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ISSN: 0976-3031

International Journal of Recent Scientific Research
Vol. 5, Issue, 5, pp.918-920, May, 2014

International Journal of Recent Scientific Research

RESEARCH ARTICLE

NASOPHARYNGEAL BURKITT'S LYMPHOMA A CASE REPORT

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ARTICLE INFO

Article History:

Received 12th, April, 2014
Received in revised form 22th, April, 2014
Accepted 15th, May, 2014
Published online 28th, May, 2014

Key words:

Nasopharynx, Burkitt's lymphoma, children, Unusual site, diagnostic challenge.

ABSTRACT

Burkitt's lymphoma is a common childhood tumour in Africa.It is a highly aggressive lymphoma characterized histologically by a diffuse infiltrate of intermediate size cells with a high mitotic rate. The lymphomas commonly have a significant spontaneous cell death (apoptosis) which results in a starry sky appearance caused by numerous macrophages that have engulfed the apoptotic debris. Affectation of the nasopharynx is unusual. We report a case of 7 year old primary school pupil who presented with Right-nasal mass, which was later, confirmed to be Burkitt's lymphoma of the Nasopharynx. The case is discussed because of its rarity and diagnostic challenges.

INTRODUCTION

Burkitt's lymphoma is a B-cell neoplasm, a non-Hodgkin lymphoma characterized by small non-un cleaved cells that are uniform in appearance and produce a diffuse pattern of tissue involvement. This neoplasm is one of the fastest growing malignancies in humans.[1, 2] The cells of Burkitts lymphoma are characterized by specific cytogenetic defect, a balanced reciprocal translocation of genetic material from the long arm of chromosomes 8 to the long arm of chromosome14s .Most cases are characterized by chromosomal translocations involving the MYC gene on chromosone8, The MYC gene most commonly translocated to the IGH gene on chromosome14, resulting t (q24, q32) [8, 14]. It can also involve the light chain genes on chromosomes2p12 (k) and22q11 (v) (3, 4).

Two variants of Burkitt's lymphomas are recognized. African variety (Endemic Burkitt's lymphoma) and non-African type (Sporadic Burkitt's lymphoma). Although very similar in histologic and cytologic features, they have very different epidemiologic patterns and clinical presentations. African Burkitt's lymphoma is closely linked to Epistein Barr virus (EBV) primary cause of infectious mononucleosis. It presents most often as a jaw or orbital tumour however other sites may be affected as this case under review. It occurs endemically in central, and West Africa. It must be noted that these areas are also endemic for malaria and yellow fever (1). In contrast non-African (sporadic) Burkitt's lymphoma occurs outside these endemic regions and presents primarily as an abdominal mass [5, 6, 7 and 8].

Treatment modalities are basically the same in terms of chemotherapy but there may be additional surgical intervention in terms of local affectation or complication.

While Burkitts lymphoma commonly affects the head region, it has been observed in other parts of the body like the abdomen. Affectation of the nasopharynx is however rare and the diagnosis can be missed.

Case Report

A seven year old male primary three pupil was referred for otolaryngological review from Paediatric ward because of persistent nasal blockage, occasional epistaxis with copious mucoid nasal discharge. Patient was said to have been reviewed by a haematologist.

On review, the child has been on admission in Paediatric ward for a week. History was that of two weeks generalized body weakness, loss of apetite, low grade fever, recurrent epistaxis and nasal blockage with blood stained mucoid discharge. There was no history of cough nor night drenching sweats, no history of vomiting, diarrhea, nor heamaturia neither was there any history of trauma to the nose or foreign body insertion. There was no history suggestive of haemoglobinopthy. There was no history of previous hospital admission neither was there any history of previous blood transfusion. Child is the 4th child of a family of five children in a polygamous setting. His mother is the second wife. There is positive history of epistaxis in the 1st sibling. And previous epistaxis in the child some years back due to trauma to the nose. The attending Paediatrician noted the chronically ill looking child with proptosis, Right greater than the left, chest was clinically clear and pulse68/min, HR80/min, HS…s1s2, no murmurs. There was no obvious jaw swelling nor abdominal organomegaly. Initial assessment was Septicaemia with Epistaxis R/o DICs, Lymphoma. Acute leukaemia,and Retinoblastoma. Haematologists review was suggestive of Intra cranial mass to R/O Lymphoma, Burkitts, stage B.

Clinical examination revealed a chronically ill looking child with slightly parlor, rhinos copy (after nasal toileting) showed fleshy non-haemorhagic mass in the right nasal cavity covered with thick mucoid discharge slightly blood stained but there was no obvious mass in the left nasal cavity which was filled with yellowish thick mucoid discharge. There was associated slight proptosis bilaterally but vision was intact. There were multiple non-tender sub-mandibular nodes but no palpable cervical, axillary, inguinal nodes nor palpable abdominal

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mass. Initial assessment of Nasoantral tumour r/o nasopharyngeal carcinoma was made. Ophthalmic review was suggestive of Antral tumour with Orbital cellulitis.

Full Blood Count(FBC)…pcv-32%,Wbc-4500, neut 55%, lymh45%, platelet-118000,Blood film…normal ,Retroviral screening(Rvs)-nonreactive to HIV1&2, Blood culture-no growth after 7 days, Bone Marrow Cytology(BMC) …not suggestive of Leukaemia or any malignant process. CSF was negative for Burkitts cells. The X-ray of the par nasal sinuses showed the sinuses to be normal. He was planned for Examination under Anaestisia (EUA) and nasal clearance which was done. At surgery naso-pharynx was filled with fleshy mass found to be continuous with the right nasal mass. Histology of specimen showed Burkitt's lymphoma of the naso-pharynx.

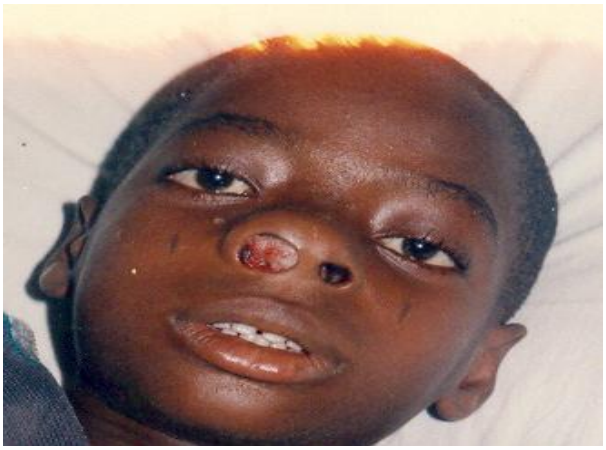

Fig .1 Child with Right Nasal Mass

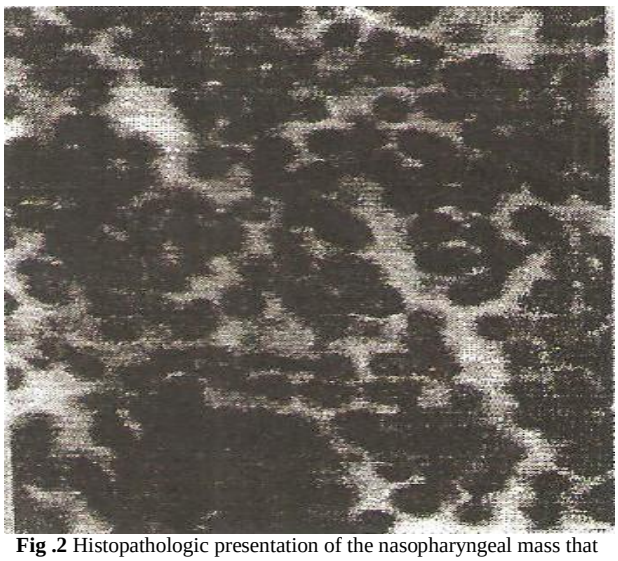

Fig .2 Histopathologic presentation of the nasopharyngeal mass that shows Burkitt's cells

He was subsequently placed on chemotherapy with the regime of 15 days intervals (5 days of active treatment plus 9 days of rest).He was scheduled to have 8 courses of the Chemotherapy.

1. Tablet Allopurinol 100 mg tds during active phase of the cycle.
2. Tablet Paludrine 100 mg daily throughout period of chemotherapy
3. Intravenous cyclophosphamide 1.2 gm /m2/day 1
4. Intravenous vincristine 1.4 mg /m2) day 1
5. Intravenous C. Arabinoside 100mg/m2days 1, 2&3
6. Intrattechal C.Arabinoside50mg day 4.
7. Intratechal Methotrxate 12.5mg days 1&8

Absolute Neutrophil Count…..above 1000
Absolute Platelet Count…….above90000.

This was accompanied by adequate hydration both orally and intravenously.

Patient improved markedly initially with this regime. The nasal mass disappeared within two weeks of Chemotherapy. However while still on chemotherapy (4th course), the disease disseminated to the lumbar spine, left elbow and loss of vision in both eyes. He had to be transfused on two occasions with one pint of packed cells due to anaemia. He was on admission throughout. Patient succumbed to death by the 6th course of chemotherapy.

DISCUSSION

Burkitt's lymphoma is one of the fastest growing malignancies in humans[3, 9, 10 and 11]. It is not an uncommon disease in the tropics among the paediatric age group with peak age incidence for the African variety (Endemic Burkitt's lymphoma) between 5 – 10 years, while that of Non-African type (Sporadic Burkitt's lymphoma) is usually 11 year and above [3,4]. Common sites affected are the jaws, orbit and abdomen.[5, 7, 8, 9, 12, 13,, 14]. Other not so common sites are extradural tissues, Para-pharyngeal space, genitourinary system, nasal alae, bone, testes, breasts, thyroid, parotid and gland and skin[15, 16, 17,18,19,20]. Involvement of the nasopharynx (post-nasal space) and nasal cavities is rare and scanty in literature.

Nasopharyngeal malignancies are not common tumours. Incidence varies from region to region with incidence of 1 per 100,000 in low incidence region to 15 – 30 per 100,000 at high incidence regions [21, 22]. It can occur at any age group but very rare below the age of 10 (10). Peak age is between 3rd – 4th decades. It is three times commoner in males than females.

Clinical features include nasal symptoms especially epistaxis, nasal blockage, blood stained nasal discharge. Nasal mass is uncommon. Neuro-ophthalmic manifestations include proptosis, diplopia, ophthalmoplegia blindness, dysphagia or drooling of saliva while about 60% of the patients presents with cervical mass [10, 11, 23].

The commonest histological type in Naesopharyngeal malignancy is squamous cell carcinoma – (80 – 90%). [4,10,11]. Therefore this particular case, Nasopharyngeal Burkitt's lymphoma (NBL) because of its unusual presentation, calls for high index of suspicion. A relationship between Burkitt's lymphoma and nasopharyngeal carcinoma has been reported with both lesions being commonly associated with Epstein Barr Virus (EBV) infection (.Prognosis of Burkitt's lymphoma depends on the histologic type and stage of the disease but good if detected early and prompt treatment initiated. Ninety percent respond to chemotherapy but all die within six months if there is no treatment [24, 25, 26]. However recurrence and dissemination do occur occasionally while treatment is ongoing as the case was in this report.

Finally, it is necessary to emphasize here that a high index of suspicion is needed for prompt diagnosis of nasopharyngeal malignancies. There is urgent need for more clinical training and extensive public awareness on this clinical entity.

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