

Case Report

Primary Signet Ring Cell Carcinoma of the Appendix: A Rare Case Report

(Appendiksin Primer Taşlı Yüzük Hücreli Karsinomu: Olgu Sunumu)

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ÖZET

Apendikte primer taşlı yüzük hücreli karsinom oldukça nadir görülmekte olup, tüm appendiks tümörlerinin %0,5'den azını oluşturur. Sunduğumuz olguda akut apendisit ön tanısı nedeniyle operasyona alınan ve apendikte kitle görülmesi üzerine sağ hemikolektomi yapılan hastanın, sağ hemikolektomi materyalinin patolojik incelemesinde apendiksi diffüz tutan kas tabakaları arasında infiltrasyon gösteren iri hiperkromatik nükleuslu taşlı yüzük görünümünde atipik epitelyal hücrelerden oluşan taşlı yüzük hücreli karsinom izlenmiştir. Olgumuzun apendikte oldukça ender görülmesi ve prognozunun appendiksin klasik adenokarsinomlarından kötü olması nedeni ile sunulmaya değer bulunmuştur.

Anahtar kelimeler: Apendiks; primer; taşlı yüzük hücreli karsinom

ABSTRACT

Primary signet ring cell carcinoma is rarely seen in appendix and accounts for less than 0.5% of all tumors of the appendix. In our case due to the diagnosis of acute appendicitis, patient was operated and right hemicolectomy was performed when a mass in the appendix was observed. Pathology of right hemicolectomy revealed signet ring cell carcinoma that large hyperchromatic nuclei with signet ring appearance of atypical epithelial cells which diffusely infiltrated muscle layers. We presented the case because it's occurrence in appendix is very rare and prognosis is poorer than the classical adenocarcinomas of appendix.

Key Words: Appendix; primary; signet cell carcinoma

INTRODUCTION

Primary carcinoma of the appendix is rare, and accounts for less than 0.5% of the entire gastrointestinal tumors. Primary signet ring cell carcinoma of the appendix is so rare and creates only 0.43% malignancies of appendix¹. Primary appendiceal adenocarcinoma is defined by Berger in 1882 for the first time, it consists of 0.1% to 1.35% of appendectomy samples and number of cases reported so far is less than 450². In a large epidemiologic study 1645 cases of malignancy in patients which performed appendectomy were identified. 613 cases as mucinous adenocarcinoma, 411 cases as colonic adenocarcinoma, 324 cases as malignant carcinoid tumor, 227 cases as goblet cell carcinoid tumor and signet ring cell carcinoma were observed only in 70 cases³. Clinical findings are non-specific and usually occurs in the statement of acute appendicitis.

Therefore, preoperative tumor diagnosis is difficult. Diagnosis is usually done by histopathological examination after surgical removal of the inflamed appendix⁴.

In our case, 61 years old man due to the right lower quadrant abdominal pain with the diagnosis of appendicitis invoiced surgery and right hemicolectomy was performed due to suspected tumor of the appendix. In the examination of the hemicolectomy material, cecum was dilated in a diameter of 4 cm, surrounding the appendix gray-white solid tumor mass was observed in the form of linitis plastica. Hematoxylin-eosin-stained sections of the appendix revealed that appendiceal tumor infiltrated mucosa, submucosa and muscularis propria and serosa (Figure 1).

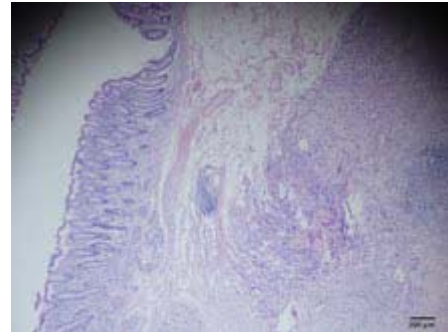


Figure 1. Tumor has damaged the appendix's submucosa in most areas (H&Ex40)

In tumoral tissue, we observed large mucin infiltration pools formed in the case of single cells

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and clusters and that make up adenoid-like sequences in a few areas, hyperchromatic nuclei located peripherally, large vacuolated cytoplasm is composed of signet ring cells (Figure 2).

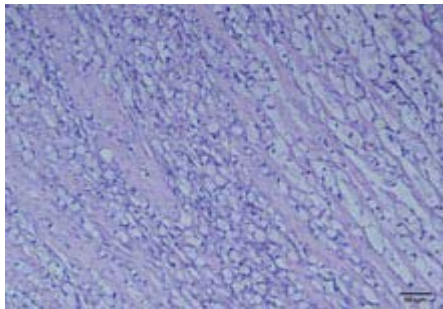


Figure 2. Muscular layers in the signet ring cells and mucin pools (H&Ex200)

Immunohistochemical examination of tumor is pan-CK diffusely positive staining was observed (Figure 3). Histochemical examination is positive for PAS ve musikarmen positive were showed.

Peritoneal tumoral infiltration called as pseudomyxoma peritoni was identified in the peritoneal surfaces.

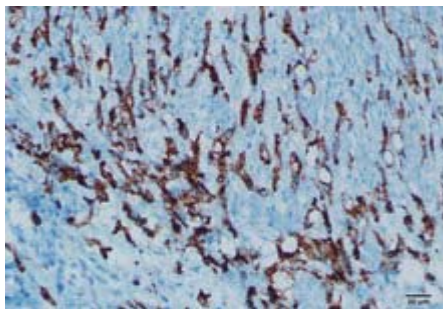


Figure 3. Pan-CK positivity in the signet ring cells (x400)

Tumoral serosal involvement, common peripheral nerve invasion, lymphovascular invasion was observed, the 16 number of dissected lymph node metastasis was found in 3. Appendix of primary signet ring cell carcinoma tumors are very aggressive disease course and have poor prognosis. Generally, patient is in the advanced stage when diagnosis is done.

DISCUSSION

The International Classification of Disease for Oncology (ICD-O) divides the tumors of appendix into five categories: colonic type adenocarcinoma, signet cell adenocarcinoma, mucinous adenocarcinoma, goblet cell adenocarcinoma, and malignant carcinoid/adenocarcinoid^{1,4}. Most appendiceal tumors are low-grade neoplasm which are typically relatively indolent. Mucinous appendiceal adenocarcinomas reported by Park et al⁵ is 20.5%. Appendiceal mucinous adenocarcinomas are overall 5-year survival rate. According to report of Mc Cusker et al³, except for malignant carcinoid and signet ring cell carcinoma, the histologic type does not have a important impact on survival. Prognosis of the tumor is the extent of disease more important predictor of survival than histology. Right hemicolectomy is considered the optimal treatment for most histologic types of primary appendiceal carcinoma even in the presence of perforation and in Dukes A Tumors. Metastatic disease treatment options are systemic chemotherapy with hyperthermic intraoperative intraperitoneal chemotherapy and peritonectomy with cytoreductive surgery^{3,4}.

Appendix of signet ring cell carcinoma tumors the differential diagnosis are clinically important since the therapeutic approach. Ovarian tumors with signet ring cell histology are sometimes Krukenberg metastases from appendiceal cancers. Kiyokawa et al.⁶ reported 120 cases of Krukenberg tumors of the ovary and mentioned that the primary tumor was diagnosed only 1 of 6 cases of primary appendiceal tumor with a Krukenberg tumor. Our patient was excluded of ovarian tumors for male. In addition, pathologic examination of the appendix demonstrated that the mucosal surface was totally involved by tumor.

In conclusion, signet ring cell carcinoma which has an extremely poor prognosis is usually diagnosed in advanced stage, in the diagnostic procedure exclusion of the carcinoma subtypes distinction is of particular importance.

Conflict of interest statement : None declared.

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