Case Report of a Burkitt's Lymphoma Arising from Meckel's Diverticulum in Childhood

Çocukluk Çağında Meckel Divertikülünde Gelişen Burkitt Lenfomalı Bir Olgu Sunumu

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ABSTRACT Meckel's diverticulum (MD) is one of the most common inherited anomalies of the gastrointestinal tract with an incidence up to 3%. MD is generally silent and becomes symptomatic when complicated. The most common complications of MD are bleeding, obstruction and inflammation due to its ectopic mucosa. The standard treatment of symptomatic MD is the surgical excision of the diverticulum or resection of the ileal segment and end-to-end anastomosis. But when found incidentally in the patients with no risk factors, the prophylactic resection of the Meckel's diverticulum is controversial. We report the second case of lymphoma in the MD. The MD in this case was complicated and treated as symptomatic MD, uneventfully. Even though the neoplastic degeneration of MD mucosa is a rare entity and occasionally exists, the long term disease free follow up of the patient made us consider prophylactic diverticulectomy in patients with MD.

Key Words: Meckel diverticulum; burkitt lymphoma

ÖZET Meckel divertikülü (MD) en sık karşılaşılan kalıtsal gastrointestinal sistem anomalilerinden biri olup %3 oranında görülür. Genellikle sessiz seyreden ve komplike olduğunda semptomatik hale gelen MD'nin en sık karşılaşılan komplikasyonları ektopik mukozasına bağlı olarak görülen kanama, obstruksiyon ve inflamasyondur. Semptomatik MD'nin standart tedavisi divertikülün cerrahi olarak eksizyonu veya ileal segmentin rezeksiyonu sonrası uç-uca anastomozdur. Rastlantısal olarak tanılandırılan ve herhangi bir risk faktörünün varlığı bilinmeyen çocuklarda MD'nin cerrahi tedavisi ise halen tartışmalıdır. Bu sunumun amacı, komplike olduğu için semptomatik MD olarak sorunsuz şekilde tedavi edilmiş 3 yaşında bir hastada konulan "divertikülde lenfoma" tanısını bildirmektir. Cerrahi sonrası lenfoma tanısı ile kemoterapi almış olan hastanın 10 yıllık takibi sorunsuzdur. MD mukozasının neoplastik dejenerasyonu nadir görülmekle beraber bu hastanın uzun süreli takibinde hastalık olmaması, rastlantısal olarak bulunan sorunsuz MD'lerinde de divertikül eksizyonuna yaklaşım konusundaki tartışmalara farklı bir bakış açısı ile yaklaşmamıza neden olacak türdendir.

Anahtar Kelimeler: Meckel divertikülü; burkitt lenfoması

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CASE REPORT

A 3-year-old boy complaining of intermittent generalized abdominal pain and nausea, admitted to the department of pediatrics. The family had complained moderate attacks of pain for the last 6 months. The boy was good in health and there was no history of weight loss, change in bowel habits and rectal bleeding. The physical examination done by pediatricians revealed generalized abdominal tenderness. Laboratory find-

ings were unremarkable except for a mild anemia. The patient was consulted to our department after the abdominal ultrasound designated an ileocolic intussusception at the right iliac fossa. The physical examination performed in our department revealed generalized abdominal tenderness with a suspected right-upper quadrant mass. Neither abdominal mass nor blood was observed at the digital rectal examination. The patient received the trial of hydrostatic reduction with barium enema. As the trial had failed, the patient has undergone a laparotomy with a diagnosis of intussusception. After a successfully completed manual reduction of ileocolic intussusception, an exploration for a pathologic leading point was done. A MD of 2.5 cm width and 5 cm length, localized on the antimesenteric border of the ileum was detected 60 cm proximally to the ileocecal valve. The segment of ileum with MD was resected and primary anastomosis was performed. The microscopic studies revealed a Burkitt's lymphoma in the MD. On microscopic examination, continuing with normal intestinal mucosa, the tumor that is composed of medium-sized lymphoid cells shows a diffuse monotonous pattern of infiltration (Figure 1). The tumor cells are uniform in size and in shape with deeply basophilic cytoplasm. The nuclei are round with clumped chromatin and contain multiple nucleoli (Figure 2). The tumor has an extremely high proliferation rate with many mitotic figures (Figure 3). On the 4th postoperative day, the patient transferred from the department of pediatric surgery to the department of pediatric oncology. The patient diagnosed as primary lymphoma of MD after further researches revealed no other origins for the malignancy. He received three regimens of chemotherapy for two years and is disease free after ten years follow up. Informed consent was taken from the parents of the child.

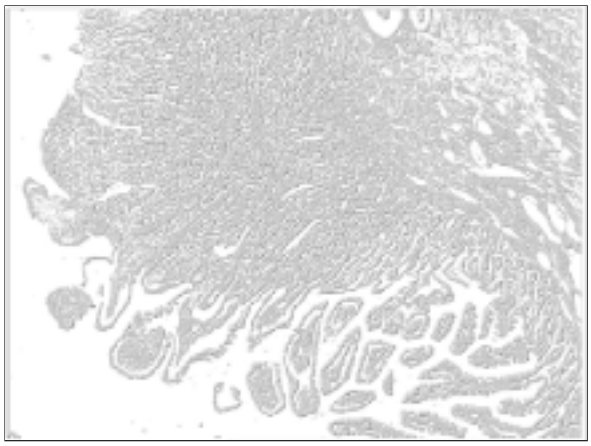

FIGURE 1: The tumor mass continuing with the normal intestinal mucosa (H&E, x12.5).

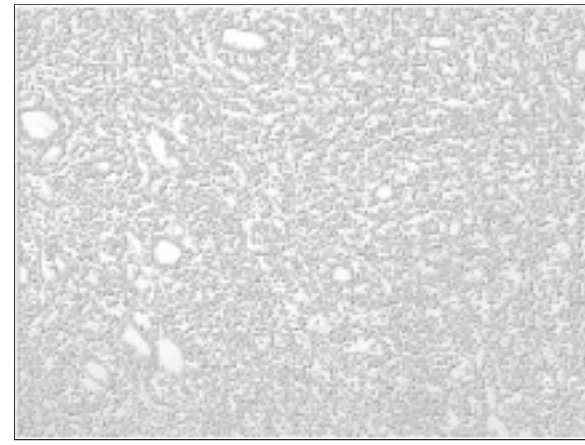

FIGURE 2: The starry-sky histiocytes, characteristic of Burkitt lymphoma (H&E, x40).

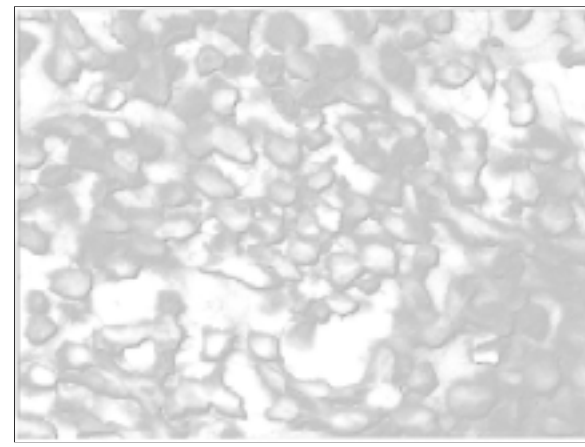

FIGURE 3: Numerous mitotic figures showing the high proliferation rate of tumor (H&E, x400).

DISCUSSION

Meckel's diverticulum (MD), the remnant of the omphalomesenteric duct, is the most common congenital anomaly of the gastrointestinal tract. It is a real diverticulum arising from the antimesenteric border of the small intestine. The majority of Mec-

kel's diverticuli is asymptomatic but becomes evident when complicated.[1-3]

The frequent cause of intestinal obstruction in infants and toddlers is intussusception due to MD as the most common anatomical leading point. The first choice of treatment is the reduction of intussusception by hydrostatic or pneumostatic method under fluoroscopic or ultrasonographic controls with a success rate up to 90%. The surgery is indicated whether this trial fails.[1-3] The patient received an unsuccessful trial of hydrostatic reduction with barium enema, operated because of a MD causing intussusception, diagnosed as Burkitt's lymphoma and after chemotherapy he is now disease free. If the trial of hydrostatic reduction with barium enema was succeeded in this patient, the diagnosis of Burkitt's lymphoma would be delayed.

Tumors in MD are not common with an incidence of 0.5-1.4%. The ratio of malignant tumors of MD (leiomyosarcoma, carcinoid tumor, adenosarcoma) to benign mesenchymal tumors of the MD (leiomyoma, angioma and lipoma) 1:3 and these tumors tend to appear in later life.[4,5] Carcinoid tumors are the most common malignancy of MD.[6,7] The primary lymphoma of the MD is an extremely rare entity. The literature review, up to date signed only one case.[8] This is the second case of primary lymphoma of the MD in the literature.

Burkitt's lymphoma is a highly aggressive lymphoma. Sporadic type of Burkitt's lymphoma, apart from endemic or immunodeficiency types, occurs worldwide and accounts up to 40% of lymphoma in children. The abdomen, especially the ileocecal area, is the most common site of involvement. These tumors have an extremely high rate of proliferation, in addition to a high rate of cell death (apoptosis). Short-duration, high-intensity chemotherapy, sometimes combined with CNS prophylaxis, yields excellent survival in children. A localized, sporadic Burkitt's lymphoma in younger age (<15 years) is related to a better prognosis.[9,10]

In this case, MD, as the leading point of the intussusception, was resected. Resection is clearly indicated in pediatric patients having the past history of unexplained abdominal pain, palpable thickening consistent of a heterotopic mucosa or presenting omphalomesenteric remnants and abdominal wall attachments.[1] But for some authors, pediatric age itself is said to be an indication for the prophylactic resection of MD because of its high complication rate.[4,11] The complication rate of MD is 4-6.4% in early childhood and 3.7% at the age of 16.[12] These rates decrease after adolescent years.

By the advancement of the minimally invasive surgical techniques, diagnosis and treatment of various intraabdominal pathologies become easier with excellent outcomes like lower morbidity and mortality rates. In the same manner, it is probable that much more MDs are now diagnosed and prophylactic resection of an incidentally found MD poses a dilemma to the surgeons more than usual. Bona et al presented a case of a MD found incidentally at laparoscopy for bilateral inguinal hernia repair.[13] Prophylactic diverticulectomy revealed a carcinoid tumor within the diverticulum. With the more common usage of laparoscopic techniques and equipments, viable and safe approaches can be offered to the patients.

The limited knowledge of the tumors of MD in the literature signs that the adjuvant therapies following the surgery can be curative for the patients.[2,5,9] We believe that, the complications of MD are higher and more severe in the pediatric age, and the resection of an incidentally found MD with no risk factors can carry the patients through better conditions. We also think that the prophylactic diverticulectomy in childhood should be considered because of the unpredictable nature of MD.

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