Primary malignant neoplasms of the cecal appendix: classification, forms of presentation and MDCT findings.

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Learning objectives

- To review the different types of primary malignant neoplasias of the cecal appendix.
- To assess the usefulness of MDCT in the diagnostic imaging of the appendiceal malignant neoplastic disease.
- To correlate clinical, radiological and pathological findings.

Background

Those primary malignant neoplasms of the cecal appendix constitute 0.2 to 0.5% of all the tumors of the digestive tube. We can divided them fundamentally into 2 groups:

- Adenocarcinomas.
- Carcinoid Tumors.

Approximately 30%-50% of all appendiceal neoplasms will manifest clinically with signs and symptoms of acute appendicitis but others manifestations include intussusception, a palpable mass, gastrointestinal bleeding...
Recognition of these neoplasms is important for appropriate patient treatment. CT appears to be the modality of choice whenever an appendiceal mass is suggested at physical examination.

Findings and procedure details

Malignant appendiceal tumors account for only 0.5% of all gastrointestinal tumors and there are usually diagnosed during the pathological study of excised appendices.

With the exception of carcinoid tumors, most appendiceal neoplasms are seen in adults who are middle-aged or older.

Although uncommon those tumors often result in clinical symptoms that may lead to abdominal imaging and acute appendicitis from luminal obstruction is the most common manifestation.

Others manifestations include intussusception, a palpable mass, gastrointestinal bleeding, increasing abdominal girth (from pseudomyxoma peritonei).
Detection of these neoplasms at preoperative imaging is important because it may change the surgical approach and obviate additional surgery.

Carcinoid tumors are common and account for 85% of epithelial appendiceal tumors followed in prevalence by mucinous adenocarcinomas and colonic-type (non mucinous) adenocarcinomas.

1. CARCINOID TUMOR:

- Classic carcinoid tumors of the appendix derive from subepithelial neuroendocrine cells and represent up to 80% of all appendiceal neoplasms.

- Its discovery at surgery or pathologic examination is most often casual because even in the setting of acute appendicitis this tumor cause an obstruction only in 25% of all cases, reflecting the fact that they are found in the distal third (over 70%) of the appendix and are less than 1 cm in size.

- The age is also unique: often seen in young female adults (fourth decade of life).

- Metastatic disease and carcinoid syndrome with an appendiceal primary site are exceedingly rare.

- Tumor size correlates with prognosis and a simple appendectomy is sufficient for most carcinoid tumors less than 1.5-2 cm in size. Tumors greater than 2 cm require right hemicolectomy.

CT findings: (fig.1)

- Relative paucity of imaging findings (relative small size, confinement - to the distal third, low complication rate).

- If it is near the base of appendix it will manifest at CT as acute appendicitis.

- Mucocele formation may occur (rare finding).

- The tumor itself may be discernible when it is of a sufficient size or demonstrates calcification (mimics a non tumoral calcification).

- Sometimes diffuse mural thickening of the appendix.

- Metastasic disease (although rare): characteristic irregular soft tissue mass near the mesenteric root.
2. MUCINOUS ADENOCARCINOMA:

Mucinous adenocarcinomas represent about 25% of all appendiceal adenocarcinomas and the gross morphology of these tumors is indistinguishable from that of mucocele. That patients present a right lower abdominal pain and/or a mass suggesting appendicitis. It is usually a well differentiated, slowly progressive neoplasm that shows a mass effect. Perforating of the affect organs results in pseudomyxoma peritonei. The term of pseudomyxoma peritonei describes intraperitoneal accumulation of gelatinous material with scalloping effect on solid organs (fig.5).

**CT findings: (fig.2, fig.3, fig.4)**

- Likely to form mucocele.
- Well-defined right low quadrant cyst mass (near water density) usually > 2cm diameter.
- Calcification (curvilinear) within wall or lumen (seen in less than 50% of cases).
- Thickened nodular wall.
- Non specific findings that suggest malignancy and/or secondary inflammation: soft-tissue thickening, irregularity of mucocele and surrounding fat.

3. COLONIC-TYPE (NONMUCINOUS) ADENOCARCINOMA:

Some studies reported up to 2:1 ratio of mucinous to colonic type adenocarcinomas. The latter tend not to form mucoceles. They mostly manifest clinically with appendicitis related with malignant tumoral obstruction. In the setting of suspected appendicitis in an older individual.

**CT findings:**

- Focal soft-tissue mass that involves the appendix but without mucocele formation.
- Subtle appendiceal mass with surrounding periappendiceal inflammation (may be mistaken for non tumoral appendicitis).

- Direct invasion of adjacent organs (mass in urinary bladder).

**Images for this section:**

![Fig. 1: Axial contrast material-enhanced CT shows a hypodense lesion with thickened enhanced wall and eccentric intraluminal calcification in the expected region of the appendix. Histologic analysis showed these findings to represent carcinoid tumor.](image-url)
**Fig. 2:** Axial contrast enhanced CT shows adenocarcinoma of the appendix: the wall of the appendix is thickened, the periappendicular fat is infiltrated and the appearance is that of a soft tissue density mass.
Fig. 3: Axial contrast-enhanced CT of adenocarcinoma (mucinous type): Nodular thickening of the appendiceal wall that enhanced after contrast administration with periappendicular fat stranding.
Fig. 4: Adenocarcinoma tumor of appendix (the same patient of fig.3)
**Fig. 5:** Axial CECT shows the classic scalloped appearance of the surface of the liver, due to the peritoneal gelatinous metastases, in this 80-year-old man with pseudomyxoma peritonei due to ruptured mucinous carcinoma of the appendix.
Conclusion

- Carcinoid tumors are the primary malignant neoplasms more frequent (80 %) of the cecal appendix.
- Clinic of these types of tumors generally simulate acute appendicitis or its complications.
- Cross-sectional imaging, particularly computed tomography (CT), is effective in the evaluation of these neoplasms.

- CT appears to be the modality of choice whenever an appendiceal mass is suspected, help to rule out or confirm an appendiceal tumor and may suggest a more specific diagnosis.

Personal information

References


