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Bilateral Orbital Rhabdomyosarcoma mimicking Burkitt's Lymphoma

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Summary

Falade AG. Osinusi K, Baiyeroju-Agbeja AM, Ogunbiyi JO, Owoeye JFA, Thomas J. Bilaterial orbital rhabdomyosarcoma mimicking Burkitt's lymphoma *Nigerian Journal of Paediatrics* **1998; 25:25.** Bilaterial orbital rhabdomyosarcoma mimicking Burkitt's lymphoma in an 11-year old girl is reported. The patient presented with bilateral proptosis and right cervical lymphadenopathy. Despite consultations at a number of hospitals, a definitive diagnosis was not made until presentation at the University College Hospital, Ibadan, where cytological examination of a fine needle aspiration biopsy specimen of the tumour and the lymph node revealed changes that were consistent with those seen in rhabdomyosarcoma. This case emphasises the fact that rhabdomyosarcoma is an important differential diagnosis of orbital tumours in childhood, especially in Burkitt's lymphoma endemic areas.

Introduction

THE early diagnosis of rhabdomyosarcoma requires that the physician exercises keen clinical judgement when the patient is first seen. The median delay between the onset of symptoms and signs and biopsy has been estimated to be about two months.[1] This delay can be further prolonged when rhabdomyosarcoma presents as orbital tumours in areas where commoner tumours like Burkitt's lymphoma,[2] neuroblastoma,[3] and retinoblasoma[4] are more likely to be thought of first. Yet, it is important to make a correct diagnosis of this tumour because the chemotherapeutic protocol is different from those for the commoner tumours. Furthermore, diagnosis of undifferentiated rhabdomyosarcoma, like other small, round, blue cell tumours of childhood, is often difficult without ancillary techniques like immunohistochemistry or electronmicroscopy.[5] The present communication highlights the diagnostic problems posed by this tumour and some clinical features that may help in distinguishing it from other childhood orbital tumours.

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Case Report

AB was an 11-year old girl who was referred from an Eye Centre, with a clinical diagnosis of retinoblastoma. She presented at the Eye Clinic, University College Hospital (UCH), Ibadan, with a three-month history of loss of sight in both eyes and two-month history of bilateral proptosis. The illness started with frontal headaches, fever and vomiting which did not respond to antimalarials, and was later associated with progressive visual impairment. She subsequently developed proptosis of both eyes within a month of the onset of symptoms. The right cheek also became swollen, with associated dental mal-alignment and occasional bleeding from the mouth. There was associated weight loss. No definitive diagnosis had been made after previous consultations at two hospitals.

Physical examination on presentation at UCH,

revealed a wasted girl (weight 20kg, surface area 1.0m^2) with bilateral proptosis, the right more than the left. There was facial asymmetry with right maxillary swelling (Fig. 1). The upper lids were stretched over firm tender orbital masses (right 15cm x 10cm; left 8cm x 5cm) with both eyes pushed superiorly, the right slightly laterally. The masses were not retropulsible. The right orbital rim could not be felt due to the size of the mass. There was marked conjunctival hyperaemia with corneal necrosis, precluding any further view of the anterior or posterior segment. Detailed ophthalmological examination confirmed that there was no light perception in both eyes. The intraorbital masses had pushed the eyeballs so far forwards that the eyelids could no longer cover the cornea. This led to exposure keratopathy and necrosis with resultant sloughing off of the cornea. Although no view of the fundi could be obtained because of the necrosed cornea, the eyeballs were not enlarged and there were no intraocular masses. Once the corneas sloughed off, the intraocular contents were forced out by pressure from the intraorbital masses. The right cervical jugulo-digastric node was enlarged (3cm x 2cm), firm, mobile and non-tender. The liver edge was palpable 3cm below the right costal margin.

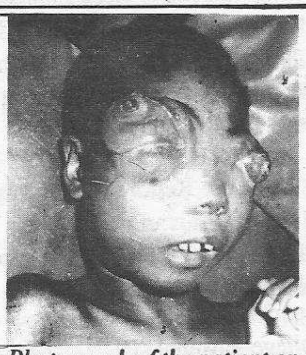

Fig 1. ***Photograph of the patient on admission, showing bilateral proptosis.***

The initial differential diagnoses were bilateral retinoblastoma and Burkitt's lymphoma. A full blood cell count revealed a packed cell volume of 32 percent, platelet count of 186 x 10^9/L and total white blood cell count of 12.5 x 10^9/L (neutrophils 85 percent, lymphocytes 14 percent, monocytes 1percent). She was mildly hyponatraemic with a serum sodium of 130mmol/L; the potassium was 3.3 mmol/L and uric acid, 4.9mmol/L. Chest radiograph was normal but radiographs of the maxilla and mandible showed "dental anarchy" and loss of dental lamina dura. She was started on chloramphenicol eye ointment, but subsequently developed complete corneal sloughing with loss of intraocular contents. There was therefore, no further need for surgical evisceration. However, cytological examination of the fine needle aspiration biopsy (FNAB) specimens obtained from the destroyed right eye and the enlarged right jugulo-digastric lymph node revealved spindle-shaped malignant cells mainly in singles and small clusters; these had pleomorphic, elongated and oval nuclei with prominent chromatin and abundant cytoplasm. These features were reported to be consistent with a malignant spindle cell sarcoma, most likely rhabdomyosarcoma. The FNAB report ranked rhabdomyosarcoma top of the list because the cellular features were consistent with a spindle cell tumour and cross striations were present together with abundant cytoplasm as with rhabdomyoblasts. Bone marrow aspiration revealed erythyroid hyperplasia.

She had five weekly cycles of intravenous (i.v.) cyclophosphamide 300mg/m^2), i.v. vincristine 1.5mg (1.5mg/m^2), and actinomycin D, 0.06mg (0.06mg/m^2). Supportive management included oral allopurinol 100mg thrice a day, transfusion of one unit of packed red blood cells for anaemia, i.v. cloxacillin (100mg/kg/24hr) and intramuscular gentamicin (7.5mg/kg/24hr) for presumed septicaemia, although blood culture results obtained later, showed no growth. She lost 3.0kg in weight during chemotherapy, and the sixth course was therefore, withheld. Palliative radiotherapy at a dosage of 20 GY in 10 fractions, was also given to

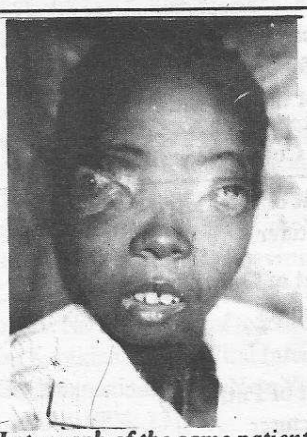

Fig 2. ***Photograph of the same patient after treatment. Note the considerable recession of the proptosis.***

both facial fields after completion of chemotherapy. She subsequently regained weight after two weeks of intensive nutritional rehabilitation. At discharge three months after admission, the proptosis had decreased appreciably (Fig 2) and she weighed 24.0kg.

Discussion

The commonest causes of orbito-facial tumours in Nigerian children are Burkitt's lymphoma,[2,6] retinoblastoma[4] and neuroblastoma.[3] Although one of the commonest sites of presentation of rhabdomyosarcoma is the head and neck,[1] this diagnosis is often not considered in the differencial diagnosis of orbital tumours. In retrospect, the clinical diagnostic errors in this case could have been avoided had the available clinical data been more critically assessed. For instance, the patient was aged 11 years when she was initially thought to have retinoblastoma, a tumour of young children, usually less than 6 years of age.[7] Fundoscopy which could have excluded retinoblastoma in our patient was not possible because the eyeballs had been destroyed by the tumour. The diagnosis of Burkitt's lymphoma, which was based on the presence of maxillary swelling and proptosis in the patient is also unlikely because of the long duration of the illness and the presence of cervical lymphadenopathy, an infrequent occurrence in Burkitt's lymphoma.[8] Characteristically, rhabdomyosarcoma is a relatively slow growing malignant tumour[9] compared with Burkitt's lymphoma. The radiological findings of 'dental anarchy' and loss of dental lamina dura which are considered characteristic of Burkitt's lymphoma of the jaw further compounded the diagnostic problem in our patient. It should be noted however, that these features are not limited to, and are therefore, not pathognomonic of Burkitt's lymphoma.

The present report highlights the difficulties which, because of lack of differentiation, are associated with the diagnosis of small round blue cell tumours of childhood. Identification of histogenetic origin is often necessary to ensure diagnostic accuracy. Lack of ancillary immunohistochemical stains and electronmicroscopy compounded this problem as these were not available in UCH at the time.

It is our view that rhabdomyosarcoma should be considered in the differential diagnosis of any childhood orbital tumour especially where the duration of the tumour is longer than one month. An urgent FNAB of the tumour and the regional lymph node if involved, should be carefully done as this can offer the quickest means of diagnosis and hence, prevention of destruction of vital structures such as the eyes.

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