

## On Mucinous Appendiceal Adenocarcinoma: Report of Two Cases

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### Abstract

### Case Report

Tumors of the appendix are very rare neoplasms to detect. These neoplasms represent about 1% of the samples entering a surgical pathology laboratory. Current classifications distinguish neoplasia of the appendix in epithelial tumors, mesenchymal tumors and lymphomas. Epithelial tumors include neuroendocrine carcinoma and mucinous adenocarcinoma, this last type being the most common form of neoplasm of the appendix. In the present article two cases of appendicular mucinous adenocarcinoma are reported in a 85 year-old man and in a 49 year-old man, with similar clinical presentation.

**Keywords:** Mucinous adenocarcinoma, neoplasm, appendix.

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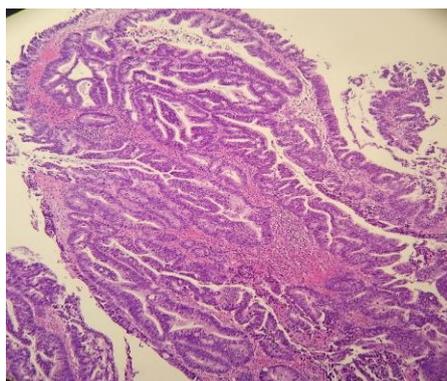
## INTRODUCTION

The appendix can be considered a rudimentary organ, representing the final part of the associated mucosal lymphoid tissue, whose initial part is found in the oral cavity. Its anatomical structure is composed by a cylindrical wall with an average length of 6 cm and a central lumen with three coats, the innermost of which presents lymphoid tissue [1]. The appendicular mucosa is of colic type associated with lymphoid tissue, seemly reactive during juvenile age. The epithelium consisting in a mix of absorbent cells, goblet cells, neuroendocrine cells and Paneth cells [2, 3]. Neuroendocrine cells can be detected with chromogranin and, occasionally, are identified by the presence in the histological samples of eosinophil cells with a showy cytoplasmic granules [3]. Many of the plasma cells present in the *lamina propria* produce immunoglobulin type IgA. It is possible that caecum carcinoma may involve the appendix in advanced stage. There are various types of neoplasms affecting the appendix, such as epithelial one, mesenchymal tumor and lymphoma [4]. Among the epithelial tumors, the most relevant are the tumors with a mucinous histotype and neuroendocrine carcinoma [5, 6]. The onset of a primitive adenocarcinoma of the

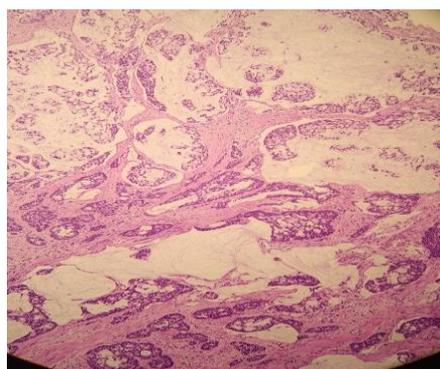
appendix is an extremely rare condition [7]. Symptoms associated with appendicular carcinoma are similar to those of an acute appendix, accompanied by inflammation of the soft tissue around the organ [8]. Immunohistochemical results of appendix carcinoma show positivity for several tumor epithelial markers such as CEA, CA 19-9, many cytocheratins, including CDX2 and Ca 125 too [9].

## CASE REPORT

In this report two rare cases of primitive adenocarcinoma of the appendix are described. The first case concerns a 85 years old man hospitalized for severe stomach ache and constant abdominal pain. Radiographic and TC scan have highlighted little peritoneal effusion, without evidence of mass enlargements. Patient was subjected to surgical excision of the appendix, with intraoperative histological examination. The histological sections performed at the cryostat showed a poorly differentiated mucinous adenocarcinoma, full-thickness infiltrating the muscular tunic and the peri-appendicular adipose tissue, with focal perforation of the serosa tunic (figures 1, 2).



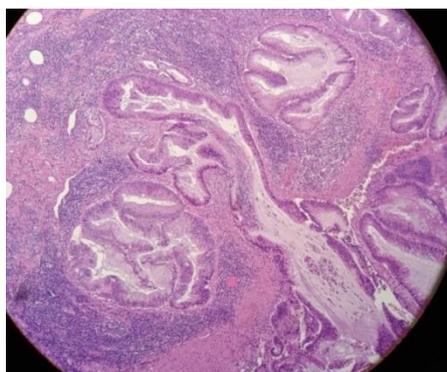
**Fig-1: E.E slide of surgical specimen**



**Fig-2: Mucinous histological aspect of the tumor**

The second case concerns a 49-year-old man who admitted to our hospital for symptoms of an acute perforated appendicitis. After the surgical removal, appendix was formalin fixed and sends in the anatomic pathology laboratory. Histopathological slides revealed a perforated primitive mucinous adenocarcinoma

(figure 3), confirmed by results of immunohistochemical tests, which showed positivity for CDX2 and CK20 (figure 4). Studies have been performed to investigate mismatch repair proteins status on neoplastic tissue and these were all expressed, so microsatellite instability should not occur.



**Fig-3: E.E. slide of appendicular carcinoma**



**Fig-4: Immunohistochemical detection of positivity for CK20 in tumor cells**

## DISCUSSION

Appendicular adenocarcinoma is a rare malignant neoplasm, characterized histologically by the presence of numerous cells in the mucus, which form flaps, and ring-shaped cells can be observed. Mucinous adenocarcinomas represent about 45% of all appendicular neoplasms [10]. This type of pathology has no well-defined clinical characteristics and the most frequent symptoms are abdominal pain, not even associated with evidence of abdominal or pelvic mass [11]. The standard tumor markers are CEA, CA19-9, and Ca1 25 [12]. The most common complication in patients with appendicular adenocarcinoma is the *pseudomyxoma peritonei*, due to the dissemination of the neoplastic mucin on the visceral and parietal peritoneum. It corresponds to the clinical involvement of peritoneal serosa from the appendicular neoplasm with widespread peritoneal mucinous tumor [13]. At the origin of the molecular changes characterizing the neoplasia, we can mention the instability of the microsatellites, caused by the DNA repair defect systems (mismatch repair MMR) [14]. The recommended treatment is surgery through right hemicolectomy and a combined adjuvant intraperitoneal chemotherapy [15].

## CONCLUSION

Malignant tumors of the appendix are very rare. They lack specific clinical symptoms or serological markers, because the most important clinical presentation mimics acute appendicitis. Then, most of the patients affected from appendicular carcinoma don't present evidence of mass enlargement on radiographic exams. Only macroscopic and microscopic examination of the appendix after surgical removal can detect carcinoma, above all on slides set up for routine histological diagnosis.

For this reason, most of cases are diagnosed in advanced stage of pathology, when tumor is perforated and disseminated in peritoneal serosa. Hence the importance of a correct clinical management of gastrointestinal symptoms, through a training of sanitaris aimed at selecting patients worthy of serological and instrumental diagnostic investigations before the increase in pain symptoms, with a consequent important worsening of the prognosis.

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