# LETTER TO THE EDITOR

## To the Editor

### Diffuse Large B-Cell Lymphoma Presenting as Bilateral Internal Auditory Canal Lesions

**Keywords:** Lymphoma, internal auditory canal

Bilateral internal auditory canal (IAC) tumors are rare and usually represent vestibular schwannomas (VS) in the context of neurofibromatosis type 2. Approximately 5% to 10 % of all IAC tumors are non-schwannomatous, but clinical differentiation from VS is challenging.[1]

Diffuse large B-cell lymphoma (DLBCL) can arise in virtually any tissue, with extranodal disease occurring in approximately 40% of cases. Central nervous system (CNS) involvement with DLBCL is rare, usually seen in relapsed disease following treatment and has a poor prognosis.[2] Here we report a rare case of DLBCL with initial presentation as bilateral IAC lesions with excellent response to treatment.

A 47-year-old, previously healthy male was referred to neurology for subacute unilateral hearing loss. He presented with a 1-week history of disequilibrium and progressive left-sided hearing loss. This was not associated with headache, tinnitus, or vertigo. Two days before our assessment, he noted weakness of the right side of his face.

On examination, vital signs were stable. Cranial nerve examination revealed normal extraocular movements and visual fields. Funduscopy was unremarkable. He had bilateral mild lower motor neuron facial palsies, more prominent on the right. Audiometry showed complete sensorineural hearing loss on the left. Motor and somatosensory examinations were unremarkable. Finger-to-nose and heel-to-shin tests were normal. During assessment of his gait, he had difficulties performing tandem gait. Systemic examination was unremarkable.

Routine blood work including blood cell count, chemistry, lactate dehydrogenase, creatinine, and liver enzymes were within normal limits. Inflammatory and autoimmune markers including ESR, CRP, ENA, ANA, c-ANCA, and p-ANCA were negative. MRI of the brain showed bilateral IAC and cerebellopontine angle masses, the right measuring 14 mm and left measuring 16 mm in length (Figure 1A-C). The images were reported as bilateral cerebellopontine angle schwannomas. Lumbar puncture obtaining cerebrospinal fluid (CSF) showed elevated nucleated cells at $28 \times 10^6$/L with 81% lymphoid predominance. Flow cytometry revealed monoclonal B-cell population representing approximately 3% of total leukocytes with cytology showing highly cellular aspirate containing a few atypical lymphocytes. Bone marrow biopsy results were not suggestive of a B-cell lymphoproliferative neoplasm. Computed tomography (CT) scan of the chest, abdomen, and pelvis demonstrated a round soft-tissue mass, measuring

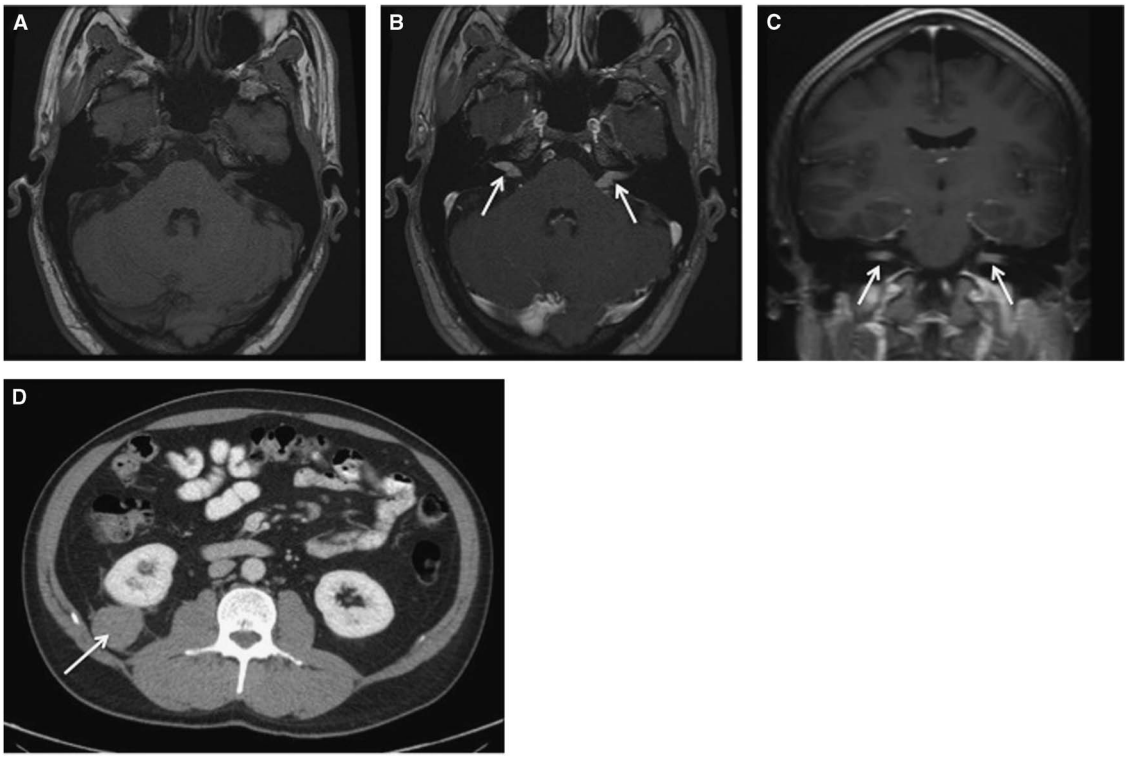


***Figure 1:*** *MRI of the brain demonstrating bilateral IAC and cerebellopontine angle masses, contrast-enhancing (arrows), the right measuring 14 mm and left measuring 16 mm in length. (A) Axial FMPSPGR (fast multiplanar spoiled gradient echo) sequence, before contrast injection. (B) Axial FMPSPGR sequence, post-contrast injection. (C) Coronal T1 sequence, post-contrast injection. (D) CT of the abdomen demonstrating a round soft-tissue mass, measuring 3.5 × 3.8 cm (arrow), abutting the right kidney.*



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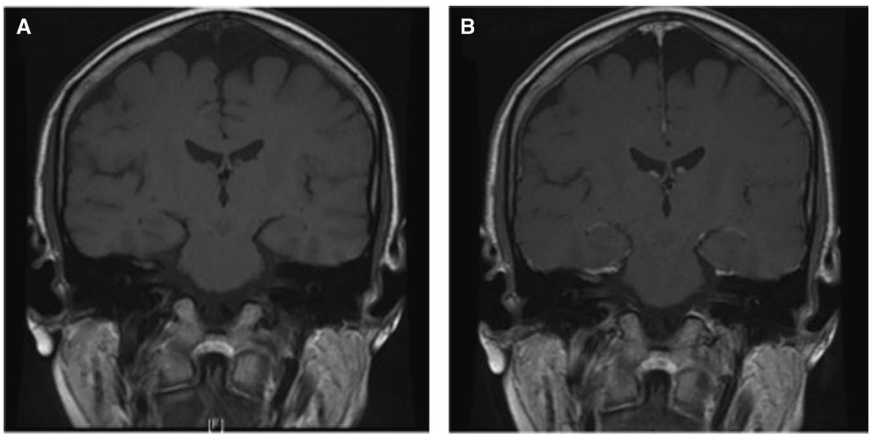


***Figure 2:*** *MRI of the brain following chemotherapy treatment, 4 months post diagnosis, demonstrating resolution of previously contrast-enhancing bilateral IAC lesions seen in Figure 1. (A) Coronal T1 sequence, pre-contrast injection. (B) Coronal T1 sequence, post-contrast injection.*

3.5 × 3.8 cm, abutting the right kidney (Figure 1D). CT-guided biopsy of the mass was performed with pathology showing DLBCL.

The patient was treated with six cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, methotrexate, and prednisone (R-CHOMP) chemotherapy with intrathecal methotrexate. At 4 months, midway through chemotherapy treatment, CT of the abdomen and pelvis showed marked improvement with only focal soft-tissue thickening remaining in the right perirenal space. MRI of the brain performed at the same time showed compete resolution of enhancement of bilateral IAC lesions (Figure 2). At 6 months, after completion of chemotherapy, a positron emission tomography scan was performed, showing no evidence of hypermetabolic lesions to suggest ongoing disease. The patient continues to show no signs of clinical recurrence at 2 years post-diagnosis. His hearing in the left ear improved to moderate loss (55-95 dB) on repeat audiology testing.

Tumors of the IAC account for approximately 10% of all intracranial neoplasms, the most common being VS occurring in greater than 90% of cases. Less common neoplasms include meningioma, lipoma, and metastases including lymphoma. Clinical symptoms of IAC lesions are not specific to the type of tumor and usually include sensorineural hearing loss, tinnitus, and vertigo. Dazert et al[1] reported a cohort of 351 cases with lesions isolated to the IAC that underwent surgery, 15 of which had a non-schwannomatous histological diagnosis. They found that preoperative differentiation of VS from other tumors based on initial clinical presentation and radiologic imaging was not reliable. Within their cohort, only one patient had clinical evidence of facial nerve involvement and was diagnosed with a facial neuroma. Symptoms of facial nerve compromise, which were also seen in our patient may be suggestive of an etiology other than VS.[1]

Bilateral IAC lesions are rare and thought to be pathognomonic of neurofibromatosis type 2. The occurrence of bilateral non-schwannomatous IAC tumors is limited to individual case reports. These include primary CNS lymphoma and myeloma,[3] lipoma,[4] and metastases from lung[5] and melanoma.[6] There have been two reported cases of DLBCL relapsing with bilateral IAC metastases.[1,7]

DLBCL is the most common subtype of non-Hodgkin lymphoma. It presents with a rapidly enlarging symptomatic mass, usually a nodal growth in the neck or abdomen, but can be anywhere in the body. Associated systemic "B" symptoms of fever, weight loss, and night sweats are found in 30% of patients. CNS involvement occurs in approximately 5% of patients and is usually seen with relapsed disease at a median duration of less than 1 year from time of diagnosis. Only 1% of patients with DLBCL have CNS involvement at the time of diagnosis. CNS relapse with DLBCL carries a very poor prognosis with median survival of less than 6 months. Literature suggests that this may be a result of some patients already having subclinical CNS disease with involvement of CSF and leptomeninges before development of neurologic symptoms. Conventional CSF cytology reveals malignant lymphoid cells in only 40% of patients with suspected CNS dissemination, making it difficult to detect these patients early.[2,8]

To our knowledge, this is the first case of DLBCL with an initial presentation of cranial nerve VII and VIII involvement representing bilateral IAC lesions mimicking bilateral schwannomas. Our patient had an excellent response to R-CHOMP chemotherapy with intrathecal methotrexate with no recurrence of disease in 2 years. This may be due to rapid diagnosis from symptom onset and prompt initiation of treatment.

## Disclosures

The authors do not have anything to disclose.

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## References

1. Dazert S, Aletsee C, Brors D, et al. Rare tumors of the internal auditory canal. Eur Arch Otorhinolaryngol. 2005;262:550-4.
2. Fletcher CD, Kahl BS. Central nervous system involvement in diffuse large B-cell lymphoma: an analysis of risks and prevention strategies in the post-rituximab era. Leuk Lymph. 2014;55:2228-40.
3. Lenarz M, Durisin M, Becker H, Lenarz T, Nejadkazem M. Primary central nervous system lymphoma presenting as bilateral tumors of the internal auditory canal. Skull Base. 2007;17:409-12.
4. Ventura E, Ormitti F, Crisi G, Sanna M, Bacciu A. Bilateral cerebellopontine angle lipomas. Auris Nasus Larynx. 2012;39:103-6.
5. Hariharan S1, Zhu J, Nadkarni MA, Donahue JE. Metastatic lung cancer in the cerebellopontine angles mimicking bilateral acoustic neuroma. J Clin Neurosci. 2005;12:184-6.
6. Jacob A, Brightman RP, Welling DB. Bilateral cerebellopontine angle metastatic melanoma: a case report. Ear Nose Throat J. 2007;86:388-90.
7. Blioskas S, Tsaligopoulos M, Kyriafinis G, et al. Bilateral secondary neurolymphomatosis of the internal auditory canal nerves: a case report. Am J Otol. 2013;34:556-8.
8. Ghose A, Kundu R, Latif T. Prophylactic CNS directed therapy in systemic diffuse large B cell lymphoma. Crit Rev Oncol Hematol. 2014;91:292-303.



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