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FACIAL NERVE PARALYSIS AND INTUSSUSCEPTION AS CLINICAL PRESENTATIONS OF LYMPHOBLASTIC LYMPHOMA: A RARE PEDIATRIC CASE REPORT

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ABSTRACT

Lymphoblastic lymphoma (LBL) is a highly aggressive lymphoblast neoplasm due to the presence of clonal hematopoietic stem cell abnormalities of B or T cell origin. LBL represents 30% of pediatric non-Hodgkin's lymphomas. We report a 2-year-11-month-old boy with complaints of a lump in the abdomen and facial paralysis on the left side. weight loss of 3 kg in 3 months, frequent nausea and vomiting, and constipation. On physical examination, the face was not symmetrical on the left and right. palpable mass from the right iliac region to the left iliac, measuring 17.5 x 10 x 5 cm, immobile, rubbery consistency. There is lymphadenopathy right inguinal region et sinistra. Abdominal ultrasound without contrast showed ileocolic intussusception, with histopathological results showing malignant non-Hodgkin's lymphoma. The therapy given was chemotherapy according to the protocol for non-Hodgkin's lymphoma. Facial nerve paralysis and intussusception are rare clinical features of LBL. It is important to consider a differential diagnosis such as LBL in a patient with a lump accompanied by facial nerve paralysis and intussusception. Histopathological and immunohistochemical examination were needed.

Keywords: Lymphoblastic Lymphoma; Facial Nerve Paralysis; Intussusception

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INTRODUCTION

The World Health Organization (WHO) classifies lymphoblastic lymphoma (LBL) as a lymphoblast tumor that mimics acute lymphoblastic leukemia (ALL) and is more frequently of T-cell origin than B-cell origin (1). Although the two conditions share a lot of molecular similarities but are not identical, LBL differs from LBL in that bone marrow is absent or just little affected, with less than 25% of blast cells present (or 20%, according to WHO) (2).

About 2% of Non-Hodgkin's Lymphoma (NHL) cases are LBL. 10% of all LBL instances are B-LBL, while 90% of these cases are T-LBL (3). LBL is a rare condition for which it is challenging to locate precise incidence data. 39 new LBL cases were found in a population-based investigation from Sweden between 2000 and 2009, but no incidence data were provided (2).

Peripheral facial nerve palsy that affects only one side of the face is a rather frequent disorder that is either idiopathic (Bell's palsy) or related to a known cause. Numerous case reports of lymphoma patients who have bilateral facial paralysis are available. Lymphoma-related facial paralysis is a medical emergency that necessitates a rigorous treatment regimen along with a thorough rehabilitation program (4).

Lymphoma can arise from any part of the gastrointestinal tract. The small and large intestines are the most frequently involved sites in children. It is most commonly found in the ileum, where the greatest concentration of associated lymphoid tissue is in the intestine. generally derived from B-cells from lymphoid tissue present in the lamina propria and submucosa (5) (6).

Through the development of intensive regimens applied according to stage, histology, or immunophenotype, the treatment of LBL has improved over the past 30 years. LBL therapy using the ALL type protocol. Previous studies have shown an improved prognosis with the ALL type protocol in LBL. In particular, the change to the ALL-type protocol has allowed the replacement of prophylactic cranial radiotherapy (PCRT) with intrathecal methotrexate and high-dose methotrexate (7).

In this journal, we will report a case of lymphoblastic lymphoma (LBL) with clinical presentation of

intussusception and facial nerve paralysis, a rare cases in children. The diagnosis in this case was made on the basis of histopathological and immunohistochemical biopsy results.

Case

A boy aged 2 years-11 months came to Dr. Hospital. Wahidin Sudirohusodo Makassar with a complaint of a painful lump in the abdomen that he had felt since 3 months before being admitted to the hospital. The lump is painful. There was weakness on the left side of the face since 1 week before admission to the hospital. The patient also complained of nausea and vomiting after every meal. Defecation is also not smooth. There is a history of weight loss in the last 3 months.

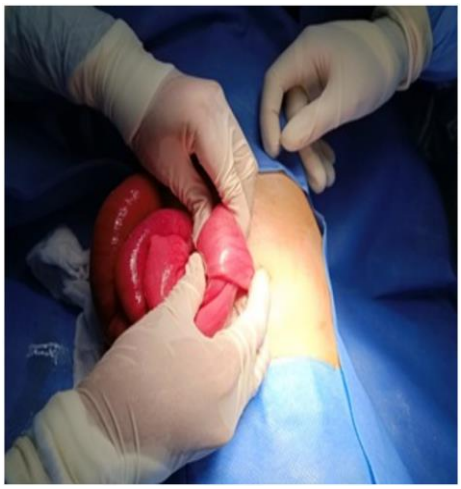

Figure 1. Operation results. Ileoileal intussusception

The general condition of the patient appeared to be seriously ill, with a GCS of 15. Poor nutritional status. Palpable mass in the right iliac region to the left iliac, through the midline shift, immobile, size 17.5 x 10 x 5 cm, There is lymphadenopathy in the right and left inguinal region. There was paralysis of the left facial nerve.

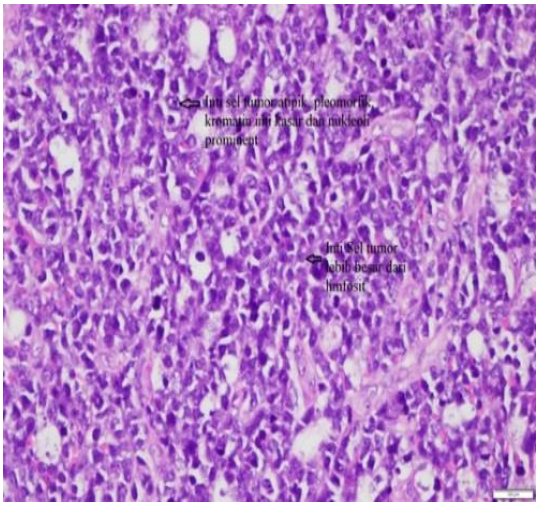

Figure 2. Histopathology, Malignant tumor with non-Hodgkin's Lymphoma

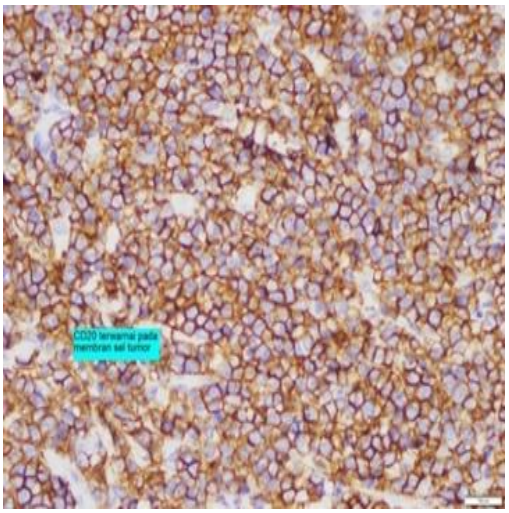

Figure 3. Positive CD 20 Result

From investigation, lactate dehydrogenase level was 4,534 U/L. Chest X-ray showed right hilar lymphadenopathy and right pleural effusion. Abdominal ultrasound

showed intussusception and ascites. Abdominal MSCT without contrast shows small bowel obstruction due to ileocolic intussusception and impacted faecal colon due to constipation. Histopathological results showed a malignant tumor with the impression of non-Hodgkin's lymphoma with the end of the resection still having a tumor. Positive CD20 result.

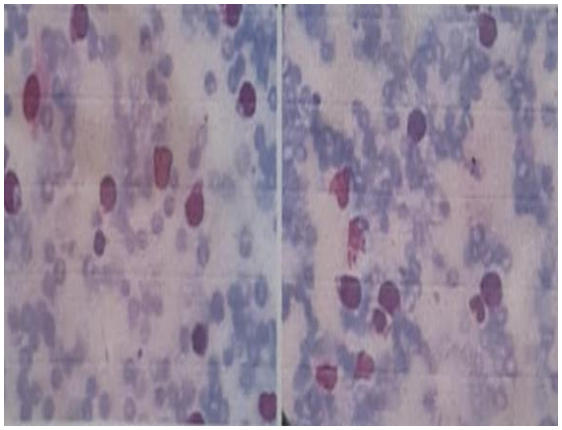

Figure 4. Bone marrow puncture, Malignant lymphoma with infiltration to bone marrow

Initially, the patient received chemotherapy according to the NHL1 protocol. After the bone marrow puncture results showed infiltration into the bone marrow, the chemotherapy protocol was changed to an acute lymphoblastic leukemia protocol.

RESULTS AND DISCUSSION

LBL is a rare disease. The total incidence of LBL, estimated at 1.3/100,000 per year, is less than 10% of the total ALL/LBL rate. LBL accounts for 2-4% of non-Hodgkin's lymphoma in adults and 30% in children (8). Only 1%–4% of all gastrointestinal (GI) malignancies are primary gastrointestinal (GI) lymphomas, making them incredibly uncommon. Lymphomas can develop in any area of the GI tract, with the stomach experiencing the highest incidence (50–60%), followed by the small intestine (20–30%), and the large intestine, rectum, and esophagus (1% of cases). The original tumor in this instance had clinical signs of intussusception and was located in the small intestine in the abdomen (Fig. 1). A pathological condition known as intussusception develops when the proximal bowel segment telescopes into the lumen of the distal segment. Less than 2% of all gastrointestinal malignancies are tiny intestinal primary malignant tumors. The ileum (60–65%) and jejunum (20–25%) were the next most frequent sites, followed by the duodenum (6%–8%) and elsewhere (8%–9%) (5). Meckel's diverticulum has been identified as the major location of intussusception most frequently. Weight loss has been found to be a prominent feature of intussusception (9). In this case, there is evidence of malnutrition associated with anorexia and prolonged vomiting. Most of these complaints appear in patients' clinically as an abdominal mass, intestinal obstruction, perforation, bleeding, or intussusception (10).

Ultrasonography is used in the diagnosis of intussusception, with sensitivity and specificity of up to 100%. Abdominal

ultrasound in this case showed the target sign and non-contrast abdominal MSCT results showed ileocolic intussusception. Surgery is the main modality of treatment, followed by adjuvant radiotherapy or chemotherapy (5). In this case, an intussusception ileal resection was performed(Fig.1). From the supporting examination, the histopathological Non-Hodkin lymphoma was found (Fig. 2), The difference between ALL and LBL is mainly based on the extent of bone marrow involvement (BM). Confirmatory biopsy is required to establish the correct diagnosis of LBL, evaluate disease morphology and immunophenotype (8) (11).

In this patient, the bone marrow puncture results showed infiltration into the bone marrow (Fig. 4). LBL patients' standard treatment choices are based on a rigorous multidrug leukemia chemotherapy program. This treatment plan includes 7–10 medications, including intense intrathecal chemotherapy on a type C basis, as well as cyclophosphamide, methotrexate, prednisone, vincristine, cytarabine, thioguanine, l-asparaginase, nitrosourea, and anthracyclines. For patients with mild or severe illness, the uterine regimen does not differ significantly. Conventional NHL regimens, aggressive NHL protocols, and ALL treatment protocols have all been used as approaches to LBL therapy. So, induction of chemotherapy, early CNS prophylaxis, consolidation, and future maintenance therapy are all included in contemporary LBL protocols (12). In the initial case, the diagnosis was made using a non-Hodgkin's lymphoma protocol. However, in our patient, CD 20 was positive (Fig. 3) with an increase in LDH (4534 IU/L). The higher the rate of proliferation of cancer cells, the higher the intracellular lactate dehydrogenase enzyme found in the blood circulation (13). A positive CD20 result is appropriate for diffuse large B-cell lymphoma (14). So the therapy given is a high-risk ALL chemotherapy protocol.

There are hardly many cases of facial nerve paralysis. Incidence among children under the age of 10 is 2.7 per 100,000 annually. Both benign and malignant neoplasms can paralyze the face in children (15). In malignant diseases, the paralysis of the facial nerve may be brought on by perineural infiltration of the temporal bone or facial nerve, invasion of the meninx by lymphoma of the central nervous system, infection and hemorrhage around the facial nerve, chemotherapy-associated toxicity to the nerve, or reactivation of a latent viral infection (4). Cytological analysis (CSF) revealed the presence of cancerous cells in this instance.

Infiltration causes nerve and blood vessel compression and injury, which results in neuropathy. Chemotherapy-induced partial improvement of facial paralysis prompted us to postulate that perineural infiltration might

have taken place. In this patient, the facial paralysis vanished within a week of finishing chemotherapy.

CNS involvement at diagnosis was significantly associated with worse outcome as well as Ann Arbor stage IV, and serum LDH levels >300 IU/L (8). In a literature review, lymphoma appeared as intussusception showed only 3 out of 10 children as long term survivors (5). This case has a poor prognosis with CNS infiltration and a significant increase in LDH.

CONCLUSION

Careful examination of patients with abdominal masses is required. Lymphoma with a primary tumor in the intestine and clinical intussusception is rare. To diagnose LBL, it is important to perform a bone marrow puncture as well as histopathological and histoimmunochemical examinations. This examination has an effect on therapy. Surgery and chemotherapy are effective therapies.

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