Apparent appendicitis in Amyand hernia with unexpected pathology

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CASE REPORT

A 70-year-old man with a history of peripheral T-cell lymphoma in complete remission since 2012, presented with a three-week history of a tender ‘knot’ in his right inguinal/suprapubic region. He denied fevers, nausea, vomiting, and constipation. Physical examination was remarkable only for a 4x5 cm firm mass in the right suprapubic region. Complete blood counts and a comprehensive metabolic panel were normal except for a chronically elevated creatinine (1.9 mg/dL) and mildly elevated lactate dehydrogenase (279 units/L). Transaxial (Figure 1A) and coronal (Figure 1B) computed tomography (CT) images, obtained without intravenous contrast, revealed marked appendiceal thickening (arrows) and periappendiceal fat-stranding (arrowheads), suspicious for acute appendicitis. As shown in Figure 1B, the appendix was located within an indirect inguinal hernia sac (bracket), an entity known as an Amyand hernia. There was no evidence of appendiceal perforation. One day later, the patient underwent a laparoscopic appendectomy and open primary right inguinal hernia repair with a post-operative diagnosis of “acute, perforated appendicitis incarcerated within an indirect right inguinal hernia sac.” In addition to acute-on-chronic inflammation in the hernia sac, pathology revealed diffuse large B-cell lymphoma of the appendix. Subsequent staging by fluorodeoxyglucose (FDG)-PET/CT revealed multiple FDG-avid liver lesions consistent with stage IV lymphoma. The patient subsequently achieved a complete response with R-CHOP chemotherapy.

DISCUSSION

Amyand hernia was first described by the surgeon Claudius Amyand in 1735. It refers to the protrusion of a vermiform appendix in an inguinal hernia sac and occurs in <1% of all inguinal hernias. Appendicitis in an Amyand hernia accounts for <0.1% of all cases of appendicitis [1–3]. Amyand hernias occur more frequently in male patients, and have a bimodal age distribution with peak incidences in neonates and in patients over 70 years of age [4–6]. Reports of Amyand hernia predominantly describe preoperative diagnoses made by ultrasound or CT, otherwise diagnosis of Amyand hernias tend to occur intraoperatively [7–9]. While almost always an indirect hernia, the phenomenon of a direct Amyand hernia has been also described [10]. A four-category classification system has been proposed by Losanoff and Basson to guide staging and management of Amyand hernias [11]. It bases surgical recommendations on the presence or absence of a normal appendix (Stage 1), localized
acute appendicitis (Stage 2), peritonitis (Stage 3), or other abdominal pathology (Stage 4). Stage 4 Amyand hernia is a small subgroup of an already rare condition, typically encompassing cases with abscess formation, or alternatively with coexistence of a malignancy, typically a primary appendiceal tumor [12, 13]. The patient described in our case report fits this category with his unexpected diagnosis of diffuse large B-cell lymphoma (DLBCL) of the appendix. The most common site of extranodal involvement of non-Hodgkin lymphoma (NHL) is the gastrointestinal (GI) tract, representing 10–15% of all NHL cases, and ~1/3 of all NHL with any extranodal involvement [14]. Involvement of the appendix is rare, as the most commonly involved sites in the GI tract are the stomach, small intestine, pharynx, colon, and esophagus [14, 15].

CONCLUSION

Computed tomography (CT) findings of acute appendicitis are nonspecific and that differential diagnoses should be considered when there is an atypical clinical presentation. In this case, the subacute presentation over three weeks and the absence of fever, leukocytosis, or gastrointestinal symptoms were unusual for acute appendicitis.

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