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Alimentary tract duplications in children: Report of 16 years' experience

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ABSTRACT

Background: Alimentary tract duplications (ATDs) are a rare condition in children, characterised by a large pathogenic, clinical, and histological polymorphism. Surgical observation and pathologic evaluation of the resected specimens are the only way to confirm the diagnosis. In this study, we want to analyse the anatomical, clinical and therapeutic aspects of this entity. **Patients and Methods:** A total of 12 cases of ATD were diagnosed over a 16-year period at paediatric surgery department. The diagnosis was evoked on clinical and radiological data. Histological study of the resected specimens confirmed the diagnosis in all cases. **Results:** The mean age of patients at diagnosis was 41 months with a peak of incidence at the 1st year of life (42%). Out of a total 12 cases, 10 were girls and 2 were boys. Abdominal pain and vomiting were the most frequent presenting features. Ultrasonography, tomodensitometry and magnetic resonance imaging were useful for diagnosis. ATDs were localised on the oesophagus in one case, the stomach in one case, the duodenum in four cases, the ileum in five cases, and the colon in one case. All these duplications were cystic, with three communicating duplications. All patients underwent surgery, and resection procedure was chosen according to duplication type and site. Histological study confirmed the diagnosis in all cases. **Conclusion:** ATDs are a rare condition in children. Diagnosis relies on histology, and treatment can only be by means of surgery. The outcome after surgery is generally favourable. Diagnosis and precocious surgery of ATDs can warn serious complications.

Key words: Alimentary tract duplications, children, surgery

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INTRODUCTION

Alimentary tract duplications (ATDs) are uncommon congenital lesions that can occur anywhere from the mouth to the anus and have a reported incidence of 1 in 4500.[1,2]

Enteric duplication cysts are hollow, epithelium-lined, cystic, spherical, or tubular structures that are basically attached to the wall of the gastrointestinal tract (often sharing the serosa) and supplied by common mesenteric blood vessels.[3]

Of the cases, 70-80% present within the 1st year of life.[4-7] The signs and symptoms depend on the type and location of the duplication. Duplication of the hindgut may be clinically silent, but it presents most often with constipation, obstruction, or abdominal distension.[4-7] Less commonly, vomiting, diarrhoea, and rectorrhagia are present.[4-7] Although ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) are frequently used pre-operatively for diagnostic imaging, the correct pre-operative diagnosis is made in <25% of cases.[8] Definitive diagnosis of ATD is based on surgical observation and histological study.

In this study, we want to analyse the anatomical, clinical and therapeutic aspects of this entity.

PATIENTS AND METHODS

It is a retrospective study of 12 cases managed at Paediatric Surgery Service in Habib Thameur Hospital, Tunis, over a 16 year period between January 1997 and December 2012.

Data collection included information on patient demographics, the location of ATD, patient signs and symptoms, the investigations used in the diagnosis, the treatment provided, and the pathologic evaluation of the resected specimens.

RESULTS

The median age was 9 months, and the mean age was 3.42 ± 4.82 years (range: 3 days to 16 years). The male-female ratio was 1:5. Nearly 42% of patients were under 12 months of age. The location and frequency of ATD are listed in Table 1. The ileal and duodenal areas were the most frequent sites of duplication, harbouring 41.7% and 33.3% of these lesions, respectively. The clinical presentation of these lesions was polymorphous and 10 children (83.3%) had gastrointestinal symptoms: Five patients (41.6%) presented with abdominal pain, three patients (25%) presented with vomiting, one patient (8.3%) presented with bowel obstruction and one patient (8.3%) presented with melena. Only one patient (8.3%) had a prenatal diagnosis suggestive of ATD. The diagnosis was made in the neonatal period in one case (8.3%); this patient was a 16-hour-old infant female neonate with "respiratory distress," which later proved to have oesophageal atresia associated with an oesophageal duplication. The radiologic studies performed were as follows: Plain abdominal radiogram (n = 4), plain thoracic X-rays (n = 1), US (n = 11), CT (n = 7), MRI (n = 1), colonoscopy (n = 1), barium enema (n = 1), upper gastrointestinal series (n = 1) and Meckel's scan (n = 1). A gasless area at the cyst location was observed on plain abdominal radiograms in one newborn patient. Imaging studies showed a cystic mass in seven patients [Figures 1 and 2]. US showed thick-walled cysts in six patients and intense, particulate cystic contents in three patients. A septated cystic appearance was detected by US in only one patient.

All patients underwent surgery. The surgical approach was a midline laparotomy for gastrointestinal duplications and a right posterolateral thoracotomy in the case of oesophageal duplication. The diagnosis of ATD was only made at the time of pathologic evaluation of the resected specimens.

All digestive duplications were cystic, with three communicating duplications. Surgical treatment consists of:

- Complete resection of the duplication followed by end-to-end anastomosis in the case of gastric duplication, the five cases of ileal duplication and two cases of duodenal duplication.
- Partial resection of the cyst wall and mucosal stripping in two cases of duodenal duplication that was close to the major duodenal papilla.
- Right hemicolectomy taking away the duplication followed by end-to-end anastomosis in the case of stenotic colic duplication. This patient had a colonoscopic examination revealing a suspicious lesion in the ascending colon.
- Complete resection of the oesophageal cystic non-communicating duplication followed by surgical treatment of oesophageal atresia in the case of oesophageal duplication with oesophageal atresia.

The pathologic evaluation of the resected specimens confirmed the diagnosis of ATD in all patients. One patient (8.3%) with ileal duplication had ectopic gastric

Table 1: Location and frequency of alimentary tract duplications

Location	No. of patients	Frequency (%)
Oesophagus	1	8.3
Stomach	1	8.3
Duodenum	4	33.3
Ileum	5	41.7
Colon	1	8.3

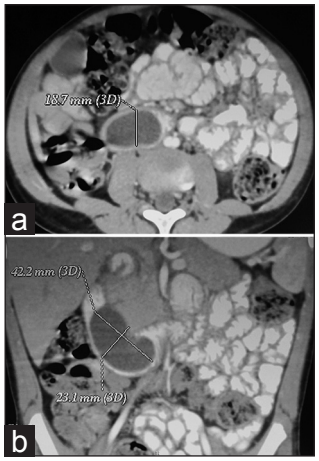


Figure 1: (a) Axial and (b) coronal computed tomography scan images showing a duodenal cystic mass in intimate contact with the head of the pancreas

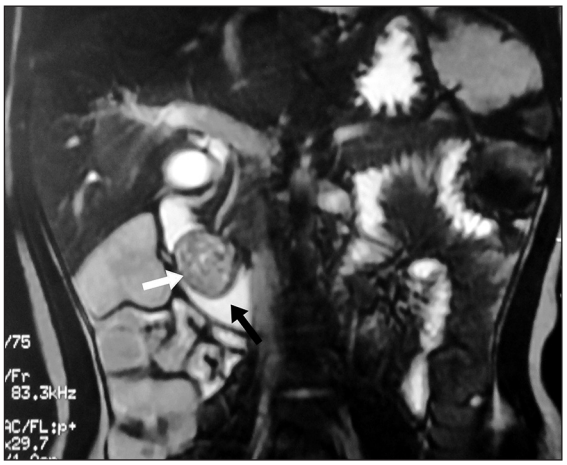


Figure 2: Coronal magnetic resonance imaging image in a 5-year-old girl showing a regular cyst (white arrow) in the duodenal lumen (black arrow), 26 mm × 29 mm in size, close to the major duodenal papilla

mucosa. The post-operative period was uneventful in all patients.

DISCUSSION

ATDs are rare congenital abnormalities. The clinical presentation or onset symptoms of this malformation range from infancy and early childhood to adulthood.[4-7] Of the cases, 70-80% presented within the 1st year of life.[4-7] Although there are some reports that duplications are more common in males, most authors agree that duplications occur more commonly in females.[9-11]

The precise aetiology of these anomalies is still unclear. Several theories have been proposed including the aberrant luminal recanalisation theory, split notochord theory, diverticular theory, and environmental factors.[12]

In our study 75% of duplications occur in the small bowel. Duplications found in the small intestine are the most frequent site of enteric duplications and most commonly occur in the ileum.[13,14] These usually share a common wall with the normal intestine and have a common blood supply so it is mandatory to remove the adjacent bowel segment along with duplication cyst. This is in contrast to Meckel's diverticulum that occurs on the antimesenteric border of the ileum.[13]

The signs and symptoms depend on the type and location of the duplication. Clinical findings in patients with a duplication include recurrent abdominal pain, vomiting, abdominal distension, intestinal obstruction, palpable abdominal mass, failure to thrive, constipation, and occasionally bleeding from ectopic gastric mucosa.[1,7,11,15] Nonetheless, duplication cysts can sometimes be detected incidentally. Complications such as extrinsic obstruction, volvulus, intussusceptions, perforation, and bleeding from ulceration may occur.[4,5,7]

Only one patient in this series had a prenatal diagnosis suggestive of ATD. The high definition provided by modern imaging techniques allows the delineation of foetal anatomy with unprecedented accuracy. The early prenatal detection of enteric duplications is possible and facilitates a prompt postnatal treatment strategy, thereby, decreasing the risk of potential complications.[16]

In this study, diagnostic studies, including US, CT, and MRI, were useful in establishing the pre-operative diagnosis. The most common modality used to image duplication cysts is US. US plays a critical role in the diagnosis of duplications and may reveal the inner hyperechogenic and outer hypoechogenic layers (double-wall sign) that are typical of duplication cysts.[17] CT is also a useful study to delineate anatomical and vascular attachment as well as cyst content. It provides better information than US regarding the exact extension and margins of cystic masses.[18] MRI is another diagnostic method that can determine the cyst and its borders clearly.[19] Although US, CT, and MRI are frequently used pre-operatively for diagnostic imaging, the correct pre-operative diagnosis is made in <25% of cases.[8] Definitive diagnosis of ATD is based on surgical observation and pathologic analysis of the resected specimens.

Duplications should be treated for local control of symptoms including obstruction, bleeding, and possible septic sequelae and to prevent the risk of malignant change. The ideal treatment of ATDs is complete resection, which is usually accomplished by resection of the adjacent bowel and mesentery. However, in the case of completely isolated duplication cysts, resection can be accomplished safely without requiring bowel resection.[3] On occasion, long tubular duplications will necessitate removal from within the mesentery by dividing vessels on one side of the mesenteric bilayer to preserve blood supply to normal bowel.[20] In some cases of duodenal duplication, due to the close proximity of the cyst to the major papilla, complete resection may not be safe because there is a possibility of damaging the biliary and pancreatic ducts. Alternatively, partial resection, marsupialisation, mucosal stripping, and internal or Roux-en-Y drainage have been reported.[21,22] This was the case for two of our duodenal duplications.

CONCLUSION

Duplications of the alimentary tract are a rare condition in children. The clinical presentation varies greatly and imaging studies may be useful in establishing the pre-operative diagnosis. Diagnosis relies on histology, and treatment can only be by means of surgery. The outcome after surgery is generally favourable. Diagnosis and surgical precocious treatment of digestive duplications can warn serious complications.

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