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Case Report

Carcinoid Tumor of the Appendix in an Adolescent Female

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Keywords

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- Neuroendocrine tumor
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Abstract

Abdominal pain, specifically right lower quadrant abdominal pain is a common presenting symptom in the pediatric emergency department (PED). While common diagnoses are more likely to be considered, sometimes, unusual causes can present with this complaint. Carcinoid tumor is the most common tumor of the appendix. It can be very difficult to diagnosis initially and is mostly discovered during surgery for misdiagnosed acute appendicitis. Prognosis is generally excellent with a 5-year survival of 94%, when tumor is confined to the appendix. For tumors <2 cm in diameter and confined to the appendix, simple appendectomy is usually curative while in larger lesions or those with extra-appendiceal invasion, an appendectomy with right hemi-colectomy and cytoreductive surgery is necessary. Here we discuss the case of a 13-year-old female who was discovered to have such a lesion during a laparoscopic appendectomy.

ABBREVIATIONS

PED: Pediatric Emergency Department; RMQ: Right Middle Quadrant; RLQ: Right Lower Quadrant; CBC: Complete Blood Count; CT: Computerized Tomography

CASE PRESENTATION

A 13-year-old Caucasian female presented to the PED with 2 days of worsening right middle quadrant (RMQ) and right lower quadrant (RLQ) pain. She admitted that over the past month she had been having intermittent left sided abdominal pain and that the current pain started two days on the left and had migrated to the RMQ and RLQ. About 3 days prior to the visit, she had an episode of non-bloody, non-bilious emesis with associated nausea. There was no history of fever, chills, constipation, diarrhea, dysuria or weight change. For this visit she was sent to an outside ED where she had lab work including a complete blood count (CBC) and a computerized tomogram (CT) of her abdomen/pelvis. She was referred to our PED for further management of suspected appendicitis.

She had no significant past medical or surgical history and was pre-menarchal. Her only medication was simethicone started by her pediatrician to help with the abdominal pain. Her vital signs in our PED included a temperature of 36.8°C, pulse of 83 beats per minute, respirations of 16 per minute and a blood pressure of 109/65. On physical exam she appeared well and in no distress. Other than tenderness on deep palpation in both the RMQ and RLQ the rest of her abdominal exam was benign including negative exams for rebound, guarding, psoas sign or obturator sign. Also, no masses or organomegaly were appreciated. Initial laboratory evaluation revealed a normal urinalysis and electrolyte panel, negative pregnancy test, white blood cell count of 6,400/mm, hematocrit of 38.5% and platelets of 167,000/mm with a normal differential. Abdominal ultrasound revealed a moderate amount of free fluid in the RLQ but the appendix itself was not visualized. A computerized tomography scan of the abdomen and pelvis with IV contrast from the outside institution was reviewed (Figure 1). This demonstrated thickening of the tip of the appendix measuring 10 mm with minimal surrounding inflammatory changes and no appendicolith. Multiple non-enlarged nodes were also noted in the RLQ.

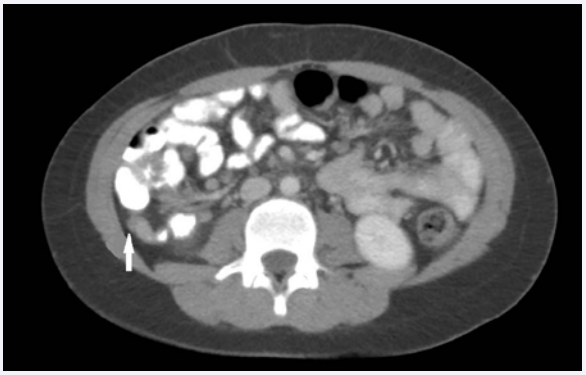

Figure 1 CT of the abdomen showing (white arrow) a dilated appendix with a distended lumen (10mm).

A diagnosis of appendicitis was made and she was taken to operating room for a laparoscopic appendectomy. The distal appendix was reported by the surgeon to be inflamed while the appendix measured 6.7 cm x 0.9 cm, however the pathology report showed no signs of appendicitis. Rather, there was a well-differentiated, low-grade (G1), neuroendocrine tumor, commonly known as a carcinoid tumor. It had extension to the serosal margin and into mesoappendiceal fat. The tumor size was 1.4 cm x 0.8 cm x 0.7 cm and proximal margins were tumor free. Based on the size and involvement it was staged T1b. She was discharged on hospital day 3 and is scheduled to follow up in 4 to 6 weeks with a referral to oncology.

DISCUSSION

Carcinoid tumors arise from neuroendrocrine cells in the foregut, midgut and hindgut with the midgut being the most common site for these tumors. It is the most common tumor of the appendix [1-3]. It is estimated that carcinoid tumors have a prevalence worldwide among all population of 1 to 2 per 100,000 although it may be higher as many cases are asymptomatic and may go unnoticed [3,4]. Caucasian females aged 13 are the most commonly affected among children [1].

Appendiceal carcinoid tumors typically have vague symptoms or are asymptomatic [4]. Many are found incidentally during surgery for suspected acute appendicitis [2]. Patients have a wide range of complaints ranging from mild, intermittent abdominal pain to complete bowel obstruction. These tumors may also be the lead point for intussusceptions [3]. The diagnosis of a carcinoid tumor is rarely thought of in the absence of carcinoid syndrome symptoms of which include flushing, diarrhea, tachycardia, hypotension, bronchospasm, telangiectasias, and potential heart failure. However, these symptoms do not occur in the absence of metastasis [1,4] which is typically to the liver and lungs [3,4]. The symptoms of carcinoid syndrome are normally induced by ingestion of tyramine or ethanol [3]. Prognosis is generally excellent with the 5-year survival when tumor is confined to the appendix being 94% [4].

Treatment for these confined tumors in the appendix is usually complete by the time the diagnosis is discovered since diagnosis is only made on biopsy [2]. The National Comprehensive Cancer Network guidelines for treatment of carcinoid tumors state that tumors <2 cm confined to the appendix can be treated with simple appendectomy with no follow-up required [5,6]. For tumors >2 cm, or those with extra-appendiceal invasion, an appendectomy with right hemi-colectomy and cytoreductive surgery is necessary [1,4].

CONCLUSION

The possibility of a carcinoid tumor should be included in the differential of a teenager presenting with long standing right lower quadrant abdominal pain. Although rare, it may require further therapy depending on staging at the time of surgery.

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