A 62-year-old male patient was admitted with fever (39 °C), chills and mild pain (localised in the right lower back and radiating to the right abdominal wall) of 7 days' duration. The patient reported that flexion of his right hip caused onset of pain in the lying position. The total blood count findings were Ht 27.6%, Hb 8.8 g/dL, WBC 21,200/μL, ESR 78 mm, serum iron 18 μg/dL, and serum ferritin 125 μg/L. Hb electrophoresis revealed a heterozygous thalassemia. Urinary sediment examination showed a few leucocytes and erythrocytes present. Blood and urine cultures were negative. Physical examination revealed no abdomen tenderness. Although the patient was treated with Flagyl, Timentin and Peflacine intravenously the fever continued. The chest CT was normal. Abdominal CT was performed before and after intravenous administration of contrast medium. Findings included a soft tissue mass, with cystic-like areas and internal septations, located at the right iliac fossa adjacent to the iliac muscle, psoas muscle and the caecum, which was focally thickened (figures 1, 2).

Comment

Primary appendiceal tumours make up only 0.5% of all intestinal tumours. Carcinoid tumours are common and account for 85% of epithelial appendiceal tumours, followed by mucinous cystadenocarcinoma (8%), colonic adenocarcinoma (4%) and adenocarcinoid (2%). Primary mucinous cystadenocarcinoma is a rare, well-differentiated neoplasm that progresses slowly. The patients are usually younger than those with adenocarcinoma. The incidence of the neoplasm is the same between the two sexes. At initial examination mucinous cystadenocarcinoma most commonly presents with acute right lower abdominal pain or a mass, or both, suggesting acute appendicitis. On ultrasound mucinous cystadenocarcinoma appears as a purely cystic mass, a cystic mass with fine echoes or as a complex cystic mass with high level echoes. CT may demonstrate a cystic mass in the expected area of the appendix of near-water or soft tissue density that has a mass effect on adjacent organs rather than infiltrating them. Contrast enhancement of a nodular lesion in the wall of the neoplasm may also be present. Other findings may be soft-tissue strands or infiltrations in the mesentery around the mass, nodular calcifications in the wall of the mass, an enlarged appendix with partial disruption of the wall and formation of an abscess, partial intussusception of the proximal appendix and disseminated intraperitoneal implantation of mucinous materials (so-called pseudomyxoma peritonei). Metastases may also be seen in other sites such as lymph nodes, liver or lung. Size, shape, wall thickness, internal septations and wall calcification of the cystic mass are not helpful in differentiating mucinous cystadenocarcinoma from benign mucinous cystadenomas. Enhancing wall nodularity and presence of pseudomyxoma peritonei are findings that can help to differentiate this neoplasm from both benign mucocele and other malignant neoplasms of the appendix and lead to a correct preoperative diagnosis. The histological features that distinguish mucinous cystadenocarcinoma from benign mucinous cystadenomas are the invasion of the appendiceal wall by atypical glands and the presence of epithelial cells in any intraperitoneal mucinous collections. The distinction is important since cystadenoma is cured by means of appendectomy while cystadenocarcinoma requires right hemicolectomy. Aggressive surgical treatment for intraabdominal metastases and pseudomyxoma peritonei can result in good long-term prognosis. All patients should be followed after surgery because they have a 15–20% chance of developing a second malignancy, usually in the gastrointestinal tract.

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