
A Case with Gaucher-Like Cells

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ABSTRACT

Gaucher-like cells (GLC) are sometimes indistinguishable from real Gaucher cells. GLC can be detected in various diseases. We present a 4.5 year old boy with massive cervical lymphadenopathy and an intraabdominal mass mimicking lymphoma. Many GLC were seen in the fine needle aspiration material of an enlarged lymph node. Ziehl-Neelsen stain of the aspirate revealed many acid-fast bacteria in the GLC. Fine needle aspiration might provide valuable information in the evaluation of enlarged lymph nodes.

Key Words: Gaucher-like cells, Lymph node aspirate, Tuberculosis.

Turk J Haematol 2001;18(4):281-283.

Received: 02.03.2001 *Accepted:* 19.04.2001

INTRODUCTION

Gaucher-like cells (GLC) have been described in a variety of disorders^[1-3]. Under the light microscope, these cells are indistinguishable from true Gaucher cells. GLC may be seen in infectious, haemolytic and myeloproliferative disorders and hematologic malignancies of lymphocytic origin^[3-11]. The presence of GLC can be explained by a greater than normal inflow of globosides and gangliosides that may overwhelm the catabolic capacity of normal cells. Electron microscopic examination of such cells shows abundant heterogeneous inclusions. In contrast, ultrastructural observation of true Gaucher cells reveals cytoplasmic sacks containing tubular formations, but no cell debris or dense bodies. This tubular formation has been shown to be composed of aggregated glucocerebroside molecules contained within

lysosomes. Electron microscopic studies and specific enzyme assays have been used in conjunction to establish definite distinctions between true Gaucher cells and GLC^[2]. Earlier reports of the association of hematologic disorders and Gaucher disease were made before these diagnostic methods were available. This implies that some reports of the association must be analyzed cautiously.

CASE REPORT

A four-year-old boy was first seen in a local outpatient clinic with a 4-month history of cervical lymph node enlargement and abdominal pain. Abdominal lymphadenopathy was detected in the CT scan, which was performed with the suspicion of malignancy, and the patient was referred to our haematology-oncology department.

Physical examination revealed bilateral cervical lymphadenopathy, the greatest being 5 x 5 cm in diameter in the left cervical region, and a mass in the left upper quadrant of the abdomen, 4 x 3 cm in size. Laboratory data showed mild anaemia (Hb: 10.4 g/dL, Hct: 33.1%, WBC: 13700/mm³ with 66% PNL, 30% lymphocytes 2% bands and 2% monocytes) and an elevated erythrocyte sedimentation rate (55 mm/h). Blood biochemistry was normal except for an increased CRP: 7.2 mg/dL (normal < 0.6 mg/dL).

Fine needle aspiration material of the cervical lymph node was stained with May-Grünwald-Giemsa and revealed GLC (Figure 1). Tuberculosis (Tbc) is one of the disorders where GLC can be encountered. Due to the high prevalence of Tbc in our country, the material was stained with Ziehl-Neelsen, which showed many acid-fast bacteria in the GLC (Figure 2).

The family history was negative for Tbc. The tuberculin skin test was nonreactive and the chest X-ray revealed no pathology. Three consecutive early morning gastric aspirates were obtained and cultures of the gastric fluid grew *Mycobacterium tuberculosis*. Antituberculous therapy consisting of isoniazid and rifampin was given for one year with streptomycin in addition during the first two months. The clinical findings responded rapidly to treatment and the patient is in good clinical condition with no complaints.

DISCUSSION

Typical Gaucher cells have one or more ec-

centric nuclei and stripped cytoplasm similar to crumpled tissue paper or jumbled tooth-picks in texture. In some instances the structure is indistinguishable from GLC by both light and electron microscopy. Gaucher cells have spiralled tubules rather than fibrils which are typical for GLC^[1-3].

Increased cellular breakdown and catabolism may play a role in the pathogenesis of GLC^[1]. These cells are macrophages. In normal bone marrow aspirates, these cells can be seen but they are few in number. In the setting of increased cellular breakdown, the insufficient digestion of cellular waste products by macrophages leads to intralysosomal storage of these waste products, creating the GLC. The storage of such material leads to deformation and angulation of the lysosomes leading to a more typical appearance. Under the light microscope, it is difficult to differentiate real Gaucher cells from GLC^[1-3]. By measuring the intracellular tubule size with the electron microscope, GLC can be more easily distinguished. The real Gaucher cells have tubules spiral in shape and wider than 60 nm while GLC have tubules 16 nm in width.

GLC have been reported in hemolytic anemia (dyserythropoietic anemia type 2, β -thalassemia), infectious diseases (Tbc, HIV), malignancies (chronic myeloid leukemia, acute lymphoblastic leukemia, nonHodgkin's lymphoma, Hodgkin's disease, plasma cell dyscrasias), immune thrombocytopenic purpura and juvenile rheumatoid arthritis when gold salts are used and after cytotoxic therapy^[1-5]. Occurrence of GLC in these disor-

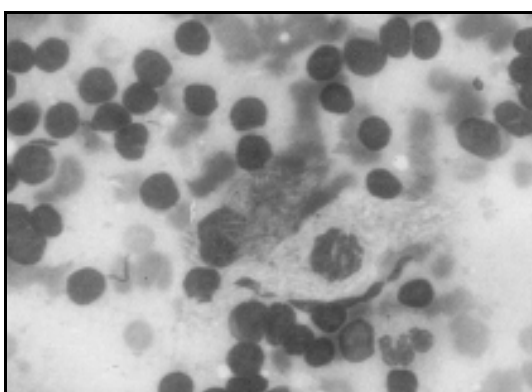


Figure 1. Gaucher-like cells (May-Grünwald-Giemsa).

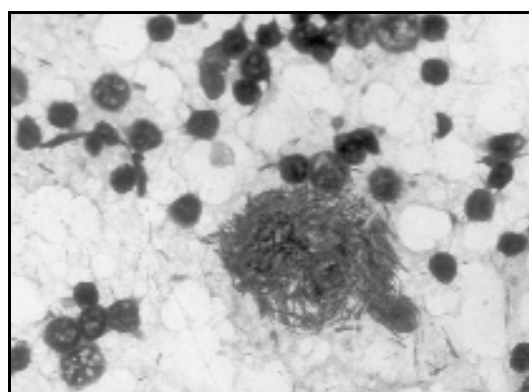


Figure 2. Ziehl-Neelsen stain, with many acid-fast bacteria in the GLC.

ders is attributed to overloading of normal enzymatic mechanisms for handling products of cellular breakdown. As we did not study the cells by electron microscopy, we could not differentiate tubules from fibrils. In our case, the Ziehl-Neelsen stain showed that the pseudo-Gaucher appearance is a result of phagocytosis of Ziehl-Neelsen positive mycobacteria (Figure 1, 2).

Cervical lymphadenopathy is a frequent clinical finding in childhood. In most instances enlarged lymph nodes represent transient proliferative responses to local or generalized infections. Although viral or less commonly nonspecific bacterial infections are responsible for cervical lymphadenopathy in most cases, Tbc is a possible etiology especially in areas with a high prevalence rate of Tbc^[13]. The rate is 4.59% for children 5-7 years of age in Turkey^[14].

Since both benign and malignant processes can cause lymph node enlargement, lymph node biopsies are frequently performed in patients with large and long-lasting lymph nodes. Fine-needle aspiration is a less invasive but cheap and practical method which enables a quick examination of the content of the enlarged lymph node^[15]. Besides being useful for the differential diagnosis of malignancies, fine-needle aspiration can provide both microbiological and cytological evidence of infectious diseases.

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