

Case Report

Primary Middle Cerebellar Peduncle Lymphoma with Primary Cervical Lymphoma: Repeated Misdiagnosis

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The coexistence of primary intracranial and extracranial lymphomas is relatively rare. In this study, we report a misdiagnosed case of primary middle cerebellar peduncle lymphoma (PMCP) with primary cervical lymphoma. This case illustrates that hearing loss may be the only manifestation of PMCP, which can be easily misdiagnosed as sudden deafness or acoustic neuroma. Patients with PMCP may also have primary extracranial lymphoma, which should not be misdiagnosed as metastatic tumor.

KEYWORDS: Lymphoma, acoustic neuroma, central nervous system, hearing loss, misdiagnosis

INTRODUCTION

Primary intracranial lymphoma (PIL) is a rare type of primary central nervous system lymphoma (PCNSL) that accounts for less than 1% of the various types of lymphoma.¹ Pathology is the golden standard for PIL diagnosis. In this case, the patient presented with sudden hearing loss only; however, 2 tumors were located in the right middle cerebellar peduncle and left neck, respectively. Pathology confirmed the diagnosis of primary middle cerebellar peduncle lymphoma (PMCP) with left cervical lymphoma, which we had misdiagnosed twice as sudden deafness and acoustic neuroma. Hence, in this report we describe the diagnosis and the differential diagnosis of this disease.

CASE PRESENTATION

A 49-year-old man suffered from sudden hearing loss in his right ear, with no obvious symptoms of intracranial nerve injury. Pure tone audiometry showed sensorineural hearing loss (Figure 1A); the results of laboratory studies and neuro-examinations were normal. With the diagnosis of “sudden deafness (right ear),” the patient accepted medical treatment. Three days later, repeat pure tone audiometry showed no significant improvement in his right ear (Figure 1B). Magnetic resonance imaging (MRI) of the brain showed an obscure shadow of a long T2 signal in the right middle cerebellar peduncle (Figure 2A). Without any obvious signs of central nervous system damage and obvious imaging features, we did not pay enough attention to it.

Forty days later, the patient accepted reexamination in the outpatient department. Complete blood count (CBC) showed a decreased lymphocyte percentage; however, the reexamination of MRI showed a 2.6 cm × 2.4 cm × 2.3 cm tumor with a long T1 signal and a long T2 signal in the right middle cerebellar peduncle (Figure 2B). Since the patient had hearing loss only and no other symptoms of intracranial nerve injury or intracranial hypertension, we considered it as acoustic neuroma. A small nodule (0.3 cm × 0.5 cm) was also found on the patient’s left neck, without pain or ulcer, which we considered as normal lymph nodes. Because the tumor was solitary and localized, the patient underwent craniotomy in the absence of stereotactic biopsy techniques. Surprisingly, the post-operative pathology reports upheld the diagnosis of PMCP: Diffuse large B-cell lymphoma (DLBCL) originates from the non-germinal center of B-cells (Figure 3A). A cervical nodule was diagnosed with DLBCL by biopsy from the germinal center of B-cells (Figure 3B). The patient received chemotherapy postoperatively. So far, there are no signs of recurrence of the tumor in the brain and neck during follow-up (Figure 1B).

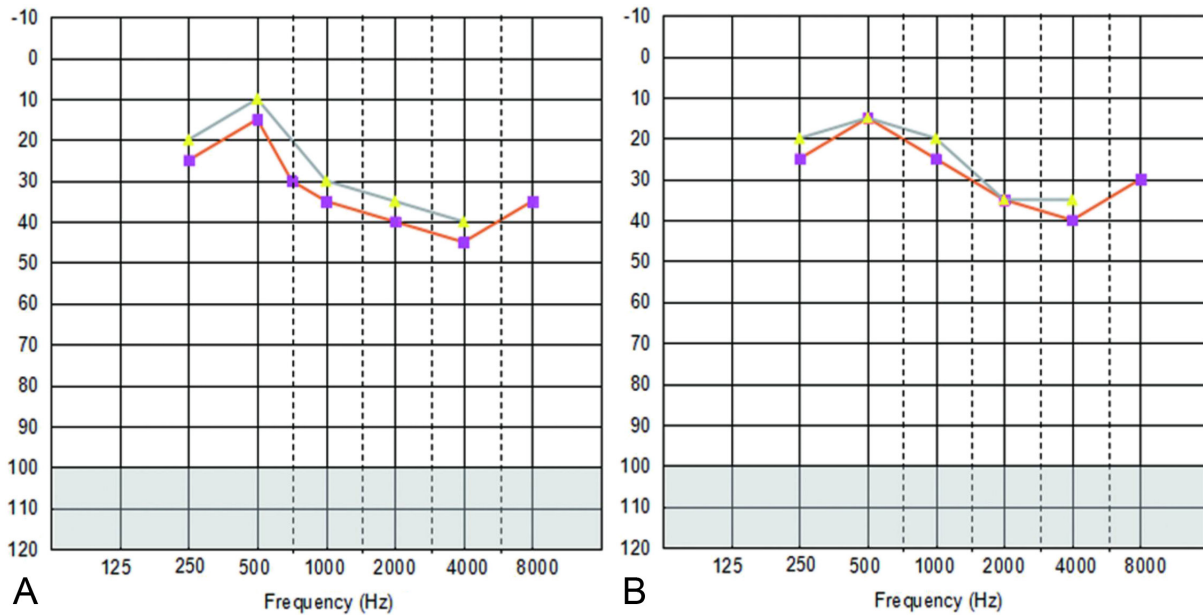


Figure 1. (A) Before treatment, pure tone audiometry showed sensorineural deafness. (B) The reexamination of pure tone audiometry showed no significant improvement in hearing.

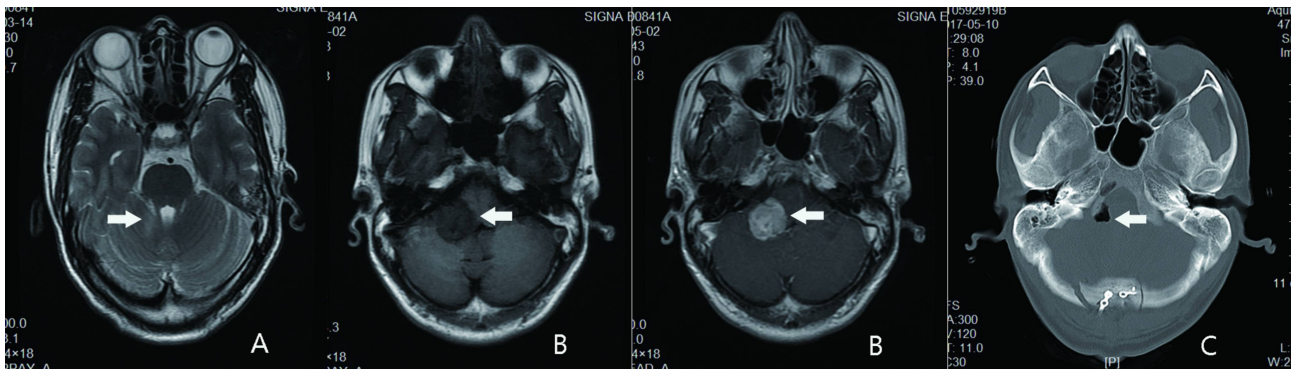


Figure 2. (A) There was a shadow of a long T2 signal in the right middle cerebellar peduncle with blurred boundary (white arrow). (B) There was a nodular shadow of a long T1 signal (white arrow) and a long T2 signal in the right middle cerebellar peduncle, accompanied with a ring edema zone over the periphery (white arrow). (C) Computed tomography (CT) scans of the brain revealed no recurrence of lymphoma (white arrow).

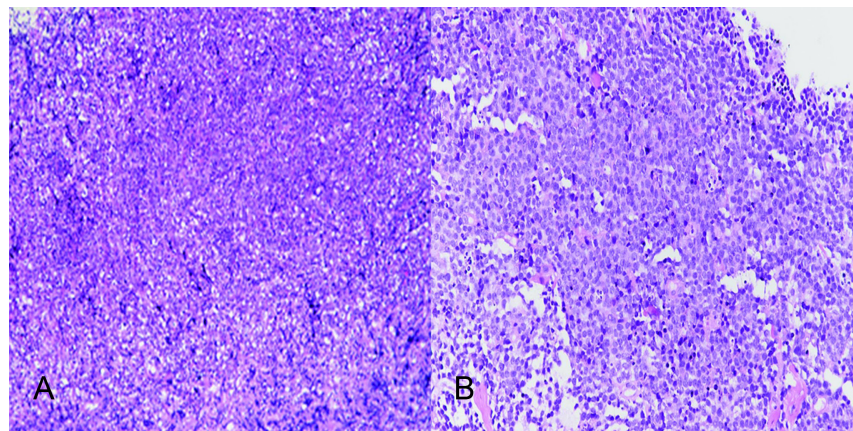


Figure 3. (A) Diffuse-type lymphoid cells with multilobulated nuclei, proliferated blood vessels. Lymph vessels and blood vessels connect to form a sleeve of hemocytes. IHC: CD3 (-), CD20 (+), CD5 s (-), CD10 (-), Bcl6 (+), MUM (+) - 1, CyclinD1 (-), Ki -67 (+ 60%), and CD117 (-), OCT3/4 (-), CD30 (-), AE1/AE3 (-). ISH: EBER (-), EBER comparison (+). (B) The meso-large lymphoid cells were distributed in a flake arrangement, and combined with immunohistochemistry, they were consistent with non-Hodgkin's lymphoma and large cell B-cell lymphoma. IHC: CD20 (+), PAX-5 (+), CD10(+), Bcl-6 (+), Bcl-2 (+), C_myc (+), Vim (f+), CD3 (+), CK (-), Mum-1 (-), Tdt (-), MPO (-), CD30 (-), ALK (-), CD21 (+), CD5 (-), CD23 (-), CyclinD1 (-), ki-67 (+,90%). ISH: EBER (-), EBER comparison (+).

DISCUSSION

Primary intracranial lymphoma is more common in the supratentorial location and is rarely presented in the middle cerebellar peduncle. Diffused large B-cell lymphoma is the most common histological type of PIL. Some studies suggest that owing to the absence of endogenous accumulation of lymphoid tissue and lymphatic circulation in the central nervous system, lymphoma invades the brain tissues through an immune escape mechanism, thus resulting in intracranial lymphoma.^{2,3} The studies confirm that the germinal center of B-cells is the main origin of PIL. In this case, post-operative pathology indicated that the origin of PMCPL was from the non-germinal center of B-cells, whereas the origin of left cervical lymphoma was from the germinal center of B-cells.

Synchronous multiple primary malignancies are defined as 2 or more primary tumors, each of which is diagnosed at an interval of less than 6 months apart.⁴ We can document the presence of a common clonal B-cell origin at the initial diagnosis and at recurrence to confirm relapse; conversely, different clonality may suggest the emergence of a new neoplasm.⁵ In this case, 2 tumors were detected at short intervals, and the centers were different, and so we speculated that PMCPL and cervical lymphoma coexist, i.e., 2 clonal diseases have occurred simultaneously. As far as we know, this phenomenon has not been reported previously in the literature, which should be verified with more clinical trials and molecular analysis in the future.

Primary intracranial lymphoma is a rare type of intracranial tumor with poor prognosis, which is easily misdiagnosed in the clinical field. Patients with PMCPL generally experience symptoms of increased intracranial pressure and intracranial nerve injury. The sole symptom in this case was hearing loss, and since the MRI feature was atypical, the condition was misdiagnosed as sudden deafness at an early stage. A tumor on the middle cerebellar peduncle was found during reexamination, but upon considering that the patient had the symptom of hearing loss only with a single lesion in the middle cerebellar peduncle, the condition was misdiagnosed as acoustic neuroma. At the same time, a contralateral neck nodule with morphology similar to that of the lymph nodes was found. Considering that acoustic neuroma had no possibility of extracranial metastasis, we mistakenly thought that it was a normal lymph node.

Repeated misdiagnosis was confirmed by pathology; however, with careful analysis, we were able to make a differential diagnosis. First, both PMCPL and acoustic neuroma can manifest as hearing loss; however, PMCPL is a malignant tumor with rapid development, whereas acoustic neuroma is a benign tumor with slow development. Second, MRI of both tumors can present long T1 and long T2 with peripheral enhancement^{6,7}; however, acoustic neuroma is often located in the cerebellopontine angle, with an enlarged inner auditory canal. In this case, primary middle cerebellar peduncle lymphoma was located in the middle cerebellar peduncle, and there was no enlargement of the inner auditory canal. Meanwhile, PMCPL can also manifest as “incision,” “angular,” “fist,” or “hard ring” signs in some cases.⁸ In a patient with a small acoustic neuroma, the internal auditory canal can be of

normal size, but we can identify the tumor with an enhanced MRI of the internal auditory canal. Last, lymphomas may be associated with metastasis, but acoustic neuroma is almost non-metastatic. In this case, it is also emphasized that even if a definite diagnosis for PMCPL has been made, when an extracranial lesion is found, especially in the neck, the lesion cannot be simply considered as an extracranial metastasis; it is better to perform a general PET-CT examination or biopsy on the suspected lesion to confirm the diagnosis.

CONCLUSION

To our knowledge, this is the first case of PMCPL with hearing loss as the sole presentation and simultaneously present contralateral primary cervical lymphoma. Since primary middle cerebellar peduncle lymphoma was manifested as hearing loss only, its diagnosis was missed, or it was easily misdiagnosed as sudden deafness or acoustic neuroma. Extracranial lesions should not be considered as metastasis of PMCPL only. The possibility of primary extracranial lymphoma should be excluded.

Informed Consent: Informed consent was obtained from the patient who participated in this case report.

Peer Review: Externally peer-reviewed.

Author Contributions: Concept – C.Z.; Analysis and/or Interpretation – C.L.; Literature Search – C.Z.; Writing – C.Z.; Critical Reviews – C.L.

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