Duplication of the vermiform appendix in an adult patient

KH Lim

Kyungpook National University Hospital, Daegu, Korea

ABSTRACT

Duplication of the appendix is extremely rare. A 69-year-old woman was admitted with a 2-day history of right lower quadrant abdominal pain. Physical examination was consistent with acute appendicitis. Ultrasonography and colonoscopy gave a clinical impression of an inflammatory appendiceal mucocoele. Operative findings were an enlarged and inflamed appendix with distal cystic changes. Laparoscopic wedge resection of the caecum was performed. A tubular structure with a true lumen was found in the appendix. Haematoxylin and eosin staining and trichrome staining showed both structures had a true mucosa and a muscular layer. The duplication in this case does not belong to any of the previously described types of duplication.

KEYWORDS

Appendix – Duplication – Mucocoele – Intussusception

Accepted 1 August 2013; published online XXX

CORRESPONDENCE TO

Kyoung Hoon Lim, Department of Surgery, Kyungpook National University Hospital, 5 Samduk-dong 2ga, Jung-gu, 702-210 Daegu, Korea
T: +82 (0)53 420 5605; F: +82 (0)53 421 0510; E: drlimkh@naver.com

Duplication of the appendix is extremely rare. The reported incidence is 0.004–0.009% (approximately 80 reported cases).^{1,2} Preoperative diagnosis of appendiceal duplication is often difficult and it is usually made at surgery. Almost all cases are detected in childhood and they are almost always associated with major malformation of the intestinal tract, genitourinary system or vertebrae. In adults, appendiceal duplication is typically an incidental finding during a laparotomy for another indication. We report a unique case of an adult who presented with appendicitis occurring in a duplicate appendix.

Case History

A 69-year-old woman was admitted with a 2-day history of right lower quadrant abdominal pain associated with nausea and diarrhoea. She had undergone no previous abdominal surgery. Physical examination revealed an afebrile patient with tenderness and rebound tenderness in the right iliac fossa, consistent with the signs of acute appendicitis. Ultrasonography showed a cystic lesion with a true wall layer and hypervascularity at the tip of the lesion (Fig 1). The white blood cell count was 16,710/ μ l (77% neutrophils). Colonoscopy revealed a protruding, hyperaemic and oedematous appendiceal orifice (Fig 2A).

With a clinical impression of an inflammatory appendiceal mucocoele, a laparoscopic exploration was undertaken. Operative findings were an enlarged and inflamed appendix with distal cystic changes. Laparoscopic wedge resection of the caecum was performed because of

oedematous changes at the appendiceal base. Grossly, there was a tubular structure with a true lumen in the appendix (Figs 2B–D). Haematoxylin and eosin staining and trichrome staining showed both structures had a true mucosa and a muscular layer (Fig 3). No postoperative complications occurred.

Discussion

Appendiceal duplication is rare. Cave was the first to describe the three types of duplication.³ This classification was modified by Wallbridge in 1962.⁴ Type A consists of various degrees of partial duplication on a normally located appendix with a single caecum. Type B comprises a single caecum with two completely separate appendices, divided into two subgroups: type B1, where two appendices are located symmetrically on either side of the ileocaecal valve, resembling the normal arrangement in birds (the ‘bird-like type’), and type B2, where a normally located appendix arises from the caecum at the usual site and a second separate rudimentary appendix is located along the line of one of the taenia (the ‘taenia coli type’). Type C consists of a duplicated caecum, each with an appendix. When anomalies are detected in childhood, they are almost always associated with several malformations; they are usually type B1 and C duplications.^{5–7}

Initially, our case appeared to be a usual, albeit enlarged, appendix. However, there was another tubular structure in the enlarged appendix. Moreover, both structures had intact mucosal and muscular layers. In a patient with a single

Figure 1 Ultrasonography showing a cystic lesion with a true wall layer and hypervascularity at the tip of the lesion

Figure 3 Haematoxylin and eosin stain (A) and trichrome stain (B) showing both structures had a true mucosa and a muscular layer (40× magnification)

appendix, our case would have been a subclass of type A but this case had two independent orifices to the caecum. As a result, it does not belong to any of the previously described types of duplication.

This entity should be categorised as separate to a caecal diverticulum or appendiceal diverticulosis.^{4,8} The wall of a caecal diverticulum does not involve any lymphoid tissue microscopically. Appendiceal diverticulosis is seen in up to 2.1% of specimens and in 1% of appendicectomies.^{8,9} The majority are acquired pseudodiverticula while true congenital diverticula are very rare. Congenital diverticula are true diverticula because they contain all the layers of the appendiceal wall, which include the mucosa, submucosa, serosa and, most crucially, the muscle layer. Acquired diverticula are pseudodiverticula because only the mucosa, submucosa and serosa are included in the diverticulum wall, without the muscle layer.¹⁰ This difference is important as congenital diverticula, owing to their thicker muscular wall, are more resistant to perforation than acquired diverticula.¹¹

This case was also distinct from appendiceal intussusceptions. Appendiceal intussusceptions have been classified into five types.¹² Our case was different on several points: the appendiceal tip was not invaginated; externally, the appendix was smooth without inflexion; and a complete tubular structure existed inside the resected appendix (Fig 2).

Figure 2 Colonoscopy revealed a protruding, hyperaemic and oedematous appendiceal orifice (A), and, grossly, there was a tubular structure with a true lumen in the appendix (B, C and D).

Conclusions

Although appendiceal duplication is rare, surgeons should be aware of the possibility, especially when clinical signs and symptoms are consistent with acute appendicitis, and the appendix appears normal at surgery. Failure to recognise this condition may have serious clinical and medicolegal consequences. In patients with appendiceal duplication, when only one of the appendices is found to be inflamed on exploration, both should be removed in order to avoid diagnostic confusion that may arise on removal of a single appendix.¹³

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