

Intralabyrinthine Presentation of CNS Lymphoma with Progressive Cerebellopontine Angle Extension

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CASE PRESENTATION

59 y/o presented with sudden right-sided hearing loss after a viral URTI. He had resolved labyrinthitis last year.

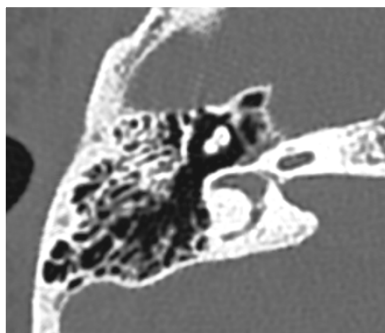
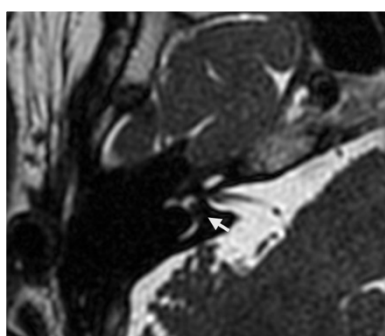
Otосcopy and tympanometry were normal. Audiometry showed mixed hearing loss on the right.

IAM MRI was suggestive of labyrinthitis ossificans thought to be sequela of previous labyrinthitis with symptoms exacerbated by recent URTI.

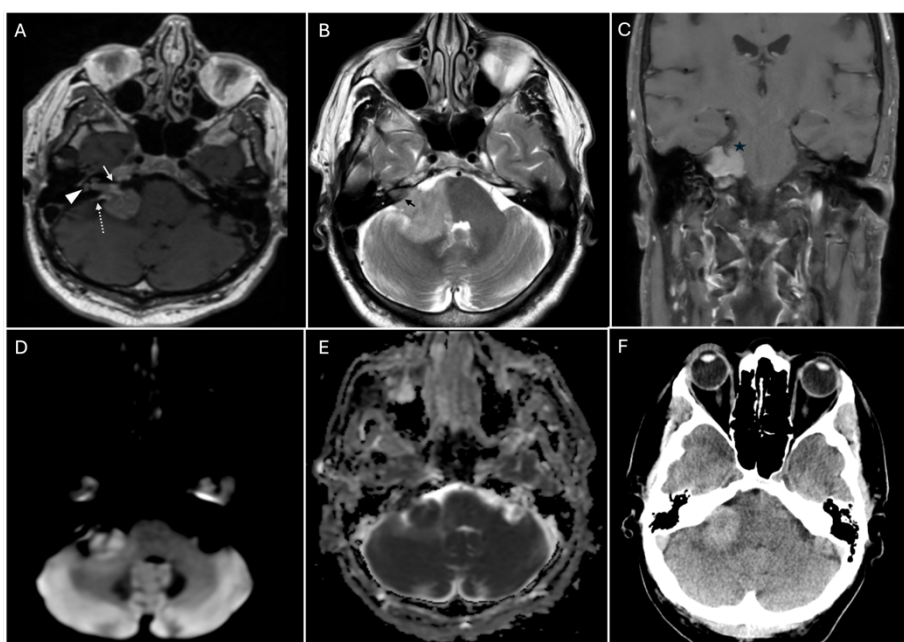
F/U CT showed no correlate ossification, suggesting early fibrous stage of labyrinthitis ossificans. Repeat imaging was planned to assess progression.

Two months later, the patient developed right-sided facial palsy, worsening hearing, ataxia, and ageusia. Repeat imaging showed progression to an enhancing CPA mass. CT chest-abdomen-pelvis excluded systemic malignancy.

Trans-labyrinthine resection was planned. Due to atypical features, intraoperative frozen section was performed and suggested lymphoma. Resection was halted. Definitive histopathology confirmed diffuse large B-cell lymphoma (DLBCL).



CISS demonstrating filling defect and loss of the high T2 signal within the right-sided vestibule (white arrow) and parts of the semicircular canals. CT did not demonstrate any correlate ossification or high-density.



Lesion is avidly and homogeneously enhancing. It occupies the right IAC and extends into the vestibule (white arrow head) and the posterior semicircular canal. There is subtle hyperenhancement of the labyrinthine segment of the facial nerve (solid white arrow) and along the right petrous dura (dashed white arrow). Lesion restricts diffusion, ADC around $0.53 \times 10^{-3} \text{ mm}^2/\text{s}$. It abuts and compresses the right middle cerebellar peduncle, right superior cerebellar hemisphere and the right lateral pons where there is associated T2 hyperintensity. Some of this enhances (black star), likely to represent infiltration. There is also the impression of subtle erosion/blunting of the porus acousticus internus when compared to the left. No haemorrhage or cystic degeneration. Lesion is CT hyperdense.

RED FLAGS

Rapid Progression

VS typically causes gradual hearing loss over months to years, growing $\sim 1 \text{ mm/year}$ [9]. Our patient had sudden hearing loss and rapid tumour growth, consistent with DLBCL's fast doubling time of weeks [10]. Even high-grade meningiomas rarely progress this rapidly.

Parenchymal Infiltration

T2/FLAIR hyperintensity in adjacent brainstem and cerebellum extended beyond expected mass effect. Some areas enhanced, suggesting direct infiltration.

Early Multiple Cranial Nerve (CN) Involvement

Facial palsy developed just eight weeks after hearing loss. Schwannomas rarely involve CN VII early, whereas lymphomas with leptomeningeal spread can affect multiple cranial nerves simultaneously.

Atypical Imaging features

The ADC was lower than expected for a schwannoma but not as low as typical PCNSL range ($0.7 - 0.9 \times 10^{-3} \text{ mm}^2/\text{s}$) (Table 1) [1]. For a VS of such large size, more heterogeneous enhancement due to cystic change and marked IAC widening would be expected. Bony erosion and CT hyperdensity favour lymphoma.

Atypical clinical and imaging features suggesting more aggressive aetiology to benign vestibular schwannoma