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Case and Review

Oesophageal Lichen Planus Successfully Treated with Budesonide Orodispersible Tablets: A Case Report

Harald Bagger-Jörgensen[a, b] Mohammed Abdulrasak[a, b]
Kevin Sandeman[c] Mohammed Binsalman[a, b] Klas Sjöberg[a, b]

[a]Department of Clinical Sciences, Lund University, Malmö, Sweden; [b]Department of Gastroenterology, Skåne University Hospital, Malmö, Sweden; [c]Division of Laboratory Medicine, Office for Medical Services, Department of Clinical Genetics and Pathology, Skåne, Sweden

Keywords
Budesonide orodispersible tablets · Lichen planus · Oesopageal lichen planus · Case report

Abstract
Introduction: Lichen planus is a relatively common inflammatory condition of the nails, skin, and mucosal surfaces. Oesophageal involvement of lichen planus is thought to be very rare, mainly described in case reports, but is associated with a high risk of oesophageal stenosis as well as squamous cell carcinoma. No evidence-based treatment recommendations exist, with the majority of described treatment regimens involving systemic immunosuppression. ***Case Report:*** In this case report, we describe a novel approach in treating oesophageal lichen planus in a patient with budesonide orodispersible tablets, a treatment normally reserved for eosinophilic oesophagitis. The patient achieved complete relief of dysphagia, with a follow-up oesophagogastroduodenoscopy 2 months after treatment commencement being macroscopically and microscopically free of inflammatory activity. This case report is to our knowledge the first to report this treatment regimen in oesophageal lichen planus. ***Conclusion:*** We consider a trial of budesonide orodispersible tablets a reasonable initial management as it's a local therapy specific to the oesophagus with a more benign side effect profile than systemic immunosuppression, but further studies need to be undertaken to corroborate our findings. Also, based on the severity and malignant potential of oesophageal lichen planus, we suggest that physicians be liberal in ordering oesophagogastroduodenoscopy with biopsy taking as part of the workup of dysphagia in a patient with known lichen planus.

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Harald Bagger-Jörgensen and Mohammed Abdulrasak are the first authors.

Correspondence to:
Harald Bagger-Jörgensen, harald.bagger-jorgensen@med.lu.se

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Introduction

Lichen planus (LP) is a chronic inflammatory condition affecting the integument (mainly nails and skin) and mucosal surfaces in locations such as the mouth, oesophagus, and vagina. There is a predisposition to the disorder occurring in middle-aged females, with an estimated prevalence of 1% worldwide, across all LP presentations [1].

Oesophageal LP (ELP) is an underdiagnosed variant of the disorder [2]. This may present as a lone presentation of LP [3], or concomitantly to LP in other locations [4]. ELP is associated with an increased risk of squamous cell carcinoma (SCC) and therefore needs to be treated aggressively [4]. Treatment includes steroids and other immunosuppressive agents, albeit there is no consensus on treatment lengths and/or follow-up intervals due to the relative rarity of the condition [5]. In this case report, we detail the diagnostic evaluation, management, and follow-up of a patient presenting with ELP and discuss potential pitfalls that may arise in that setting. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000538894).

Case Report

A 60-year-old woman presented to her general practitioner with a decade-long history of chronic dysphagia, with acute deterioration in the past 3 months. She had previously been diagnosed, through her general practitioner, with "oesophageal spasm" on spurious clinical grounds, with no oesophagogastroduodenoscopy (EGD) performed for evaluation of this dysphagia (she had undergone an EGD approximately 20 years prior to this, but on another indication). Having previously tolerated solid foods with some difficulty, the patient could now only take fluids. Her past medical history was significant for mucosal ulcerative LP in the mouth and genitalia for the past 15 years, heterozygotic α1-antitrypsin deficiency, active smoking with 40 pack years, and COPD. The oral LP had been difficult to treat, mainly managed with on-off local group IV steroids. Her hepatitis C status was negative. At the time of her presentation to our clinic, her oral LP was active with painful lesions of the oral mucosa, while her genital LP was entirely quiescent. She was urgently referred to an EGD for suspected oesophageal carcinoma.

Macroscopically, the oesophageal mucosa was rigid and trachealised with a papier-maché appearance, suspicious for long-standing eosinophilic oesophagitis (EoE) (Fig. 1). Biopsies were taken along the length of the oesophagus. Histopathological examination of the biopsy of the distal oesophagus revealed a lichenoid oesophagitis pattern of injury (Fig. 2), with a prominent band-like lymphocytic infiltrate involving the interface of the lamina propria and epithelium as well as Civatte bodies – apoptotic keratinocytes considered a hallmark of LP. Additionally, intraepithelial lymphocytes and dyskeratotic keratinocytes were observed. Several biopsies showed hyperkeratosis and granulation. No dysplasia or fungal organisms were identified. The patient was subsequently referred to our clinic at the Department of Gastroenterology, Skåne University Hospital, for further evaluation.

Given the suspicious histopathologic appearance for LP as well as the significant past medical history of mucosal LP, the patient was diagnosed with ELP over other far less likely differential diagnoses such as Crohn's disease, infections, medication-induced oesophagitis, or pill oesophagitis. A discussion was had as to the best treatment strategy, as no internationally accepted treatment guidelines for ELP exist. Several pharmacological treatment regimens were found in the literature, including oral budesonide formulations, calcineurin inhibitors, mycophenolate mofetil, PDE-4 inhibitors, as well as systemic corticosteroids [6–10]. As

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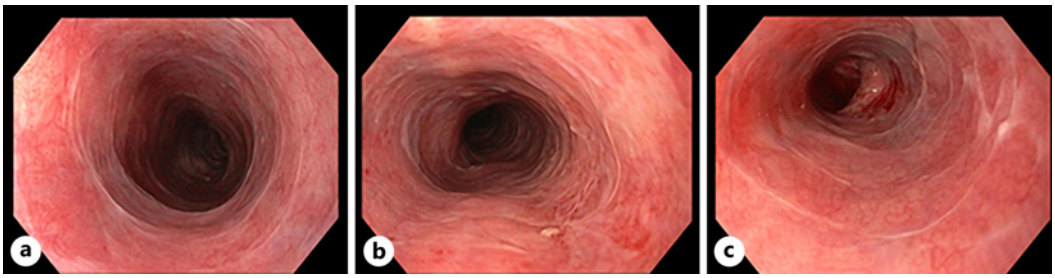

Fig. 1. EGD showing friable and borderline trachealised oesophageal mucosa, in the proximal (**a**), middle (**b**), and distal (**c**) oesophagus.

topical corticosteroids are the first line of oral LP, we opted for Jorveza™, a novel budesonide orodispersible tablet (BOT) designed for EoE highly selective to the oesophagus [11]. Prior to the introduction of BOT, EoE treatment was largely based on topical steroid therapies. These are now, however, defunct given the efficacy of BOT for EoE treatment. We found no studies on the efficacy of BOT on ELP, only nondescript topical steroid solutions now defunct in our clinical practice. We opted for BOT as this formulation seemed the most targeted and efficacious approach. The patient gave informed consent and was commenced on oral BOT (Jorveza™) 1 mg BD (twice daily) for 10 weeks.

Within days of commencement of treatment, the patient experienced a complete remission of dysphagia. She reported no side effects. Surprisingly, she also reported marked improvement in her oral LP, with near-complete healing of ulcerations. A follow-up EGD was performed 2 months post-therapy initiation, which was normal apart from incidental candidiasis (a common side effect of BOT [11]), after which the patient received fluconazole 200 mg BD for 14 days. Biopsies taken from the length of the oesophagus showed no evidence of existing oesophagitis, with an entirely normal mucosal appearance – apart from diffuse candida hyphae and spores without inflammatory response in the distal oesophagus (Fig. 3).

After 10 weeks of induction therapy, the patient remained completely asymptomatic. The patient's weight increased from 68 kg (BMI 25.9) pretreatment to a steady state of 72 kg (BMI 27.4). Her serum albumin remained unchanged, ranging between 38 and 42 g/L. We opted to cease active treatment and now plan for renewed EGD in 6 months followed by once yearly for surveillance of subclinical disease and/or SCC of the oesophagus as well as to monitor the possible need for oesophageal dilation. In the event of clinical recurrence, we plan the recommencement of induction dose BOT 1 mg BD for 6 weeks. Alcohol and smoking cessation was advised to further reduce the risk of oesophageal SCC.

Discussion

In this case report, we are, to the best of our knowledge, the first to describe BOT (Jorveza™) as a viable treatment option for ELP. Treatment with BOT was highly successful in inducing complete clinicopathologic remission, reversing long-standing, debilitating dysphagia without any need for invasive endoscopic dilation. Her quality of life improved drastically. While the BMI increase post-treatment was fairly modest, she had for many years compensated for the dysphagia by eating liquidised foods and avoiding meat. As outlined above, a wide range of treatment regimens are described in the literature, ranging from topical fluticasone formulations to systemic immunosuppressants with potentially serious side effects [6–10]. BOT seems a tempting future treatment option for ELP, given its near complete lack of systemic effect.

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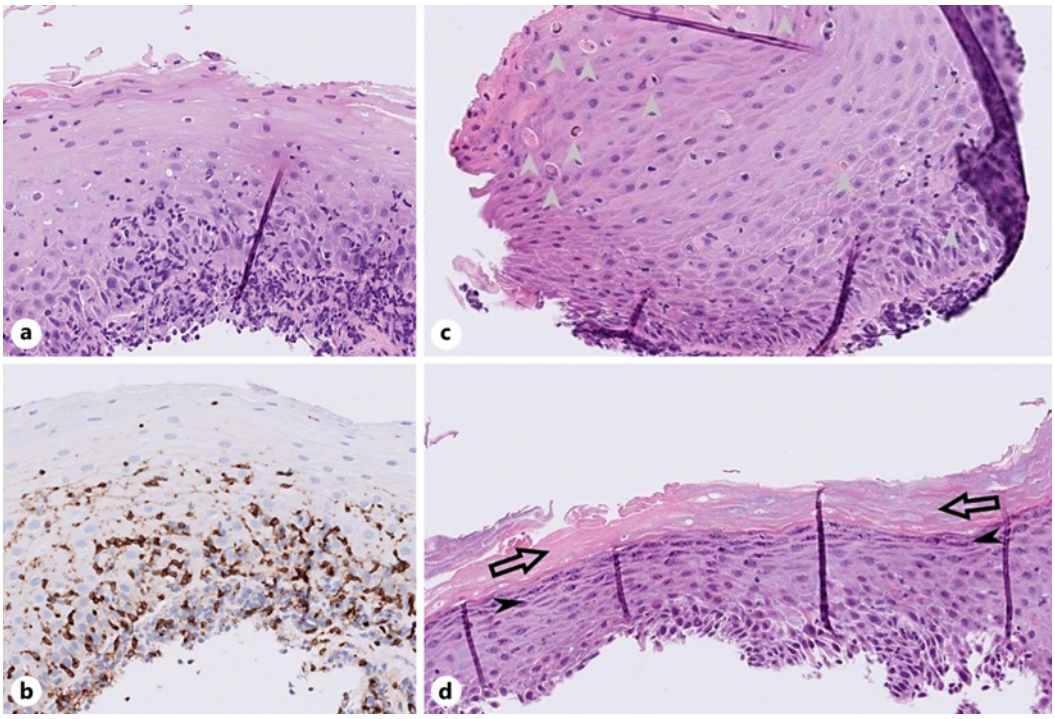

Fig. 2. Lichenoid histologic appearance of oesophageal mucosa. **a** Band-like lymphocytic infiltrate at the interface of basal oesophageal epithelium and lamina propria, using HE staining (×20; 0.5 µm/pixel). **b** Predominance of T lymphocytes (brown CD3-positive cells) in the epithelium and lamina propria, using DAB technique (×20; 0.5 µm/pixel). **c** Multiple dyskeratotic degenerative keratinocytes also known as Civatte bodies (grey arrowheads), using HE staining (×20; 0.5 µm/pixel). **d** Oesophageal hyperkeratosis with keratinisation (arrows) and hypergranulation (arrowheads) of the squamous epithelium, using HE staining (×20; 0.5 µm/pixel).

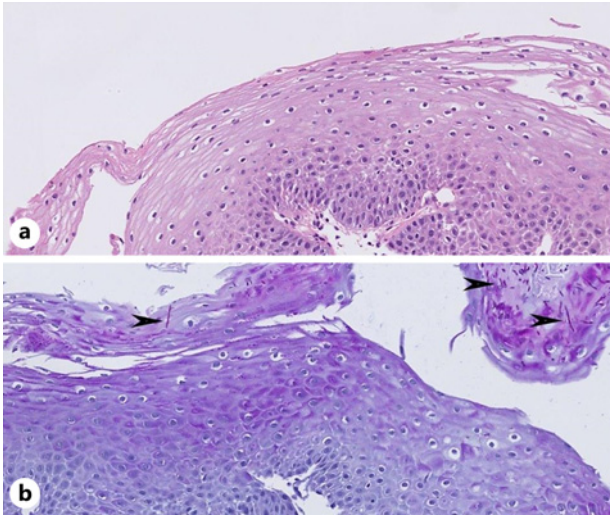

Fig. 3. **a** Post-treatment oesophageal mucosa appears normal without any significant pathological findings, using HE staining (×20; 0.5 µm/pixel). **b** *Candida albicans* spores and hyphae (arrowheads) are observed as commensal findings on the normal oesophageal mucosa, without any associated cellular reactions, using ABPAS staining (×20; 0.5 µm/pixel).

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Endoscopic findings in ELP include erosive changes, ulcerations, sloughing, strictures, and also trachealisation (potentially mimicking EoE, as seen in this patient). Histopathologically, ELP is characterised by lymphocytic infiltrates in the lamina propria and parakeratosis of the epithelium with characteristic Civatte bodies (apoptotic keratinocytes) (2).

In our patient, there was total endoscopic and symptomatic resolution of ELP, and also of her oral lesions. This was achieved using BOT, which is usually administered by the patient placing the tablet on the tongue and pressing it on the top of the hard palate for it to slowly disintegrate, thereby causing effervescence and hypersalivation and a steady "stream" of saliva loaded with BOT to go through the mouth, oropharynx, and oesophagus [11]. When analysing how this drug is administered, it is easy to understand that the potential local effect of the drug has also aided in the quick resolution of the patient's oral lesions, being a serendipitous effect. BOT may therefore be a future treatment option for severe oral LP instead of systemic immunosuppressants. This, however, may act as a two-edged sword given the risk for mucosal candidiasis, which our patient also developed. Candidiasis is easily treated with antifungals and should not be a deterrent to prescribing BOT to this patient cohort.

ELP is considered a rare presentation of LP, with only 10% of LP patients having this specific involvement [5]. However, some (albeit smaller) studies have suggested concomitant asymptomatic oesophageal involvement may be up to 50% in patients with cutaneous LP, when these patients were screened at initial presentation [12]. Current guidance suggests screening LP patients for oesophageal involvement when the development of upper GI symptoms, such as dysphagia or odynophagia, occurs [13]. It is unclear, however, if asymptomatic ELP is associated with SCC development, or if that is reserved for symptomatic cases, and further studies are needed in that regard.

The diagnosis of ELP is usually delayed by >2 years from the time patients first report their symptoms to physicians [14]. This is a significant delay that may have important therapeutic implications, given that the median time to progression to SCC in ELP patients was reported as 3.5 years from when ELP diagnosis was established [13], with the highest risk being present ≈ 5 years after diagnosis [15]. Patients with LP in other locations should therefore be actively screened for the presence of upper GI symptoms, either through questionnaires or by history, with early EGD with biopsy taken from the oesophagus of patients with upper GI symptoms. This is in order to avoid such aforementioned diagnostic delay, given the ≈ 6% SCC rate in patients diagnosed with ELP and the endoscopic treatment options available for patients with early oesophageal dysplasia/carcinoma development [13]. Our case highlights the importance of having a high degree of suspicion in LP patients presenting with dysphagia, as our patient had suffered from dysphagia for nearly a decade but was misdiagnosed with oesophageal spasm without any endoscopy being performed.

In conclusion, ELP is difficult to diagnose given its rarity. However, in the setting of LP elsewhere, there should be a low threshold to suspect the diagnosis, especially in patients presenting with upper GI symptoms, with EGD to be performed liberally. Treatment with BOT seems effective and relatively uncomplicated, as well as having a major advantage over other treatment options with its minimal systemic effects. Further studies are, however, needed to fully establish BOT as a recommended treatment option for ELP.

Statement of Ethics

Ethical approval was not required to publish this case report, nor did the retrospective review of patient data did require ethical approval, in accordance with national guidelines. Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors have no conflict of interest to declare.

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Author Contributions

All authors contributed to the writing/editing part of this paper. Harald Bagger-Jörgensen and Mohammed Abdulrasak wrote the manuscript. Mohammed Binsalman, Klas Sjöberg, and Kevin Sandeman edited and guided in writing this manuscript.

Data Availability Statement

The authors declare that data supporting the findings of this study are available within the article. Further enquiries can be directed to the corresponding author.

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