Acta Biomed 2022; Vol. 93, Supplement 1: e2022273 DOI: 10.23750/abm.v93iS1.13322

Case report

Amyand's hernia: which oncologic risk can be hidden in the sac?

Manuel Baldinu[1], *Filippo Montali*[2], *Alfredo Annichiarico*[1,2], *Edoardo Virgilio*[1,2], *Renato Costi*[1,2]

[1]Department of Medicine and Surgery, University of Parma, , Parma; [2]Department of Surgery, Azienda Sanitaria Locale di Parma, Fidenza (PR), Italy

Abstract. Amyand's hernia (AH) is a rare condition in which the appendix is found in the sac of an inguinal hernia. It occurs in only 1% of adult inguinal hernias. The herniated appendix can occasionally range varying degrees of acute inflammation up to neoplastic transformation. An appendiceal tumor can be rarely described inside the AH sac. We describe a case of gangrenous appendicitis in AH and offer a review of the literature on AH when presenting with appendicitis associated with appendiceal cancer. As of 2022, only nine cases of AH presenting with appendicitis associated with appendiceal cancer have been reported by the pertinent literature. In order of decreasing frequency, AH, AH-related appendicitis and AH-related appendicitis associated with appendiceal cancer are three rare conditions. Particular attention should be kept in each situation as diagnosis is achieved postoperatively most of the times. (www.actabiomedica.it)

Key words: Amyand hernia, inguinal appendicitis, appendiceal cancer.

Introduction

In 1735 Claudius Amyand performed and published the first successful appendectomy on a 11-years-old boy who presented with a perforated appendix in his inguinal hernia sac (1). The finding of the vermiform appendix within an inguinal (Amyand's hernia, AH) or crural hernia (De Garengeot's hernia) amounts to about 1% of cases (2-5). An acute appendicitis associated with incarceration or strangulation is even rarer with an incidence of approximately 0.13% (2). Cases of AH have been recorded in every age, from neonates to elderly, and prefer male gender (3). AH corresponds to 2% of all appendectomies during neonatal period and infancy (3). The clinical presentation can be dreadful since mortality of has been reported to range from 14 to 30% and was linked to peritoneal spread of sepsis. Preoperative diagnosis is challenging and most AH cases are detected during an urgent or emergent surgery (6). Occasionally, an appendiceal tumoral lesion can be described inside the AH sac. In the following case the clinical suspicion that the patient had an inguinal oncological mass was high. A malignancy of the appendix should always suspected when a right inguinal mass is observed leading to the spermatic cord, in order to provide the most appropriate surgical treatment.

Case report

An 84-year-old man presented to our Emergency Department with a three-day worsening abdominal pain associated with a palpable mass in his right inguinal region. The patient had a 5-year history of right-side reducible inguino-scrotal hernia. Other known diseases were chronic bronchitis under oxygen therapy and arterial hypertension under medications.

Blood tests showed leukocytosis (13130 cells/mm^3) with increased C-reactive protein (136 mg/dL) and procalcitonin (0.44 ng/mL). On physical examination, a non-reducible painful swelling of about 6 cm in diameter was observed in the right inguinal region. A clinical diagnosis of strangulated hernia was suspected. A contrast-enhanced CT scan of the abdomen was promptly carried out showing a right inguinoscrotal hernia of about 67x55x165 mm with a breach of about 30 mm containing adipose tissue as well as the last ileal tract: such anatomic structures showed radiologic signs of acute inflammation (**Fig.1**).

The patient underwent an emergency surgery with an open right inguinotomy. After dissecting the external oblique muscle's fascia, a large mass with free purulent discharge was observed tenaciously tethered to the spermatic cord. After carefully isolating and opening a thickened cremaster muscle and the peritoneal sac, the appendix, which was macroscopically barely recognizable because presenting like a solid necrotic lesion, infiltrated the posterior spermatic cord. The mass (appendix) was ligated and removed. Urology consult was carried out intraoperatively: the ipsilateral testicle appeared ischemic and was removed. We also performed a right direct hernioplasty without prothesis due to the presence of purulent contamination. One inguinal drainage was left behind. Post-operative course was uneventful. In the suspicion of a spermatic cord or appendiceal tumor, the tumor markers (CEA, Ca 19.9 and AFP) were appraised: all resulted within normal range. The culture test revealed an infection by a multisensitive strain of *Escherichia coli*: we therefore continued with the therapy already set empirically. The patient was discharged 6 days after surgery. Since, he had never undergone colon cancer screening before surgery, a postoperative colonoscopy was arranged resulting free from lesions. On histology, a conclusive diagnosis of gangrenous appendicitis with hydrocele and testicular atrophy with ischemic necrosis was achieved (**Fig. 2**).

Written informed consent was obtained from the patient before pubblication

Discussion

Amyand's hernia (AH) is a particular type of inguinal hernia characterized by the presence of vermiform appendix in the groin. It is a rare clinical condition ranging from 0.19% to 1.7% of all the cases of inguinal hernia commonly reported by the literature. AH is three times more frequent in children than in

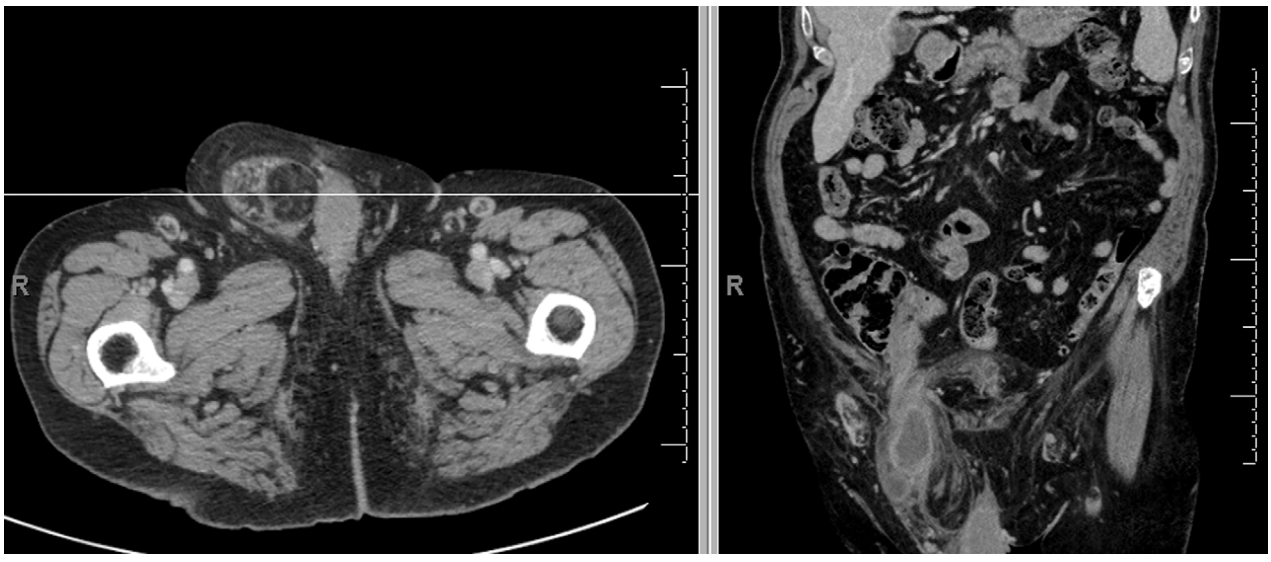

Figure 1. Contrast-enhanced CT scan of the abdomen showed showing a right inguinoscrotal hernia of about 67x55x165 mm with a breach of about 30 mm containing adipose tissue as well as the last ileal tract: both of these anatomic structures showed radiologic signs of acute inflammation (left: xial view; right: sagittal view).

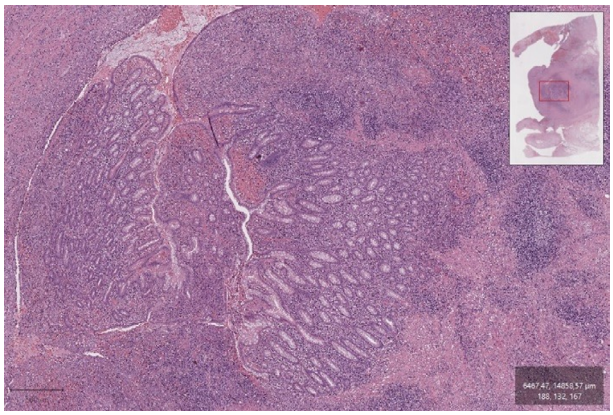

Figure 2. Pathologic verification of orchifunicolectomy showed an edematous *tunica vaginalis*, hydrocele and an atrophic testicle with a congested *rete testis*. No focal or tumor lesions were found.

adults, due to the patency of the *processus vaginalis* in this population (7). The coexistence between appendicitis and AH is even rarer with an estimated rate of 0.07-0.13% (8). In 2003, D'Alia and colleagues examined 1341 cases of inguinal hernia and determined an AH incidence of 0.6%, always on the right side and exclusively in male patients (6). Sharma et al. reported that, when herniated inflamed appendix affects females, such subjects are generally in postmenopause (8). Mortality of AH has been reported to range from 14 to 30% and it has been related to the peritoneal spread of sepsis (6). Due to the anatomical position of the appendix, most AH occurs in the right inguinal region whereas very few cases involve the left groin (7). AH, by definition, develops above Poupart's ligament (the inguinal ligament); when appendix protrudes into the femoral canal, such a condition takes the name of De Garengeot's hernia which is, however, to be considered a variant of AH and which prefers female gender paralleling the frequency of crural hernia in this sex (3-5). Achieving a clinical diagnosis of AH is not a simple task if considering only clinical presentation and laboratory results because they are aspecific; additionally, imaging techniques of the abdomen frequently miss to identify the herniated appendix and are consecutively not helpful to differential diagnosis (6). Common complaints experienced by patients include sudden-onset epigastric or periumbilical pain with localized tenderness in the right lower quadrant, combined with a tender irreducible mass in the inguinal or inguino-scrotal region (7). Clinical diagnosis is rarely achieved in the preoperative period: in fact, it is generally obtained during surgery (8). The cause-effect relationship between inguinal herniation and inflammation of the appendix has not yet been satisfactorily clarified. Two are the most popular theories (6). According to the former, the main event is represented by an appendiceal inflammation which progressively leads to a severe edema; in advanced stages, the edema initially causes venous stasis but later takes to an insufficient arterial supply until a bacterial necrosis and superinfection supervene. The latter, on the other hand, states that the appendix engaged into the hernial sac exposes itself to small traumas that give rise to an inflammatory reaction with the formation of adhesions that entrench the retention of the appendix in the hernial sac. The physiologic contraction of the abdominal muscles can suddenly increases the abdominal pressure; abdominal hypertension causes compression of the appendix resulting in a further inflammation that compromises blood supply. This pressure-dependent mechanism eventually leads to a severe appendiceal inflammation with bacterial proliferation and superinfection (9, 10). In support of this hypothesis, from a clinical point of view, it must be taken into account that attempts of manual reduction could cause edema, hematoma and lymphatic hyperplasia leading to further obstruction of the appendicular lumen. Furthermore, it is supported by several authors (9-11). Apostolidis and colleagues, for example, described a case of a 68-year-old man in whom the histological examination of the surgical specimen revealed signs of chronic inflammation affecting the hernial sac along with periappendicitis with vascular congestion, suggesting a prior incarceration of the appendix followed by subsequent inflammation (11). Although incarceration of the appendix within an inguinal hernia does not always lead to appendicitis, this is not an uncommon finding. Ash and coauthors, in fact, suggested a relationship between appendix incarceration in the inguinal canal and the development of inflammation (12). In some circumstances, clinical scenario of AH can become dreadful because herniated appendicitis can further complicate with appendiceal perforation in the inguinal canal (13), perforation with periappendicular abscess (14), perforation or strangulation with inflammation of the right

testicle and spermatic cord (15), testicular ischemia (even in newborns) (16), hyperemia and hemorrhage within the hernial sac (17), necrotizing fasciitis of the anterior abdominal wall (18) or necrotizing fasciitis of the inguinal region (19). An extremely rare complication of an inflamed AH is represented by thrombosis as observed in a case reported by Wilson et al. in 2012, in which an intra-abdominal abscess led to the formation of an *in situ* arterial thrombosis (20). Notably, in exceptionally rare cases, inflammation of an AH-related appendix can be determined by a foreign body (pin) ingestion or a penetrating abdominal trauma (1, 21).

In 2007, Losanoff and Basson proposed a IV-tier classification system in ascending order of severity: due to the presence of purulent appendicular peritonitis, our case falls within class III but, at the time of intervention, the suspicion of a malignancy prompted us to suspect a class IV (22).

When grappling with a right inguinal mass in the presence of an AH, a malignancy should always be suspected involving the spermatic cord or the appendix. Spermatic cord malignancies are very rare tumors given that less than 100 cases of cord liposarcoma have been reported in the literature so far (23). Appendiceal tumor associated with AH are even rare since, after digging into the pertinent literature, we found only 9 cases to be published so far (24-32) (Table I).

Four AH cases showed an appendicular carcinoid (two "pure" carcinoid tumor and two cases of goblet cell carcinoid tumor); three had mucinous cystadenoma; one had villous adenoma; the last had adenocarcinoma (24-32) (Table I). Carcinoids affect the gastrointestinal tract in 55% of cases: of these, appendix is involved in 17% of cases preceded by the small bowel and rectum (45% and 20%, respectively) and followed by the colon and stomach (11% and 7%, respectively) (33). As reviewed, carcinoid tumor is the most common neoplasm found not only in a not-inflamed appendix but also in the appendix involved in AH (24-27). For carcinoid or neuroendocrine tumor (NET) of the appendix, surgery always represents the gold standard treatment. Simple appendectomy is considered adequate in NETs of the appendix with a diameter of less than 10 mm or for those with a diameter of 10-19 mm but without any mesoappendix invasion (24). Among tumors with a diameter ranging between 10 to 19 mm, right hemicolectomy is (RH) the most appropriate approach if one or more of the following parameters are present: mesoappendix invasion, vascular invasion, grade II proliferation, suspected or positive surgical borders, and mixed histology (goblet cell carcinoid, adenocarcinoid) (24-27). RH is the most appropriate approach also for NETs with a diameter equal to or greater than 20 mm (33, 34). RH also represents the standardized treatment for appendiceal adenocarcinoma in every cases including the ones associated with AH: irrespective of location and size, it should be conducted as soon as possible, during the original intervention or following the pathology report (32). Even if the appearance is benign, an appendiceal mucocele should be always treated with surgical resection with clean borders, since some of them have been shown to have a potential for malignancy. Hence, while simple appendectomy is adequate for some histological subgroups (such as mucocele's retention cyst, mucosal hyperplasia and cystadenoma), RH is necessary for complicated mucoceles comprising the cecum and terminal ileum and for the cases of mucinous cystadenocarcinomas (28-30).

Table I. Review of the literature on clinicopathologic features of neoplastic appendix associated with Amyand's hernia.

References	Year	Age	Sex	Preliminary Diagnosis	Side	Type	Hernia Status	Surgery	Hernia Repair	Approach to Appendix	Histopathological Evaluation	Tm Size (mm)	Tm Location
Christodoulidis	2017	52	M	Inguinal Hernia	Right	Indirect	Incarcerated	Open	Bassini	Appendectomy	Goblet Cell Carcinoid	22	Tip
Yahya	2017	NA	NA	NA	NA	NA	NA	NA	NA	NA	Goblet Cell Carcinoid	NA	NA
Elbanna	2015	81	M	Amyand's Hernia	Right	Indirect	Incarcerated	Open	Bassini	Appendectomy	Carcinoid	15	Tip
Reynu	2015	70	M	Inguinal Hernia	Right	NA	Strangulated	Open	Primary	Appendectomy	Mucinous Cystadenoma	NA	NA
Nahmias	2013	50	M	Amyand's Hernia	Right	Indirect	Strangulated	Laparoscopic	Internal Ring Closure	Appendectomy	Carcinoid	10	NA
Shabeeb	2010	62	M	Inguinal Hernia	Right	Indirect	Incarcerated	Open	Lichtenstein Bassini	Appendectomy	Mucinous Cystadenoma	30	NA
Wu	2010	62	M	Inguinal Hernia	Right	NA	Incarcerated	Open		Appendectomy	Adenocarcinoma	25	NA
Salemis	2006	61	M	Inguinal Hernia	Right	Indirect	Strangulated	Open	Shouldice	Appendectomy	Perforated App + Villous Adenoma	3	Base
Oh	2019	37	M	Acute appendicitis	Right	Indirect	Incarcerated	Laparoscopic		Appendectomy	Mucinous Cystadenoma	2	NA

Conclusion

The concomitant existence of Amyand's hernia and appendiceal tumor inside the hernia sac is a very rare clinical phenomenon. The majority of Amyand's hernia mimic signs and symptoms of incarcerated inguinal hernia and, as a consequence, diagnosis is made incidentally during surgery most of the times. The basic principles of surgical oncology should always be followed in the management of a mass discovered inside a hernia sac (35).

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

References

1. Vincenzo Lippolis, P, Barlettai M, Filidei F, Seccia M. The Aymand's hernia. Case report and review of the literature. Ann Ital Chir 2007; 78 (2). 157-8.
2. Smith-Singares E, Boachie JA, Iglesias IM. A rare case of appendicitis incarcerated in an inguinal hernia. J Surg Case Rep 2016; 2016 (6): rjw096.
3. Rollo A, Franzini C, Casali L, Santi C, Lombardo E, Violi V. De Garengeot hernia: laparoscopic treatment in emergency. Acta Biomed 2019; 90 (4): 568-71.
4. Davies MG, O'Byrne P, Stephens RB. Perforated appendicitis presenting as an irreducible inguinal hernia. Br J Clin Pract 1990; 44 (11): 494-5.
5. Pitchaimuthu M, Dace S. A rare presentation of appendicitis as groin swelling: a case report. Cases J 2009; 2 (1): 53.
6. D'Alia C, Lo Schiavo MG, Tonante A, et al. Amyand's hernia: case report and review of the literature. Hernia 2003; 7 (2): 89–91.
7. Baldassarre E, Centonze A, Mazzei A, Rubino R. Amyand's hernia in premature twins. Hernia 2009; 13 (2): 229-30.
8. Sharma H, Gupta A, Shekhawat NS, Memon B, Memon M. Amyand's hernia: a report of 18 consecutive patients over a 15-year period. Hernia 2007; 11 (1): 31-5.
9. Abu-Dalu J, Urca I. Incarcerated inguinal hernia with a perforated appendix and periappendicular abscess: Report of a case. Dis Colon Rectum 1972; 15 (6): 464-5.
10. Logan MT, Nottingham JM. Amyand's hernia: a case report of an incarcerated and perforated appendix within an inguinal hernia and review of the literature. Am Surg 2001; 67 (7): 628-9.
11. Apostolidis S, Papadopoulos V, Michalopoulos A. Amyand's Hernia: A Case Report and Review of the Literature. Internet J Surg 2005; 6: 1.
12. Ash L, Hatem S, Ramirez GA, Veniero J. Amyand's hernia: a case report of prospective ct diagnosis in the emergency department. Emerg Radiol 2005; 11 (4): 231-2.
13. Lyass S, Kim A, Bauer J. Perforated appendicitis within an inguinal hernia: case report and review of the literature. Am J Gastroenterol 1997; 92 (4):700-2.
14. Kueper MA, Kirschniak A, Ladurner R, Granderath FA, Konigsrainer A. Incarcerated recurrent inguinal hernia with covered and perforated appendicitis and periappendicular abscess: case report and review of the literature. Hernia. 2007; 11 (2):189-91.
15. Serrano A, Ackerman NB. Perforated appendicitis in an incarcerated inguinal hernia. Arch Surg 1979; 114 (8): 968.
16. Milburn JA, Youngson GG. Amyand's hernia presenting as neonatal testicular ischaemia. Pediatr Surg Int 2006 22 (4): 390-2.
17. Ioannidis O, Styliani P, Kakoutis E et al. Strangulation of the vermiform appendix within a right inguinal hernia: a rare variation of Amyand's hernia. Am Surg 2010; 76 (11): 1305-6.
18. Marron CD, Khadim M, McKay D, Mackle EJ, Peyton JW. Amyand's hernia causing necrotising fasciitis of the anterior abdominal wall. Hernia 2005; 9 (4):381-3.
19. Osorio JK, Guzmán-Valdivia G. Ipsilateral Aymand's and Richter's hernia, complicated by necrosing fascitis. Hernia 2006; 10 (5):443-6.
20. Willson TD, Rao V, Podbielski FJ, Blecha MJ. *In situ* aortic thrombosis secondary to intra-abdominal abscess. Am J Case Rep 2012; 13: 149-52.
21. Llullaku SS, Hyseni NSh, Kelmendi BZ, Jashari HJ, Hasani AS. A pin in appendix within Amyand's hernia in a six-years-old boy: case report and review of literature. World J Emerg Surg 2010; 5: 14.
22. Losanoff JE, Basson MD. Amyand hernia: what lies beneath--a proposed classification scheme to determine management. Am Surg 2007; 73 (12): 1288-90.
23. Virgilio E, Uccini S, Marchetti P, Mercantini P. Commentary on "a case of paratesticular leiomyosarcoma successfully treated with orchiectomy and chemotherapy". Cancer Res Treat 2016; 48 (1): 422-4.
24. Nahmias NC, Kaplan LA, Muddasani KP. Image of the month. Incarcerated inguinal hernia. JAMA Surg 2013; 148 (6): 573-5.
25. Elbanna KY, Alzahrani HA, Azzumeea F, Alzamel HA. Neuroendocrine tumor of the appendix inside an incarcerated Amyand's hernia. Int J Surg Case Rep 2015; 14, 152-5.
26. Yahya Z, Wang CI, Hong B. Amyand's hernia containing an appendiceal goblet cell carcinoid tumour. BMJ Case Rep 2017; 2017: bcr2017219428.
27. Christodoulidis G, Perivoliotis K, Diamantis A, Dimas D, Spyridakis M, Tepetes K. An appendiceal carcinoid tumor within an Amyand's hernia mimicking an incarcerated inguinal hernia. Case Rep Surg 2017; 2017: 5932657.
28. Shabeeb F, Hairol AO, Jarmin R. Amyand's hernia with mucinous cysadenoma of the appendix. Indian J Surg 2010; 72 (Suppl 1): 341-3.

29. Reynu R. A rare presentation of caviar appendix in an Amyand's hernia. Brunei Int Med J 2015; 11 (4): 209-13.
30. Oh HB, Chen Y, Soon Y. Low-grade appendiceal mucinous neoplasm within an Amyand's hernia. ANZ J Surg 2019; 89 (6): E286-7.
31. Salemis NS, Nisotakis K, Stavrinou P, Tsohataridis E. Perforated appendix and periappendicular abscess within an inguinal hernia. Hernia 2006; 10 (6): 528-30.
32. Wu CI, Yu Cc. Amyand's hernia with adenocarcinoid tumor. Hernia 2010; 14 (4): 423-5.
33. Pinchot SN, Holen KH, Sippel RS, Chen H. Carcinoid tumors. Oncologist 2008; 13 (12): 1255-69.
34. Ellis L, Shale MJ, Coleman MP. Carcinoid tumors of the gastrointestinal tract: trends in incidence in England since 1971. Am J Gastroenterol 2010; 105 (12): 2563-9.
35. Sarici B, Akbulut S, Piskin T. Appendiceal carcinoid tumor within Amyand's hernia: a case report and review of the literature. Turk J Emerg Med 2019; 19 (2): 73-5.

Correspondence:
Received: 7 June, 2022
Accepted: 5 July, 2022
Filippo Montali
Department of Surgery
Azienda Sanitaria Locale di Parma
Vaio Hospital, Via Don Tincati 5, 43036 Fidenza (PR)
+39 3471811416
fmontali@ausl.pr.it