

## *Burkitt's Lymphoma and Guillain-Barré Syndrome: a possible relationship?*

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### Summary

**Bademosi, O. and Ajayi, O. O. (1979).** *Nigerian Journal of Paediatrics*, 6 (1), 15. **Burkitt's Lymphoma and Guillain-Barre Syndrome: A possible Relationship?** Burkitt's Lymphoma (BL) appears to have peculiar geographical distribution and clinico-pathological features. The protean clinical presentation poses diagnostic pitfalls. We report a case of a Nigerian child who presented initially with cranial neuropathy, meningism and cerebrospinal fluid (CSF) findings of cyto-albuminic dissociation consistent with Guillain-Barré syndrome (GBS), who, 8 months later, developed a jaw tumour due to BL. This association suggests similar pathogenesis with a common initiating factor.

THE clinico-pathological features and the poor prognosis of central nervous system involvement in Burkitt's lymphoma (BL) has been well documented (Burkitt and O'Connor, 1961; Clifford *et al.*, 1967; Frank, 1968; Zeigler *et al.*, 1970; Zeigler, 1972; Odeku, Adelaye and Osuntokun, 1973). The commoner presenting features are cranial neuropathy and paraplegia. Although abnormalities of the cerebro-spinal fluid (CSF) do not invariably accompany clinical involvement of the neural axis, it is rare for patients with features of meningism, encephalopathy and raised intracranial pressure not to have tumour cells in the CSF (Zeigler *et al.*, 1970; Zeigler, 1972; Odeku, Adelaye and Osuntokun, 1973). Neuro-

logical deficits attributable to BL are easily recognisable when associated with facial or extra-facial tumour masses whether the patients are on chemotherapy or in remission. We report here a case in which cranial neuropathy, raised intracranial pressure, and CSF findings, which are consistent with a Guillain-Barré syndrome (GBS), antedated the appearance of BL of the jaw and clinical diagnosis by 8 months.

### Case Report

A nine-year old Nigerian schoolboy presented with a one week history of febrile illness and severe, persistent, generalised, pulsatile headache worsened by change of posture, straining and coughing associated with blurred vision and retro-orbital discomfort. His symptoms were accompanied by troublesome non-projectile vomiting and neck pain with stiffness.

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Clinical examination revealed an ill-looking, anaemic boy with bilateral complete ophthalmoplegia, a left infranuclear facial palsy, neck stiffness and positive Kernig's sign. The motor system revealed global muscle weakness worse proximally especially in the lower limbs. There was no associated sensory deficit. No tumour mass was palpable, and there was no ascites.

The CSF showed a white cell count under 5 per cu. mm., normal sugar content of 50mg. 100 ml but elevated protein of 100 mg%. No Burkitt's lymphoma cells were found on repeated and detailed phase cytology and tissue cultures. The cranial neuropathy, muscle weakness, and signs of raised intracranial pressure subsided on no specific therapy. He was discharged after five weeks with persistently raised CSF protein.

He presented with an increasing jaw mass eight months later, by which time there was a median mandibular tumour and massive ascites. The clinical diagnosis of Burkitt's lymphoma was confirmed histologically from biopsy of the jaw tumour. The tumour and ascites regressed completely on two courses of intravenous cyclophosphamide (Endoxan) at 15 mg/kilo/day for five days. He was still on remission ten months after the last course of Endoxan. Unfortunately he was subsequently lost to follow-up.

### Discussion

Burkitt's lymphoma (BL) continues to attract considerable interest on account of its distinctive geographical distribution, clinico-pathological characteristics, tumour cell kinetics, immunobiology and remarkable chemosensitivity (O'Connor, 1961; Cooper, Hughes and Topping, 1966; Iversen *et al.*, 1972; Zeigler, Magrath and Bluming, 1976). The clinical presentation in this case was consistent with GBS, a relatively common neurological problem in this environment (Osuntokun and Agbebi, 1973). The diagnosis of BL could not be sustained at the onset in this patient because of the absence of tumour cells in the CSF. The clinical evidence of meningism, cranial neuropathy and raised intracranial

pressure in patients with BL suggest diffuse involvement of the meninges and are commonly associated with the presence of tumour in the CSF (Zeigler, 1972).

The temporal relationship between the neurological features and BL jaw tumour in this patient raises some speculations. The neurological presentation in this patient may be unrelated to the lymphoma but simply reflect the clinical course of GBS (Henson and Ulrich, 1972; Osuntokun and Agbebi, 1973). On the other hand, the initial GBS-type presentation followed by BL in this patient might suggest a possible parallel in the aetio-pathogenesis of the two diseases. The pathogenesis of GBS is now accepted as a cell-mediated auto-immune process probably with a viral initiator (Henson and Ulrich, 1972). The aetio-pathogenesis of BL is still uncertain but several factors including malarial endemicity, viral infections and host-immune factors have been suggested (Ngu, 1967; Wright, Bell and Williams, 1967; Anderson, 1973; Le Quesne, 1973).

The prognosis of BL is poor in patients developing CNS involvement and its treatment remains a therapeutic challenge (Zeigler *et al.*, 1970; Klein, 1971). It is suggested that in environments where BL is endemic, children who present with bizarre neurological syndromes for which no aetiological factor is identified should be under close surveillance for subsequent manifestation of the disease.

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