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Hypereosinophilic Syndrome with Hepatic Involvement in a Young Child[1]

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Hypereosinophilic syndrome, whose etiology is unknown, involves the infiltration of various organs by a large number of eosinophils. The sites of involvement are the heart, skin, lung, liver, nervous system, and gastrointestinal tract. The disorder occurs mostly in middle-aged men and is characterized by striking peripheral eosinophilia. There have been few reports of hypereosinophilic syndrome in patients younger than 15 years and the disease also shows a predilection for males. We report a case of hypereosinophilic syndrome with hepatic involvement in a 17-month-old girl, and correlate the imaging features with the pathologic findings.

Index words : Liver, abnormalities
Liver, CT
Liver, US

Hypereosinophilic syndrome is a spectrum of disorders characterized by marked eosinophilic leukocytosis and dysfunction of the involved organ. There is no identifiable cause (1, 2). The syndrome, which occurs mostly in middle-aged men, is characterized by striking peripheral blood eosinophilia. There have been few reports of the condition in patients younger than 15 years, and 91 - 92% of patients are male, an overwhelming preponderance (3). Our case is unusual not only because of the patient's very young age, but also her gender. The criteria for the diagnosis of hypereosinophilic syndrome include persistent eosinophilia of more than 1500 eosinophils/ml for longer than 6 months, or death before 6 months; the absence of parasitic, allergic, or other known causes of eosinophilia; and evidence of organ involvement (3). We report the imaging findings of hypereosinophilic syndrome with hepatic involvement in a 17-month-old girl.

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A Case Report

A 17-month-old girl presented with irritability, fever and cough, which had developed 10days earlier. Previously she had been healthy, and there was no past or familial history of allergy or asthma. Physical examination revealed mild hepatosplenomegaly. On admission, the leukocyte count was 32,600/ml, with 56% of eosinophils. Serum aspartate aminotransferase was 37 IU/L, alanine aminotransferase 67 IU/L, alkaline phosphatase 474 IU/L, and bilirubin 0.3 mg/dl. The results of a stool test for ova and parasites were normal. Bone marrow biopsy revealed hypercellular marrow and an increased number of eosinophils (48%), with no evidence of hematologic malignancy. Chest radiography indicated nothing abnormal. Abdominal sonography using a 3.5-MHz transducer revealed multiple, poorly marginated hypoechoic nodules, about 1 cm in diameter throughout the liver (Fig. 1A); a 7-MHz linear probe showed that some had a bull's eye or target appearance (Fig. 1B). Portal-phase CT demonstrated multiple hypodense lesions distributed diffusely in the periportal area

(Fig. 1C), and under the sonographic guidance, percutaneous needle-gun biopsy of the liver was performed. The pathologic specimen obtained showed several portions of eosinophilic infiltration in the periportal area, and piecemeal necrosis (Fig. 1D). The patient was treated with prednisolone and followed up for two months. During this time her leukocyte count decreased to 12,400/ml, with 6% of eosinophils in peripheral blood. Follow-up imaging studies were not performed.

Discussion

Hypereosinophilic syndrome is defined by the presence of peripheral and bone marrow eosinophilia and by the infiltration of multiple organs by mature eosinophilic cells (3). Among reported cases, patient age has ranged from 5 to 80 years (6), the most frequent age of occurrence being the fifth decade. Men account for about 85% of patients, and in 50 - 90% of cases there is hepatic involvement (6). To our knowledge, our case involves the youngest hypereosinophilic syndrome patient on record. The clinical manifestations depend on

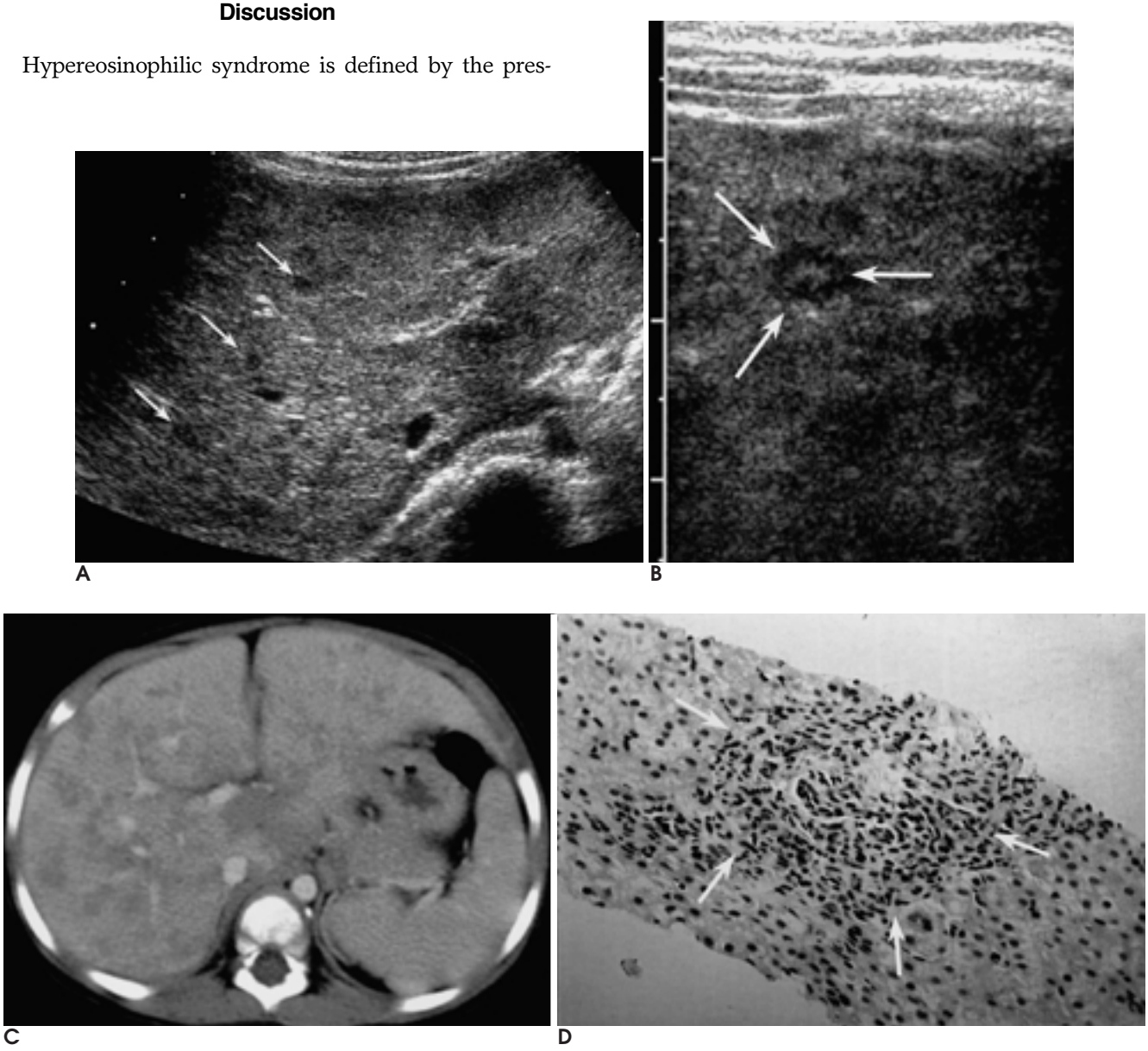

Fig. 1. A 17-month-old girl with hypereosinophilic syndrome.
A. Abdominal sonography using a 3.5 MHz transducer shows multiple, round, poorly marginated hypoechoic nodules (arrows) of about 1 cm in diameter.
B. Abdominal sonography using a 7 MHz transducer demonstrates bull's eye or target-appearing nodule (arrows).
C. Portal phase CT scan depicts multiple hypodense lesions distributed diffusely in the periportal area.
D. The pathologic specimen shows eosinophilic infiltration in the periportal area (arrows) of the liver (Hematoxylin and Eosin stain, × 100).

the organ involved and include weight loss, recurrent abdominal pain, fever with night sweats, nonproductive cough, various neurologic abnormalities, pruritic rash, and congestive heart failure (1 - 3).

Several reports have described the sonographic, CT, or scintigraphic findings of hepatic involvement of hypereosinophilic syndrome in adults (5 - 9). Sonography, for example, has disclosed multiple poorly defined, small, round or oval scattered focal lesions in both lobes of the liver (6), and in our case, multiple, round, poorly defined, hypoechoic or target - appearing lesions less than 1cm in diameter were apparent at sonography. CT findings of multiple small hypoattenuating foci without contrast enhancement have been reported (8), and lesions were most conspicuous during the portal phase (7); in our case, as in previous reports (5 - 7), CT revealed multiple poorly defined hypodense lesions in the periportal area. Although scintigraphy was not performed in our case, abnormal scintigraphic findings on scintigrams are thought to be due to circulatory disturbance caused by eosinophilic infiltration of the periportal area or abnormally functioning Kupffer's cells (5). Some authors have indicated that the number and extent of eosinophil-related necrotic foci correlate closely with eosinophil counts in peripheral blood (8, 9).

Pathologic specimens revealed eosinophilic infiltration in the periportal or centrilobular area of the liver in the present case, and in terms of distribution, the findings correlated closely with the radiologic findings.

Because radiologic findings of eosinophil-related liver necrosis are nonspecific, it may be difficult to differentiate these foci from metastasis, lymphoma, leukemia, or microabscess. Hypereosinophilic syndrome should, therefore, be diagnosed only where other supportive clinical and laboratory findings are present. In addition, it should be remembered that malignant neoplasms such as lymphoma, leukemia, and carcinoma are often associated with eosinophilia (8).

The prognosis of hypereosinophilic syndrome is variable and the most serious complications arise from involvement of the heart or central nervous system. Congestive heart failure is the most common cause of death (1 - 3). Our patient showed no evidence of involvement of the heart or central nervous system, and as expected, responded gradually to corticosteroid therapy.

In summary, although imaging findings of hepatic involvement are nonspecific, the possibility of hypereosinophilic syndrome should be considered, even in a young child, if the clinical setting is appropriate.

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정미란 · 구현우 · 홍성숙 · 윤종현

과호산구 증후군은 원인을 알 수 없이 많은 수의 호산구가 여러 장기에 침윤되는 질환이며, 주로 침범되는 장기는 심장, 피부, 폐, 간 , 신경계, 그리고 소화기관이다. 이 질환은 주로 중년의 남자에서 생기며 특징적으로 말초혈액의 과호산구증을 보인다. 15세 이하의 어린이에서는 거의 보고된 적이 없고 보고된 증례들도 남자에 편중되는 경향이 있다. 저자들은 유소아에서 과호산구 증후군이 간을 침범한 예를 경험하였기에 방사선학적 소견과 조직학적 소견을 보고하고자 한다.