

# Central nervous system primary lymphoma of the cerebellum

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## ABSTRACT

**Introduction and importance:** A rare cranial malignant hematological malignancy is Primary Central Nervous System Lymphoma (PCNSL). Compared to other encephalic regions, PCNSL in the cerebellum is less frequent. Cerebellar PCNSL has a variety of imaging presentations, making a diagnosis rather challenging. The purpose of this case series study is to examine the impact of surgery on cerebellar PCNSL and whether surgery might be utilized to confirm the diagnosis.

**Methods:** We present three examples of cerebellar PCNSL treated with general anaesthesia by neuronavigation microsurgery. The procedure was carried out by authors. Due to postoperative obstructive hydrocephalus, one patient received left lateral ventricular drainage on the fourth and tenth

days after the procedure. Following histology confirmation, all patients received chemotherapy or radiation treatment.

**Results:** The malignancies in every patient were entirely eliminated. One patient underwent surgery; experienced perioperative obstructive hydrocephalus twice, was treated with drainage, and eventually left the hospital after making a full recovery. The other two patients made a full recovery and were released without incident. Nine months after the surgery, one patient passed away, but the other two patients lived.

Three patients' prognoses were influenced by the size of the tumour and prompt follow-up chemo-radiation therapy. All patients' histologies revealed diffuse large B-cell lymphoma (GCB phenotype). Patients with suspected cerebellar PCNSL should have surgery to make the diagnosis, and then get radiotherapy and chemotherapy.

**Keywords:** Primary central nervous system lymphoma; Cerebellum; Neuronavigation; Tumour

## INTRODUCTION

Primary Central Nervous System Lymphoma (PCNSL), a malignant primary central nervous system illness that accounts for 2.4%-3% of all brain tumours and around 3% of Non-Hodgkin Lymphomas (NHL), is uncommon and aggressive. The majority of PCNSL cases are diffuse large B-cell lymphomas histologically; as a result, this tumour is highly aggressive and has a bad prognosis. Although it can enter the parenchyma, PCNSL is restricted to the central nervous system. The most frequent location for lesions is the brain hemispheres, and the majority of these are found in the deep frontal lobe. You can also get within the callosum. However, only about 9% of all PCNSLs are found in the cerebellar area, making it a rare site of PCNSL discovery. PCNSL has no distinct symptoms and is clinically readily misdiagnosed. Primary Central Nervous System Lymphoma (PCNSL), a malignant primary central nervous system illness that accounts for 2.4%-3% of all brain tumours and around 3% of Non-Hodgkin Lymphomas (NHL), is uncommon and aggressive. The majority of PCNSL cases are diffuse large B-cell lymphomas histologically; as a result, this tumour is highly aggressive and has a bad prognosis. Although it can enter the parenchyma, PCNSL is restricted to the central nervous system. The most frequent location for lesions is the brain hemispheres, and the majority of these are found in the deep frontal lobe. You can also get within the callosum. However, only about 9% of all PCNSLs are found in the cerebellar area, making it a rare site of PCNSL discovery. PCNSL has no distinct symptoms and is clinically readily misdiagnosed.

## LITERATURE REVIEW

A 22-year-old junior high school student complained of headache and dizziness for a month, which became worse and were accompanied by nausea and vomiting for a week. He had no history of surgery, medication, or any pertinent illnesses. He had smoked 20 cigarettes per day for the previous 7 years, occasionally drank, never used drugs for fun, and had no allergies or family history. He worked in the transport industry. Mixed signals in the bilateral cerebellum were detected using Magnetic Resonance Imaging (MRI), primarily on the right side. Compression of the fourth

ventricle and brain stem was seen. A 22-year-old junior high school student complained of headache and dizziness for a month, which became worse and were accompanied by nausea and vomiting for a week. He had no history of surgery, medication, or any pertinent illnesses. He had smoked 20 cigarettes per day for the previous 7 years, occasionally drank, never used drugs for fun, and had no allergies or family history. He worked in the transport industry. Mixed signals in the bilateral cerebellum were detected using Magnetic Resonance Imaging (MRI), primarily on the right side. Compression of the fourth ventricle and brain stem was seen. The patient's recovery went without any problems. Three weeks following surgery, the patient began eight rounds of chemotherapy. The prescribed regimen was vincristine (VCR), 1.8 mg, 1.2 mg/m<sup>2</sup>, day 1, cyclophosphamide (CTX), 1.2 mg, 0.8 mg/m<sup>2</sup>, day 1, and prednisone (PDN, 100 mg, po, and day 1-5, q3w). Radiotherapy was not administered to the patient. Unfortunately, this therapeutic approach failed to reduce PCNSL, and the tumour spread to other parts of the brain. Within 9 months, this patient passed away.

A junior high school student who is 26 years old and unemployed presented with dizziness and an unsteady gait for more than ten days. He had no history of surgery, medication, or any pertinent illnesses. He consumed alcohol infrequently, smoked 20 cigarettes per day for 10 years, had no prior history of using recreational drugs, and had no allergies or family history. A strong signal was found in the right cerebellum using enhanced MRI. Damage to the cerebellar nerve system was present in the patient. Right cerebellar tumour removal under general anaesthesia with a surgical microscope by neuronavigation was carried out to restore cerebellar neurological function after the preoperative evaluation was completed and no contraindications were discovered.

Complete excision of the tumour was carried out during the 497-minute procedure under a microscope. Due to postoperative obstructive hydrocephalus, the patient had left lateral ventricular drainage on the fourth and tenth days after the procedure. This tumour has non-Hodgkin B-cell lymphoma as its histology (DLBCL, diffuse large B-cell lymphoma, GCB phenotype). According to molecular analysis, this PCNSL displayed the following characteristics: CK, CD3, CD20 (+), Ki-67 (95 %+), CD10 (+), CD79a (++) , C-myc (30%+), BCL-6 (+), and P53 (+). Due to intolerance, the

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patient underwent one round of chemotherapy after surgery that included methotrexate and cytarabine but no radiation. The patient has lived and is currently doing well overall.

A 54-year-old junior in college came in with an occipital sore that had been present for a week. He didn't smoke, drink, or use drugs recreationally in the past, and he didn't have any allergies or personal medical history either. He observed the surroundings. In both cerebellar hemispheres, MRI revealed many nodular signals with low T<sub>1</sub> and marginally high T<sub>2</sub>. An annular hyperintensity was visible in the FLAIR sequence, and the enhanced scan clearly demonstrated enhancement. We performed a right cerebellar tumour resection under general anaesthesia using a surgical microscope by neuronavigation to examine its histology and direct subsequent therapy after the preoperative assessment was completed and no contraindications were discovered. Complete removal of the tumour was carried out during the 291 minutes procedure under a microscope. The non-Hodgkin B-cell lymphoma was the histology (diffuse large B-cell lymphoma, GCB phenotype). After that, the patient underwent six rounds of radiotherapy with no chemotherapy. This patient is currently in good health and has not yet had tumour progression.

### DISCUSSION

The age of disease beginning in our instances was quite young, with an average incidence age of 34 years across the three patients. In two cases, the patients were under 30 years of age. The surgery was carried out by authors 3 and 5, who are both skilled academics and surgeons. After surgery, a histopathological diagnosis was made, and all cases were determined to be diffuse large B-cell lymphomas, which was consistent with the histology of the majority of PCNSLs. One patient, who underwent surgery and chemotherapy, passed away nine months after their diagnosis (CHOP protocol, 8 rounds). The two additional patients are still surviving. One patient underwent surgery and six rounds of radiotherapy, while the other underwent surgery and chemotherapy (only once). These three patients' varying survival times may be related to the size of the tumour. Immunodeficiency, including acquired or congenital immunodeficiency, is the most frequent aetiology of PCNSL since the patients' leukocytes and lymphocytes were normal prior to presentation, the instances we described here did not, however, and have a clear immunodeficiency. With about 3% of all cranial tumours being PCNSL, it is a very uncommon malignant tumour. Every year, about 1600 patients in America are impacted by PCNSL. Patients who are older than 65 appear to experience it more frequently. With ages of 22, 26, and 54, the three patients in this case were all reasonably young about 80% of PCNSL cases occur in the supratentorial region, which includes the lobe, callosum, and basal ganglia. Compared to supratentorial PCNSLs, cerebellar and spinal PCNSLs are less common, with cerebellar PCNSLs making up only about 9% of all instances. The pan-B-cell markers CD19, CD20, CD22, and CD79a are expressed by diffuse big B cells, which make up the majority of PCNSLs and are found in almost 95% of patients. PCNSL is difficult to identify due to the variety of symptoms that might be shown on a CT or MR scan. To diagnose this condition, tissues from biopsy or resection-obtained lesions must be examined using cytology and histology. Additionally, radiotherapy, chemotherapy, and surgery are the key components of the PCNSL

therapeutic strategy. Since PCNSL is extremely susceptible to radiation, patients should receive radiation therapy after a diagnosis. Additionally, some chemotherapeutic drugs such as methotrexate, procarbazine, cytarabine, and pharomubicin are toxic to PCNSL. Surgery is typically not the first option for PCNSL unless the tumor's mass effect is so significant that it endangers the patient's life. Following a diagnosis, radiation and chemotherapy are frequently used as treatment options. However, the prognosis for PCNSL is still dismal even with prompt therapeutic intervention. More of these examples are being gathered at this point to inform clinical care.

### CONCLUSION

Particularly when the cerebellum is the location of the lesion, PCNSL is an uncommon form of malignant cranial tumour. Patients with suspected cerebellar PCNSL should have surgery to make the diagnosis, then get radiotherapy and chemotherapy.

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