CASE REPORT

Amyand's Hernia in a 9-Month-Old Infant: Case Report & Review of the Literature

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ABSTRACT:

Amyand's hernia is defined as the presence of an appendix inside the sac of inguinal hernia whether inflamed or not, it is very rare occurring in less than 1% of patients of inguinal hernia surgery. Although it was first described three centuries ago in an 11-year-old boy by Claudius Amyand, it still continues to possess some fuzziness regarding its diagnosis & treatment because of the scarcity of its occurrence. It has variable clinical presentation according to the extent of appendix inflammation & related complications. The diagnosis of an Amyand's hernia is difficult to settled clinically. However, imaging studies are beneficial for both diagnosis and recognition of the associated complications. Here, we report a case of Amyand's hernia in a 9-month-old male infant discovered incidentally during surgery for right sided inguinal hernia in the department of pediatric surgery in the Central Child Teaching Hospital in Baghdad in which we did reduction of a normal appendix into abdominal cavity & herniotomy with high up transfixation of hernia sac.

The purpose of presenting this case report is the rare occurrence of Amyand's hernia & the possibility of encountering an unusual content of hernia sac during the repair of inguinal hernia, that is why Amyand's hernia should be taken in consideration while operating on obstructed or strangulated inguinal hernia. The surgeon need to be aware of all clinical presentations with which the Amyand's hernia may present and the suitable, individualized option of surgical treatment applied. In this paper we review the literature on presentation of Amyand's hernia and discuss the diagnostic modalities with the current options of its surgical treatment.

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INTRODUCTION:

The repair of an inguinal hernia in children is very common operation done in surgical practice, but even experienced surgeons may encounter technical difficulties & a critical decision need to be done when they faced with an unexpected peroperative finding in the form of uncommon contents of hernial sac. In majority of cases, bowel or omentum is the content of hernial sac but in some cases unusual contents can be found intraoperatively like ovary, fallopian tube, urinary bladder, colonic diverticula, Meckel's diverticulum or very rarely appendix. The presence of an appendix in the sac of an inguinal hernia is called ''Amyand's hernia'' irrespective of its situation (normal, inflamed, perforated, or gangrenous) [1,2]. This very rare condition offers variation in their clinical presentation and respective management.

Garengoet was the first who report a case of an appendix in a hernia sac in 1735, he identified an appendix within a femoral hernia, this was followed by the detection of an appendix within the inguinal hernia by Claudius Amyand in the same year [1]. Amyand was a French surgeon working in St George's Hospital in England. On December 1735, he performed the first reported appendectomy on a boy aged 11 years who had a perforated, severely inflamed appendix within the sac of right inguinal hernia. The appendix was perforated by a previously swallowed pin, leading to development of an enterocutaneous fistula in the scrotum. The patient survived, but the hernia recurred [1].

The prevalence of Amyand's hernia in general poulation is 0.4–0.6 % among all cases of inguinal hernias but higher in children reaching approximately 1 % [2]. Most cases of Amyand's hernia occur on right side due to the normal anatomical position. Nevertheless, left-sided Amyand's hernia also have been reported & is usually attributed to the associated congenital anomalies like situs inversus, malrotation of

midgut & mobile cecum [3]. Amyand's hernia is more common in male patients [4] & commonly an indirect hernia, although direct Amyand's hernia has been also reported infrequently [5]. The incidence of appendicitis within an inguinal hernia in children is extremely rare; with an estimated rate at 0.07–0.13% [6].

The clinical features of an Amyand's hernia is variable depending on the degree of inflammation and whether it presents with or without a perforation. Hence, it may be presented as normal or complicated inguinal hernia like obstruction, strangulation or acute scrotum. since there are no specific findings with Amyand's hernia it may mimic epididymo-orchitis, testicular torsion and inguinal lymphadenitis [3]. Preoperative diagnosis of Amyand's hernia is not straight forward & often difficult due to indistinct clinical features and a lack of clear diagnostic radiological features, so it is generally an incidental finding during surgery [3, 7]. However, imaging studies like ultrasound & CT scan may be valuable for both diagnosis and detection of the associated complications [5].

Treatment of this condition involves primary surgical repair of the hernia with or without appendicectomy [3,5,7]. Surgical options for the appendix in Amyand's hernia depend on its inflammatory state. While some studies believe in the necessity of an appendectomy even when the appendix is not inflamed to avoid future complications [8], others encourage for doing appendectomy only in case of inflamed appendix [9,10].

CASE REPORT:

Herein we present a case of 9-month-old male infant with incarcerated irreducible right sided inguinal hernia, his family gave a history of recurrent swelling in the right inguino-scrotal region dated back 6 months ago, it was diagnosed as simple inguinal hernia & surgical repair was offered on elective basis but this was delayed because his family was immigrant for long time due to military conflict. The hernia was reducible until the last week when the swelling became irreducible but without history of vomiting, fever, abdominal distension or constipation. The infant was admitted to the ward of pediatric surgery in the Central Child Teaching Hospital in Baghdad. He was stable & his vital signs were within normal limits at time of presentation, his abdomen was flat with no tenderness. Local examination revealed a firm, non-tender swelling in the right inguinal region extending into the scrotum, it was irreducible with normal overlying skin. The right testis was seen high up in upper scrotum, the left scrotum and testis were normal. The swelling couldn't be reduced manually so the patient was planned for inguinal exploration. No radiological studies were required as usual in cases of inguinal hernia & surgical exploration was performed after routine laboratory tests like Hb level, chest X-ray & hepatitis screen. During surgery, a classic right inguinal crease incision was created and a big hernial sac was identified. On opening the hernial sac, an appendix was found adherent to the sac (Fig.1) with a fibrous cord connecting its tip to the testis (Fig.2). Since the appendix was normal, an appendectomy was not performed, therefore we released it from its attachment & reduced to abdominal cavity (Fig.3) then the hernial sac was dissected free from the spermatic cord, and a right herniotomy was completed with high ligation of the sac (Fig.4).

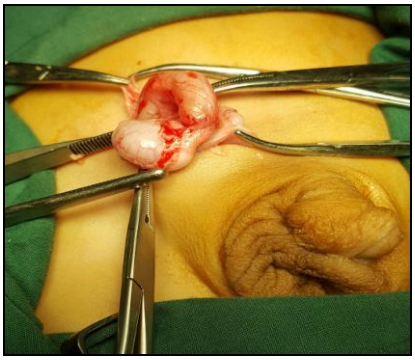

Fig.1: Appendix attached to testis.

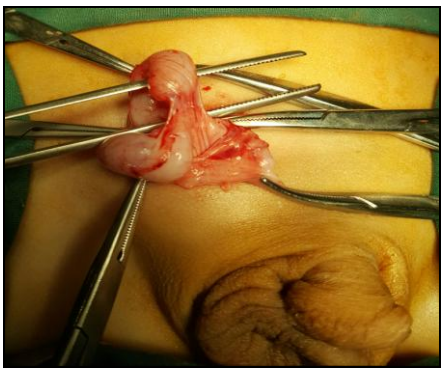

Fig.2: Fibrous band connecting appendix to testis.

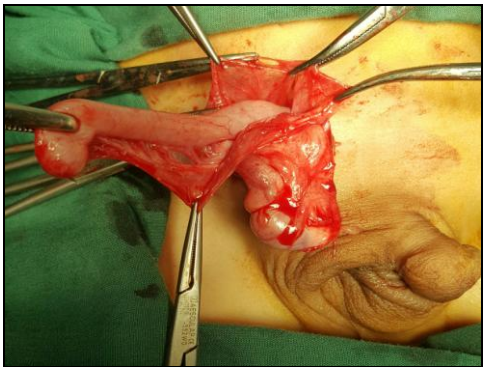

Fig.3: Release of appendix inside hernia sac. **Fig.4: Herniotomy after reduction of appendix.**

The testis then easily reached the base of scrotum as it was attached to the appendix preventing its complete descent. Postoperative course was uneventful. The patient was discharged on the second day with good general condition, normal vital signs & normal bowel motion.

DISCUSSION:

The age of patient with Amyand's hernia has been reported in the range from 3 weeks to 92 years [3]. Its incidence in children is three times more than adult due to patency of the processus vaginalis in the pediatric population [11]. Additionally, Amyand's hernia in children less than 1-year-old represents 2% of the total cases of appendicitis [12]. Moreover, the incidence of perforated appendix incarcerated within an inguinal hernia is too rare constituting less than 0.1% of all cases of appendicitis [13]. D'Alia et al. in their study examined 1341 cases of inguinal hernias and they find out that the incidence of Amyand's hernia is 0.6%, always on the right side, and only in males [14]. We hereby present a case of Amyand's hernia discovered incidentally at the time of repair of right sided inguinal hernia in male infant.

The pathophysiology of Amyand's hernia and its relationship with appendicitis are unknown [12]. However, many authors believe that there is an association between incarceration of the appendix in the inguinal canal and its inflammation [3,5,15]. It is thought that the appendix of Amyand's hernia in contrast to an anatomically normal appendix is more likely to become inflamed, since it is vulnerable to trauma, adhesions and jeopardized blood supply, followed by inflammation and bacterial overgrowth [3]. Additionally, the appendix may also be compressed by the contraction of the abdominal muscles resulting in further inflammation [5]. A congenital band extending from the appendix into the scrotum and attached to the right testis & the funnel-shaped tapering cecum of the neonate & infants are two more possible pathogenetic factors [16]. We noticed such a fibrous band connecting the tip of appendix to right testis in our case (Fig.2). Many authors reported this fibrous connection [5,16] & they believed that this could lead to guidance of the appendix and passage through the inguinal canal especially in the presence of patent processus vaginalis.

Appendix in an Amyand's hernia may develop inflammation, infection, perforation, or it may be just incarcerated but entirely healthy [3]. Common complaints in adult patients include sudden-onset of central abdominal pain with localized tenderness in the right iliac fossa, combined with tender irreducible mass in the inguinoscrotal region [6], but in children & infants, appendicitis is rarely seen [5]. This could be justified by the fact that the inguinal orifice is large in younger patients, and thus the appendix can easily herniate but not so easily incarcerate. Also, the appendix's lumen is relatively wider in neonate, meaning that it is more difficult for a foreign body to block its lumen and cause appendicitis [17].

In spite of the rarity of left sided Amyand's hernia, it should be included in the list of differentials when a patient present with left-sided inguinal pain because of the potentially serious sequelae of a missed or delayed diagnosis [12,18].

Different complications of Amyand's hernia have been reported in the literature like abscess formation [19], inflamed right testis & spermatic cord [3], testicular ischemia [20], or more severe

complication; necrotizing fasciitis of the lower abdominal wall firstly reported by Marron et al. [21]. An extremely rare presentation of a foreign body as a cause of appendicitis in Amyand,s hernia has been reported roughly once per century. In addition to the initial report by Amyand, another report of a swallowed pin causing appendicitis in a child was reported by Llullaku et al. in 2010 [22]. Amyand's hernia can also present with appendicular adenocarcinoid tumor [23].

The Preoperative diagnosis is very rare due to absence of distinct presenting clinical features. Inguinal hernias are typically diagnosed clinically even when complicated. Although a preoperative imaging study like ultrasound & CT scan may be helpful in securing the correct diagnosis of complicated hernia, this is not a routine practice after the clinical diagnosis [24]. Sonography has been reported to be informative in differentiating Amyand's hernia and other acute scrotal conditions [25]; It can differentiates hernias from other structures, detect content of hernias whether there is a blind ending tubular structure of appendix or Meckel's diverticulum in the inguinal canal**,** sacculations and moving content of the colon, soft tissue structure, recognize normal testes and epididymis & exclusion of testicular tumors and epididymitis as these conditions may have similar symptoms. However, preoperative diagnosis based on sonographic study alone is mainly depend on the technical skill & experience of the operator, and in itself remains a relatively unreliable modality [3]. Tycast et al. suggest using laparoscopic surgery as diagnostic & therapeutic tool for those patients in which diagnosis of Amyand's hernia was unclear [26].

There are no fixed principles for the management of Amyand's hernia. However, there are some factors which may have impact on the outcome, these are age of the patient, anatomic condition of the tissue, whether appendix is inflamed or not and contamination of surgical field during operation [2].

Losanoff and Basson in 2008, proposed a classification of Amyand's hernias with their suggested treatment guidelines [7]. (Table.1)

Table 1: Losanoff and Basson suggested classification & treatment of Amyand's hernia.

Type of hernia	Feature	Surgical treatment
1	Normal appendix.	Reduction or appendectomy (depend on age), hernia repair.
2	Acute appendicitis localized in the sac.	Appendectomy through hernia, hernia repair.
3	Acute appendicitis, peritonitis.	Appendectomy through laparotomy, hernia repair.
4	Acute appendicitis, other abdominal pathology (abscess or malignancy).	Appendectomy, diagnostic workup, other procedures as appropriate.

Our case was type I according to Losanoff and Basson classification so we did reduction of appendix into abdominal cavity & herniotomy. Really there is some argument among authors about doing appendectomy in Amyand's hernia; some advocate appendectomy even when the appendix is normal in an effort to avoid future complications [10]. Manipulation of a normal appendix during surgery may induce inflammation and stimulate secondary appendicitis [5]. Johari et al. recommended performing an appendectomy in cases of left sided Amyand's hernia even if the appendix is normal, because of the atypical presentation of appendicitis in the future which may delay its diagnosis & result in serious complications [27]. While other suggest appendectomy only if the appendix is inflamed [10], Baldassarre et al. suggested reducing the appendix into abdominal cavity without resection if it is not inflamed, because removal of appendicular lymphoid tissue may compromise the development of immune system in pediatric patients [11]. The other reasons for hesitation in the resection of the normal appendix may be as follows: (1) appendectomy may add to the risk of a surgical site infection [2]; (2) the incision may need to be enlarged to dissect the base of appendix, leading to the weakening of tissues and increasing the probability of a recurrence of inguinal hernia [28] ;(3) The appendix may also be useful in the future for anterograde bowel enemas, urinary diversion, or biliary tract reconstruction. However, the decision to do appendectomy or not will most likely remain the surgeon's personal choice.

Hernia repair most often is accomplished during primary surgery with few exceptional cases in which hernia repair is delayed due to complications and presence of severe inflammation [3].

Recently, a variable modality have begun to appear in the literature in managing Amyand,s hernia. Vermillion et al. reported the first use of laparoscopy for appendectomy in a case of Amyand's hernia with appendicitis [29]. Several reports nowadays indicate that the incidence of laparoscopic surgery in such conditions is on the rise [30,31]. However, development of complications may necessitate a conversion from laparoscopic to open hernia repair.

Mortality of Amyand's hernia has been reported in the range of 14–30% [3], indeed Amyand's hernia is not a cause of death, and all deaths are attributed to other comorbid situations [6,32] like peritoneal spread of sepsis or necrotizing fasciitis as a dangerous and rare complication [21,33]. In comparison with classic appendicitis, the prognosis of Amyand's hernia is better because of earlier diagnosis and limitation of inflammation inside the hernia sac [6].

CONCLUSION:

Amyand's hernias are rare, but should be considered in the differential diagnosis of patients with incarcerated inguinal hernias particularly on the right side. It has a challenging diagnosis due to its low incidence, vague clinical features, and ambiguous appearance on radiologic studies, hence surgery is often diagnostic & therapeutic at the same time. Removal of the appendix is not always necessary since it may be not inflamed when found inside the hernia sac, this decision is actually a surgeon's choice. Because of the rarity of Amyand's hernia & the wide range of its presentation, each case study and review of an article highlights a useful information regarding its diagnosis and treatment.

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