Clinical Oncology: Case Reports

Case Report

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# Central Nervous System Burkitt Lymphoma Presenting as a typical Guillain-Barre Syndrome

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**Abstract**

A previously healthy 25-year-old man presented with a 3-weeks history of frontal headache, right-sided ptosis and binocular horizontal diplopia. The diagnosis of right 3rd nerve palsy was made. Magnetic Resonance Imaging/angiography (MRI/A) of the brain was interpreted as normal. Two days later, right facial droop and weakness developed along with lower back pain, paresthesias of both legs and left leg weakness. On exam, he had bilateral upper lid ptosis, bilateral adduction deficits and areflexia of the left patella with bilaterally decreased ankle reflexes. It was concluded that he now has bilateral partial pupil-sparing left 3rd nerve palsies and right peripheral 7th nerve palsy.

**Keywords**

Central nervous system; Guillain-barre syndrome; Burkitt lymphoma

## Introduction

MRI of the brain and spine demonstrated enhancement and thickening of cauda equina nerve roots, enhancement within the right internal auditory canal with nodularity of the right facial nerve and enhancement in left Meckel's cave (Figure 1). Two high volume lumbar punctures were then performed 1 week apart revealing very elevated protein levels (3.3 and 5.5 g/L, normal up to 0.45 g/L), positive oligoclonal banding and pleocytosis (100 cell/um, 98% lymphocytes). Flow cytometry, lymphoma panel and serum protein electrophoresis were negative on both Cerebrospinal Fluid (CSF) samples. Extensive infectious and inflammatory workup of serum and CSF which included hepatitis B and C serologies, HIV antibodies, Lyme titers, Polymerase Chain Reaction (PCR) for mycobacterium, syphilis testing, Anti-Nuclear Antibodies, Antineutrophilic Cytoplasmic Antibodies, complement component 3 and 4, rheumatoid factor, and Angiotensin-Converting Enzyme levels were all negative. A provisional diagnosis of atypical Guillain-Barre syndrome (Miller Fisher variant) was made based on the clinical and radiological features and intravenous immunoglobulin was administered with minimal symptomatic improvement. Methylprednisolone was then administered intravenously (1 gram for 5 days) which lead to significant improvement in the lower back pain and resolution of the paresthesias and leg weakness. Two months after treatment with methylprednisolone commenced all symptoms had resolved.

Nine months after the initial presentation, new progressive right-sided weakness developed. MRI of the brain and spine demonstrated a new large enhancing lesion in the left centrum semiovale with a surrounding halo of restricted diffusion (Figure 2). No cauda equina enhancement was seen and previously seen enhancement in the right internal auditory canal and left Meckel's cave resolved. Four expert neuro-radiologists debated the diagnosis of Balo's concentric sclerosis versus Central Nervous System (CNS) lymphoma. Three additional high volume lumbar punctures 5 days apart were performed. Cytology of all CSF samples was negative for malignancy and flow cytometry was negative for monoclonality. Computed Tomography (CT) imaging of the thorax, abdomen, and pelvis was performed and was normal.

## Results

Treatment with 50 mg of oral prednisone was initiated, however, while on treatment, new left facial nerve palsy developed. MRI of the brain and spine was repeated and demonstrated the left frontal mass to be larger in size, and now with the enhancement of the facial nerve and auditory canal. A decision was made to proceed with biopsy of the lesion in the left centrum semiovale. The biopsy confirmed the final pathological diagnosis of Burkitt CNS lymphoma (Figure 3).

The patient underwent multiple rounds of chemotherapy (cyclophosphamide, vincristine, and doxorubicin, high-dose methotrexate alternating with ifosfamide, etoposide, and high-dose cytarabine) and radiation over the next 6 months. Two years after the initial presentation he remained in complete remission clinically and radiologically.

## Discussion

Burkitt CNS lymphoma is extremely rare with a recent literature review citing only 36 reported cases worldwide. While the incidence of primary CNS lymphoma constitutes only about 7% of all newly diagnosed central nervous system tumors, primary CNS Burkitt lymphoma constitutes only a fraction (3%-5%) of these cases. These statistics speak to the rarity of this disease [1].

Most patients present with intraparenchymal brain involvement of the cerebral hemispheres and rarely with the involvement of the pituitary gland, cerebellum, or brainstem. Only a few cases with primary spinal or epidural involvement have been described [2,3].

Our case is unique in that it demonstrated several extra parenchymal manifestations of Burkitt lymphoma: enhancing lesions of the facial nerves within the internal auditory canal, Meckel's cave, and spinal nerve roots. Another striking feature was glucocorticoid responsiveness with complete resolution of symptoms for nine months before relapsing. Glucocorticoids are known to be potent inducers of apoptosis in lymphoid cells and in primary CNS lymphoma, the benefits of glucocorticoids are not only rapid lysis of neoplastic cells but also reduction in cerebral edema. In the literature, there are reports of long-term remission of primary CNS lymphoma following administration of glucocorticoids alone lasting from 6-60 months [4]. In many of these cases of long-term remission, glucocorticoids were administered prior to biopsy or tumor resection, the diagnosis was obscured and missed and eventually confirmed when the tumor recurred upon withdrawal of the glucocorticoids. Thus any patient suspected of having primary CNS lymphoma should not be treated with glucocorticoids prior to biopsy. Osmotic agents should

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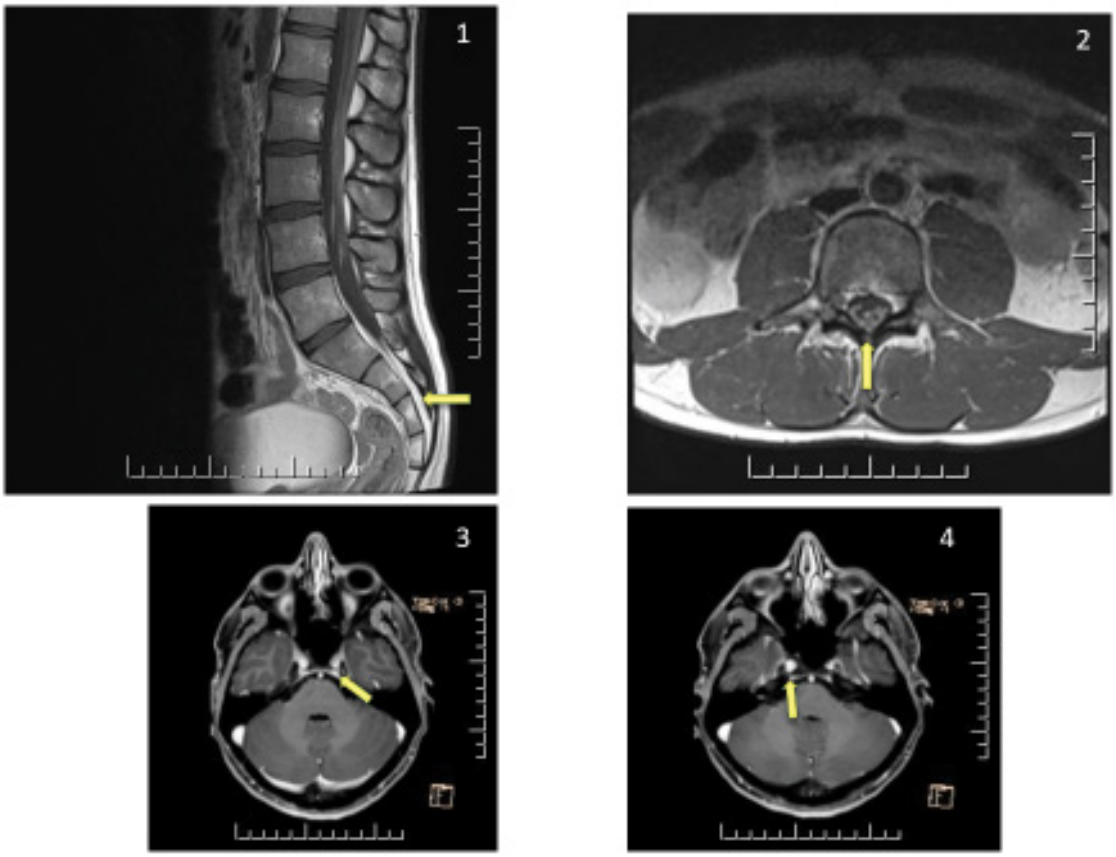


**Figure 1:** MRI of the brain and spine demonstrating enhancement and thickening of cauda equina nerve roots (1,2) enhancement in left Meckel's cave (3) and enhancement within the right internal auditory canal with nodularity of the right facial nerve (4).

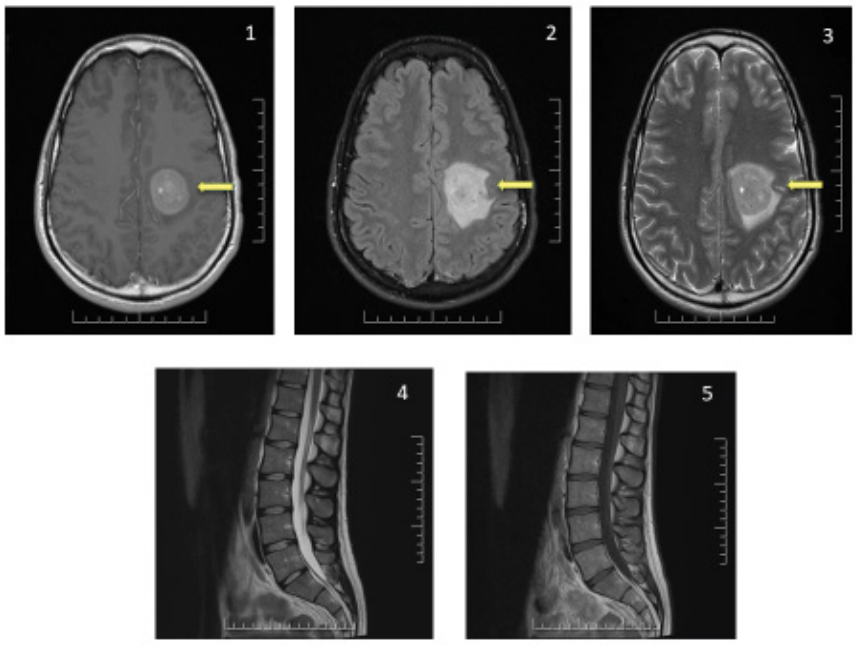


**Figure 2:** MRI of the brain and spine demonstrating a large enhancing lesion in the left centrum semiovale (1,2,3). No cauda equina enhancement was seen and previously seen enhancement in the right internal auditory canal and left Meckel's cave resolved (4,5).

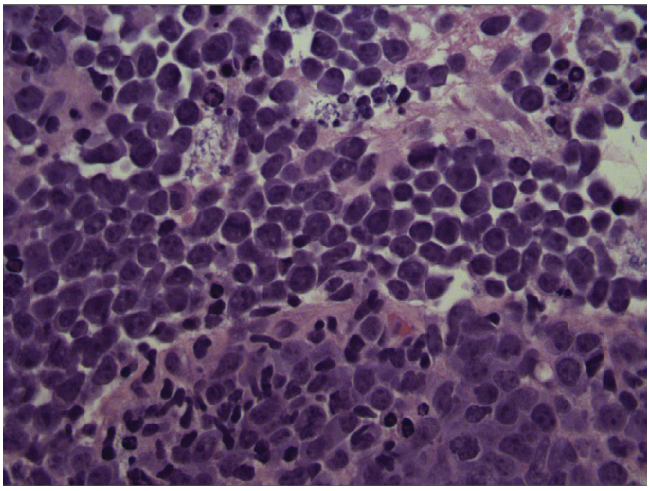

**Figure 3:** Pathological appearance of Burkitt CNS lymphoma.

be considered instead of control of increased intracranial pressure. In our case, glucocorticoids were used as the diagnosis of atypical Guillain-Barre syndrome was initially suspected based on imaging and clinical findings.

While it is generally accepted that the yield of three consecutive large volume lumbar punctures in the diagnosis of leptomeningeal metastasis is around 95%, in our case all 5 high volume lumbar punctures were negative for malignancy. Serial CSF samples may be necessary to make the diagnosis due to the low number of recognizable malignant cells found in the CSF of primary CNS lymphoma patients. More recently, PCR examination of the CSF has become an important adjunct to cytology [5].

## Conclusion

In summary, we have described a unique case of a young man with primary Burkitt CNS lymphoma initially presenting with clinical and radiological symptoms of atypical Guillain-Barre syndrome. Five high volume lumbar punctures had normal cytological examinations and high dose intravenous glucocorticoids led to complete resolution of symptoms for 9 months. This case emphasizes the importance of maintaining a high index of suspicion in all cases of atypical Guillian-Barre syndrome, the fact that glucocorticoids can suppress clinical manifestations of CNS lymphoma for a very long time, and that cytological examination of multiple high volume CSF samples can be negative in CNS lymphoma.

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