



Multiple Primary Malignant Neoplasms in the Presence of Concomitant Chilaiditi Syndrome

Chilaiditi Sendromu ile Eş Zamanlı Çoklu Birincil Malign Neoplazmlar

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ABSTRACT

Chilaiditi syndrome, jejunal diverticulosis, and multiple primary malignant neoplasms (MPMN) are all relatively rare entities. In this study, clear cell renal cell carcinoma together with adenocarcinoma arising in the rectum were confirmed in an 82-year-old patient. Jejunal diverticulosis and Meckel diverticulitis were detected intraoperatively. Following radiotherapy with 25 Gy delivered in 5 fractions over 1 week, the patient underwent partial nephrectomy as well as abdominoperineal rectal resection and cholecystectomy due to cholelithiasis. The immediate postoperative period was uneventful, but the patient died of myocardial infarction on postoperative day 4. Malignancy may accompany Chilaiditi syndrome. In the presence of MPMN, appropriate treatment must be chosen individually on a case-to-case basis. Although there is no treatment for asymptomatic jejunal diverticulosis and Meckel diverticulitis, it should always be kept in mind that they may cause potentially serious complications.

Keywords: Multiple, primary neoplasms, Chilaiditi syndrome

ÖZ

Chilaiditi sendromu, jejunal divertikülozis ve multipl birincil malign neoplazmlar (MBMN) sıklıkla nadir antitelerdir. Bu, 82 yaşında olan bir hastanın berrak hücreli renal karsinomunun rektum kökenli adenokarsinom ile birlikteliğinin teyit edildiği bir çalışmadır. Ameliyat esnasında jejunal divertikülozis ve Meckel divertikülü tespit edilmiştir. Bir haftalık sürede beş seansta verilen 25 Gy radyoterapiyi müteakiben, parsiyel nefrektomi, abdomino-perineal rezeksiyon ve kolelithiazis nedeni ile kolesistektomi uygulanmıştır. Ameliyat sonrası 4. gün herşey normal seyrinde giderken hasta miyokardiyal enfarktüs nedeniyle kaybedilmiştir. Chilaiditi sendromuna maligniteler eşlik edebilir. MBMN varlığında her hastaya göre uygun tedavi yöntemleri seçilmelidir. Asemptomatik jejunal divertikülozis ve Meckel divertikülü için herhangi bir önlem alma gerekliliği yok iken bunların ciddi komplikasyonlara yol açabileceği akılda tutulmalıdır.

Anahtar Kelimeler: Çoklu, biricil neoplazm, Chilaiditi sendromu

Introduction

Chilaiditi syndrome is a rare disease in which the bowel (usually transverse colon or hepatic flexure) or the small intestine is found interposed in between the diaphragm and the liver, and was first described in 1910 by the Greek radiologist Demetrius Chilaiditi.¹ The frequency of this anomaly in proportion to the general population is 0.025-0.28%. The incidence of increasing multiple malignant tumors is a critical challenge for the clinician and great attention should be paid to avoid misdiagnosis. Besides, early diagnosis is of crucial importance for scheduling a radical treatment method. Treatment approach should attentively be

selected for each patient based on structural discrepancies.² Though typically asymptomatic jejunal diverticulosis is a rare disease, it may give rise to chronic symptoms and acute complications.³

Case Report

Eighty two years-old male patient suffering from rectal bleeding for the last 6 weeks. In the history of the patient, he was diagnosed with hypertension and cardiac arrhythmia. In colonoscopic examination, a mass lesion starting on the 2nd cm of the rectum where the lumen was found to narrow down significantly was detected. Extensive diverticulosis existed



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along the entire colon. Colonoscopic biopsy confirmed the existence of adenocarcinoma. X-ray of the chest showed free air under the right diaphragm pertaining to intestinal loops (Figure 1a). On thoraco-abdominal computed tomography (CT) evaluation, it was observed that the lumen became narrowed due to a mass lesion affecting approximately 12 cm of the rectum (Figure 1b). In addition, a mass lesion of 33x28 mm giving the impression of renal cell carcinoma (RCC) in the inferior pole of the right kidney was seen (Figure 1c). Again on CT, structures pertaining to the intestine lain between the right diaphragm and liver were found. Multiple calculi were observed in the gallbladder. Blood values were detected as follows: Hemoglobin 12.4 g/dL, hematocrit 38.8%. Other blood values were within the reference ranges. Twenty five Gy in five fractions in one week radiotherapy was planned for the patient who was envisaged unable to receive chemotherapy as a result of the multidisciplinary oncology consultation.⁴ The patient was operated on the 3rd day after the radiotherapy was finalized. Abdominoperineal rectal resection along with cholecystectomy was applied. Additively, partial nephrectomy for right-sided RCC was performed by the urology team. Uncomplicated extensive jejunal diverticulosis and meckel diverticulitis were seen (Figure 2a, 2b). The mesentery of the colon and the small intestine appeared to be mobile and the intestinal loops were located between the liver and the right diaphragm (Figure 2c). On pathological examination, two separate primary

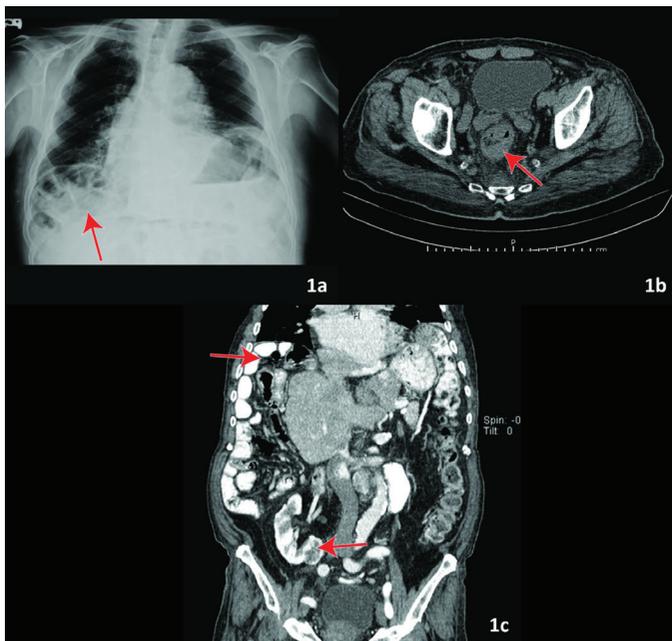


Figure 1. a) On anterior-posterior chest X-ray, the elevation of the right hemi-diaphragm and air under the diaphragm (indicated by the arrow), b) pelvic cut of the computed tomography (CT) scan showing thickened rectal wall (indicated by the arrow), c) CT scan coronal view showing the right-sided renal cell carcinoma and Intestinal loop transposed between the diaphragm and the liver (indicated by the arrow)

malignant tumors were reported. Adenocarcinoma in the rectum and clear cell RCC in the kidney were confirmed. On the post-operative 4th day, the patient died because of myocardial infarction.

Discussion

Typically asymptomatic Chilaiditi sign is named Chilaiditi syndrome in the presence of concomitant clinical symptoms. The frequency of Chilaiditi syndrome rises with increased age.⁵ Conservative treatment methods and surgical intervention are practiced for the management of this disease. Since Chilaiditi syndrome is a rare entity and highly associated with malignancies, it is generally misdiagnosed in clinical practice.¹ Treatment and diagnosis principles of multiple primary malignant neoplasms (MPMN) differ from metastasis or recurrent cancers. On patients with MPMN, the treatment method should carefully be chosen and planned based on the clinical status of each individual patient.² Results from the Swedish Rectal Cancer Trial demonstrate that the application of short-term pre-operative radiotherapy with a high-dose regimen lowers the rate of local recurrence by roughly 65% after at least two years of follow-up.⁴ Due to the occurrence of rectal bleeding with our case, the risk of blockage by the lesion where the rectum was found to narrow down and the existence of two separate primary

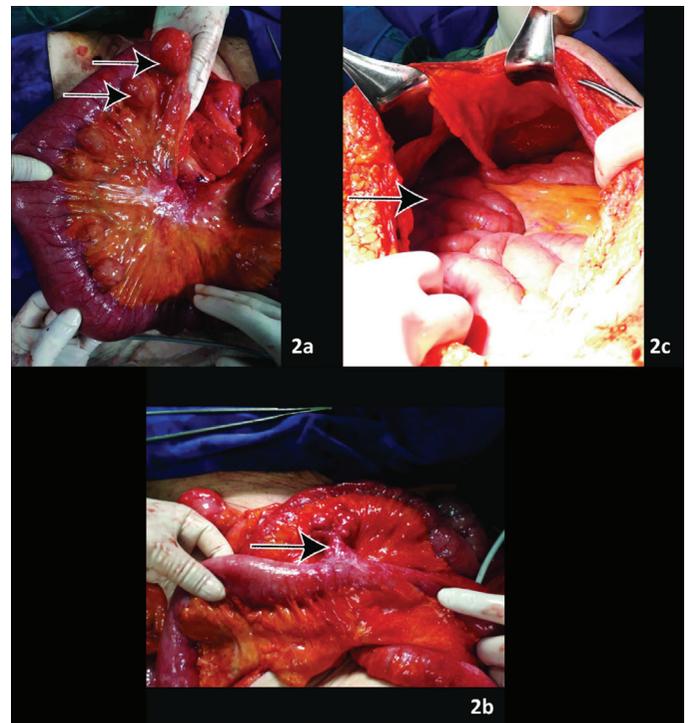


Figure 2. a) Multiple diverticula arisen at the mesenteric border of the jejunum (indicated by the arrow), b) meckel diverticulum arising from terminal ileum (indicated by the arrow), c) intestinal loop transposed between the diaphragm and the liver (surgical images) (indicated by the arrow)

tumors in the right kidney, early abdominoperineal resection for rectum cancer following a short-term radiotherapy was preferred in order not to cause a delay in initiation of the treatment.⁶ The technique of nephron-sparing surgery has evolved over the past two decades and has become an established surgical treatment for small renal masses rather than the traditional method of radical nephrectomy for clear cell RCC. In progress of time, improvements in the techniques and indications for partial nephrectomy have made it an alternative to radical nephrectomy.⁶ There is no requirement for intestinal resection on patients with asymptomatic jejuno ileal diverticulosis.³ Through imaging or laparotomy, when proximal jejunal diverticula are coincidentally identified, close follow-up is necessary as the diverticula may cause serious complications. In conclusion, although Chilaiditi syndrome is a rare entity, malignancies may accompany with this disease. In the event of MPMN, a special treatment method peculiar to each individual patient should be chosen. As with our case, when uncomplicated jejunal diverticulosis and meckel diverticulitis are verified, no action can be taken for these conditions. However, during follow-up, it should be taken into consideration that these situations can cause such serious complications as rectal bleeding, infection and perforation.⁷

Ethics

Informed Consent: Obtained.

Peer-review: External and internal peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.E., Concept: A.P., Design: A.E., Data Collection or Processing: A.P., Analysis or Interpretation: A.E., Literature Search: A.P., Writing: A.E., A.P.

Conflict of Interest: No conflict of interest was declared by the authors.

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