SHORT REPORT

Unusual presentation of a primary spinal Burkitt’s lymphoma

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Abstract

Primary CNS lymphomas are detected with increasing frequency in immunocompetent and immunodeficient persons. Primary involvement of the spinal roots has only rarely been reported. The unusual history is described of a patient with a primary spinal Burkitt’s lymphoma initially presenting as an S1 syndrome showing lymphocytic pleocytosis in the CSF, leading to the misdiagnosis of meningo-radiculitis. Repeated spinal MRI disclosed a spinal mass lesion and histological and immunohistological examination of the tumour confirmed the diagnosis of spinal Burkitt’s lymphoma.

Keywords: primary spinal lymphoma, meningo-radiculitis

Primary CNS lymphomas account for about 5% of primary brain tumours and their incidence in immunocompetent and immunodeficient patients has rapidly increased in the recent years. They usually present as multifocal brain tumours with focal signs according to

Figure 1  (A) First lumbar MRI was normal. (B) Six weeks later MRI showed an epidural mass lesion (arrowheads). The white line within this lesion corresponds to the puncture of the 4th spinal tap (arrow).
their site, such as headache and seizures, and visual disturbance in patients with ocular involvement. Primary spinal lymphomas—a subgroup of primary CNS lymphomas with isolated spinal manifestation—are very rare. They are thought to originate in the epidural space and examination of the CSF usually shows a normal cell count and increased protein. We describe a patient with primary spinal lymphoma initially presenting as an S1-syndrome, in whom a meningoradiculitis—for example, from neuroborreliosis—was considered because of lymphocytic pleocytosis in the CSF. However, repeated MRI disclosed development of an epidural mass lesion and finally histology led to the diagnosis of primary spinal lymphoma of Burkitt’s type.

**Case report**

A 43 year old white woman was admitted in October 1997 with lumboischialgia which had suddenly started 4 days previously. She had previously been healthy. She complained about slight hypaesthesia of the right leg and severe radicular pain both corresponding to the right S1 root. Neurological examination showed a positive Lasègue sign and diminished right ankle reflex but was otherwise normal. A herniated lumbar disc affecting the S1 root on the right side was suspected. Lumbar CT, however, was normal. On subsequent days the patient complained about itching and hypaesthesia in the dermatomes Th10 to L1 on the right and developed slight paresis of flexor muscles of the right foot. Lumbar MRI was normal (fig 1 A), but examination of the CSF showed 150 cells/µl with a heterogenous population of cells with small lymphocytes, some activated lymphocytes, and monocytes. Oligoclonal bands were negative and protein, glucose, and lactate in the CSF were normal. Despite normal CSF protein, neuroborreliosis with meningoradiculitis was suspected and treatment with intravenous ceftriaxon started. The patient did not improve and antibodies against *Borrelia burgdorferi* were negative in five consecutive serum samples as well as in CSF. A second lumbar tap showed 200 cells/µl. These consisted mainly of lymphocytes of which 10% showed atypical features (fig 2 A). These cells were immunopositive for the B cell markers CD19 and CD20 and for IgM (fig 2 B). The normal appearing lymphocytes consisted of a mixed population of T cells positive for CD3, CD4, and CD8. The CSF cytology was compatible with reactive meningoradiculitis but meningeal lymphoma could not be excluded. Therefore an extensive investigation

![Image A](https://www.jnnp.com)

![Image B](https://www.jnnp.com)

![Image C](https://www.jnnp.com)

![Image D](https://www.jnnp.com)

**Figure 2.** (A) Routine cytology of CSF from the second spinal tap showing two atypical cells (arrows) (Giemsa-May-Grünwald, originally×3400). (B) These cells were positive for IgM (R1/69, Dako) (APPAP, originally×200). (C) Light microscopy of the tumour showing cohesive medium sized blasts with numerous mitotic figures and interspersed macrophages (Giemsa, originally×540). (D) Immunohistochemistry showing an intense and homogeneous expression of B cell antigen CD20 (L26, Dako, Hamburg, Germany) on the tumour cells (ABC method, originally×270)
including CT of the chest and abdomen, MRI of the head, bone marrow examination, gynaecological and ophthalmological evaluation (including slit lamp examination) was performed without any pathological findings. Erythrocyte sedimentation rate, a complete blood count with differential, thrombin time and partial thromboplastin time, electrolytes, glucose, creatinine, liver enzymes, LDH, autoimmune screening, and tumour markers CEA, CA 19–9, CA 15–3 were normal, and serological studies for Listeria, Mycoplasma pneumoniae, Toxoplasma, Treponema pallidum, Leptospira, Legionella, hepatitis, neurotropic viruses including HSV, varicella zoster, EBV, CMV, and HIV, and microbiological examination of the CSF were negative. After the second spinal tap oral steroids were started. The patient improved and a third spinal tap in November 1997 showed only 100 cells/µl. The patient was discharged and returned for reassessment 3 weeks later. She reported exacerbation of pain and hypesthesia and complained about gait problems. In neurological examination the abdominal reflexes were absent, the paresis of the right leg was increased, and all reflexes of the right leg were missing. The patient reported hypesthesia in the dermatomes Th10 to L1 and S1 on the right. In a fourth spinal tap 90 cells/µl were found containing about 30% atypical large cells as seen before. Spinal MRI was repeated and, 6 weeks after the first spinal MRI, an epidural mass lesion at the L2-L3 level was found (fig 1B). The cauda equina seemed to be clumped but the paraspinal area was free of tumour on axial images. Surgery was performed and the epidural tumour at L2-L3 could be excised. The dura was indurated and seemed to be infiltrated by the tumour as well as the roots of the cauda equina. No intradural mass lesion was found. Histological and immunohistological examination of the tumour was done.

HISTOLOGY
Histological examination disclosed a lymphoid lesion, which was diagnosed as Burkitt’s lymphoma. The tumour consisted of a single population of medium sized blast-like cells with basophilic cytoplasm and round nuclei with few centrally placed prominent nucleoli. Many mitotic figures were seen. The tumour cells formed cohesive masses infiltrating adipose tissue. Intermingled with the tumour cells were numerous tangible body macrophages, giving the tumour the typical starry sky appearance (fig 2 C). Immunohistologically the tumour cells demonstrated an intense and homogeneous expression of B cell antigen CD20 (L26, Dako, Hamburg, Germany) (fig 2 D). Only a few CD3 positive T lymphocytes were seen scattered between the tumour cells, preferentially at the border (anti-CD3, Dako). No Epstein-Barr virus antigen was detected immunohistochemically (LMP, EBNA2, Dako).

FOLLOW UP
After surgery the patient was treated by local radiation and polychemotherapy (intrathecal with methotrexate and systemic with cyclophosphamide, vincristin, methotrexate, ifosfamide, adriamycin, and dexamethasone). During therapy she developed severe paraparesis and became unable to walk. Thereafter, she recovered slowly. Two years after initial presentation she has not relapsed and is able to walk for about 300 m despite a moderate paraparesis.

Discussion
We present a case of a primary spinal lymphoma (Burkitt’s type) in a formerly healthy immunocompetent white woman. At first a herniated disc was suspected, because of typical history with acute radicular lumboischialgia with signs of an S1 syndrome. However, this diagnosis was easily ruled out by negative lumbar CT and MRI. Herniated lumbar disc has been described as a differential diagnosis to spinal lymphoma.4 When spinal tap showed lymphocytic pleocytosis we had to deal with the differential diagnosis of meningoradiculitis (table 1). The main cause of meningoradiculitis, especially in spring and autumn, is acute neuroborreliosis. The CSF in neuroborreliosis typically shows lymphocytic pleocytosis, increased protein, and intrathecally produced IgG or IgM antibodies against Borrelia burgdorferi.5 Lymphocytic pleocytosis is indicated by a heterogeneous population of lymphocytes with many plasma cells and lymphoblastic-like cells, often with atypical features which sometimes mimic meningeal lymphoma. Therefore in every case when neuroborreliosis is suspected meningeal lymphoma is an important differential diagnosis.6 8 ImmunocytoLOGY is helpful in this situation, because usually meningeal lymphoma consists of a monoclonal population of B cells.9 10 However, in our patient the situation was more complicated. The initial CSF showed pleocytosis with a heterogeneous population of small lymphocytes and activated cells. In the second CSF atypical B cells homogenously positive for CD20 were found, but these cells accounted for only 10%–15% of cells and otherwise a mixed lymphocytic population showing immunocytological features of reactive pleocytosis was found. Extensive staging showed no systemic or primary cerebral lymphoma and thus we decided not to start aggressive chemotherapeutic treatment and initiated only steroids. Three weeks later the correct diagnosis was made by follow up MRI: despite continuous steroid therapy, which usually leads to dramatic reduction of cerebral lymphomas,1 a rapidly growing lumbar epidural tumour which was classified histologically as primary spinal lymphoma of Burkitt’s type was found.
More than 90% of primary CNS lymphomas are histologically classified as high grade B cell lymphomas and when subtypes are differentiated about 5%–15% of these are of Burkitt’s type. By contrast, in the small series of epidural primary spinal lymphomas reported by Lyon et al none were classified as Burkitt’s lymphoma. By comparison with this series where CSF cell count was always normal another unusual aspect is that in our patient atypical cells were found in the CSF before a solid tumour became manifest. So leptomeningeal lymphoma was followed by an epidural lymphoma. An entity of primary leptomeningeal lymphoma has been described, but those patients never developed lymphomatous mass lesions, neither spinal nor cerebral. Thus our patient might show a new subgroup of primary spinal lymphomas with the combination of leptomeningeal and epidural manifestation. However, it cannot be ruled out that our patient had primary leptomeningeal lymphoma and that leptomeningial tumour cells secondarily invaded the epidural space using rents due to the three prior lumbar taps. The question arises whether all primary spinal lymphomas originate in the epidural space as has been proposed or if a subgroup of primary spinal lymphomas, as in the patient presented here, originate from tumour cells grown in the CSF which invade the dura, which seems to be infiltrated in our patient, and migrate to the epidural space where they finally form solid tumours.

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