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Review Article

The Small Intestinal Duplications in Children

Lassaad Sahnoun*, Kechiche N, Ksia A, Mekki M, Belghith M and Nouri A

Department of Pediatric Surgery, Fattouma Bourguiba Hospital, Faculty of Medicine, Monastir, Tunisia.

***Corresponding Author**: Lassaad Sahnoun, Department of Pediatric Surgery, Fattouma Bourguiba Hospital, Monastir, Tunisia.

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Abstract

The small intestinal duplication is rare congenital malformation that may vary greatly in presentation, size, location, and symptom.

Objective: To review the clinical presentation, etiopathogeny and treatment modalities, through our observations and an overview of literature.

Study Design: retrospective chart review including twenty observations of duplications of the jejuno-ileum collided between 2000 and 2015 in Monastir department of pediatric surgery. Medical charts were examined for clinical presentation and treatment modalities.

Results: Our cohort was 20 children (11 male, 9 female) with small intestinal duplication with a median age of 10 months (ranged from 1 day to 4 years). The clinical features were polymorphous but remained dominated by a digestive symptomatology. The abdominal ultrasound was conducted in 18 patients and made a significant contribution to the diagnosis by showing in 12 cases pictures of cystic with double tunic strongly suggestive of the diagnosis. All our patients were operated, an open surgical procedure was undertaken in 75% of the cases, where a laparoscopic assisted procedure for a first look was practiced in 25% of cases. The surgical treatment consisted in a total mass resection with an end-to-end anastomosis in 15 patients, where enucleation of the cystic mass was undertaken in 4 patients. In one patient, the excision of the duplication was subtotal and the residual mucosa was peeled off. The postoperative recoveries were uneventful in 85% of the patients. The average follow-up was 2 years and 8 months; it was satisfying in all cases.

Conclusion: The small intestinal duplication is a rare congenital lesion. Surgical resection must be undertaken before the occurrence of the complications.

Keywords: *Enteric Duplication; Intestinal Obstruction; Surgery*

Introduction

The small-intestinal duplication is a rare congenital malformation, which is characterized by a very large clinical and histopathological polymorphism.

It represents about the half of the digestive duplications. Most authors excluded the duodenal duplications from the small-intestinal duplications, regarding their clinical features as well as their therapeutic particularities.

The diagnosis of the small-intestinal duplication is suspected on the clinical status and strongly evocated on the radiologic investigations data especially at the abdominal ultrasound. The diagnosis is often done per-operatively and requires histological confirmation.

This disorder raises a diagnostic problem, seeing the diversified symptomatology, and a therapeutic controversy concerning the timing of surgery and the operative technique notably for the stretched tubular forms.

Through our observations and an overview of literature, we propose to remind the etiopathogenic, epidemiological.

Patients and Methods

This is a retrospective study including twenty observations of duplications of the jejuno-ileum collided between 2000 and 2015 in Monastir department of pediatric surgery.

A chart review was conducted to collect the different clinical features, the imaging data, as well as the operative procedures and the histopathological reports.

The small-intestinal duplications were double in five cases, achieving a total of 25 duplications with 20 patients.

Results

Our patients age varied between one day and four years old with 75% of children aged less than one year. The average age was of 10 months.

A slight male dominance was noted with 11 boys and 9 girls.

The prenatal ultrasound suspected the diagnosis in 5 children belonging to our list.

The clinical features were polymorphous but remained dominated by a digestive symptomatology. Thus 9 cases presented an occlusive syndrome made of bilious vomiting and abdominal distension. Two patients had isolated abdominal pain while two others had profuse proctorrhagia. The diagnosis was fortuitous for two other patients.

The clinical examination showed an abdominal mass of variable size and consistency in six patients. It revealed an abdominal bloating in three patients (15%) with an occlusive syndrome and an abdominal sensibility in two other children (10%).

The additional explorations were requested according to the clinical context. The abdominal radiography was realized with 14 patients (70%) and showed hydroaeric levels in four patients and repressed small intestines in three other patients.

The abdominal ultrasound was conducted in 18 patients (90%) and made a significant contribution to the diagnosis by showing in 12 cases pictures of cystic mass either plain or bilobate with double tunic strongly suggestive of the diagnosis (Figure 1).

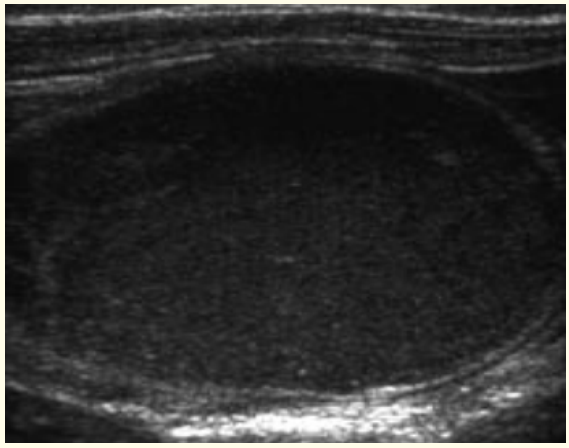

Figure 1: *Abdominal ultrasound showing anechogen mass with double tunic.*

Elsewhere, the abdominal ultrasound showed liquid formation without suspecting the intestinal duplication in two cases, a digestive distension and a peritoneal effusion in two other cases and an acute intestinal intussusception in one case. It showed no anomalies in the last case.

The abdominal CT scan was conducted in 9 patients. It objectified intra-peritoneal cystic or tubular masses of different size and helped suspect the diagnosis in these 9 cases. The CT scan showed a thickened intestinal wall in 4 cases, an intimate contact with intestinal loops in 4 other cases and a wall enhancement with the injected scan in one patient.

A digestive scan with injection of marked technetium (99m) was practiced in two patients having rectal bleeding of high abundance and which objectified in both cases increased uptake focal points of an early development and an increasing intensity; one projecting under the umbilicus and the other to the left flank.

The digestive opacifications were not commonly used in our series. We had recourse to it with only one patient; yet this examination showed the absence of communication between the cystic mass and the adjacent intestinal segment.

Associated malformations were observed with 5 patients (25%). A pyelo-ureteral junction syndrome at a minimal level was noted with an infant, a volvulus complicating an intestinal malrotation was found with two children, an inguinoscrotal and umbilical hernia affected one child and a type III small-intestinal atresia affected another.

All our patients were operated; an open surgical procedure was undertaken in 75% of the cases, where a laparoscopic assisted procedure for a first look was practiced in 25% of cases (Figure 2).

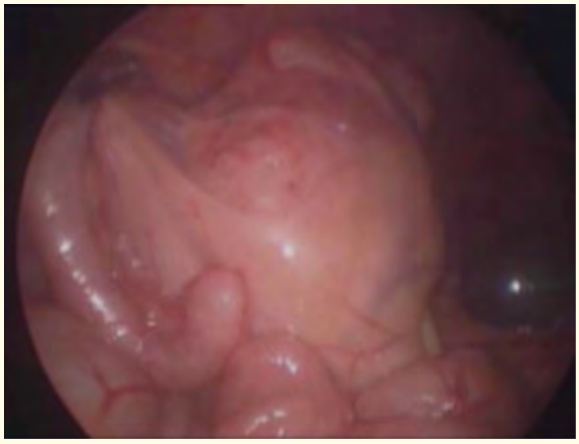

Figure 2: *Laparoscopic exploration of cystic ileal duplication.*

Among our 20 patients, 80% had duplication of ileal seat and only 20% had jejunal duplication.

There were no communication between the normal and the duplicated loops in all cases and the duplications were located on the mesenteric side of the loop in 90% of the cases. Two duplications were located in the antimesenteric portion of the loop.

The duplications were cystic in 19 cases (including 4 jejunal cystic duplications); in the other 6 cases the duplications were tubular (Figure 3).

The surgical treatment consisted in a total mass resection including a portion of the duplicated ileal loop with an end-to-end anastomosis in 15 patients (18 duplications), where enucleation of the cystic mass was undertaken in 4 patients (6 duplications). In one patient, the excision of the duplication was subtotal and the residual mucosa was peeled off.

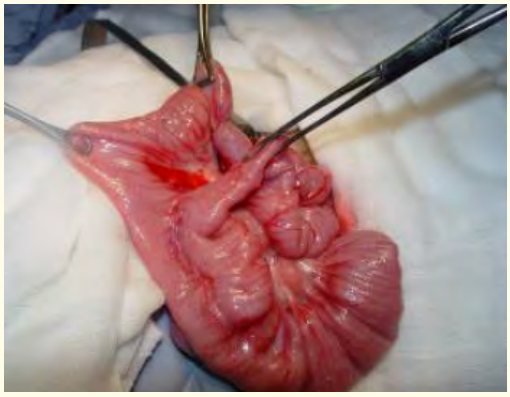

Figure 3: *Tubular ileal duplication.*

The postoperative recoveries were uneventful in 85% of the patients. An anastomotic leakage was observed with two children and an acute postoperative intestinal intussusception appeared in one case. The three patients were reoperated and their evolution was favorable.

The average follow-up was 2 years and 8 months; it was satisfying in all cases.

Discussion

The small-intestinal duplication is a rare pathology that occurs at an early age without gender selection. Its embryopathology remains very controversial. The small-intestinal seat (duodenum excluded) constitutes the most frequent localisation of the entire digestive duplications [1-5]. It represents half of the complied digestive duplications in our department during the same study period.

The intestinal duplication symptomatology is often polymorphous. It varies according to the seat, the size, the communicating character or not of the duplication and the possible heterotopias of the gastric or pancreatic lining. The antenatal discovery is possible when it concerns a voluminous cystic form enabling thus an early management before the installation of complications [6]. This was noted with 25% of our patients.

This pathology can be revealed by trite signs or by a noisy symptomatology in case of a complication. Furthermore, the discovery can be fortuitous [7-9]. In our study, an occlusive syndrome was revealing in 45% of the cases and proctorrhagia in 10% of the cases.

The radiologic additional examinations are non-specific and no exploration can allow a certain diagnosis on its own [10-13].

Sometimes, the diagnosis of intestinal duplication is only made during the surgical exploration and confirmed after a histopathological examination.

Nevertheless, the abdominal ultrasound remains the main radiologic examination; especially in the cystic forms where we can see an anechogenic mass with a wall made of a hyperechogenic internal layer (mucous membrane) and a hyperechogenic external layer (muscular coat) [11].

This aspect is highly evocative of the diagnosis but non-specific. The abdominal ultrasound was practiced in 18 patients of our series.

The rest of the radiological explorations (tomodensitometry, scintigraphy, MRI and digestive opacifications) can bring additional informations enabling to guide the diagnosis.

The confrontation of the different radiological examinations together with the clinical elements can highly evocate the diagnosis of small-intestinal duplication [11,14,15].

The association of the intestinal duplication with other malformations is possible but rare. The small intestinal atresia is the most common inter-related malformation [16,17].

Some complications like the intestinal volvulus, an acute intestinal intussusception, a perforation or a digestive haemorrhage can be seen during the evolution of the duplication or be reveal it [18]. The digestive perforation and haemorrhage are often related to the acid secretions due to the existence of heterotopic mucous membrane in half of the cases of intestinal duplication.

The DG treatment is surgical. The progress made in laparoscopic surgery allowed a significant shift in their diagnostic and therapeutic care [19].

The resection should be as radical as possible. It depends of the seat, of the morphology, of the type, of the vascularization and of the relations of the duplication with the underlying digestive tract. The most frequently used gesture is the duplication as well as the concerned intestinal portion excision with end-to-side anastomosis. If the gesture is revealed to be damaging or at risk of complication, and if there is a cleavage plane between the DG and the carrying intestinal loop, we can opt for an enucleation but it's not always possible to implement. The mucosectomy is advised when the complete excision might pose serious problems. It is the case for the extended tubular duplications with risk of short bowel syndrome [9,15].

In our series, the laparoscopy was practiced in 5 patients for the first look, and then conversion with a trans-umbilical incision (in four cases) and mini-laparotomy (in one case) was realized. The surgical procedure consisted in a resection of the duplication and an end-to-end anastomosis in 18 cases, an enucleating of the duplicated cyst in 6 cases and in a subtotal resection with mucosectomy in one case.

The histopathological study of the duplication allows us to confirm the diagnosis [20]. The results of the surgery of this pathology are generally good, with a favorable evolution.

The mortality rates depend on the occurrence of complications, and on the associated malformations, especially when they are severe.

Conclusion

The small intestinal duplications are rare malformations with a large anatomoclinical polymorphism.

The diagnosis must be evocated in case of intestinal occlusion or abdominal mass during the neonatal period, and the treatment undertaken before the occurrence of the complications.

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