	Single stricture traversed with 10 mm scope (1)
Sloughing or peeling (2)	Multiple strictures or narrowing (2)
Exudate (1-mild, 2-moderate, 3-severe)	Ant stricture requiring downsizing to 5 mm scope (2)
Score range: 1-10. Interpretation: Mild (1-2), Moderate (3-4), Moderate-Severe (5-7), Severe (8-10).	

the distal esophagus are more likely to be caused by GERD. Furthermore, any observed strictures - which can be caused by both GERD and ELP - should be carefully evaluated: the presence of surrounding erosive esophagitis and/or linear ulcers are not seen in ELP and therefore are more suggestive of GERD (13).

Histologically, a lymphocytic infiltrate can be found in both GERD and ELP. However, dilated intercellular spaces (22) and intra-epithelial eosinophils (21) are not common in ELP and rather support a diagnosis of GERD. Conversely, the presence of Civatte bodies or a typical lichenoid infiltrate are indicative of ELP.

• Eosinophilic Esophagitis (EoE)

Both conditions can present with predominant symptoms of dysphagia. Endoscopically, EoE can resemble ELP quite closely. Trachealization of the esophagus, an endoscopic finding typically associated with EoE, has also been described in ELP (1, 23). Moreover, esophageal strictures can be caused by both EoE and ELP (24).

However, these 2 conditions occur in demographically different patient populations. ELP occurs almost exclusively in middle-aged women, whereas EoE is mainly diagnosed in young men (25). Further ways to differentiate between EoE and ELP can be subtle clues from a patient's history, such as history of asthma or food allergies, which would make EoE a more likely diagnosis (24).

Ultimately, however, histology is necessary to make a firm distinction between the two. According to the 2018 Updated International Consensus

diagnostic criteria for EoE, a definitive diagnosis of EoE requires histological evidence of esophageal eosinophilic inflammation (more than 15 eosinophils per high power field) in the absence of alternative causes of esophageal eosinophilia (16). Other causes of esophageal eosinophilia include hypereosinophilic syndrome, pill esophagitis, Crohn's disease, etc. The aforementioned guidelines, however, are unclear about how far clinicians must go to exclude these alternative diagnoses.

• Pemphigus vulgaris and mucosal membrane pemphigoid

Pemphigus vulgaris and mucosal membrane pemphigoid are auto-immune blistering diseases with predominant mucosal involvement (26). Both diseases can very rarely present with esophageal abnormalities even in the absence of cutaneous or oral lesions and can therefore resemble ELP in the form of erosions and strictures.

Differential diagnosis between ELP and pemphigus/pemphigoid requires direct immunofluorescence: unlike pemphigus or pemphigoid, ELP will not show intercellular or basement membrane zone IgG deposition (3).

• Candida esophagitis

This fungal form of esophagitis can occur in both immunocompromised and immunocompetent patients. Endoscopically, Candida esophagitis may mimic ELP as it can also show mucosal areas with

white discoloration. In *Candida*, however, these areas are typically white, cottage cheese-like plaques or exudates that cannot be rinsed off by irrigation (27). A definitive distinction between the two can be made by demonstrating yeasts and hyphae invading the mucosal cells on biopsy (28).

- Viral esophagitis

Viruses such as herpes simplex virus (HSV) and cytomegalovirus (CMV) are other infections that can cause inflammation of the esophageal mucosa. Although much more common in immunocompromised patients, HSV can rarely occur in immunocompetent individuals (29). Like ELP, viral esophagitis can cause symptoms of dysphagia or odynophagia.

Endoscopy may reveal mucosal abnormalities that are uncharacteristic of ELP, such as erythema, erosions, and ulcers (30).

A viral etiology of esophageal abnormalities can be ruled out by the absence of multinucleated giant cells with ground-glass nuclei in case of HSV (31), and by the absence of intranuclear and intracytoplasmic inclusions in case of CMV (32).

- Esophagitis dissecans superficialis (EDS)

An important endoscopic term to mention when discussing the differential diagnoses of ELP is esophagitis dissecans superficialis (EDS). EDS describes an excessively sloughed-off appearance of the esophageal mucosa, in some cases forming a tubular cast within the esophageal lumen (18). While EDS-like findings can occur in ELP as an extreme form of mucosal denudation, they are not unique to ELP and can also result from trauma (33), certain medications (34), or desquamating dermatological conditions such as pemphigus vulgaris and pemphigoid (18). Therefore, EDS should be viewed as an endoscopic finding rather than a distinct diagnosis.

- Lymphocytic esophagitis

Another relevant term in this context is lymphocytic esophagitis. This description may be used by pathologists to describe a setting where peripapillary lymphocytes and intercellular edema are observed in the relative absence of granulocytes (35). Currently, there is insufficient data to declare lymphocytic esophagitis a distinct clinical entity (36). In the past, the presence of Civatte bodies was viewed as a feature specific to ELP and a way to differentiate between ELP and lymphocytic esophagitis (37). However, recent findings indicate that Civatte bodies can also be observed in lymphocytic esophagitis (36). Therefore, the distinction between these two does not seem to be so clear-cut. For now, it is best to view lymphocytic

esophagitis as a histological pattern of injury rather than a standalone diagnosis.

Complications and natural history

Esophageal stricture/stenosis

In cases of esophagitis resulting in strictures that are not eosinophilic in nature, ELP is a common underlying cause. Marin et al. investigated a cohort of 38 patients with this disease pattern and found that lichen planus was the cause in 47% (n=18) of patients (38).

Conversely, in patients with known ELP, formation of strictures is very common. A recent multicenter descriptive report of 78 ELP patients found that esophageal strictures were present in more than half of cases (53.8%, n=42) (39). Moreover, in one third of these patients, multifocal strictures were observed. Additionally, 21 patients were described as having a “narrow caliber esophagus”, although it is unclear whether this term describes a separate characteristic or a label that can coincide with single or multifocal strictures.

Given the rarity of ELP, it is not surprising that no large-scale studies have been conducted to accurately describe the natural history of this condition. However, case reports and retrospective case series suggest a pattern of long diagnostic delays, frequent need for stricture dilatation, and, in very severe cases, the need for surgical esophagectomy due to inability to safely perform dilatation (40), or perforation and pneumomediastinum (41).

Malignant transformation

As mentioned above, one reason for the dearth of knowledge about the natural history of ELP is the paucity of large-scale observational analyses. However, a large cohort study of 13,100 female lichen planus patients in Finland demonstrated that patients with lichen planus, whether cutaneous, oral, genital or esophageal, were significantly more likely to develop esophageal cancer (42). As a standalone finding, this indicates a correlation between lichen planus and esophageal cancer but does not necessarily prove the existence of a hypothetical ELP-to-SCC sequence.

The development of squamous cell carcinoma (SCC) in patients with known ELP has been described in multiple case studies (13, 43-45). A retrospective cohort study published in 2019 on 132 patients with known ELP found that SCC developed in 8 patients, correlating to an alarming 6.1% of cases (46). Another concerning finding from this study was the seemingly rapid malignant transformation, with half of these cancers observed within 1 year after ELP diagnosis. However, it cannot be fully excluded that the cancer

was already present at the time of diagnosis and contributed to symptoms necessitating the endoscopy.

Currently, no guidelines for the surveillance of ELP exist. Although many different treatment modalities for ELP are available and possibly symptom-reducing, there is currently no evidence that any of these treatment regimens prevent malignant degeneration (43).

The authors of the previously mentioned cohort study have suggested an initial endoscopic follow-up and SCC screening 6 months after diagnosis, with yearly endoscopic follow-up thereafter (46).

Management

General mucosal lichen planus management

For this indication, a variety of treatments is available. However, there does not seem to be a clearly defined treatment algorithm: choice of therapy is mainly guided by the physician's assessment of disease severity; their familiarity with certain medications; and patient preference (47).

Topical corticosteroids are first-line treatment for moderate to severe mucosal lichen planus(2). Formulations for oral application containing high-potency corticosteroids are available, and do not appear to cause any of the known side effects of systemic corticoids.

The use of **systemic steroids** should be reserved for very severe mucosal lichen planus (erosive, multisite lesions). The initial dose (for example, prednisone 40mg/day, or 0.5mg/kg/day) should be tapered over the course of several weeks to months.

For maintenance therapy, **systemic retinoids** have shown to be an effective treatment with an acceptable safety profile. Among the class of retinoids, acitretin (25-50mg/day) is the preferred agent (15).

Systemic **cyclosporine** has been found to be effective in multiple studies. The literature mentions a dosage of 3-10 mg/kg/day, though in clinical practice lower doses are generally prescribed, out of concern for systemic toxicity. This unfavorable safety profile is also the reason why cyclosporine should generally be reserved for severe cases.

Enoxaparin sodium, although mostly known for its anticoagulant properties, has been evaluated for its anti-inflammatory effects in lichen planus, typically in a once or twice weekly low-dose regimen. A 2019 evidence-based review of treatments used in lichen planus found inconsistent results (47). To formally advise the use of enoxaparin sodium for the treatment of lichen planus, more research is needed with better designed randomized controlled trials (RCTs). However, as its longstanding use for other indications (and in higher doses) has shown that this drug is very safe, the usage of enoxaparin as a "trial" add-on therapy can be considered, while awaiting more data regarding its efficacy.

Hydroxychloroquine sulfate has been studied in a cohort of 10 patients (open label) with oral lichen planus(48). Pain relief and disappearance of focal oral erythema were observed in 9 patients after 6 months of therapy. Much like enoxaparin, the favorable safety profile of hydroxychloroquine should allow some freedom to consider adding this therapy to the treatment regimen, even in the absence of robust efficacy data.

Management of esophageal lichen planus

Because the prevalence of ELP is quite low and esophageal involvement of lichen planus has only become the subject of academic interest some 20 years ago, robust data from placebo-controlled trials are lacking. Indeed, when a 2012 Cochrane systematic review tried to summarize all existing evidence for the treatment of erosive mucosal lichen planus and set out to include all trials investigating oral, genital, and esophageal lichen planus, not a single RCT studying treatments in ELP-populations was identified (49). Neither have we found any RCTs on the treatment of ELP after the publication of this systematic review in 2012. Therefore, the following description of possible therapeutics and their efficacy is based on case studies and retrospective case series, each including relatively few patients (Table 2; ref. (50-66)). There is still no standardized treatment for ELP, firstly because of its rarity, but also due to the often-lengthy period between onset of symptoms and diagnosis. During this interval, many medications as well as dilatations of strictures may have already been tried. This further complicates our understanding of the efficacy of potential therapies (3).

The role of **proton pump inhibitors (PPI)** in the treatment of ELP, although frequently empirically prescribed as first-line treatment, seems to be limited. In a recently published retrospective multicenter report, PPI monotherapy did not seem to improve ELP, although endoscopic and histological records pre- and post-treatment could only be obtained from 4 and 2 patients, respectively (39). An older case series on 6 ELP patients reported that PPI monotherapy had failed in all 6 patients (67). Furthermore, during our review of case studies/series mentioning ELP treatment, we found multiple reports where PPI were empirically started as first-line treatment, without clinical benefit (68-70).

A case series published in 2019 reported that in 7 out of 16 patients later diagnosed with severe ELP, and in 7 out of 18 patients diagnosed with mild ELP, acitretin was already part of the ongoing therapy. With these – very limited – data in mind, it seems less likely that systemic retinoids are a potent treatment for severe ELP (1). The role of acitretin in milder forms of ELP is unclear.

Effective treatment with **topical corticosteroids** (e.g. swallowed fluticasone or budesonide viscous

Table 2. — Therapies investigated for ELP.

Study	Year of Publication	Study Design	Number of Patients	M:F ratio	Intervention	Dose	Frequency of intervention	Duration of follow-up	Outcome
Eustace et al.	2015	Case report	1	0	Adalimumab	40 mg	q2weeks (after initial induction dose of 80mg)	6 weeks	No symptomatic improvement
Hafner et al.	2016	Case report	1	0	Apremilast	20mg	bid	4 weeks	Complete clinical response (dysphagia); Endoscopic improvement
Chaklader et al.	2009	Case report	1	0	Cyclosporine	Not specified	Not specified	Not specified	“Successful” (not otherwise specified)
Manna et al.	2013	Case report	1	0	Cyclosporine A	1.5 mg/kg/day	bid; for 24 weeks	12 months	Improvement in dysphagia; weight gain; endoscopic improvement
Keate et al.	2003	Retrospective case series	3	0	Intralesional injection of triamcinolone + oral tacrolimus in 2 out of 3 patients	5 to 20 mg	Ranging from once to 7 times over course of 3 years	Not specified	Decrease in esophageal symptoms; decrease in need for endoscopic dilatation
Wedgeworth et al.	2009	Retrospective case series	5	0	Intralesional injection of triamcinolone + dilatation	40 to 60 mg	Every 3 to 4 weeks; intervention only as needed	18 to 54 months	Good response, need for intervention became less frequent
Acar et al.	2021	Case report	1	0	Intralesional injection of triamcinolone acetate	40mg	5 times, q4w	1 month	Improvement in dysphagia; complete resolution of stricture
Bagger-Jørgensen et al.	2024	Case report	1	0	Budesonide orodispersible tablet (Jorveza™)	1mg	bid	10 weeks	Complete clinical response (including oral LP!) + Endoscopic improvement after 2 months
Westbrook et al.	2008	Case report	1	0	Mycophenolate mofetil	750 mg	bid	Not specified	Decrease in oral pain but no effect on dysphagia and need for dilatation
Parmentier et al.	2008	Case report	1	0	Rituximab	375 mg/m ²	weekly, over the course of 4 weeks	6 months	Symptomatic and endoscopic complete response (no dysphagia or remaining stenosis)
Donnellan et al.	2010	Retrospective case series	5	0	Swallowed fluticasone propionate	500 µg	bid	Not specified	Symptomatic improvement in 3/5 patients
Hou et al.	2011	Case report	1	0	Swallowed fluticasone	440 µg	bid	6 months	Complete clinical and endoscopic response

Table 2. — Therapies investigated for ELP - part 2.

Study	Year of Publication	Study Design	Number of Patients	M:F ratio	Intervention	Dose	Frequency of intervention	Duration of follow-up	Outcome
Ynson et al.	2013	Case report	1	0	Swallowed fluticasone propionate	220 µg	bid	15 weeks	Symptomatic and endoscopic improvement
Podboy et al.	2017	Retrospective cohort analysis	32	0,25	Swallowed budesonide	3 mg	bid	Not specified	Decrease in dysphagia (P<0,001) + Endoscopic Improvement in 72% of patients (pooled)
Podboy et al.	2017	Retrospective cohort analysis	8	0,25	Swallowed fluticasone	880 µg	bid	Not specified	Decrease in dysphagia (P=0,029) + Endoscopic Improvement in 72% of patients (pooled)
Kooybaran et al.	2022	Case report	1	0	Tofacitinib (+ swallowed fluticasone)	5 mg (+ 880 µg)	bid initially, later reduced to qd (+ bid)	7 months (recurrent symptoms after discontinuation; after restart of therapy: symptomatic improvement)	Decrease in dysphagia and odynophagia + weight gain; endoscopic improvement

solutions) has been described in multiple reports (60-63). However, cases refractory to these therapies have also been reported, prompting some clinicians to try tyrosine kinase inhibitors such as tofacitinib (64, 65) and upadacitinib (66) in off-label use, with promising results.

In severe cases of ELP, **systemic corticosteroids** can be needed to achieve rapid symptomatic relief. These are not an optimal therapy long-term maintenance, however, as high doses are needed, leading to systemic side effects. Furthermore, reduction or cessation of systemic steroids often results in recurrence of symptoms (3).

We found one case report evaluating the effect of anti-TNF agent **adalimumab** for ELP, without a beneficial effect on symptoms (50).

In a small-scale retrospective cohort study of 22 patients with ELP where 11 patients were treated with **systemic tacrolimus** (1-2mg bid) and 11 patients received no tacrolimus, there was a significant endoscopic improvement quantified by the esophageal lichen planus severity score (Table 1) as well as a longer time to necessity of dilatation (71).

Esophageal dilatation

Even before a formal ELP diagnosis, many patients have already undergone endoscopic dilatation for esophageal strictures (3). Endoscopic dilatation is a generally safe intervention that can provide symptomatic relief (39, 54, 65). Considering medication alone is unlikely to treat existing stenoses, dilatation is seen as an important adjunctive to anti-inflammatory medications. Nevertheless, caution is needed, as there have been reports of perforation after dilatation (41).

Cases of refractory esophageal strictures, not amenable to dilatation, have also been described, in extreme cases resulting in esophagectomy (40).

As mentioned before, the rarity of ELP has made it difficult to perform prospective studies or RCTs. As such, robust data regarding the efficacy and safety of endoscopic dilatation in ELP are lacking.

Discussion

In this narrative review of the literature regarding esophageal lichen planus, we found that overall, it is a rare disease, although there is reason to assume that its occurrence may be higher due to lack of standardized screening approach in patients with extraesophageal lichen planus, as well as the possibility of esophageal involvement of lichen planus without classical symptoms such as dysphagia. Diagnosis of ELP is often preceded by many years of symptomatic disease and repeated dilation therapy for presumed GERD-induced strictures. Endoscopists should consider ELP when faced with proximal esophageal strictures, especially in middle-aged women. Endoscopic criteria for ELP include white

discoloration, mucosal sloughing, and trachealization. Histological findings include the characteristic dense band-like T-cell lymphocytic infiltrate at the subepithelial level (the lichenoid infiltrate) with the presence of Civatte bodies. Furthermore, epithelial detachment, a lymphocytic infiltrate, and dyskeratosis can be seen on histopathology. Differential diagnoses to consider are GERD, eosinophilic esophagitis, bullous dermatological diseases such as pemphigus/pemphigoid, candida esophagitis, and viral esophagitis. The natural history of ELP is that of a chronic inflammatory condition, of which the most relevant complications are (i) the formation of esophageal strictures, and (ii) malignant transformation to squamous cell carcinoma. Management of ELP consists of anti-inflammatory medication, and endoscopic dilatation of chronic strictures. Unfortunately, recommendations regarding optimal practice based on high-powered studies are lacking. Both topical and systemic corticosteroids are often used. In cases refractory to steroids, a number of anti-inflammatory agents have been proposed. Given the rarity of ELP, however, only small-number studies and case reports about the use of these alternative anti-inflammatory agents are available. More research regarding the optimal anti-inflammatory agent for ELP is needed. In conclusion, we urge gastroenterologists and endoscopists to be aware of this condition and consider its diagnosis, especially in middle-aged female patients with esophageal strictures refractory to PPI treatment, and in patients with known extraesophageal forms of lichen planus with esophageal symptoms such as dysphagia. More research is needed to guide clinicians towards optimal management of this rare disease.

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