An Infant Case of Neutropenia Developed Repeated Omphalitis : A Case Report

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ABSTRACT. We report on a case of autoimmune neutropenia of infancy (ANI). A seven-month-old girl was referred to our hospital because of repeated omphalitis. Her reddish and swollen umbilicus stiffened and drained a little pus. Ultrasonograhy revealed a subcutaneous mass caused by inflammation. She was admitted because of moderate fever and appetite loss. However, a blood exam disclosed a decrease in neutrophils which reached zero the next day. Detection of antineutrophil antibody led us to a diagnosis of ANI. Although the inflammation was resolved and she left the hospital, she suffered from omphalitis again three months later. At that time we resected the urachus under a diagnosis of urachal remnant. The postoperative course was favorable and her neutrophil level has gradually been returning to normal with age. ANI is relatively rare disease and patients with it experience repetition of infectious disease, but it is not a particularly grave. We recognized once again that care should be taken regarding the possibility of an immunodeficient disorder in every infant with repeated infection.

Key words ① **Autoimmune neutropenia of infancy (ANI)** ② **Omphalitis** ③ **Urachal remnant**

Neutropenia occurs due to varied causes. Among primary disorders, autoimmune neutropenia of infancy (ANI) is a relatively rare disease[1-6]. Patients usually experience trouble due to repeated infection but their condition does not become serious. We reported on a case of ANI which was characterized by the symptom of repeated omphalitis.

Case

A seven-month-old girl was referred to our hospital with a complaint of omphalitis lasting two months. She had no past or familial history. Her umbilicus had been reddish and granulation had formed, and her local doctor had been unsuccessful in healing it with a few trials of cauterization using silver nitrate. Then a subcutaneous mass was palpable. On her first visit, her umbilicus was reddish and swollen without a cave. It stiffened and drained a little purulent fluid (Fig.1). Ultrasonography disclosed a low echoic mass in the

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Fig. 1. Umbilical finding at the time of the first exam
The umbilicus was reddish and swollen. It stiffened and drained a little pus and omphalitis was suspected.

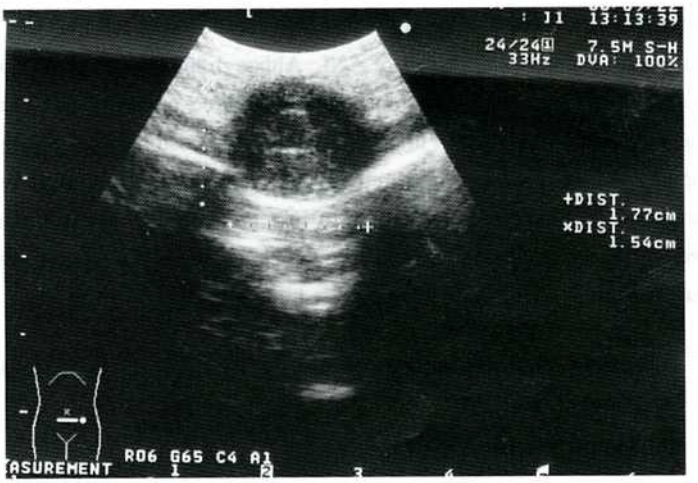

Fig. 2. Ultrasonography
Ultrasonography demonstrated a low-echoic mass in a subcutaneous lesion of about 15mm in diameter.

subcutaneous lesion which was about 15 mm in diameter (Fig. 2). A small quantity of fluid was drained by puncture with needle and afterwards staphylococcus was detected in the culture.

Because of moderate pyrexia and loss of appetite, she was hospitalized for treatment. A blood exam at that time revealed serious neutropenia (Table.1), and she was transferred to a room with HEPA filter. On the next day, her neutrophil count reached zero. Then under the status of increasing neutrophils with G-CSF, a bone marrow aspiration was performed. Based on the result of that aspiration and detection of antineutrophil antibody in her peripheral blood, we made a definite diagnosis of autoimmune neutropenia of infancy (ANI).

The inflammation completely regressed with antibiotics and she left the hospital. However, three months later, her umbilicus became inflamed again. Serious neutropenia was seen as before, and she was given antibiotics and G-CSF. This time, since we suspected the presence of an urachal fistula, we operated and removed a funicular structure which was consecutive from the umbilicus to the bladder. Although neither

Table.1 WBC count and classification on the day of admission and on the next day
Serious neutropenia was seen on the day of admission and her neutrophil count fell to zero the next day.

	admission	day 2
WBC (/μl)	6060	6030
Neutrophils (/μl)	303	0
Lymphocytes (/μl)	4181	5186
Blasts (/μl)	0	0
Band (%)	5	0
Seg (%)	0	0
Mon (%)	17.0	8
Lym (%)	69.0	86.0

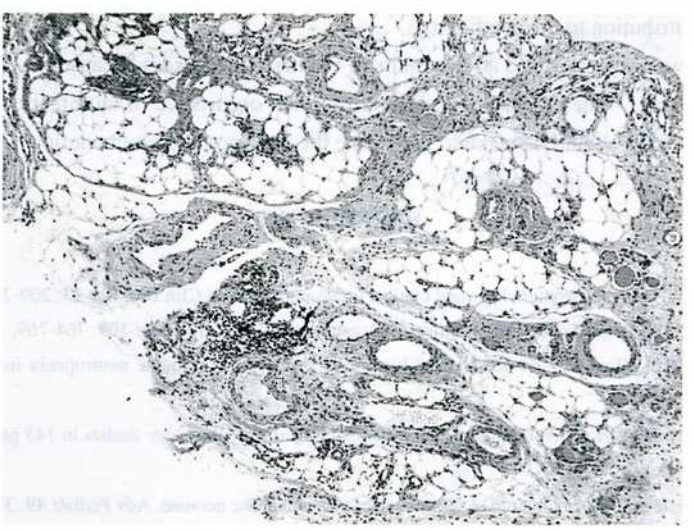

Fig. 3. Pathological finding of resected specimen
Fat or fibrous tissue and lymphocytic infiltration were seen in this specimen, but probe patency and aberrant epithelium were not detected.

probe patency nor aberrant epithelium were detected in the specimen (Fig.3), it was concluded that the duct was closed or lost due to intense inflammation. Since then, she has not suffered from omphalitis for two years and her neutrophil level is gradually returning to normal without treatment.

DISCUSSION

Secondary neutropenia occurs for varied reasons, such as viral infection or medication. Neutropenia caused by bone marrow suppression by anticancerous drugs is a well-known and sometimes lethal cause. Primary neutropenia may develop in pediatric groups. Among these conditions, autoimmune neutropenia of infancy (ANI)[1~6] is a relatively rare disease with a sporadic rate of 1 in 100,000 in children 1 to 10 years old. Patients usually experience repeated but not so grave infection such as cellulitis, otitis media or gingivitis. Although most patients do not become serious if the other immune system is normal, some may experience severe infections such as pneumonia or sepsis. The detection of antineutrophil antibody provides the diagnosis with high reliability[7),8)]. The neutrophil level increases with age without treatment, but G-CSF is useful for serious infection or surgical management.

We suppose that there are three causes for childhood omphalitis. The first is persistence of the umbilical cord in a neonate. This can be treated easily with silver nitrate even if granulation forms on the surface. However in our case, there no umbilical symptoms until the infant was five months old or the treatment by a former doctor had no effect. The second cause is inflammation due to a continuous fistula from other organs, e.g., urachal or omphalomesenteric duct remnants. Most of these tend to form an abscess, but a granuloma could also form. The last cause is inflammation from a wound. If a patient begins to move in a prone position, mechanical irritation can result in a wound and repeated chronic inflammation, which could lead to a granuloma. In our case, we suspected the existence of an urachal remnant rather than inflammation from a wound. Although there was no the proof of this in the pathologic exam, her good prognosis after surgery could suggest its contribution to the condition.

In present case, we were surprised at the results of the first blood exam because we had not suspected any immunodeficiency at all. When a child experiences repeated occurrence of an infectious disease, regardless of the type or degree, care should be taken to determine the presence of immunodeficiency with this disease.

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