# Neuroblastoma occurring in a 38-year old Nigerian man: a rare finding

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## Abstract

Neuroblastoma (NB) is a common malignancy in children, but rarely occurs in adults. Accepted unfavorable prognostic factors include age over one year, low histological grade and advanced stage, MYCN amplification, chromosomal aberrations, elevations of neuron specific enolase and lactate dehydrogenase, and increased catecholamine metabolites in urine or serum. In adults, abdomen/retroperitoneum are the primary sites and in children the adrenal gland. We report a 38-year old civil servant who presented at our urology clinic on the 21st of December 2007 with a six month history of right flank dull pain which was worse on walking and relieved by rest, hypertension and a large right retroperitoneal mass. Tumor resection revealed a grade III NB. Chemotherapy using a combination of vincristine, adriamycin and cyclophosphamide was started. Follow-up showed regression of the mass initially with a relapse after patient absconded for three months. He resurfaced with new masses and he had a repeat chemotherapy with disappearance of the masses and is currently undergoing further treatment. To our knowledge this is the only report of NB in an adult registered so far in Nigeria and perhaps the whole of Africa. Currently, there are no standard treatment guidelines for patients with NB in adulthood. This study emphasizes the need for a standard treatment regime for adult onset neuroblastoma and its recognition as a possible differential in intra-abdominal mass in adults.

## Introduction

Neuroblastic tumors, of which neuroblastomas are the most common, are embryonic neoplasms of the sympathetic nervous system, and are the most common solid neoplasms of childhood other than central nervous system tumors.[1,2] Neuroblastoma rarely occurs in adults, although abdomen and retroperitoneal region are reported to be the primary sites in these individuals.[3] There are no documented reports of this lesion in adults from Nigeria or anywhere else in Africa. It has been suggested that the behavior of this disease may be different in older patients than in young children with a longer course, ultimately resulting in a poor outcome regardless of the stage.[4,5]

## Case Report

A 38-year old civil servant came as a referral to our urology clinic on the 21st of December 2007 with a six month history of right flank dull non-radiating pain which was worse on walking and relieved by rest. Three months later, the abdominal swelling became more generalized and tense. He had an intravenous pyelogram which showed bilateral hydronephrosis with a possible retroperitoneal malignancy. Neither family history nor social history was significant. On examination, a grossly distended abdomen was found tense and non-tender. Tumor resection revealed a grade III NB. The abdominal mass measured 34x28 cm and was hard to touch. His hemoglobin was 88 g/L and leukocyte count 7.3x10$^9$/L. Chest X-ray was normal and HIV status was negative for HIV1 and 11. Abdominal ultrasound showed normal liver, both kidneys were covered by a mass 85x65 cm with little ascites. Urinary meta-epinephrine was mildly elevated. Figures 1, 2 and 3 show the morphological description and appearance of the lesion, cut surface and microscopy. A diagnosis of an abdominal invasive neurofibroma was made whose origin may be either from the adrenal or from the sympathetic ganglia. In an adult, this is a hitherto unreported lesion not only in Nigeria but perhaps in the whole of Africa. Hence the importance of this report is to highlight the possibility that neuroblastoma, though a childhood malignancy, can present as an abdominal mass in an adult. The patient was placed on combination chemotherapy using vincristine, adriamycin and cyclophosphamide. He made a dramatic recovery but later absconded only to resurface three months later with what looked like a recurrence. He was again counseled and a repeat course of the same cytotoxic drugs was given with disappearance of the masses. He has remained compliant and relatively stable since then.

## Discussion

Neuroblastoma is essentially a tumor presenting in childhood, usually occurring before the age of four.[2] There are only a few isolated reports of its occurrence in adults.[1,3,4]

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The diagnosis in this case report was easy; however, no diagnosis could be made from the gross and clinical presentation until the histological tests had been performed. These showed sheets of small round blue proliferating cells forming circular or rosette like struc-

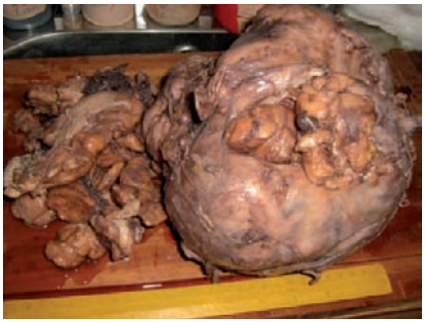

**Figure 1. Shows partly encapsulated grayish white lobulated seemingly encapsulated hemorrhagic lesion altogether measuring 34x28 cm. Firm in consistency and superficially resembling the brain.**

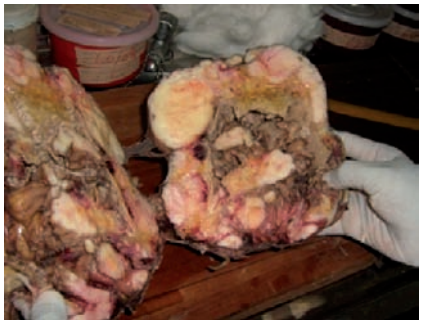

**Figure 2. Cut surface shows a cystic and solid lesion with several areas of necrosis. The lesion remains grayish white in color.**

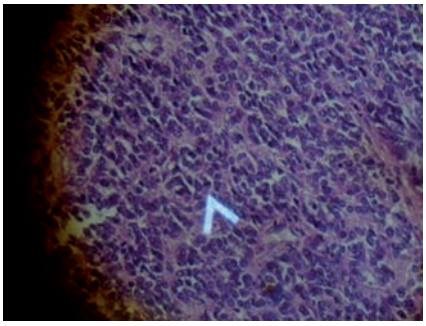

**Figure 3. Shows the microscopic description. Sections show sheets of small round blue cells proliferating and forming circular or rosette like structures with a central arranged neurofibrils directed inwards (Homer Wrights rosettes). These are disposed in a background of a scanty myoxid stroma. The lesion shows a brisk mitosis per high-power field POINTER (magnification x 40).**

tures with central arranged neurofibrils directed inwards (Homer Wrights rosettes). These are disposed in a background of a scanty myoxid stroma. The lesion also showed a brisk mitosis per high-power field. In case of doubt and in more experienced centers, routine immunohistochemistry tests would also be performed for S-100, neurone specific enolase, neurogranin and bombesin to further establish the diagnosis and differentiate them from other lesions like rhabdomyosarcoma and lymphomas. Electron microscopy may also be useful in centers where it is available and will show dendritic processes, dense core granules and desmosomes.

The successful diagnosis of NB for those in Africa, where ancillary investigative processes are not well established, requires awareness and a high index of suspicion so as to see this lesion as a possible differential of a malignant abdominal lesion in an adult. Furthermore, a revision of the treatment protocol of the lesion needs to be put in place globally as in the case of the same lesion occurring in children, as the disease indeed appears to differ in its biological behavior. This patient is still under observation and has just concluded another radiological survey to ascertain the extent of the disease.

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