Case Report

Appendiceal Carcinoid Tumor Contained in a Large Inguinal Hernia: A Case Report

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We report the first case of a clinically diagnosed incidental appendiceal carcinoid tumor contained in an incarcerated inguinal hernia without inflammation of the appendix. A 67-year-old white man underwent elective open repair of an incarcerated right inguinal hernia containing small bowel, cecum, appendix, and ascending colon. He was found to have a grossly abnormal mass at the tip of his appendix. Appendectomy was performed, and histopathologic analysis confirmed the presence and adequate resection of a neuroendocrine tumor. It is important for general surgeons to be aware that unusual presentations of appendiceal tumors may not be associated with appendiceal inflammation or related symptoms.

Key words: Inguinal hernia – Amyand’s hernia – Appendiceal carcinoid

Inguinal hernia repair is one of the most common general surgery procedures. The inclusion of intra-abdominal structures within the contents of the hernia sac, most commonly omentum, bowel, or intestine, requires careful dissection and repair. Rarely, unexpected contents are encountered in the hernia sac, including intestinal diverticula (Littre’s hernia) or the vermiform appendix (Amyand’s hernia). Amyand’s hernia is defined as the presence of the appendix within an inguinal hernia sac and was named in honor of Claudius Amyand (1680–1740). Dr Amyand was a surgeon in the British Army who performed the first recorded appendectomy while repairing an inguinal hernia of an 11-year-old boy in 1735. Discovery of the appendix in an inguinal hernia sac is very rare (reportedly less than 1% of all inguinal hernias), with coincidental findings of abnormalities of the appendix in this setting being even more unusual.1–6

Neoplasms of the appendix in general are also rare, found incidentally in less than 1% of appendectomy specimens and rarely diagnosed preoper-
The occurrence of these 2 conditions together has been reported only 3 times, and all previous cases were associated with appendiceal inflammation and diagnosed on postoperative histopathologic analysis and not at the time of surgery.7,8

Case Report

A 67-year-old man presented to clinic with a large right scrotal mass that he noticed on waking a few weeks prior to presentation. He denied any recent trauma or straining. He experienced discomfort with movement but denied significant pain. He reported weak urine stream but denied any changes in bowel movements. His medical history included an ascending aortic dissection, type II diabetes mellitus, atrial fibrillation, hypertension, and urinary retention. His prior surgeries included emergent repair of the aortic dissection with aortic valve replacement and a tonsillectomy. On examination, he was noted to have a large right inguinal hernia extending into the scrotum, measuring approximately 30 cm in length and 17 cm in diameter. The scrotum was nontender but moderately tense with nonblanching erythema. His vital signs and laboratory studies were within normal limits. He was scheduled for elective inguinal hernia repair. He underwent preoperative medical clearance.

He presented 2 weeks later for anterior inguinal hernia repair under general endotracheal anesthesia. The very large indirect hernia sac was opened and found to contain 60 cm of small bowel, omentum, cecum, and approximately half of the ascending colon, as well as thin serous fluid. The small and large bowels were incarcerated but appeared otherwise healthy and viable, with no signs of ischemia. The appendix was noticed and found to have a 1-cm firm whitish-tan mass on the distal end. Because of this obvious abnormal pathology and clinical concern for a carcinoid or other neoplasm, the decision was made to perform an appendectomy. The appendix was resected at the base and the appendiceal stump was invaginated into the cecum. There was a 3- to 4-cm margin, and the appendix was sent for intraoperative pathologic consultation. The hernia sac contents were then able to be reduced. The neck of the sac was elevated off the underlying cord structures and divided. The distal end was left open and not dissected out of the scrotum. Proximally the sac was dissected back to the internal ring and high ligation carried out. There was no inguinal floor present, making any attempt at primary repair likely to be futile. The increased risk of infection with prosthetic mesh was considered, and biological mesh was an option, but was not considered a durable way to repair this large hernia. The decision was made to proceed with Lichtenstein tension-free hernia repair with polypropylene mesh. Instruments used in appendectomy were passed off the field prior to the hernia repair. The frozen section was consistent with carcinoid tumor. The patient was notified of the operative findings the evening of surgery. The final pathology revealed a grade 1 neuroendocrine tumor with an 8-mm radial margin and without lympho-vascular invasion (pT1b tumor).

The patient’s postoperative recovery was complicated by urinary retention requiring discharge with a Foley catheter and urology follow-up. He was seen in the clinic 2 weeks after discharge and was found to be recovering well with no signs or symptoms of infection or hernia recurrence. He was referred to an oncology clinic for follow-up, although it was not anticipated that he would require further treatment of his carcinoid tumor. Unfortunately, before he was seen by oncology, he had a fall and was admitted to the hospital with a right femoral neck fracture 1 month after his hernia repair. He was re-examined by the surgical team at that time and again found to be without evidence of infection or hernia recurrence. He did not require additional surgical follow-up but has been followed by internal medicine without hernia or carcinoid recurrence for 2 years postoperatively.

Discussion

Losanoff and Basson12,13 developed a classification system for Amyand’s hernias based on the degree of appendiceal inflammation. Type I signifies the finding of a normal appendix within the hernia sac. Type II involves localized appendicitis in the hernia sac commonly secondary to incarceration. Type III includes localized peritonitis and type IV extends to generalized peritonitis. This patient presented without preoperative signs or symptoms or intraoperative findings indicative of appendicitis or peritonitis; therefore, this is considered a type I Amyand’s hernia.

The suggested management for each type of Amyand’s hernia is variable. In type I Amyand’s hernia, some sources support incidental appendectomy, whereas others elect to reduce and leave the normal appendix.1-6,13 Use of mesh in repair of type I Amyand’s hernia in which appendectomy is also performed remains controversial because of the
conversion to a contaminated case with the removal of the appendix.\textsuperscript{2,4,12–14} In this case, mesh hernia repair was performed because the large size of the hernia would have resulted in a technically challenging, if not impossible, primary closure with an extremely high risk of recurrence.

Current recommendations state primary mesh repair is contraindicated in cases that include appendiceal inflammation/perforation given the increased risk of complications such as prosthetic mesh or wound infection; thus, proposed management for type II includes appendectomy and primary hernia repair.\textsuperscript{1–6,13} Management of types III and IV should include laparotomy, appendectomy, and primary hernia repair without mesh, as well as treatment of any other associated pathologies including abscess.\textsuperscript{2,12,13} There is a lack of discussion in the literature regarding management of an appendix with an obvious noninflammatory abnormality presenting within a hernia sac.

Independent of an associated hernia, neoplasms of the appendix are found in less than 1\% of appendectomy specimens.\textsuperscript{7,8} The most common type, neuroendocrine tumors, has been reported in 0.3\% to 0.9\% of patients at the time of appendectomy.\textsuperscript{15,16} These lesions are typically asymptomatic and found incidentally during appendectomy for acute appendicitis or other abdominal surgery. The majority of carcinoid neoplasms are located at the distal tip of the appendix. Occasionally, the tumor can obstruct the lumen, causing appendicitis. Appendiceal carcinoid tumors measuring less than 2 cm in diameter and treated with standard appendectomy rarely require further treatment because of the low risk of metastases of these lesions (less than 2\%).\textsuperscript{17} Although prognosis is good for appendiceal carcinoid with a 5-year survival rate greater than 90\%, patients are at a 33\% increased risk of developing colorectal neoplasm in the future, and thus routine screening for colorectal cancers is important in this population.\textsuperscript{15–17}

We believe this is the first reported case of a clinically recognized carcinoid tumor associated with an Amyand’s hernia. The intraoperative findings of an abnormal appendix would seem to qualify as a subcategory of type I, in which pathology of the appendix other than appendicitis is revealed. The fact that the appendix was noted to be grossly abnormal intraoperatively differentiates this case from other cases reported in the literature, in which neoplastic findings of the appendix were noted via routine postoperative histopathology following appendectomy for acute appendicitis.\textsuperscript{9,31,15}

Wu \textit{et al}\textsuperscript{9} were the first to report a clinical experience with this coincidental diagnosis. They found a goblet cell adenocarcinoid tumor of the appendix on pathologic analysis following open appendectomy and inguinal hernia repair. The preoperative diagnosis was as incarcerated inguinal hernia, and the tumor was not recognized during the operation.

Nahmias \textit{et al}\textsuperscript{10} and Elbanna \textit{et al}\textsuperscript{11} recently published similar reports. The patient treated by Nahmias \textit{et al} was taken to the operating room for laparoscopic primary repair of a type II Amyand’s hernia diagnosed on preoperative imaging. Postoperative tissue analysis demonstrated a T1a appendiceal carcinoid tumor. The patient was taken back to the operating room for transabdominal preperitoneal hernia repair with mesh and was not found to have any evidence of remaining malignancy. Elbanna \textit{et al} described a patient with a right incarcerated inguinal hernia, found on preoperative imaging to contain distal ileum, cecum, and a thickened appendix. Open appendectomy and primary right inguinal hernia repair was performed. A T1b well-differentiated low-grade neuroendocrine tumor was found incidentally on histopathology.

There is little discussion in the literature of appendiceal pathology other than inflammation in the setting of an Amyand’s hernia, but carcinoid tumors, although rare, require proper management to prevent complications and avoid the need for additional procedures. This unique case demonstrates the variety of intraoperative findings and management decisions encountered during hernia repair and the role of individualized management on a case-by-case basis. It also highlights the importance of a careful examination of the involved and surrounding anatomical structures at the time of surgery to facilitate the discovery of unexpected but clinically significant pathology.

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