Abdominal Presentation In Burkitt's Lymphoma With Mortality From Renal Failure: Report Of Two Cases

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Citation

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Abstract

Burkitt's lymphoma, the commonest childhood malignancy in Tropical Africa, although highly chemosensitive and now curable, still causes high morbidity and mortality in Nigeria because of various adverse factors militating against its effective management. We report two cases of Burkitt's lymphoma with massive tumour load who died from acute renal failure. The patients' case files were retrieved and reviewed. We also reviewed the literature on Burkitt's lymphoma.

Death from acute renal failure in these patients could have been prevented if treatment was commenced early. The various adverse factors affecting management outcomes of BL in Nigeria including poor education, poverty, poor environmental living conditions, massive tumour load at presentation and acute tumour lysis syndrome are well illustrated by these two cases.

INTRODUCTION

Burkitt's lymphoma (BL) is the commonest childhood malignancy in tropical Africa[1]. It is a rapidly proliferating tumour, which is highly chemosensitive. In developed centres of the world, about 90% of Burkitt's lymphoma patients go into complete remission at induction and 80% are cured[2]. In fact some developed centres are now facing the challenges of taking care of the late effects of treatment in adult long-term survivors of Burkitt's lymphoma. Complete remission rates in Nigeria have been variously reported as 48%[3], 70%[4], 67%[5] and 22.8%[6], with mortality rates of 60% within 4 months[7] and 70% within 3 months [8]. What is even worrisome now is the declining overall complete remission rates [6] . Various factors are responsible for the poor outcome of management of Burkitt's lymphoma in Nigeria. Abject poverty in the community which leads to high default rate from hospital treatment and deterioration of hospital facilities for monitoring all aspects of the chemotherapy [3,7] are high on the list of adverse factors affecting management. Other factors include late presentation, multiple tumour sites, and high tumour burden at presentation. Most of our patients are from the low socioeconomic group living in rural areas, associated with a combination of life styles such as poor nutrition and poor hygienic living conditions. The pattern of presentation also appears to be changing. Initially most cases presented with facial symptoms but currently in Nigeria more studies are showing predominant abdominal presentations in Burkitt's lymphoma [4,6] . This has implications both in tumour detection, management, and outcome. We present two cases of Burkitt's lymphoma with predominant abdominal presentation and death occurring from renal failure following massive tumour invasion of the kidneys. Detailed postmortem histopathological findings are discussed.

CASE ONE

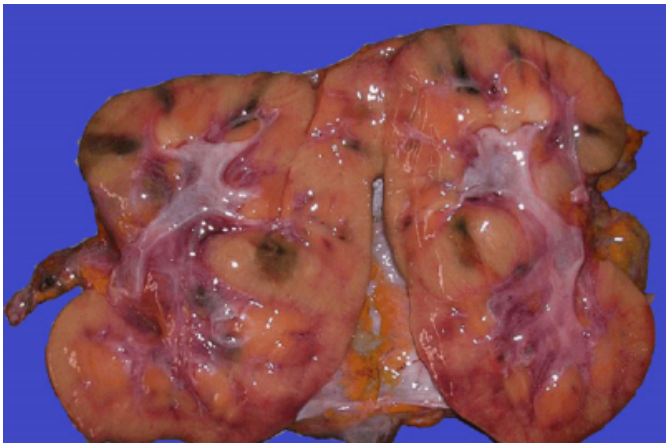
O.U., an 11 year old female Nigerian primary six pupil was first seen in October 2003 at the University of Nigeria Teaching Hospital Enugu, Nigeria with a two week history of a right sided jaw swelling and a one week history of inability to open the right eye. The jaw swelling rapidly increased in size, extended to the left jaw and later into the mouth with subsequent loss of teeth and inability to close her mouth. Another swelling was also seen on the bridge of the nose. One week after the onset of the illness, the right eye became permanently closed. There was associated low grade intermittent fever, swelling and bleeding from the gums, difficulty with swallowing and weight loss. She lived with her mother and siblings in a 2-room apartment. She was the 4 th child in a monogamous family with 3 boys and 3 girls. The father had died 3 years previously from hypertensive disease. The mother did not have any formal education.

Physical examination revealed an asthenic malnourished young girl who was pale, anicteric, and febrile. She had no peripheral lymphadenopathy. There was an obvious right mandibular mass with extension to the intra-oral cavity (hard palate) and left mandible. Another mass was also seen on the bridge of the nose. She had bilateral proptosis, much greater on the left than on the right. There was an associated paralysis of the 3 rd , 4 th , and 6 th cranial nerves. The abdomen was full with a firm nodular suprapubic mass, 14cm above the pubic symphysis. The spleen was palpable 6cm below the left costal margin while the liver was palpable 10cm below the right costal margin. A clinical diagnosis of Burkitt's lymphoma stage D was made and the patient was scheduled for investigation including Fine Needle Aspiration Cytology (FNAC) which could not be carried out immediately because of financial constraints.

On the second day of admission, the patient did not make any urine. An urgent serum electrolytes and urea showed potassium 5.6 mmol/L (3.5 – 5.0 mmol/L), urea 24.3 mmol/L (2.5 – 8.5 mmol/L), and creatinine 564 mmol/L (44.2 – 194.5 mmol/L). Full Blood Counts showed Packed Cell Volume (PCV) of 0.37L/L, White Blood Cell (WBC) count of 13.6 x 10 ^{9} /L and Platelet count of 21.5 x 10 ^{9} /L. The patient was commenced on intravenous fluids and frusemide to challenge the kidneys. FNAC of the jaw mass was done on the 6 th day of admission and was subsequently reported as “positive for malignant cells, compatible with small noncleaved cell lymphoma (Burkitt's lymphoma)”. Her condition deteriorated progressively until she died on the 9 th of the admission while still being resuscitated. Post-mortem examination showed extensive maxillary (palatal) and mandibular masses measuring 6cm and 11cm respectively in their longest axes which destroyed the affected bones. The masses were soft and had a variegated appearance with necrotic bones. The thorax showed 2 tumour nodules measuring 2cm and 0.5cm respectively. On opening the abdomen, 600ml of serous ascitic fluid was evacuated. There were tumour masses in the liver (multiple nodules measuring between 1cm and 6cm and involving 30% of the parenchyma), spleen (4cm nodule), both kidneys (diffusely invaded and enlarged with 90% of total renal tissue destroyed) (see figure 1), right adrenal gland (totally destroyed by retroperitoneal tumour), left adrenal (1cm nodule), both ovaries (diffusely invaded and enlarged), and retroperitoneum. The stomach contained about 200ml of altered blood and showed a bleeding 2cm superficial ulcer – Curling's ulcer. The urinary bladder mucosa had reddish areas and contained pussy urine – acute cystitis.

Figure 1

Figure 1: Showing the right kidney which is diffusely infiltrated by the tan coloured tumour mass.



Histological assessment of the various organs confirmed the pre-mortem cytological diagnosis of Burkitt's lymphoma with involvement of the maxilla, mandible, pericardium, liver, spleen, adrenal glands, kidneys, retroperitoneum, and ovaries. It showed sheets of monotonous intermediate-sized tumour cells with hyperchromatic nuclei and scanty cytoplasm. Many macrophages with abundant pale cytoplasm which frequently contain phagocytosed cellular debris were seen among the intermediate-sized tumour cells imparting a characteristic “starry-sky” appearance. There were extensive areas of coagulative necrosis.

CASE TWO

A 9-year-old girl was admitted on account of two weeks history of peri-orbital puffiness, generalised body pains, two day history of inability to walk, and fever. Physical examination revealed bilateral masses in the lower abdomen measuring about 6cm by 5cm each. They were non-tender, firm, and not fixed to skin or underlying structures. No facial mass or any other abnormality was identified. A diagnosis of Burkitt's lymphoma Stage D was made and the patient was commenced on cytotoxic therapy consisting of vincristine, methotrexate, cytosine arabinoside, predinisolone, and cyclophosphamide. The masses regressed with the first course of chemotherapy. However, she subsequently developed features suggestive of pneumonia. Patient suddenly became breathless, pale, with a high grade fever. She died after 17 days of admission.

Post-mortem examination showed patchy foci of consolidation on both lungs corresponding to bronchopneumonia. There was a whitish nodular mass measuring 3cm in the spleen. Both kidneys were enlarged

and completely destroyed by a whitish mass. Two nodular masses measuring 2cm each with a whitish 'fish-flesh' cut surface were isolated from the para-aortic lymph node region. All the other organs were essentially within normal limits.

Histological assessment of the retroperitoneal lymph nodes, splenic mass, and kidneys showed involvement by Burkitt's lymphoma. The sections had an appearance similar to that described in Case 1.

DISCUSSION

The first patient presented with only facial symptoms. There was hepatosplenomegaly and a suprapubic mass indicating abdominal involvement but the extent was not known. The patient, however, did not have any abdominal complaint. The second patient did not have any facial swelling besides peri-orbital oedema. She however had abdominal masses. These findings are similar to some Nigerian reports which showed that abdominal masses are noted on presentation in most of the patients with Burkitt's lymphoma [4,6]. The autopsy findings in Case 1 revealed very massive abdominal tumours. The liver weighed 1,750g with multiple intrahepatic nodules, accounting for 30% of the liver, which corresponds to approximately 525g of tumour. Since the liver has a large functional reserve which requires the destruction of about 75% of the liver mass before being overwhelmed, it is not surprising that this patient did not present with jaundice or any of the features of hepatic insufficiency.

The same goes for the kidneys. Compensatory single nephron hyperfiltration can maintain normal glomerular filtration rate in spite of 50% nephron loss (i.e., unilateral nephrectomy) for 2-3 decades without any serious consequences. Renal decompensation commences only when more than 50% of nephron mass is lost. This patient did not have any feature of renal failure until about 24 hours prior to demise, yet autopsy showed about 90% tumour destruction of the kidneys which had a combined weight of 920g (600g and 320g). This gives a total renal tumour mass of about 830g. This, likewise, is not surprising giving the immense compensatory capacity of the kidneys. Both ovaries had a total mass of 480g and about 80% tumour involvement, which corresponds to about 380g of ovarian tumour mass. This patient, therefore, had a total abdominal tumour load of 1,735g and yet presented only with facial masses.

In Case 2, both kidneys were totally destroyed by the tumour and the patient succumbed to renal failure that rapidly deteriorated and led to her death.

As noted above, recent works from Nigeria show that abdominal involvement is being noted with higher frequency than facial involvement [4,6]. Patients usually present earlier when there is facial manifestation because it is striking and disfiguring [5]. Patients with only abdominal tumours may present very late (terminally) or not at all because of the large hepatic and renal functional reserves. The autopsy findings in these patients clearly show that the patients died from the large tumour load (mostly abdominal) and terminal renal failure. The facial tumour in Case 1 contributed minimally to the tumour load and eventual outcome.

Late presentation of Burkitt's lymphoma in our environment can be partly attributable to the late manifestation of abdominal tumours as against the early manifestation of facial disease. This is something that should be borne in mind when managing Burkitt's lymphoma even when only a facial tumour is noted clinically. We recommend that at least, an abdominal ultrasound examination should be done to ascertain the level of involvement of the abdominal cavity. Renal and hepatic function tests will only reveal very advanced tumours. The low socioeconomic status of the general population is another crucial factor in our environment.

In conclusion, we have presented two cases of Burkitt's lymphoma with predominant abdominal presentations and death from destruction of both kidneys with ensuing renal failure.

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