Use of upadacitinib in refractory esophageal lichen planus: Endoscopic improvement in planus sight

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Introduction

- Lichen planus (LP) is an inflammatory skin condition that affect oropharyngeal and esophageal mucosa
- Prevalence of 0.19% but is thought to be underdiagnosed
- Presents with ulceration, exudates, rings and/or strictures
- Caused by a T-cell mediated response against basal keratinocytes
- JAK signaling pathway is thought to play a role in pathogenesis
- Upadacitinib is a selective JAK1 inhibitor
- **We present a case of refractory esophageal LP (ELP) with endoscopic and histologic response to upadactinib after failing multiple other treatment regimens.**

Clinical Presentation

- 68 year-old female
- History of Sjogren's syndrome, lichen planus, and prior peptic stricture in gastroesophageal junction (s/p dilation 4 years prior)
- Presented with recurrent dysphagia to both liquids and solids, intermittent reflux, regurgitation, and impactions

Work-up

- Initial upper endoscope (Figure 1 and 2):
  - Severe esophagitis seen throughout the entire esophagus, crepe paper esophagus noted, multiple strictures, narrowest with 8mm luminal diameter
  - Passage of standard upper endoscope resulted in dilation
- She was started on pantoprazole 40 mg twice daily and budesonide oral slurry with limited response and recurrent fibrostenotic disease.
- Started on mycophenolate 500 mg daily→ no significant histologic improvement
- Mycophenolate increased to 2000 mg twice a day→ again minimal histologic improvement
- In the interim, patient was seen by an oral surgeon and diagnosed with oral lichen planus.
  - Her medications were reviewed and hydrochlorothiazide was discontinued due to its association with lichen planus.
  - Hepatitis C was negative.

Imaging

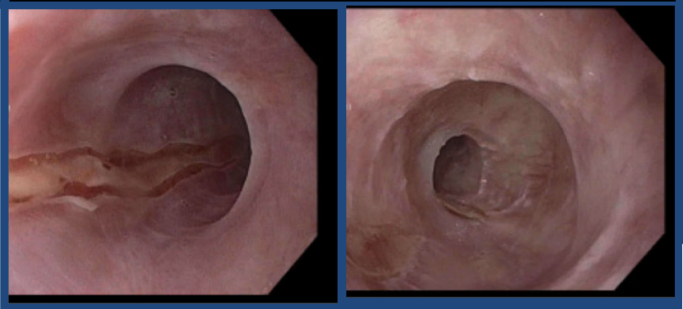

Figure 1 and 2: Esophageal stenosis, severe esophagitis, and mucosal ulceration

Follow-up

- She was then started on upadacitinib 30 mg twice daily.
- Repeat endoscopy 3 months (Figure 3-4):
  - Showed dramatic macroscopic and histologic improvement, with only some residual fibrostenotic disease remaining
  - Dramatic symptomatic improvement

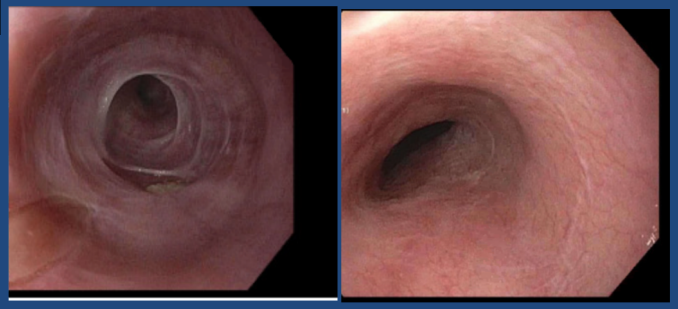

Figure 3 and 4. Repeat endoscopy showing improvement of esophagitis and ulceration; residual fibrosis noted but biopsies show resolution

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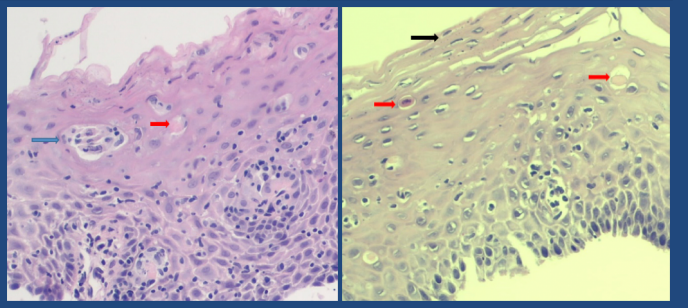

Figure 5. Lichenoid esophagitis. Lymphocytic infiltrates, focal histiocytic aggregate (blue arrow), dyskeratosis (Civatte body, red arrow), and parakeratosis (black arrow).

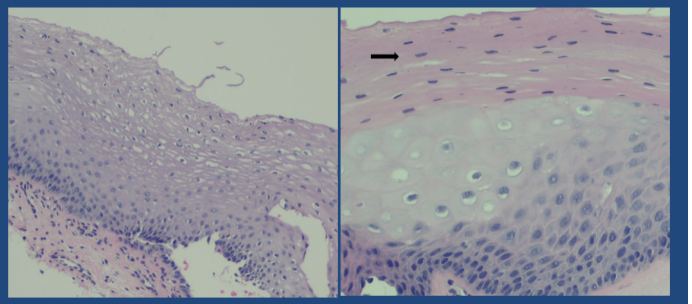

Figure 6. Esophageal mucosa after treatment. Most biopsy fragments show near normal esophageal mucosa (left panel). Focal areas show parakeratosis (black arrow).

Discussion

- Multimodal treatment is typically employed in ELP with medical therapy as well as endoscopic dilation.
- Systemic or topical steroids are typically first line.
  - Efficacy of topical steroids in ELP is estimated to be 60-70%.
- Other medical options include tacrolimus, cyclosporine, mycophenolate, rituximab, and adalimumab.
- Data of efficacy is limited and mainly anecdotal with these therapies.
- Limited data on tofacitinib
- To date, there are only two cases reported in the literature of successful treatment of oro-esophageal LP with upadacitinib.
- Given profound improvement seen in our patient, upadacitinib may offer another alternative for these difficult to treat patients.