CASE REPORT

BURKITT'S LYMPHOMA

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ABSTRACT:

A case of Burkitt's Lymphoma is reported here in a patient who had disuse swelling of the mandible and right maxilla. It almost obliterated the buccal cavity and invaded the orbits resulting in bilateral proptosis and blindness.

INTRODUCTION:

Burkitt's Lymphoma is a rare monoclonal B cell neoplasm. It was first described in 1958 by Denis Burkitt, who reported rapidly growing jaw tumours and abdominal masses in Uganda children. According to Keil's classification, Burkitt's lymphoma has been divided into:

- i) Endemic or African type.
- ii) Non-endemic or South American type.

In the African type, in the first decade of life, different clones of cells e.g. centroblasts proliferate and their DNA express receptors for C# and Epstein Barr viruses (EBV). These centroblasts cells are triggered to proliferate by infections such as malaria and EBV infection. These cells appear frozen in functional immaturity. This frozen state accounts for the morphological monomorphism found in the African type.

The EB virus is present in ninety person of the African type but in less than half of the non-endemic cases⁵. Typically, primary EBV infection precedes the development of Burkitt's lymphoma by at least seven or more months. Chromosomal translocation has been identified in Burkitt's lymphoma.

CASE REPORT:

A 13-year-old male child was admitted in ENT Unit in DHQ Hospital Abbottabad on December 10, 1990. He had severe respiratory distress for which an emergency tracheostomy was performed and the patient was shifted to the intensive care unit, being in a precarious condition. The history of the patient dated twelve days back, when his parents noticed a swelling on the neck which increased progressively and one day before admission he developed difficulty in breathing along with dysphagia

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EXAMINATIONS:

The patient had bilateral proptopsis with swelling of the mandible and maxilla on the right side, almost obliterating the buccal activity. The patient was dehydrated and in a state of peripheral circulatory failure. He was running a fever of 100 degree Fahrenheit. A solitary lymph node was palpable in the neck. He was pale and confused. Other systems were normal.

Blood picture showed a haemoglobin of 8.6 g/dl and a total leucocyte count of 6,7000/cm³. The patient collapsed on the second day of admission. Keeping in view these typical findings a provisional diagnosis of Burkitt's lymphoma was made and the following treatment was given:

- i) Cyclophosphamide injection, 300 mg intravenously.
- ii) Vincristine injection, 0.5 mg intravenously as continuous infusion.

The patient improved in 36 hrs after the start of treatment. The swelling disappeared, he become fully conscious, started talking and was able to eat, but he had lost his eye-sight.

The patient was also given other supportive measures like blood transfusion and antibiotic. He was given a second course of therapy, after ten days. He was sent home and advised for a revisit after two weeks for the third cycle of treatment, but he arrived late due to personal reasons. He redeveloped the swelling of the jaw along with sever hematemesis.

Blood picture then showed a haemoglobin of 7.3 g/dl, a total leucocyte counts of 6,500/cm³. A differential count showed lymphocytes 50%, monocytes 2%, polymorphs 17%, eosinophils 1% and immature blast cells 30%. Bone marrow showed myeloid cells 36%, immature blast cells 28%, lymphocytes 28%, monocytes, 8%, immature cells were of lymphoid type, some of them were having cytoplasmic vacuolations similar to these seen in Burkitt's lymphoma. The patient was given another cycle of treatment but he had severe hematemesis which proved fatal.

DISCUSSION:

In this patient who come as an emergency to the ENT Department, the tumour progressed so quickly that an emergency tracheostomy had to be done. Keeping in view the typical features of Burkitt's lymphoma and his rapidly deteriorating condition, the appropriate treatment was begun after a provisional diagnosis of the condition and the treatment proved fortuitous. The bone marrow was strongly suggestive of Burkitt's lymphoma.

Burkitt's lymphoma is classified as a high grade, small non-cleaved cell malignant lymphoma. It is composed of intermediate sized lymphoid cells. Larger number of mitotic figures are seen an 'starry sky' appearance is characteristics.

In the African type, also called endemic Burkitt's lymphoma, there is a strong association with malaria, because repeated attacks of malaria lead to stimulation of lymphoid tissue and the patients become more susceptible to neoplasia in the presence of EB virus. This may result in the development of Burkitt's lymphoma. The endemic type has distinct epidemiological features and gross clinical presentation that differs sharply from the American (non-endemic) Burkitt's lymphoma. In the endemic type, 72 percent of the

patients present with jaw swelling, 50 percent with abdominal swelling and 30 percent with CNS disease at the time of diagnosis. Kidneys are most frequently and commonly affected. Patients may present with paraplegia due to infarction of lower thoracic segments of the spinal cord. Involvement of other parts of CNS including the cranial nerves especially the optic nerve may be a presenting feature in relapses. Patients may present with bilateral ovarian tumours in the females and testicular tumours in the males. Breast tumours are frequent and occur during pregnancy and lactation.

Lymph nodes, spleen and lungs are rarely affected in the African (endemic) type as against the non-endemic type. The non-endemic type occurs sporadically in other parts of the world. It may be associated with immune dysfunction. It is similar in presentation with endemic Burkitt's lymphoma but jaw involvement is very rare, present only in 1-2 percent of the cases. There is high incidence of involvement of lymph nodes, bone marrow, lungs and spleen.

Burkitt's tumours are extremely sensitive to chemotherapy. Tumour regression is clinically evident in 35-48 hrs after initiation f treatment, partial remission, however is usually followed by evidence of further tumour growth.

Combination chemotherapy is used in all the patients and may be highly effective. High dose cyclophosphamide may be curative as a single agent, but complete response rates and remission durability of combination regimens is superior.

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