

AN UNUSUAL PRESENTATION OF BURKITT'S LYMPHOMA

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ABSTRACT

Burkitt's lymphoma grows very rapidly and a person who appeared in good health a month or 6 weeks ago may now be critically ill. I present a case of Burkitt's lymphoma that was having symptoms for one year, but she was not looking like a patient and was missed by the clinicians for one year.

KEY WORDS: Burkitt's lymphoma, Lymphadenopathy, Lymphoma.

INTRODUCTION

Burkitt's lymphoma is an uncommon type of non Hodgkin Lymphoma (NHL). Burkitt's lymphoma commonly affects children. It is a highly aggressive type of B-Cell lymphoma, that often starts and involves body parts other than lymph nodes. In spite of its fast-growing nature, Burkitt's lymphoma is often curable with modern intensive therapies.

There are two broad types of Burkitt's lymphoma – the sporadic and the endemic varieties. There is a very high incidence of this disease in equatorial Africa and disease in this region is called endemic Burkitt's lymphoma. Disease in other regions of the world is much less common and is called sporadic Burkitt's lymphoma. Though they are the same disease, the two forms are different in many ways.

Endemic Burkitt's lymphoma: In equatorial Africa, about half of all childhood cancers are Burkitt's lymphoma. The disease involves children much more than adults and is related to Epstein Barr virus (EBV) infection in 95% cases. The male to Female ratio is 2:1. The commonest site of disease presentation in eBL is the face with multiple facial bone involvement. It involves the maxilla and mandible.^{7,9,10}

Sporadic Burkitt's lymphoma affects the rest of the world, including Europe and America. It is also mainly a disease in children. The link between EBV is not as strong as with the endemic variety, though direct evidence of EBV infection is present in one out of five patients. More than the involvement of lymph nodes, it is the abdomen that is notably affected in more than 90% of the children. Bone marrow involvement is more common than in the endemic variety. Jaw involvement is extremely rare.⁷

In children, symptoms may appear as soon as four to six weeks after the Burkitt's lymphoma begins to grow. The more common symptom pattern is a large tumor in the child's abdomen accompanied by fluid buildup, pain and vomiting. If the lymphoma begins in the bone marrow, the child may bleed easily and become anemic. In adults the first symptoms of Burkitt's lymphoma may be generalized lymphadenopathy with pressure effects.

There may be unexplained itching or significant weight loss. Other patients may have more general symptoms such as fever and lethargy.

CASE HISTORY

A 16 years old girl presented with pain abdomen for one year. She was also giving history of vomiting and loose motions for two months. She had anorexia and weight loss during this period.

On examination she was a young fair looking girl. General examination was normal. A mass could be seen in the upper abdomen. On palpation a hard mass could be felt extending from the epigastrium down to below the umbilicus. It was fixed to posterior structures. The peripheral lymph nodes were not palpable. Rest of the systemic examination was normal.

Ultrasonography of abdomen showed enlarged matted mesenteric and para aortic lymph nodes with ascites.

Her routine investigations showed: Haemoglobin 12.6 g/dl, TLC 8,700/mm³ Neutrophils 64%, Eosinophils, 3%, Lymphocytes 32%, Monocytes 1%, Blood group A+ve, ESR 38 mm 1st hour, HBsAg & HCV negative. X-Ray chest was unremarkable.

This patient was subjected to exploratory laparotomy. There were enlarged mesenteric Lymph nodes matted together and forming abdominal mass of around 10 x12 cm in the midline with adherent small gut loops. These were fixed and hard. Two small lymph nodes were excised for histopathology. Liver and spleen were normal. Biopsy report came out as Burkitt's lymphoma.

Patient was subjected to chemotherapy and is showing improvement.

DISCUSSION

The case is unusual in the sense that the patient was having pain in upper abdomen for the last one year and the mass had reached to such a huge size that it could be seen while patient was lying or standing but the symptoms were very mild and even her look was not like a patient having cancer in her belly and the Hb was 12g/dl.

Burkitt's lymphoma is a type of B-Cell lymphoma. In 1956, a British surgeon called Dennis Burkitt, described this unusual type of lymphoma (1). In Africa, this tumour accounts for approximately 50 % of all childhood cancers. Outside Africa, it accounts for less than 2 % of all cases of Non Hodgkin' lymphoma. In an Indian series of solid malignant tumours in children, paramanik et al, in 1997 studied 263 cases over 10 year period and found 02 cases. (0.76%) of B.L (5). Many etiologic theories have been espoused. The role of EBV in BL is not well understood. The virus may be a prime etiologic agent, a co-carcinogen, or just an innocent passenger. This virus preferentially infects B cells via the C3d complement receptor, CD21 (9). Other cofactors may include. Chromosomal abnormalities, Immune defects, and protein energy deficits. Although the most important prognostic features have yet to be determined, some features that have been associated with adverse outcome in adults and children include older age, advanced stages, poor performance status, bulky disease, high LDH, and CNS or marrow involvement (2-8). Staging is performed using the Ann Arbor or, more often, the St.Jude/Murphy staging system (4). Approximately 30% of patients present with limited stage disease. (I or II), while 70 % present with wide spread disease (stage III or IV) (3). Burkitt's lymphoma usually starts in the abdomen but it may also affect other organs, such as the eye, the Ovaries, Kidneys and glandular tissue as the breast, thyroid and tonsil. Burkitt lymphoma usually affects the jaw bone. It can also affect the bone marrow. Diagnosis is done by removing the enlarged lymph node or part of it. Biopsy may be taken from other tissues.

Chemotherapy is used to treat this type of Cancer. Commonly used medicine includes Pred-

nisolone, cyclophosphamide, vincristine, cytarabine, doxorubicine, methotrexate and others. More than half of those treated with Burkitt's lymphoma can be cured with intensive chemotherapy. The rate may be lower if the cancer spread to the bone marrow or spinal fluid.

Radiation therapy is used to treat lymphomas that affect the jaw and the area around the eyes. New method of treatment for Burkitt's lymphoma includes bone marrow or stem cell transplantation and monoclonal antibodies (antibodies produced by cloned mouse cells grown in a Laboratory).

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