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Barrett's esophagus and eosinophilic esophagitis in a young pediatric patient with esophageal atresia

Li-Zsa Tan [a], Andrew J. Gifford [b,c], Cathy M. Clarkson [a], Guy M. Henry [d], Usha Krishnan [a,*]

[a] Department of Gastroenterology, Sydney Children's Hospital, Randwick, New South Wales 2031, Australia
[b] Department of Anatomical Pathology (SEALS), Prince of Wales Hospital, Randwick, New South Wales, Australia
[c] The School of Women's and Children's Health, University of New South Wales, New South Wales, Australia
[d] Department of Pediatric Surgery, Sydney Children's Hospital, Randwick, New South Wales, Australia

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ABSTRACT

We report the case and follow up of a 4 year old girl who was born with esophageal atresia and was found to have concomitant Barrett's esophagus and eosinophilic esophagitis. Development of metaplasia has generally been regarded as a chronic, long term complication post esophageal atresia repair, however this case highlights the need for early and regular endoscopic surveillance in this vulnerable patient population. In addition to this, eosinophilic esophagitis has only rarely been reported in patients with concomitant Barrett's esophagus. Our patient is the youngest in reported literature with Barrett's changes post esophageal atresia repair, and the first such reported patient with both eosinophilic esophagitis and Barrett's esophagus.

Esophageal atresia (EA) affects 1:2400 live births, and is therefore a relatively common congenital anomaly [1]. Expectant management of long term complications such as strictures, esophageal dysmotility, gastroesophageal reflux disease (GERD) and an increased risk of Barrett's and cancer [2] has become increasingly relevant with improved survival rates. Eosinophilic esophagitis (EoE) is an emerging entity as a chronic inflammatory condition associated with esophageal dysfunction [3] which has only rarely been reported in EA patients [4]. Here we report the case and follow up of a young girl with EA diagnosed with EoE and Barrett's esophagus (BE) 4 years after her primary repair.

1. Case report

Our patient was born with long gap, Type A esophageal atresia. A gastrostomy was created shortly after birth and sutures were placed as per the esophageal growth augmentation technique first described by Foker [5]. Delayed primary anastomosis was performed at 3 months. Fundoplication was not performed as it is not routine practice at our center following a Foker long gap EA repair. She was prescribed proton pump inhibitor (PPI) therapy but was non compliant. Her gastrostomy was closed at age 2. She had recurrent anastomotic strictures requiring thirteen radiological dilatations, mostly in her first year of life. The patient also had eczema and asthma, but no food allergies were detected on skin prick testing.

In August 2013 she was referred to the newly established multidisciplinary EA-TEF clinic at our hospital by her treating surgeon for increasing dysphagia, regurgitation and food impaction. She had been recommenced on PPI therapy for her symptoms shortly before but was still symptomatic. She underwent a barium swallow which showed a mild anastomotic stricture with no hold up of contrast.

Her first gastroscopy at age 4 years 5 months showed a thickened distal esophagus with multiple erosions, mild furrowing and white exudate (Fig. 1).

There was a slight ledge at the site of previous EA repair but no strictures. The lower esophageal sphincter (LES) appeared to be lax. There was no hiatal hernia. Biopsies from the proximal esophagus showed spongiosis and parakeratosis, patchy basal zone hyperplasia and elongation of some lamina propria papillae, accompanied by intraepithelial eosinophils numbering up to 55/High Power Field (HPF). Distal esophageal biopsies showed similar changes with eosinophils numbering up to 80/HPF in addition to

* Corresponding author. Tel.: +612 9382 1752; fax: +612 9382 1787.
E-mail address: Usha.Krishnan@sesiahs.health.nsw.gov.au (U. Krishnan).

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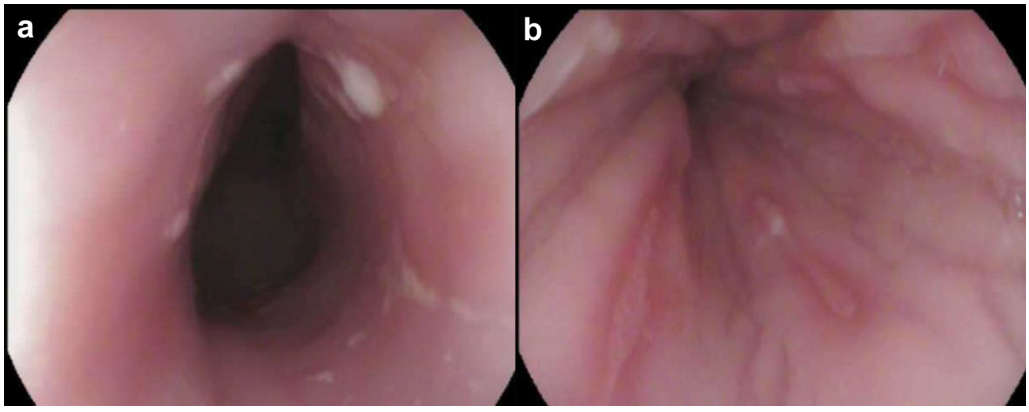

Fig. 1. Initial gastroscopy findings. (**a**) White exudates in the distal esophagus. (**b**) Furrowing with erosions in the distal esophagus.

hyalinization of subepithelial stroma. She was formally diagnosed with EoE. Her PPI was optimized to regular oral pantoprazole 20 mg BD (weight 14 kg) and commenced on budesonide slurry 0.5 mg BD with marked improvement in symptoms.

A progress gastroscopy was performed after 3 months. This showed abnormal salmon colored mucosa suggestive of BE arising 1 cm from the LES at the 12, 6 and 9 o'clock positions, which had not been present previously (Fig. 2).

The furrowing and exudates seen previously had resolved. Four quadrant biopsies taken from the abnormal sites confirmed the histological presence of goblet cells, consistent with a diagnosis of BE intestinal metaplasia (Fig. 3). There was no evidence of dysplasia.

The number of intraepithelial eosinophils however had reduced proximally to 10/HPF and distally to 50/HPF (Table 1).

LK also had her first impedence study which showed an acid reflux index (ACI) of 2.1% on her current treatment. This, along with the detection of BE intestinal metaplasia, led to further optimization of LK's acid suppression from pantoprazole to esomeprazole 30 mg each morning and 20 mg each evening.

Three months later, biopsies from her third follow up gastroscopy showed residual EoE changes with 35 eos/HPF proximally and 24 eos/HPF distally. Gastric metaplasia was reported but this time there was no intestinal metaplasia. The budesonide slurry was increased from 0.5 mg BD to 1 mg BD. Despite adequate fasting there was a significant amount of bile and food in the stomach at time of gastroscopy. Hence LK was commenced on domperidone 5 mg BD as a prokinetic.

Her subsequent progress gastroscopy after three months revealed a moderate hiatal hernia for the first time. In addition, there was macroscopic erosive esophagitis. Biopsies revealed that her EoE was in remission (eosinophil count $<$ 6 per HPF both proximally and distally). Biopsies taken above the Z-line showed only gastric metaplasia, with no sign of intestinal metaplasia. LK also had a repeat impedence study which again showed an elevated acid reflux index of 3.1% (Table 2) despite an increase in her acid suppression therapy three months prior. There was no improvement in the number of retrograde bolus movements (RBM), and LK continued to have prolonged acid reflux episodes and prolonged acid clearance time.

A High Resolution Esophageal Manometry (HREM) was performed. Significant esophageal dysmotility has been reported in all EA patients but HREM has only recently been used to delineate motility patterns [6]. The HREM was performed using a 36 channel solid state catheter (MD Solutions). This showed a hypotensive LES (6 mm Hg) and a moderate sized hiatal hernia. There were three different motility patterns seen with the wet and bread swallows. Peristalsis was seen only in the distal esophageal segment with 69% of the wet swallows and 60% of the bread swallows. Complete absence of peristalsis was seen with 31% of the wet swallows. Pressurizations were seen with 40% of the bread swallows (Figs. 4–6). Based on these results, and the presence of a moderate hiatal hernia and erosive esophagitis, LK is planned for a floppy Nissen fundoplication.

2. Discussion

EoE has only recently been described in patients post EA/TEF repair [7]. This patient required frequent dilatations for her symptoms in the first years of her life; more than would normally be expected even for a post Foker technique long gap repair. This raises the possibility that she may have already developed eosinophilic esophagitis earlier on. Unfortunately as she only had her first gastroscopy at age 4, there are no baseline biopsies to confirm or rule this out. A condition termed PPI-responsive Esophageal Eosinophilia (PPI-REE) has been described [8] however as our patient was already on PPI at the time of diagnosis of EoE it is more likely that she had true EoE rather than PPI -REE.

There have been recent reports in literature of a higher incidence of EoE in EA when compared to the general population. Our own group recently reported a 17% incidence of EoE in EA patients in a cohort of 103 EA patients [4]. One theory is that chronic

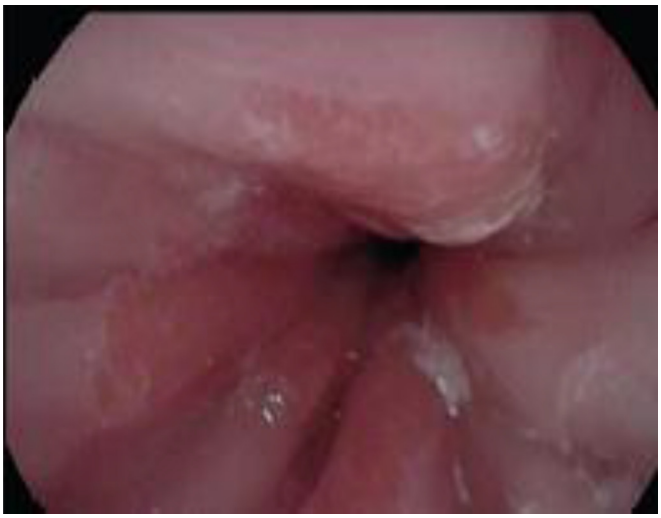

Fig. 2. Second gastroscopy showing abnormal salmon colored mucosa arising from LES.