

# Adenocarcinoid tumour of the appendix masquerading as acute appendicitis: a word of caution

## Apendiksin akut apandisit şeklinde maskelenen bir adenokarsinoid tümörü: Uyarı notu

Jawad AHMAD,<sup>1</sup> Syed I. H. ANDRABI,<sup>2</sup> D. K. THEKKINKATTIL,<sup>1</sup> Munir A. RATHORE<sup>3</sup>

Adenocarcinoids are rare and aggressive tumors with histological features of both carcinoid tumor and adenocarcinoma. We report a case of a 32-year-old man with diffuse infiltration of the appendix wall and mesoappendix by an adenocarcinoid. Due to the involvement, a hemicolectomy was performed at a later date. We suggest that patients with diffuse appendiceal involvement require an aggressive surgical therapy and follow-up.

**Key Words:** Adenocarcinoid tumour; adenocarcinoma; acute appendicitis; carcinoid tumor.

Adenokarsinoid tümörler, karsinoid ve adenokarsinomaların histolojik özelliklerini taşıyan ve nadir görülen agresif tümörlerdir. Bu yazıda, apandiks duvarı ile mezoapandiksin bir adenokarsinoid tümör tarafından difüz şekilde infiltre edildiği, 32 yaşındaki bir erkek hasta sunulmaktadır. Tutulum nedeniyle, daha sonraki bir tarihte bir hemikolektomi ameliyatı gerçekleştirildi. Biz, difüz apandiks tutulumu olan hastaların agresif bir cerrahi tedavi ve takip gerektirdiğini düşünüyoruz.

**Anahtar Sözcükler:** Adenokarsinoid tümör; adenokarsinom; akut apandisit; karsinoid tümör.

Appendiceal tumours constitute about 0.5% of all gastrointestinal malignancies.<sup>[1]</sup> Adenocarcinoid tumours of the appendix are extremely rare<sup>[2]</sup> and are more aggressive than conventional carcinoid tumours.<sup>[3]</sup>

We present our experience about appendiceal adenocarcinoid with a brief literature review.

### CASE REPORT

A 32-year-old man was admitted with one day history of severe abdominal pain. Pain started in the periumbilical region and then shifted to the right iliac fossa. It was constant and severe. His pain was associated with anorexia, nausea and one episode of vomiting.

On examination, he had low grade pyrexia of 37.7°C with a pulse rate of 100 beats per minute. Abdominal examination revealed tenderness mainly in the right side of the abdomen with guarding and rebound tenderness. His blood investigation showed a raised white cell count of 16.700 per mm<sup>3</sup> with neutrophilia. C-reactive protein (CRP) was 135.

A clinical diagnosis of acute appendicitis was made and he underwent an appendicectomy. Per-operatively a large amount of mucoid fluid around a gangrenous and perforated appendix was found. Appendicectomy was performed with a generous abdominal lavage. The patient made a smooth post operative recovery and was discharged on third post operative day.

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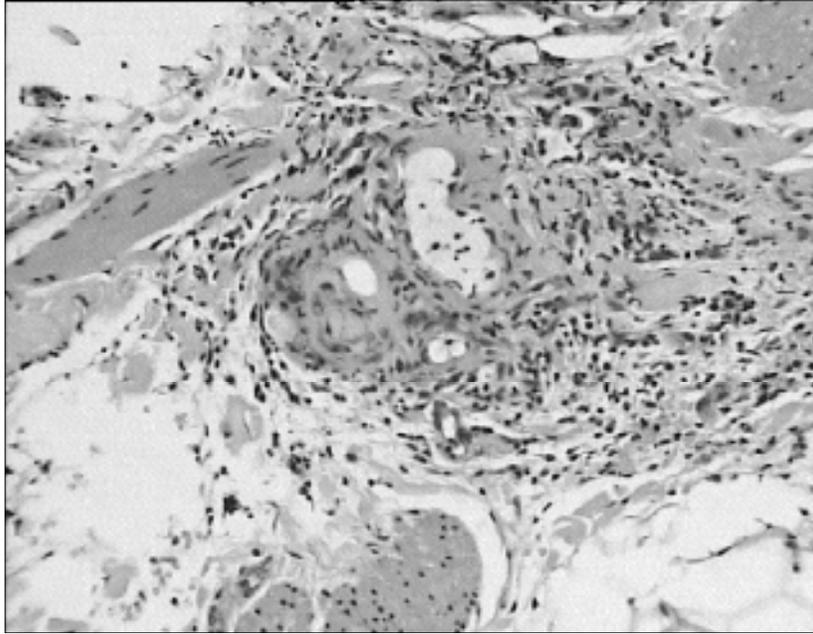
<sup>1</sup>Friarage Hospital, Northallerton, North Yorkshire, <sup>2</sup>Craigavon Area Hospital, Portadown, <sup>3</sup>Antrim Area Hospital, Antrim, U.K.

<sup>1</sup>Friarage Hastanesi, Cerrahi Kliniği, Northallerton, North Yorkshire;

<sup>2</sup>Craigavon Hastanesi, Cerrahi Kliniği, Portadown; <sup>3</sup>Antrim Hastanesi, Cerrahi Kliniği, Antrim, İngiltere.

Correspondence (İletişim): Syed I. H. Andrabi, FRCS. 73 Sicily Park, Belfast, United Kingdom.

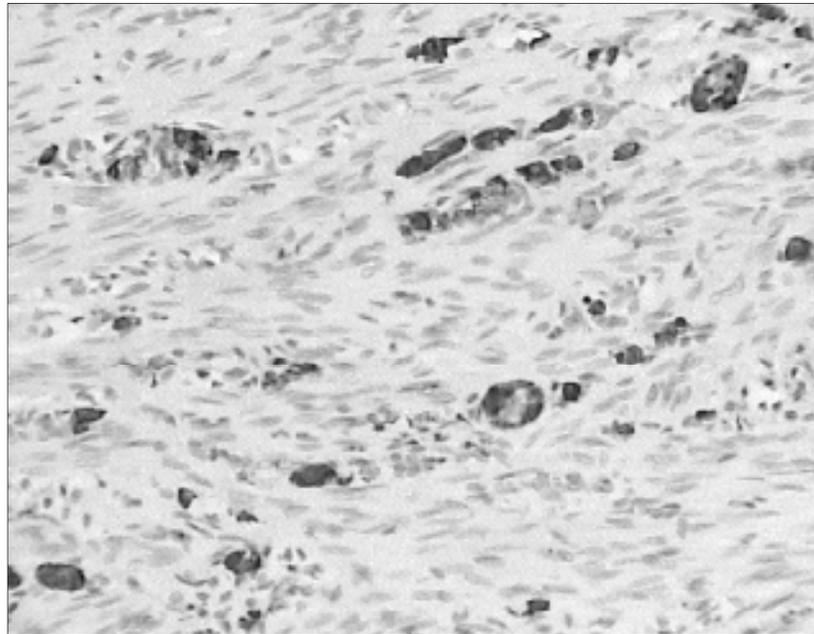
Tel: +44 - 7878358260 e-mail (e-posta): imranandrabi@gmail.com



**Fig. 1.** Adenocarcinoid of appendix with haematoxylin-eosin slide showing tumour cells infiltrating through the muscularis propria. Note surrounding inflammatory change.

Microscopic examination of the specimen showed acute suppurative appendicitis with gangrenous changes. Middle third of appendix showed thickening of the wall with goblet cells (Fig. 1).

Stains were positive for mucin and carcinoid granules (Fig. 2). The final pathological diagnosis was an 18x20 mm adenocarcinoid of the appendix with invasion into the surrounding mesoappendix.



**Fig. 2.** Adenocarcinoid of appendix with special immunohistochemical staining for cytokeratin 20 (CK20).

Following this pathological report a staging abdominal and thoracic computerized tomography (CT) did not reveal any metastatic disease. In view of the aggressive pathology and mesoappendiceal invasion, he underwent a right hemicolectomy 6 weeks following his initial appendicectomy. The histological examination of hemicolectomy specimen showed no metastatic tumour in the mesenteric lymph nodes or any evidence of a residual tumour. At 12 months the patient remains disease free.

## DISCUSSION

Appendiceal tumours are rare, contributing 0.5% of all gastrointestinal malignancies.<sup>[1]</sup> Most are carcinoid tumours (85%). They are usually incidental findings after appendicectomy.<sup>[4]</sup> Most tumours (75%) occur at the tip of the appendix and are <2 cm in diameter. Goblet cell tumours (adenocarcinoid) are distinct from carcinoid and adenocarcinoma. These are extremely rare and few than 600 have been reported in the world literature.<sup>[5]</sup> Like carcinoid tumours, goblet cell carcinomas probably develop from APUD (amine precursor uptake and decarboxylation) cells. However, they contain nests of goblet cells and exhibit a different pattern of spread (intramural) than carcinoids. Typically they spare the lining mucosa and stain positive for carcinoembryonic antigen and cytokeratin.<sup>[6]</sup> The mean age of diagnosis is 59 years and incidence is equal for men and women. Most patients present with signs and symptoms of an acute appendicitis due to luminal obstruction. The tumour cells proliferate sparsely and the appendiceal wall thickens diffusely with fibrous proliferation. This narrows the appendix lumen eventually leading to obstruction and acute appendicitis. Rare presentations include bleeding, intussusception and the presence of a mass.

Appendectomy alone could be used for appendiceal adenocarcinoid provided that the tumor is less than 1 cm, does not extend beyond the appendix adventitia, has less than 2 mitoses/10 high power fields and has surgical margins that are tumor free.<sup>[7,8]</sup> However, as 20% of patients will have metastasis noted on post appendicectomy staging, the tumours clearly have malignant poten-

tial. Clinicians have to decide if further adjuvant surgical therapy (right hemicolectomy) or chemotherapy<sup>[9]</sup> is required following the initial appendicectomy. Survival following resection is 60% at 10 years.<sup>[10]</sup>

## CONCLUSION

Adenocarcinoid tumours are rare tumours of the appendix. They differ from more common carcinoid tumours in their position, mode of presentation, metastatic potential and local involvement. This case highlights that tumours arising from the appendix may present as acute appendicitis. Careful pathological examination of appendicectomy specimens is always warranted as further surgery and chemotherapy may be necessary.

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