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A case of diffuse nodular lymphoid hyperplasia

Diffüz nodüler lenfoid hiperplazili bir olgu

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Diffuse nodular lymphoid hyperplasia represents a rare disease that is grossly characterized by the presence of numerous visible mucosal nodules measuring up to, and rarely exceeding, 0.5 cm in diameter. These may involve the entire small intestine, the large intestine, or both. The etiology is unknown. When diffuse nodular lymphoid hyperplasia is found predominantly in the colon, it can mimic a variety of polyposis syndromes and this may cause difficulties in diagnosis. The disease may be associated with other pathologies, especially gastrointestinal malignancies. This causes controversy when deciding the treatment options. Following patients without any treatment may lead to malignant progression, while surgical treatment may result in unnecessary radical resections because of obscurity in the diagnosis. We report here a diffuse nodular lymphoid hyperplasia case who underwent a radical resection because of obscurity in the diagnosis.

Diffüz nodüler lenfoid hiperplazi, oldukça ender rastlanan, etiyolojisi bilinmeyen, nadiren 0,5 cm'den büyük olan sayısız mukozal poliplerin oluşturduğu bir patolojidir. Lezyonlar tüm ince barsaklarda, kolonda veya her ikisinde birden görülebilmektedir. Diffüz nodüler lenfoid hiperplazi tanısı özellikle kolonik lezyonlarda polipozis sendromları ile karışabileceği için zorluklar göstermektedir. Hastalık, başta gastrointestinal maligniteler olmak üzere, başka patolojilerle birliktelikler gösterebilmektedir. Bu durum tedavi seçeneği hakkında karar vermeyi zorlaştırmaktadır. Hastalığın tedavisiz takibi malignite gelişme riski taşımaktayken, cerrahi tedavi de tanıdaki yanılma payı nedeni ile gereğinden fazla radikal rezeksiyonlara yol açabilmektedir. Bu olgu sunumunda ameliyat öncesi endoskopik ve patolojik tanılardaki belirsizlik sonucu radikal rezeksiyon yapılmak zorunda kalınmış bir diffüz nodüler lenfoid hiperplazi olgusu tartışılmıştır.

INTRODUCTION

Diffuse nodular lymphoid hyperplasia (DNLH) is a very rare disease of unknown etiology. The characteristic of DNLH is the presence of numerous visible mucosal nodules in the small intestine, colon, or both. Mucosal polypoid lesions rarely exceed 0.5 cm in diameter. The presence of the lesions in the colon may mimic a variety of polyposis syndromes, and this may cause some problems in diagnosis and treatment.

Histologically in DNLH, there is enlargement of the mucosal B cell follicles caused by hyperplasia of the follicle centers. These hyperplastic follicles are confined to the mucosa and surrounded by a normal appearing mantle zone. DNLH may be associated with intestinal lymphoma with or without dysgammaglobulinemia (1-6). In children, it is reported to be related to a condition of delayed type food hypersensitivity (7).

CASE REPORT

An 18-year-old male presented with an 11-year history of recurrent lower gastrointestinal bleedings. Physical examination revealed no significant features. All laboratory tests were normal including blood count, erythrocyte sedimentation rate, and biochemical and immunological parameters. Colonoscopy showed innumerable polyps in the rectum and sigmoid and descending colons, while transverse and ascending colons were reported as normal.

The histopathological diagnoses of the multiple materials obtained from colonic polyps were acute nonspecific inflammation and edema. An extended left hemicolectomy was planned due to recurrent gastrointestinal bleeding and polyposis of the colon. Multiple polyps were observed in the resection margin in the operation, in conflict with the colonoscopy report, and thus total colectomy with an

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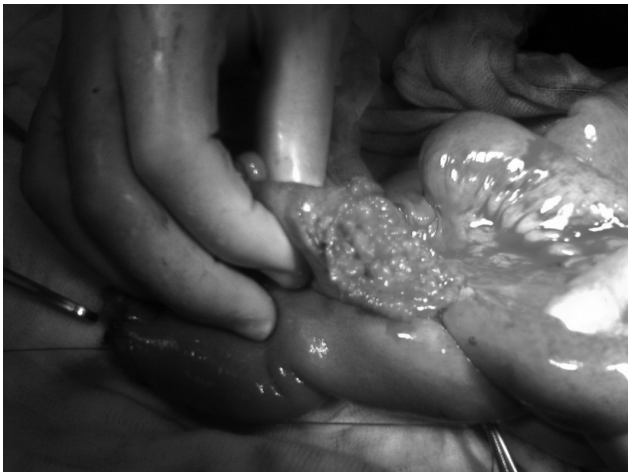

Figure 1. Macroscopic appearance of the multiple polypoid structures in the ileum.

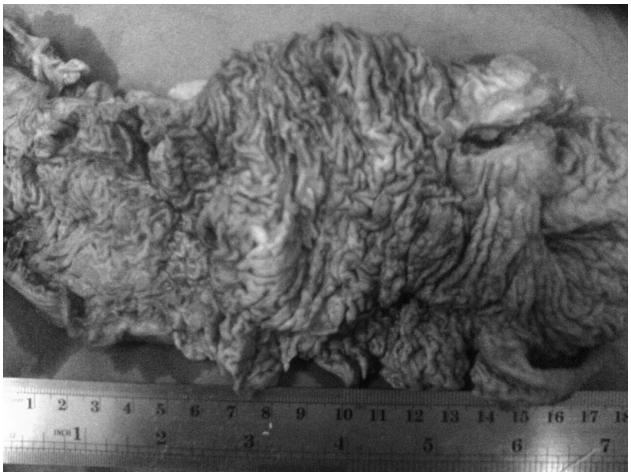

Figure 2. Macroscopic appearance of the multiple polypoid structures in the colon.

ileo-anal pouch was performed. Multiple millimetric polyps were noted in the ileal segment also (Figure 1). The macroscopic examination of the whole colon revealed innumerable polyps located in all segments including the cecum, and final histopathological diagnosis was DNLH (Figures 2, 3). At 12 months follow up, the patient was doing well and had no bleeding clinically.

DISCUSSION

Diffuse nodular lymphoid hyperplasia is a rare condition frequently associated with increased risk of gastrointestinal tumors, mainly gastrointestinal lymphoma (1, 8). The pathogenesis of DNLH is unknown, but is likely related to plasma-cell precursors due to a maturational defect in the development of B-lymphocytes in order to compensate for functionally inadequate intestinal lymphoid tissue (2, 8, 9). Intestinal lymphoid hyperplasia has been divided into DNLH and focal forms involving the terminal ileum or rectum (2). The present case belongs to DNLH type, but it revealed an unusual clinical presentation that included recurrent gastrointestinal bleedings that started 11 years ago. There is only one report about massive gastrointestinal bleeding caused by DNLH, and the diagnosis was made by ruling out other pathologies (10). In our case, bleeding was not massive but recurrent.

Diffuse nodular lymphoid hyperplasia has been described in healthy adults and children as well as in patients with common variable immunodeficiency syndrome, gastrointestinal lymphoma, Giardia infection and chronic diarrhea (1, 3, 11, 12). Our patient had no signs of any of the associated disorders. This variability in clinical symptoms usually causes delays in diagnosis or misdiagnosis.

The endoscopic diagnosis can be problematic. When DNLH is found in the colon, the endoscopic appearance can be strikingly similar to that of polyposis syndromes including familial adenomatous polyposis, multiple lymphomatous polyposis, juvenile or hamartomatous polyposis and hyperplastic polyposis, among others (9). In addition, the 2-3 mm polyps can be misdiagnosed at colonoscopy and may lead to over-treatment. Therefore, the histopathological diagnosis is the most important factor to determine DNLH. Although the disease is expected to have millimetric lesions, Chandra

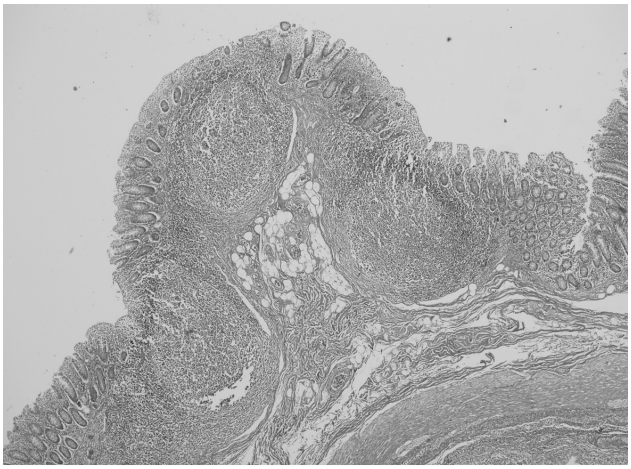

Figure 3. Hyperplastic lymphoid follicles with large germinal centers are seen in the lamina propria and superficial submucosa of the colon (hematoxylin-eosin stain, x40).

(13) reported two cases in which nodular lymphoid hyperplasia caused intestinal obstruction.

The clinical significance of DNLH lies in the possibility of these nodules serving as a nidus for prolapse and intussusception and in the association with immunosuppressive states (14). Spontaneous remission and resolution following chemotherapy for lymphoma is also reported (11,14). In addition, the probability of association with malignancies makes the decision of therapeutic modality controversial. DNLH is especially reported to be associated with colonic adenocarcinoma and lymphoma (1,4,6). Koren et al. (6) reported a case with small bowel lymphoid hyperplasia who had two different adenocarcinomas in the colon, leading us to consider DNLH as a systemic problem. On the other hand, when clinically symptomatic, the therapeutic modality is not clear. If the disease has a systemic basis, then local resections would increase the morbidity without sufficient benefits.

In conclusion, DNLH is a rare but problematic disease with respect to diagnosis and treatment. However, it must be distinguished from a variety of polyposis syndromes to avoid misdiagnosis and unnecessary radical treatments. The potential of malignancies associated with DNLH appears to be the most conflicting factor when deciding on the therapeutic modality.

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