

CASE REPORT

T. Hasegawa · J. Sumimura · S. Mizutani
Y. Tazuke · S. Okuda · T. Dezawa

The doughnut sign: an ultrasound finding in pediatric intestinal Burkitt's lymphoma

Accepted: 12 November 1996

Abstract This paper describes a doughnut-like ultrasound (US) finding in pediatric intestinal Burkitt's lymphoma. A 9-year-old boy had a fist-sized, hard, non-movable mass in the lower abdomen. US showed a thickened, layered ring like a doughnut. The outer, low-level echoes seemed to be consistent with mucosa and muscle layers and the inner, high-level echoes seemed to be intraluminal air or mucus. The serum lactic dehydrogenase level was high. At surgery, a solid, hard tumor 15 × 10 cm in size was found in the jejunum. The intestinal wall was diffusely thickened with an intact mucosa. From this experience, the US doughnut sign may be a helpful diagnostic finding in pediatric intestinal Burkitt's lymphoma.

Key words Intestinal lymphoma · Burkitt's lymphoma · Doughnut sign · Ultrasonography

Introduction

Pediatric Burkitt's lymphoma (BL) often affects the gastrointestinal tract, and has a poor prognosis because of very rapid progression and difficulty in diagnosis [1]. The symptoms and laboratory data lack specificity and radiologic findings are not pathognomonic. We present a case of pediatric intestinal BL with a doughnut-like ultrasound (US) finding.

Case report

A 9-year old boy developed vomiting, low-grade fever, and mild abdominal pain with a fist-sized, hard, non-movable mass in the lower abdomen. Laboratory data showed mild anemia and an elevated serum lactic dehydrogenase (LDH) level of 1,447 (normal 150–450) IU/l. On US, a longitudinal section of the abdominal tumor showed several layers with low- and high-level echoes (Fig. 1) and a transverse section showed a thickened, layered ring like a doughnut (Fig. 2). The outer, low-level echoes seemed to be consistent with mucosa and muscle layers and the inner, high-level echoes appeared to be intraluminal air or mucus. Computerized tomography (CT) of the tumor also showed a large, thickened ring. Intravenous pyelography and barium enema studies were normal. The differential diagnosis included chronic intussusception and a tumor originating from the intestine or mesentery.

At laparotomy, a solid, hard tumor 15 × 10 cm in size was found in the jejunum as well as multiple enlarged lymph nodes in the mesentery. The tumor and mesenteric lymph nodes were resected. Section of the tumor showed a diffusely thickened intestinal wall and an intact mucosa and was diagnosed as BL by histologic, immunohistochemical, and chromosomal studies. The tumor was already in stage 4 based on metastases to the mediastinal and submandibular lymph nodes detected by gallium scintigram and CT. It was aggressively treated with a combination of cyclophosphamide, vincristine, methotrexate, adriamycin, VP-16, cytosine arabinoside, and steroids, despite which the patient died 6 months later.

Discussion

Burkitt's lymphoma isolated in the intestine can be resected in early stages, and the early administration of aggressive chemotherapy may lead to a good outcome [4]. Because progression of the disease is very rapid [1], the diagnosis should be made as early as possible. The present case was already in an advanced stage when diagnosed by the surgical specimen, and the patient subsequently died despite the aggressive chemotherapy. Pediatric patients with intestinal BL present with abdominal pain, anorexia, weight loss, nausea, vomiting, an abdominal mass and fever and often develop intussusception [4], all of which, however, are not specific for BL. Although elevation of serum LDH levels is seen in many malignancies, the level in intestinal BL correlates

T. Hasegawa
Division of Pediatric Surgery,
Kinan General Hospital,
510 Minato, Tanabe City,
Wakayama, Japan 646

J. Sumimura · S. Mizutani · Y. Tazuke
Department of Surgery,
Kinan General Hospital,
510 Minato, Tanabe City,
Wakayama, Japan 646

T. Hasegawa (✉)
Department of Pediatric Surgery,
Osaka University Medical School,
2-2 Yamadaoka, Suita, Osaka,
Japan 565

S. Okuda · T. Dezawa
Department of Pediatrics,
Kinan General Hospital,
510 Minato, Tanabe City,
Wakayama, Japan 646

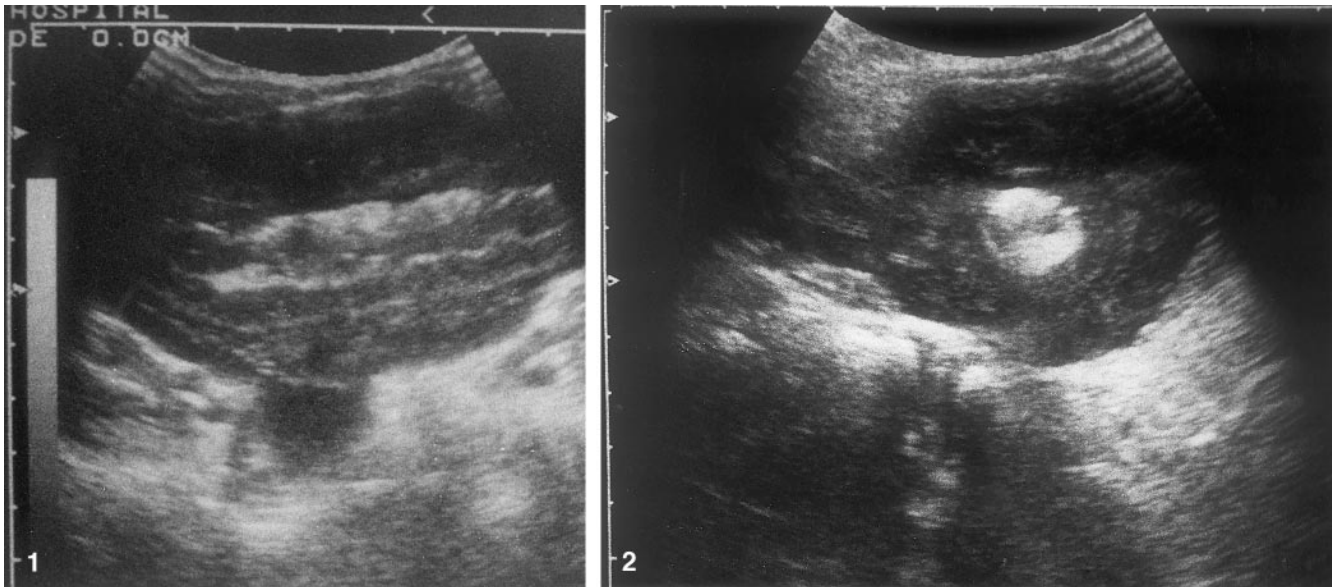


Fig. 1 Ultrasound scan of abdominal tumor: longitudinal section shows several layers with thickened high- and low-level echoes

Fig. 2 Transverse section shows doughnut-like finding including high- and low-level echoes

well to the prognosis [1], and the elevation is more specific to lymphoma than to other intestinal tumors, intussusception, etc., in children.

US is an initial diagnostic method for abdominal tumors. The US finding in the present case was a doughnut-like, layered ring. The doughnut and pseudokidney signs have been recognized as US findings of intussusception [4]; intestinal lymphoma has also been described as a cause of these signs [6, 8, 9]. Intestinal BL usually invades the submucosal layer [6, 9] causing diffuse wall thickening, which produces low-level echoes [6, 8, 9] as seen in the present case. A central echogenic level is caused by intraluminal air or mucus [6, 9] when the lumen is not obstructed, as was seen in the present case, and this combination causes the doughnut-like appearance, which was consistent with the

CT finding and the surgical specimen. However, this doughnut sign is seen in other gastrointestinal pathology invading the submucosal layer, such as intussusception, ischemic bowel disease, intramural hematoma, radiation enteritis, etc. [2, 3, 5, 7].

Based on this experience, the US doughnut sign combined with an elevated serum LDH level may be a helpful diagnostic finding in pediatric intestinal BL.

References

1. Arseneau JC, Carellos GP, Banks PM, Berard CW, Gralnick HR, De Vita VT (1975) American Burkitt's lymphoma: a clinicopathologic study of 30 cases. I. Clinical factors relating to prolonged survival. *Am J Med* 58: 314
2. Bluth E, Merritt CR, Sullivan MA (1979) Ultrasound evaluation of the stomach, small bowel and colon. *Radiology* 133: 677-680
3. Fakhry JR, Berk RN (1981) The "target" pattern: characteristic sonographic feature of stomach and bowel abnormalities. *Am J Radiol* 137: 969-972
4. Fleming ID, Turk PS, Murphy SB, Crist WM, Santana VM, Rao BN (1990) Surgical implications of primary gastrointestinal lymphoma of childhood. *Arch Surg* 125: 252-256
5. Lee TG, Brickman FE, Avecilla LS (1977) Ultrasound diagnosis of intramural intestinal hematoma. *J Clin Ultrasound* 5: 423-424
6. Miller JH, Hindman BW, Lam AHK (1980) Ultrasound in the evaluation of small bowel lymphoma in children. *Radiology* 135: 409-414
7. Sener RN, Alper H, Demirci A, Diren HB (1989) A different sonographic "pseudokidney" appearance detected with intestinal lymphoma: "hydronephrotic pseudokidney" *J Clin Ultrasound* 17: 209-212
8. Swischuk LE, Hayden CK, Boulden T (1985) Intussusception: indications for ultrasonography and an explanation of the doughnut and pseudokidney signs. *Pediatr Radiol* 15: 388-391
9. Teele RL, Share JC (1991) Anterior abdominal mass. In: Teele RL, Share JC (eds) *Ultrasonography of infants and children*. Saunders, Philadelphia, pp 274-285