JBR–BTR, 2011, 94: 63-65.

PRIMARY APPENDICEAL TUMORS: CLINICAL IMAGING AND PATHOLOGICAL FINDINGS. REPORT OF FOUR CASES

A. Koktener[1], K. Akin[1], D. Kosehan[1], C. Dener[2]

Primary appendiceal tumors are rare clinical and radiological entities. We report 4 cases including muscinous cystadenoma associated with pseudomyxoma peritonei, muscinous cystadenoma associated with mucocele, low-grade muscinous neoplasia associated with appendicitis in elderly patients, and carcinoid tumor associated with appendicitis in a young adult.

Key-word: Appendix, neoplasms.

Primary appendiceal neoplasms are rare clinical and pathological entities. Most common manifestation of these tumors is acute appendicitis due to the luminal obstruction. Intussusception, palpable mass, gastrointestinal bleeding, abdominal distention (because of pseudomyxoma peritonei), genitourinary symptoms may be also seen. On the other hand, they may be asymptomatic and incidentally diagnosed (1, 2).

We present four primary appendiceal neoplasm with different clinical, pathological, and radiological findings.

Case 1: A 75-year-old man presented with the symptoms of disuria, dyspepsia, epigastric pain. His physical examination determined abdominal distension, ascites. Laboratory investigation showed high level of CEA: 20.9 mg/ml (0-5 mg/ml), PSA: 29.4 ng/ml (0-4 ng/ml), CRP: 19 mg/L (0-8 mg/L) and low level of hemoglobin 12.8 g/dL. Abdomen ultrasonography (US) showed ascites with peritoneal thickening. Colonoscopy revealed tubuler adenoma at the rectum. Gastroscopy found eosaphageal ulcer, erythmatous gastritis, bulbitis. Abdomen computed tomography (CT) determined large amount of ascites (Fig. 1). Parasynthesis found mucinous ascites without any cell. Right hemicolectomy was performed. Final pathologic analysis reported rupture of appendiceal mucinous cystadenoma.

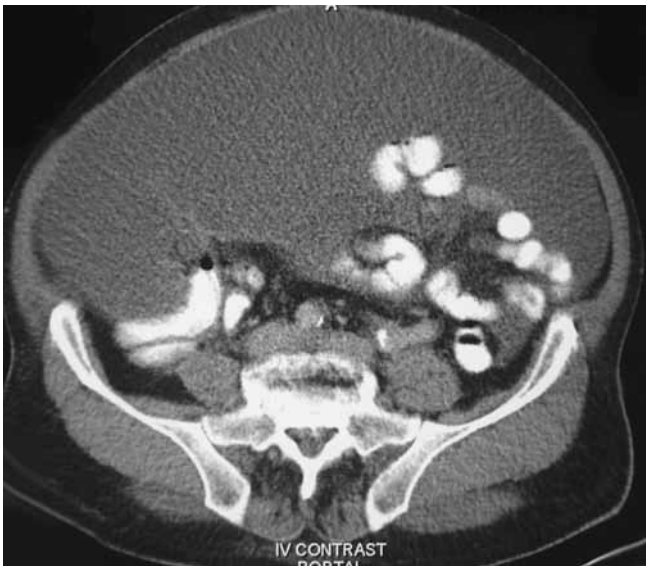

Fig. 1. — Axial contrast-enhanced CT scan shows ascites associated with pseudomyxoma peritonei.

Case 2: An asymptomatic 68-year-old man followed-up for prostate cancer admitted to the hospital for routine controls. On US examination paracecal localized a bilobulated cystic mass with 20 × 10 mm dimension and minimal peritoneal fluid were seen (Fig. 2). Laboratory examination including tumor markers found no abnormality. Colonoscopic examination showed colitis, diverticulosis at the sigmoid colon and tubuler adenoma at the rectum. Abdomen CT determined no abnormality except the findings on US (Fig. 3). Laparotomy including appendectomy was performed. Pathologic examination determined mucinous cystadenoma.

Case 3: A 59-year-old man presented with right lower quadrant pain lasting 3 days. Physical examination showed right lower quadrant tenderness. Laboratory investigation found elevated level WBC: 17600/mL (3600-9600/mL), ESR: 65 mm/hr, CRP: 78 mg/L. Abdomen US examination determined acute appandicitis features with peritoneal fluid (Fig. 4). Appendectomy was performed and pathology found low-grade mucinous neoplasia with peritonitis.

Case 4: A 38-year-old man admitted to the hospital with the symptoms of emesis, and abdominal pain. Physical examination determined tenderness of the lower quadrants. Laboratory examination showed leukocytosis (WBC: 13800/mL). Abdomen US examination showed edema of the mesenteric fat and peritoneal fluid in the right lower quadrant. Abdomen CT determined appendicitis with mesenteric inflammation (Fig. 5). Appendectomy was performed. Pathologic examination reported the diagnosis of tubular carcinoid tumor.

Discussion

Primary appendix tumors are not common, and seen 0.5%-1% of

From: 1. Department of Radiology, 2. Department of Surgery, School of Medicine, Fatih University, Ankara, Turkey.
Address for correspondence: Dr A. Koktener, M.D., Department of Radiology, School of Medicine, Fatih University. Alparslan Turkes Cad. No: 57 06510 Emek – Ankara Turkey. E-mail: akoktener@yahoo.com

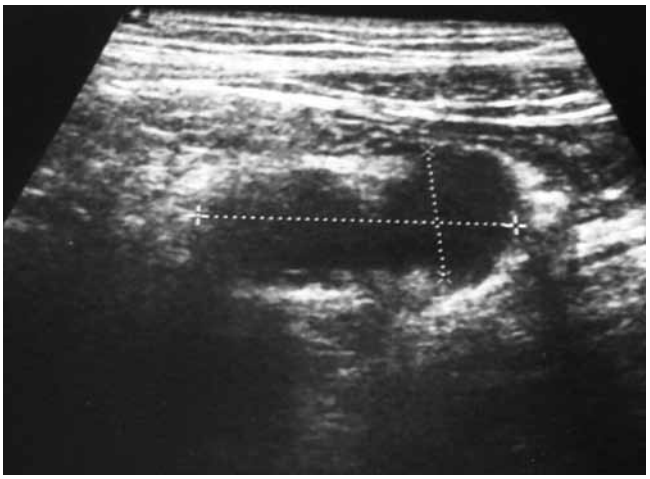

Fig. 2. — Transabdomainl US image demonstrates a bilobulated cystic lesion-mucocele.

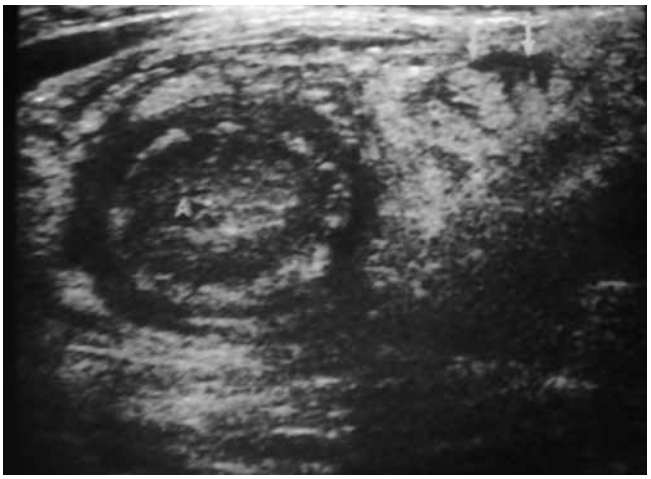

Fig. 4. — US image showes thick-walled, dilated appendix (A) with mesenteric inflammation (arrows).

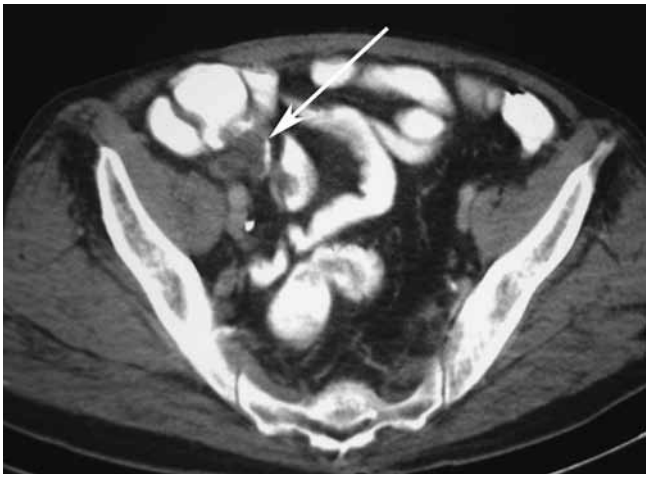

Fig. 3. — Axial contrast enhanced CT scan demonstrates fluid-filled lesion; mucocele near the cecum (arrow).

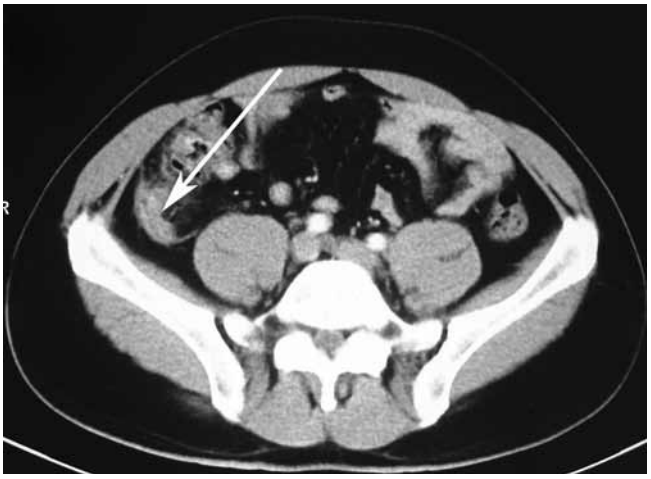

Fig. 5. — Axial contrast-enhanced CT scan showes the retrocecal appendix with wall thickening and mesenteric inflammation (arrow).

appendectomy specimens. Epithelial tumours of the appendix include low-grade mucosal tumours as well as invasive adenocarcinoma. The other neoplasms are carcinoid tumor, lymphoma, ganglioneuroma, phechromocytoma, mesenteric tumors. Retention cysts, hyperplastic polyps, and diffuse mucosal hyperplasia are not truly neoplastic (1, 2). Of these, mucocele is the most common one. Mucocele is a mucinous distention of the appendiceal lumen caused by mucosal hyperplasia, cystadenoma, and cystadenocarcinoma (3). They are relatively asymptomatic and found incidentally during palpation or abdominal imaging. When inflammation occurs, acute appendicitis, abscess may develop. The other presentations of mucocele may be intussusception or pseudomyxoma peritonei. In the treatment, benign tumor are cured with appendectomy, malign lesions need right hemicolectomy (1, 4).

In the diagnosis of mucinous neoplasms, abdominal radiography may show a soft-tisue mass in the right lower quadrant with curvilinear mural calcification in less than 50% of cases (5). At US, a cystic mass with or without calcification is seen. The differential diagnosis includes periappendiceal abscess, duplication cyst, mesenteric cyst, and hydrosalpinx (1). Francica et al. described the features of giant mucocele of the appendix at B-mode and contrast-enhanced sonographic examinations. They determined a thin hyperechoic border without either solid vegetan or sign of infiltration of surrounding tissue, a complex internal echo structure without anechoic lacunae, including curvilinear, wavy bands of echogenic material (onion skin sign), and avascularity on contrast-enhanced sonography (6). CT shows the anatomic relation and mural calcification more sensitively. Pseudomyxoma peritonei, and intussusception may be also diagnosed by US or CT (1).

CT findings of pseudomyxoma peritonei include a nonspecific ascites and a diagnostic signs of visceral scalloping which distinguishes mucinous from fluid ascites on CT (7, 8). Poorly adhesive mucin producing cells pool in the sites of relative statis like pouch of Douglas/rectovesical pouch, right and left subphrenic spaces and surface of the liver and spleen.

Mucosal hyperplasia and mucinous cystadenoma may be associated with hyperplastic poylps and adenomatous polyps (9). Mucinous

cystadenoma and colon cancer have been diagnosed with an incidence of up to 20% (10). That is why colonoscopy is needed when appendiceal mucinous cystadenoma is determined.

Appendiceal neuroendocrine tumors (carcinoids), represent up to 80% of appendiceal neoplasia, are found 1/100-300 appendectomies. Peak age is within the fourth decade. Epithelial tumours are seen in the adults older than the carcinoid group. Most of the carcinoids are asymptomatic and diagnosed after pathological examination. Although it has a malign potential, metastases and carcinoid syndrome are very rare (1). Because of their small size, carcinoid tumors of the appendix are not typically diagnosed by imaging. If they are metastatic, mesenteric or liver masses may be seen (11). Rioux et al showed sonographic finding of carcinoid tumor as a distal hypoechoic mass with a normal proximal wall of appendix in two cases (12). Imaging features show the signs of obstruction and appendicitis at CT and US. Mucocele rarely develops (13). Calcification may be seen but mimics appendicolith. Appendectomy is sufficient for the tumours less than 1.5-2 cm in size. When it is greater, right hemicolectomy is warranted (2, 14).

On the other hand, CT findings suggesting acute appendicitis could be seen in other inflammatory or neoplastic processes including Crohn's disease, right sided colon cancer (15, 16).

Except for carcinoid tumor, all our cases were in elderly population. Patient with mucocele was asymptomatic and incidentally diagnosed at US as a cystic mass. Patient with abdominal distention had pseudomyxoma peritonei result of the rupture of appendiceal mucinous cystadenoma. Both the patient with low-grade mucosal neoplasia and the patient diagnosed carcinoid tumor had symptoms and imaging features of acute appendicitis. However, we could not determine the carcinoid tumor of retrocecal localized appendix. Except this, our findings were correlated with the literature.

In conclusion, primary appendiceal neoplasms are rare clinical and pathological conditions. Especially in elderly group, neoplasms should be kept in mind, besides a simple obstruction and inflammation of appendix vermiformis by the radiologist and the surgeon as it may alter the surgical approach or it may give a way to diagnose the associated abnormalities (peritoneum, colon, over). After the operation the patient should be followed-up for any developing pathology including the colon, and peritoneum.

References

1. Pickhardt P.J., Levy A.D., Rohrmann C.A., Kende A.I.: Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. *Radiographics*, 2003, 23: 645-662.
2. Barry M., Collins C.G., McCawley N., McGunness J., Leahy A.L.: Synchronous appendiceal tumours. *Surgeon*, 2007, 5: 111-113.
3. Higa E., Rosai J., Pizzimbono C.A., Wise L.: Mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendiceal 'mucocele'. *Cancer*, 1973, 32: 1525-1541.
4. Rutledge R.H., Alexander J.W. Primary appendiceal malignancies: rare but important. *Surgery*, 1992, 111: 244-250.
5. Madwed D., Mindelzun R., Jeffrey R.B. Jr.: Mucocele of the appendix: imaging findings. *AJR*, 1992, 159: 69-72.
6. Francica G., Lapiccirella G., Giardiello C., Scarano F., Angelone G., De Marino F., et al.: Giant mucocele of the appendix: clinical and imaging findings in 3 cases. *J Ultrasound Med*, 2006, 25: 643-648.
7. Sulkin T.V., O'Neill H., Amin A.I., Moran B.: CT in pseudomyxoma peritonei: a review of 17 cases. *Clin Radiol*, 2002, 57: 608-613.
8. Dixit A., Robertson J.H.P., Mudan S.S., Akle C.: Appendiceal mucoceles and pseudomyxoma peritonei. *World J Gastroenterol*, 2007, 13: 2381-2384.
9. Qizilbash A.H.: Mucoceles of the appendix. Their relationship to hyperplastic polyps, mucinous cystadenoma, and cystadenocarcinomas. *Arch Pathol Lab Med*, 1975, 99: 548-55.
10. Lino H., Jass J.R., Simms L.A., Joung J., Leggett B., Ajioka Y., et al.: DNA microsatellite instabilty in hyperplastic polyps, serrated adenomas, and mixed polyps: a mild mutator pathway for colorectal cancer? *J Clin Pathol*, 1999, 52: 5-9.
11. Bittle M.M., Chew F.S.: Radiologic reasoning: recurrent right lower quadrant inflammatory mass. *AJR*, 2005, 185: S188-194.
12. Rioux M., Duchesne N., Langis P.: Carcinoid tumor of the appendix: ultrasound findings in two cases. *J Clin Ultrasound*, 1994, 22: 129-133.
13. Hermans J.J., Hermans A.L., Risseeuw G.A., Verhaaar J.C., Meradji M.: Appendicitis caused by carcinoid tumor. *Radiology*, 1993, 188: 71-72.
14. Moertel C.G., Weiland L.H., Nagomey D.M., Dockerty M.B.: Carcinoid tumor of the appendix: treatment and prognosis. *N Engl J Med*, 1987, 317: 1699-1701.
15. Checkoff J.K., Wechsler R.J., Nazarian L.N.: Chronic inflammatory appendiceal conditions that mimic acute appendicitis on helical CT. *AJR*, 2002, 179: 731-734.
16. Watchorn R.E., Poder L., Wang Z.J., Yeh B.M., Webb E.M., Coakley F.V.: Computed tomography findings mimicking appendicitis as a manifestation of colorectal cancer. *Clin Imaging*, 2009, 33: 430-432.