Case report

Oesophagectomy for achalasia in a child


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Abstract. A 12-year-old girl with a long history of achalasia and numerous unsuccessful attempts at treatment is discussed. Investigation and management by oesophagectomy are described, with a discussion of achalasia in childhood and of the role of oesophagectomy in achalasia.

Key words: Achalasia – Oesophagectomy

Case report

A 12-year-old girl whose symptoms had been present for 10 years presented to us. Her mother recalled that when the patient was 2 or 3 years of age she was not eating properly, was not thriving, and required repeated admissions for "bronchitis". Two years later she developed vomiting and her weight dropped far below the 3rd centile. At this stage achalasia was diagnosed and at age 6 an oesophageal myotomy was performed. Unfortunately, 9 months after surgery the symptoms recurred and at age 7 the procedure was repeated. Once again, after initial relief of symptoms the patient became unwell, and until the age of 11 years repeated dilatations of the oesophagus were required at monthly intervals. The patient's mother reported that her daughter spent almost half her life in hospital. At age 11 a third myotomy was attempted. This was complicated by an oesophageal leak requiring thoracotomy, drainage, and long-term total parenteral nutrition. Approximately 4 months after discharge from hospital symptoms recurred and the patient was transferred to this unit.

When presenting to us, the patient's main complaints were dysphagia with regurgitation and substernal and severe epigastric pain. Multiple surgical scars were present. There had been a 2-kg weight loss over the last 6 months. Barium swallow showed a moderately dilated oesophagus (5 cm) with no peristalsis. Smooth narrowing at the gastro-oesophageal junction was obvious, as was the presence of a Nissen fundoplication. Emptying of the oesophagus was very delayed, with virtually no progress being made in 20 min (Fig. 1). Gastroscopy was performed and showed a dilated oesophagus. The mucosa appeared normal but histology revealed mild oesophagitis. The gastro-oesophageal junction was easily negotiated. Motility studies showed a normal upper oesophageal sphincter and completely amotile oesophageal body. Lower oesophageal sphincter pressure was only 11 mmHg, but there was complete failure of relaxation of the sphincter (Fig. 2). Oesophageal pH monitoring revealed no gastro-oesophageal reflux (Fig. 3).

The above findings indicated persistent achalasia, and in view of this transhiatal oesophagectomy with cervical oesophago-gastric anastomosis was performed. As the patient's oesophagus and stomach had been operated upon several times, surgery was not easy. However, there were no complications and her progress was good. Nasogastric decompression and mediastinal drainage were used for 3 days. On day 7 anastomotic patency and integrity were confirmed by contrast radiology; thereafter feeding was commenced. On day 8 the cervical suction drain was removed and the patient discharged. Surgery provided prompt relief of both pain and dysphagia, and the patient has continued well thus far.

Discussion

When considering our patient's case history, two points may be of interest: firstly, the special features of achalasia in childhood, and secondly, the role of oesophagectomy in achalasia.

Achalasia is a rare disease (roughly 1 case per 100,000 population) that is particularly rare in children. Approximately 5% of patients present before the age of 15 years [1], and most paediatric surgical centres report experience of only 1 case per year. This alone makes diagnosis difficult, but the difficulty is compounded by the atypical presentation of achalasia in childhood. Particularly in young infants, pulmonary symptoms due to aspiration as well as failure to thrive dominate the clinical picture. Once the physician's suspicion has been aroused, however, the diagnosis of achalasia can be confirmed easily. Barium swallow is usually diagnostic and oesophageal manometry can be performed in older children.

The aetiology of achalasia in childhood may be of interest. Whereas the vast majority of cases are idiopathic, two rare familial syndromes may present with childhood achalasia: familial dysautonomia (Riley-Day syndrome) and Allgrove's syndrome. In the former achalasia is associated with hyperhidrosis, failure of lacrimation, labile blood pressure, instability of temperature control, relative
insensitivity to pain, and emotional lability. In the latter achalasia is combined with failure of lacrimation and glucocorticoid deficiency and frequently results in hypoglycaemia with convulsions and mental retardation.

In the treatment of childhood achalasia, the same modalities that have been used in adults have been tried. Bouginage has failed in both children and adults, and is particularly unsuitable as general anaesthesia is usually required [1, 9, 11]. Pneumatic dilatation has been used in children. Technical problems may be experienced in the very young, but those centres that have overcome the problems intrinsic to the apparatus have reported good results in the majority of cases [1, 5]. The most commonly used treatment modality in children remains the modified Heller myotomy [1, 2, 4–6, 10, 11]. Most authors prefer the abdominal to the thoracic approach in paediatric patients [2, 6, 10].

Fig. 1. a X-ray film showing dilated oesophagus with smooth, tapered narrowing of its lower end, characteristic of achalasia. b Oesophagus narrows distally. A diverticulum is seen at junction of middle and lower thirds. c Fundus of stomach is seen encircling lower end of oesophagus, evidence of a previous Nissen fundoplication.