

Primary Appendiceal Lymphoma Presenting as Acute Appendicitis : A Case Report¹

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Because primary lymphoma of the appendix is a very rare disorder and commonly presents as acute appendicitis, it is seldom diagnosed by preoperative imaging studies. We encountered a patient with pathologically proven primary appendiceal lymphoma associated with acute and chronic appendicitis. Ultrasonography revealed a non-compressible sausage-shaped hypoechoic mass with a linear hyperechoic center caused by the mucosa-lumen interface in the right lower quadrant. Post-contrast CT examination showed a markedly enlarged target-like appendix with obliteration of the lumen; the outer layer showed higher attenuation than the central portion. There were also multiple strands in the periappendiceal fat and thickening of adjacent lateroconal fascia and the colonic wall, and this suggested acute appendicitis associated with appendiceal lymphoma.

Index words : Appendix, neoplasms
Appendix, US
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Primary lymphoma of the appendix is a very rare disease that may cause clinical signs of appendicitis (1-3). Because preoperative imaging studies are seldom performed in this disease, the diagnosis is usually made by a pathologist after examination of an appendectomy specimen. Consequently, appendiceal lymphoma has received little mention in the imaging literature and radiologic textbooks(2-4). With the increasing use of imaging modalities, particularly ultrasound and CT, for the evaluation of patients with right lower quadrant pain, the imaging findings of this disease, which may clinically mimic acute appendicitis, are becoming important to radiologists. We report a case of primary lymphoma of the appendix presenting as acute appendicitis, and describe the findings of

ultrasonography and CT.

Case Report

A 54-year-old male was admitted after a week of diffuse abdominal pain and fever of up to 38.5 °C. Physical

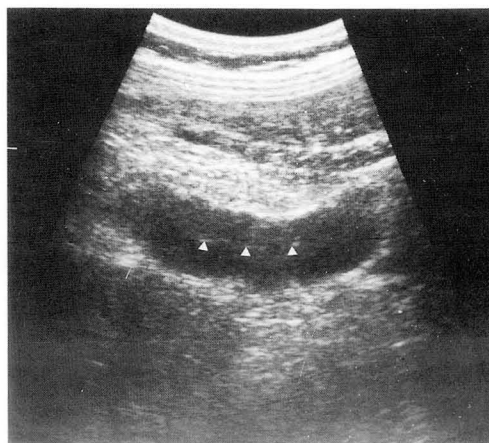


Fig. 1. Ultrasonogram of appendix. Longitudinal section shows a sausage-shaped hypoechoic mass with linear hyperechoic center representing mucosa-lumen interface (white arrow heads) in right lower quadrant of abdomen.

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examination revealed diffuse abdominal and rebound tenderness, which clinically suggested peritonitis. Laboratory examination showed a WBC of $17.6 \times 10^9/l$, with a predominance of segmented neutrophils(89%). Ultrasonographic examination identified a noncompressible sausage-shaped hypoechoic mass with a linear hyperechoic center representing the mucosa-lumen interface in the right lower quadrant(Fig. 1). CT examination following intravenous contrast administration revealed a markedly enlarged target-like appendix in which the outer layer showed higher attenuation than the central portion. There was no discernible lumen

within the massively thickened appendiceal wall. In addition, multiple linear strands were noted within periappendiceal fat, and both adjacent lateroconal fascia and colonic walls were mildly thickened(Fig. 2A, B). Appendectomy and wedge resection of the cecum were performed. Grossly, the appendix, measuring 15cm in length and 2cm in diameter, was seen to have an irregular granular mucosal surface and was diffusely thickened by neoplasm(Fig. 3A). Microscopically, the muscularis propria had been diffusely infiltrated by lymphoma cells. The subserosa and serosa were heavily infiltrated with neutrophils and showed fi-

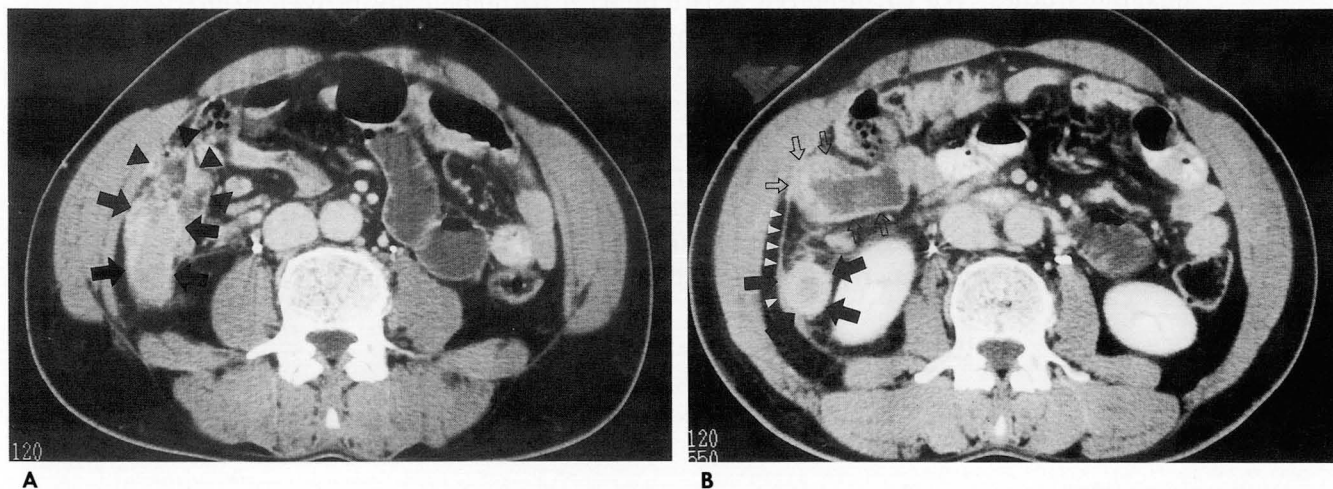


Fig. 2. A. Post-contrast CT scan shows a longitudinal section of markedly enlarged appendix (arrows) at its proximal portion which protrudes from cecum. The wall of cecum is also irregularly thickened (arrow heads).

B. Post-contrast CT scan (3.2cm upper level) shows an axial section of markedly enlarged appendix (arrow). The appendix is target shaped, in which the outer layer showed higher attenuation than the central portion. There is no discernible lumen within it. In addition, multiple linear strands were noted at periappendiceal fat, and both lateroconal fascia (white arrow heads) and wall of ascending colon (open arrows) were mildly thickened.

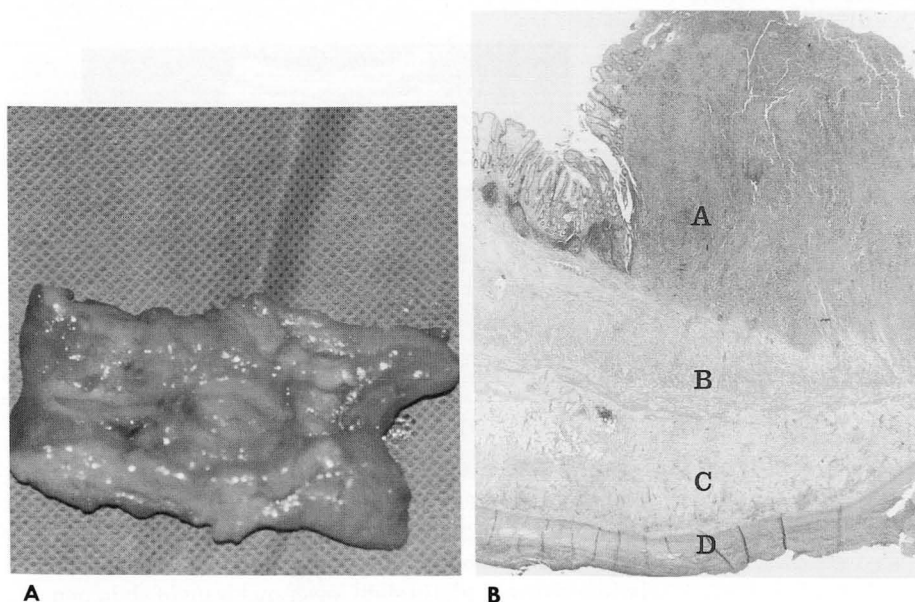


Fig. 3. A. Longitudinal section of the resected specimen shows irregular mucosal surface with several small ulcerations. The appendiceal wall is diffusely thickened by tumor infiltration.

B. Photomicrograph of the resected specimen (Hematoxyline-Eosin stain, $\times 10$). There are massive infiltrations of lymphoma cells (A), which extends to the muscularis propria (B). The subserosa (C) and serosa (D) are heavily infiltrated with neutrophils and show severe fibrinosuppurative changes.

brinosuppurative change(Fig. 3B). Pathologic diagnosis was malignant lymphoma of diffuse, mixed small and large cell-type associated with acute suppurative and chronic inflammation. Except for post-operative change in the cecum, barium enema, performed 21 days later, showed no abnormal colonic findings and Gallium-67 citrate scan revealed no additional lymphomatous bodily infiltration.

Discussion

The gastrointestinal tract is the most common site of involvement of extranodal lymphoma. The commonest gastrointestinal site for lymphoma is the stomach, followed by the small intestine and the ileocecal region(5). Primary appendiceal lymphomas are very rare and comprise about 1-3% of all gastrointestinal lymphomas(5, 6). Schmutzer et al. (6) reported 101 tumors of the appendix in 8699 appendectomy specimens: three cases were lymphoma. Primary appendiceal lymphoma, compared to other gastrointestinal lymphoma, may have a better outcome, as it gives rise to symptoms early in the disease and allows diagnosis at an early stage(1, 5). The most common clinical manifestation is that of right lower quadrant pain suggesting appendicitis. According to a review of the literature by Muller et al.(1), however, only one-fifth of appendiceal lymphomas have presented as acute appendicitis. Less common clinical presentations include a palpable mass, intussusception, peritonitis secondary to appendiceal perforation and lower gastrointestinal bleeding(4).

Our case had very similar radiographic findings to those described in previous articles in which diffuse intramural wall thickening of the appendix with luminal obliteration, has been described(2-4).

Krepel et al.(3) called the luminal obliteration of the appendix "the empty lumen sign". In our case, ultrasonography revealed a non-compressible sausage-shaped hypoechoic mass with a linear hyperechoic center caused by the mucosa-lumen interface in the right lower quadrant. Post-contrast CT examination showed a markedly enlarged target-like appendix, with obliteration of the lumen, in which the outer layer showed

higher attenuation than the central portion. In accordance with the pathologic report, the central portion suggested lymphoma itself, and the outer layer suggested inflammatory hyperemia caused by appendicitis. Acute and chronic appendicitis associated with appendiceal lymphoma were manifested as multiple strands of periappendiceal adipose tissue, and thickening of the lateroconal fascia and colon wall.

In view of the comparable clinical characteristics, distinguishing appendiceal lymphoma from acute appendicitis is difficult. In our opinion, helpful differential features are appendiceal wall thickness and whether or not an appendiceal lumen is obliterated. In acute appendicitis, the inflamed appendix appears as a distended, fluid-filled tubular structure about 0.5 to 2cm in diameter, the cause of this being obstruction of the appendiceal lumen followed by infection. The inflamed wall is thickened, but rarely exceeds 3mm. In contrast, in appendiceal lymphoma, pathologic thickening of the submucosal lymphoid layer is followed by circumferential diffuse infiltration of the total appendix, resulting in considerable thickening of the appendiceal wall and luminal obliteration. These findings can help differentiate appendiceal lymphoma from acute appendicitis.

In summary, we have described a case of appendiceal lymphoma presenting as acute appendicitis. Diffuse appendiceal wall thickening and luminal obliteration might be helpful for differentiating lymphoma from acute appendicitis.

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급성 충수염으로 발현된 충수의 원발성 림프종 : 1예 보고¹

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충수의 원발성 림프종은 매우 드물며 임상적으로 급성 충수염으로 발현하는 경우가 많아 수술전 방사선학적 검사로 진단되는 경우는 거의 없다. 저자들은 최근에 병리학적으로 증명된 급성, 만성 충수염과 병발한 충수의 원발성 림프종 일례를 경험하였기에 초음파 및 전산화 단층촬영 소견과 함께 보고한다. 초음파 검사상 충수는 쏘세지 모양의 매우 큰 저에코 종괴로 보였고 그 내부에 점막과 내강의 계면을 시사하는 선상의 고에코가 있었다. 조영제 주입후 전산화 단층촬영상 충수는 과녁 모양의 두 층으로 이루어진 비후를 보였고 바깥층이 중심부보다 더 높은 감쇄를 보였으며 내강은 완전히 폐색되어 있었다. 또한 충수 임파종과 병발한 급성 충수염을 시사하는 소견으로 충수 주위 지방조직에 다수의 선상음영과 함께 인접한 대장벽 및 lateroconal 근막의 비후소견이 보였다.