Lichen planus: Five variants presenting in one patient

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INTRODUCTION

Lichen planus (LP) is a chronic disorder of unknown origin that causes mucocutaneous inflammation.[1] Classic lesions of LP are flat-topped, polygonal, violaceous papules and plaques with fine white lines called *Wickham striae*[2] most commonly affecting the skin and oral mucosa.[1] Variants of LP may involve the genitals, esophagus, conjunctivae, scalp, and nails.[3] Although oral involvement of LP is common (30% to 70%),[2] few have reported multiple variants occurring in an individual simultaneously. Here we present a case of LP occurring in 5 forms at once: cutaneous, oral, vulvovaginal, and esophageal (ELP) and lichen planopilaris (LPP) of the scalp.

Abbreviations used:

EGD:	esophagogastroduodenoscopy
ELP:	esophageal lichen planus
LP:	lichen planus
LPP:	lichen planopilaris
SCC:	squamous cell carcinoma

CASE REPORT

A 72 year-old woman with a 20-year history of extensive LP including cutaneous, oral, vulvovaginal, and LPP was referred to the gastroenterology department for evaluation of a 5-year history of intermittent solid food dysphagia and choking episodes. Her dysphagia had progressed to include soft foods and pills resulting in a 40-lb weight loss over a year. She denied symptoms of gastroesophageal reflux disease or proton pump inhibitor use. During this period, her cutaneous LP and oral lesions were well controlled with clobetasol cream and benzocaine and fluocinonide gels, respectively. She has never been on systemic medications or phototherapy for her LP and has never received chemopreventive agents such as acitretin to avert the development of mucosal squamous cell carcinoma (SCC). She denied taking any medications that are known to cause drug-induced LP before the onset of her skin lesions; she was only taking levothyroxine for hypothyroidism at that time. The patient does not have a history of hepatitis C or HIV infection.

Initial esophagogastroduodenoscopy (EGD) with dilation to 10 mm noted a proximal esophageal stricture with sloughing mucosa (Fig 1, *A*). Stricture pathology findings showed ulceration, degenerated squamous cells, and dyskeratosis without evidence of herpes simplex virus, candida, or eosinophilic esophagitis. The patient was started on swallowed fluticasone and underwent 2 subsequent EGDs with dilation with slight improvement of dysphagia. Three months later, repeat EGD with dilation to 16 mm showed marked endoscopic improvement of her stenosis (Fig 1, *B*). Clinically, the patient's dysphagia improved and she was able to tolerate solid foods and pills.

Physical examination of her other LP lesions found erythematous macules across the inframammary chest with lichenoid papules on the back and

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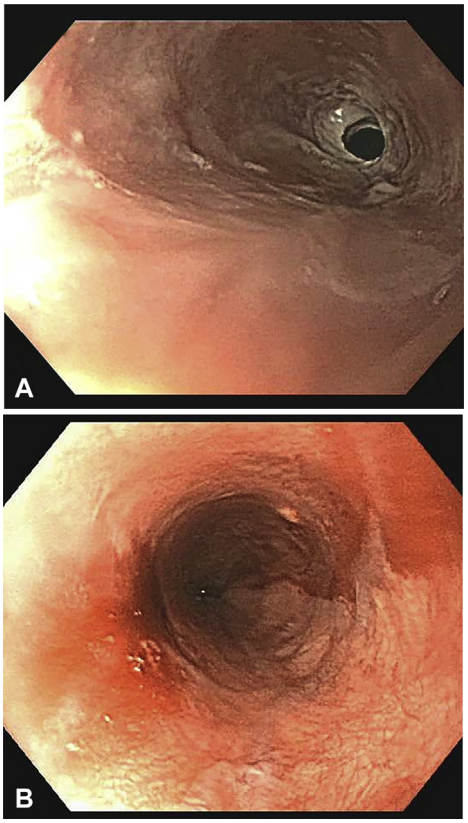

Fig 1. Endoscopic images of esophageal LP lesions. These endoscopic images show improvement of esophageal stenosis after multiple balloon dilations. **A**, Proximal stricture on initial dilation. **B**, Proximal stricture after dilation to 16 mm.

bilateral lower extremities (Fig 2, *A*). The oral cavity showed gingival erythema and white, lacy plaques with one erosion on the right buccal mucosa. Although adherent patches were noted in the vaginal orifice, the vulva was fused around the clitoral hood. The vertex scalp demonstrated an erythematous patch with scarring hair loss (Fig 2, *B*).

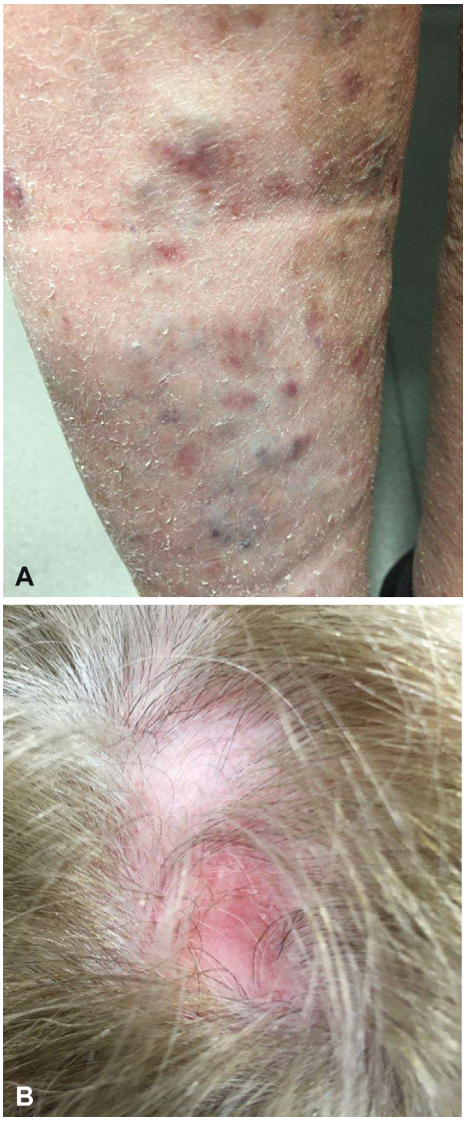

Fig 2. Clinical presentation of multiple LP variants at once. These images show the various LP lesions of a woman with a 20-year history of extensive LP including cutaneous, oral, and vulvovaginal and LPP of the scalp. **A**, Cutaneous LP of the right lower leg. **B**, LPP of the scalp.

DISCUSSION

Several clinical subtypes of LP have been described occurring concurrently or sequentially.[3] In a large study of patients with oral LP, 16% had previous cutaneous disease, 19% had vulvovaginal-gingival syndrome, and others had rare variants involving the esophagus, nails, or conjunctiva.[3] Despite the common overlap of LP manifestations, a review of the literature found few cases that report even 4 forms of LP occurring at once.

Fewer than 100 cases of ELP occur in the literature; however, it may be underreported.[4] Histologic features of ELP include lichenoid inflammation and Civatte bodies. Endoscopic findings include white papules, pinpoint erosions, desquamation, pseudomembranes, and stenosis most commonly affecting the proximal or mid-esophagus.[5] Continued esophageal inflammation with subsequent stenosis may cause dysphagia and carries a risk for esophageal SCC.[5] Although this patient never received any chemopreventive treatment, retinoids such as acitretin may prevent SCC from developing within LP lesions through gene transcription regulation and their interaction with various transcription factors.[6] Esophageal biopsies may not capture ELP histologically; therefore, ELP is not well recognized by pathologists or gastroenterologists. The patient's history and progressive nature of disease suggest that her esophageal abnormalities and associated stricture

were likely a sequela of long-standing inflammation and fibrosis from underlying lichen planus.

Although correct diagnosis of ELP is difficult, it bears important therapeutic implications, as it is a chronic, relapsing condition that may require systemic or local immunosuppressive therapy and repeated endoscopic dilations for strictures. In this case, the patient had good response to serial esophageal dilations and did not require immunosuppressive therapy. As multiple LP forms can present simultaneously in one individual, clinical vigilance and histopathologic examination are critical for the early detection and proper management of various LP manifestations.

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