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Vestibular schwannoma causing normal pressure hydrocephalus

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ABSTRACT

Vestibular schwannoma is a common benign tumour that may cause local complications. However, vestibular schwannoma has a known association with communicating hydrocephalus presenting with symptoms of normal pressure hydrocephalus and requiring treatment by ventricular shunting or tumour resection. We report a 79-year-old woman who presented with subacute gait apraxia, cognitive impairment and urinary incontinence. CT and MR imaging identified a 20 mm vestibular schwannoma and communicating hydrocephalus; her cerebrospinal fluid (CSF) protein was elevated. Her symptoms improved following ventriculoperitoneal shunt insertion. The mechanism by which non-obstructing vestibular schwannoma causes hydrocephalus is unclear, but hyperproteinorrachia is probably important, likely by impeding CSF resorption.

INTRODUCTION

Vestibular schwannoma is a benign Schwann cell-derived tumour originating from the vestibulocochlear nerve. Large vestibular schwannomas have a mass effect on neighbouring intracranial structures, particularly with fourth ventricular compression leading to obstructive hydrocephalus. This may be treated neurosurgically either by primary tumour resection or by permanent cerebrospinal fluid (CSF) diversion via ventriculoperitoneal shunt or endoscopic third ventriculostomy. Vestibular schwannomas comprise 80% of all cerebellopontine angle tumours and usually are diagnosed by gadolinium-enhanced MR brain imaging.

Vestibular schwannoma may rarely be associated with communicating hydrocephalus with no obstructive features on brain imaging,[1] but presenting with symptoms of normal pressure hydrocephalus (NPH). This condition usually presents in people aged over 60 years with the characteristic symptom triad (Hakim's triad) of gait apraxia, cognitive impairment and urinary incontinence caused by aberrant CSF dynamics causing communicating hydrocephalus.[2] MR imaging findings include ventriculomegaly (raised Evans' index), reduced callosal angle, disproportionate enlargement of the Sylvian fissures, temporal horns and periventricular white matter changes.[3] Most cases are idiopathic with no clear underlying structural cause such as a tumour or haemorrhage. The only definitive management is permanent CSF diversion, usually by ventricular shunt insertion. Initial short-term CSF diversion involves either a large volume lumbar puncture or a 72-hour lumbar drain with monitoring for symptom improvement to identify likely shunt responders. A growing body of literature is identifying communicating hydrocephalus in people with vestibular schwannoma who have associated symptoms of NPH which resolve after CSF diversion.

The mechanism of communicating hydrocephalus in people with vestibular schwannoma (without obstruction) is unclear. However, the CSF protein, often elevated in people with vestibular schwannoma, probably contributes to the development of hydrocephalus[4 5] potentially through impaired CSF resorption.

CASE REPORT

A 79-year-old right-handed white British woman presented to the Emergency Department with a 6-week history of unsteadiness, urinary incontinence and confusion. She had progressive balance impairment causing reduced mobility, recent onset urinary urgency causing incontinence and slowed thinking. She had no falls, weakness, sensory change, headache, visual or bulbar symptoms. There was a 7-year history of sensorineural hearing loss, initially left-sided but with progression to bilateral loss and of

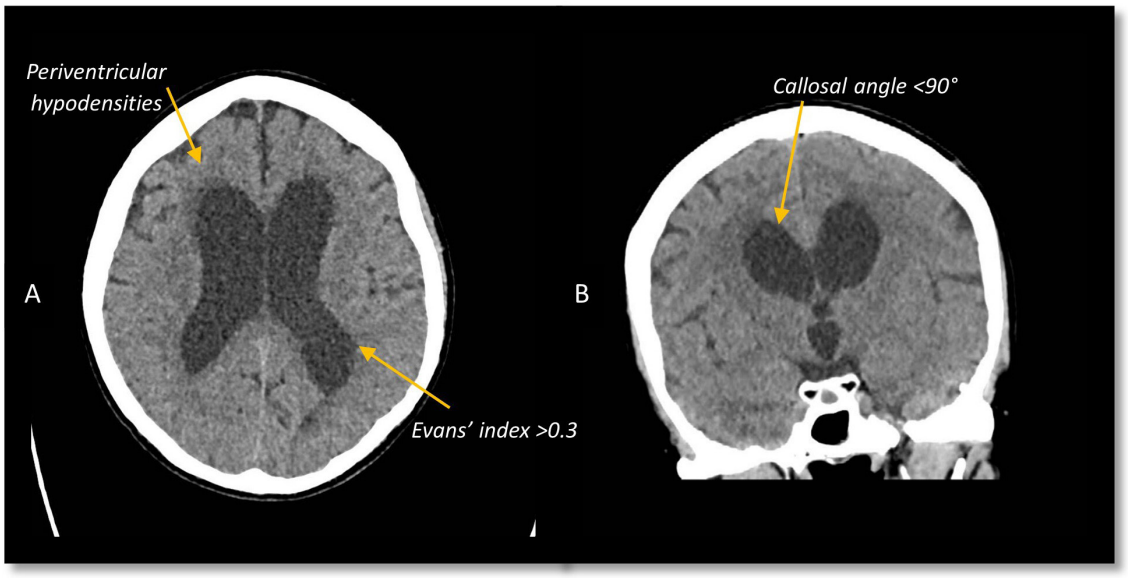

Figure 1 CT scan of head showing hydrocephalus with NPH features. NPH, normal pressure hydrocephalus.

benign paroxysmal positional vertigo. There was no history of head injury or of childhood illness such as meningitis and no family history of hydrocephalus.

On examination, she had a slow, broad-based apraxic gait with short stride length and shuffling steps. There was first-degree, gaze-evoked, persistent horizontal nystagmus bidirectionally; she had pathological hyperreflexia with finger jerks and crossed adductors. Plantars were flexor. Neurological examination was otherwise normal with normal limb tone, power, coordination and sensation.

Routine laboratory tests were normal. CT scan of the head (figure 1) showed ventriculomegaly with a raised Evans' index of 0.42. The callosal angle was reduced at 60 degrees with 5 mm diameter temporal horns and periventricular hypodensities suggesting white matter disease. These findings, along with the clinical presence of Hakim's triad, suggested possible NPH, although the short duration and rapid symptom progression seemed unusual for the idiopathic form.

Subsequent MR scan of the brain (figure 2) confirmed the imaging features of NPH with a calculated idiopathic normal pressure hydrocephalus (iNPH) Radscale score of 8. There was no disproportionate enlargement of the Sylvian fissures. The imaging also identified a cisternal and intracanalicular left vestibular schwannoma indenting the middle cerebellar peduncle and measuring 20 mm posterior to the petrous ridge. There was no ventricular system obstruction as the fourth ventricle was not compressed, indicating the hydrocephalus was communicating in nature.

We treated this as communicating hydrocephalus due to NPH given the characteristic symptoms and absence of obstruction by the schwannoma. We performed a large volume (40 mL) lumbar puncture as per protocols for NPH. The opening pressure was 15 cm H_2O and CSF protein was 1.5 g/L (normal 0.15–0.45). Neurodegenerative biomarkers were normal. Her walking speed did not significantly improve following lumbar puncture and we subsequently asked the neurosurgery team to insert a 72-hour lumbar drain. This significantly improved her mobility and walking speed indicating a possibly shunt-responsive condition, although patients with no response to tap test or lumbar drain may still improve after shunt insertion. The neurosurgeons inserted a proGAV 2.0 ventriculoperitoneal shunt under general anaesthesia. There were no postoperative complications and she was discharged home. At 10-week follow-up, her mobility had returned close to her premorbid baseline with a narrow steady gait and fast stride, while her cognition and bladder symptoms had resolved completely. Her iNPH grading scale score had improved from 7 (cognitive impairment 1, gait disturbance 3, urinary disturbance 3) to 1 (0, 1, 0).[6]

We include videos of the patient following large volume lumbar puncture and post-shunt insertion. Following large volume lumbar puncture, there was persistent gait impairment and a 10 m time of 34 s (figure 3 and online supplemental video 1). Following shunt surgery, her gait improved significantly and 10 m walking time was reduced to 10 s (figure 3 and online supplemental video 2). Gait assessments were carried out on a ProtoKinetics Zeno Walkway, and the videos include step analysis from ProtoKinetics Movement Analysis Software (figure 3 and supplementary video 3).

DISCUSSION

Vestibular schwannoma is a common benign brain tumour that can cause local complications through mass effect, notably hydrocephalus. The mechanism for obstructive (non-communicating) hydrocephalus is well understood with local compression of the fourth

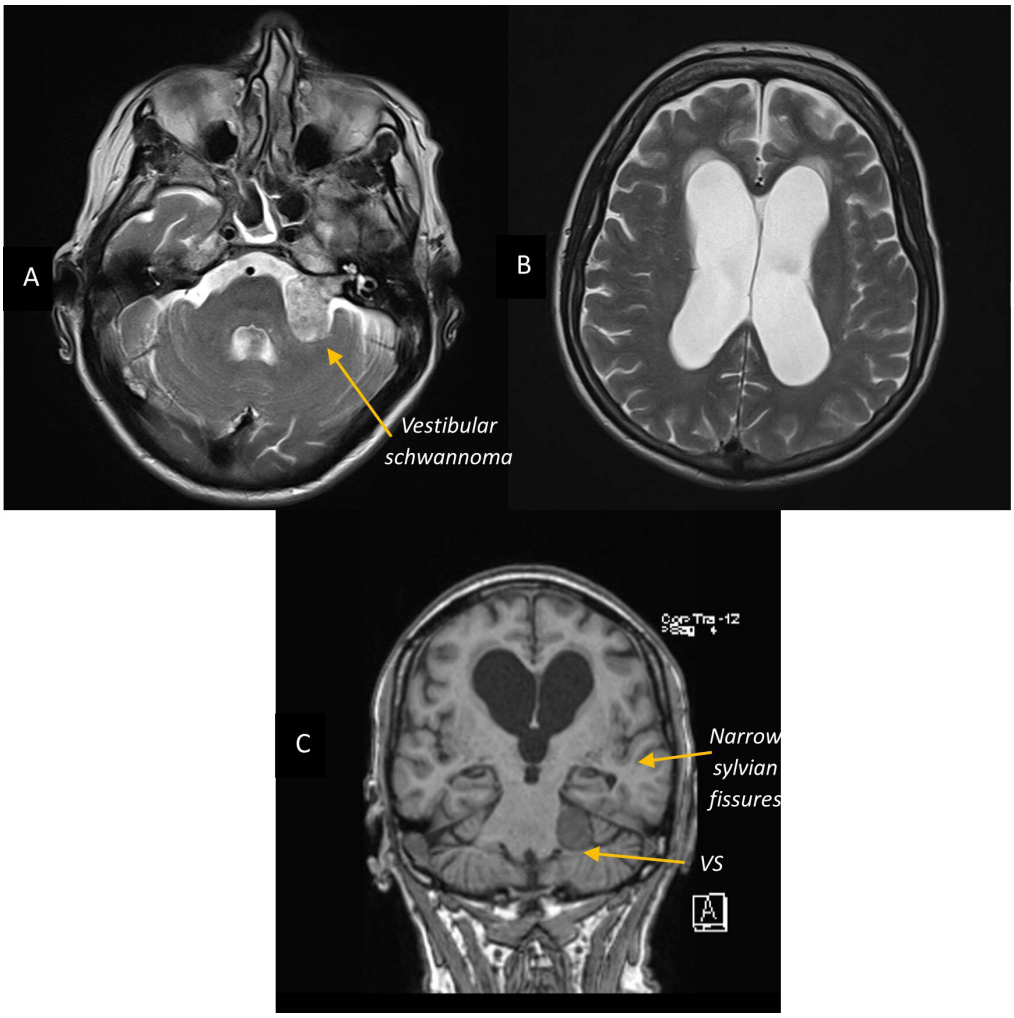

Figure 2 MR scan of brain showing a 20 mm left vestibular schwannoma on axial T2 WI (A). This also confirmed the presence of hydrocephalus and showing periventricular white matter disease (B). C showing a coronal view on T1 WI of both hydrocephalus and vestibular schwannoma.

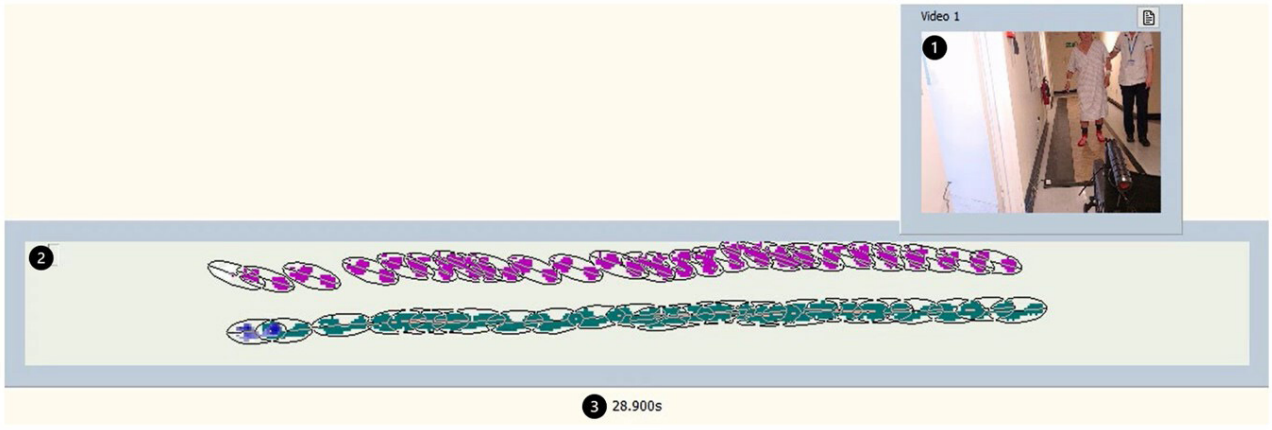

Figure 3 Video still from online supplemental video 1 showing gait analysis before shunt insertion on the Protokinetics Zeno Walkway. 1) Video recording of the patient's 10m walk on the ProtoKinetics Zeno Walkway, allowing qualitative analysis of gait. 2) Individual step data as recorded by the ProtoKinetics Zeno Walkway, showing improvement in step length, gait stability and step speed after shunt insertion. 3) Time elapsed.

ventricle impairing CSF outflow causing an increase in intracranial pressure. A less common but previously described complication is communicating hydrocephalus where hydrocephalus arises despite a lack of radiological evidence of fourth ventricle obstruction.[7] This may present in a way similar to NPH with characteristic gait apraxia, cognitive impairment and urinary incontinence that improves after ventricular shunting.

Communicating hydrocephalus associated with vestibular schwannoma may be treated with tumour resection, ventricular shunting or a combination of both. In this patient, shunting alone gave a near resolution of gait impairment. Tumour resection was not indicated as there were no threatening local complications. The presence of bidirectional nystagmus might have suggested local impairment of central brainstem pathways by the tumour, but the clear improvement following shunting demonstrates that this sign should not be regarded as a contraindication to shunting. This is consistent with previously described cases in which gait impairment either resolved or greatly improved following ventriculoperitoneal shunt insertion and without tumour resection.[4 8] Alternatively, patients presenting in this way have been successfully treated by resection of the tumour alone.[1 9] However, occasionally patients receiving gamma knife radiosurgery to treat vestibular schwannoma have subsequently developed symptomatic hydrocephalus that improved after shunt insertion.[10] While there is limited evidence for either approach, the evidence suggests that vestibular schwannoma causing communicating hydrocephalus but no other local effects may be effectively treated by ventriculoperitoneal shunt insertion alone with primary resection only indicated if there are other complications.

The mechanisms by which a small, non-obstructing vestibular schwannoma cause communicating hydrocephalus are debatable. A consistent finding in these patients is elevated CSF protein, as in this case, and this is probably a causative factor.[4] While the measured protein concentrations in these patients are not high enough to cause CSF accumulation by osmotic mechanisms alone, the hyperproteinorrachia may interfere with CSF resorption leading to altered CSF dynamics and ultimately to hydrocephalus. It was originally suggested that CSF hyperviscosity due to high protein concentration might block resorption at the Pacchionian (arachnoid) granulations, but studies have shown that the protein concentrations in these cases have little effect on CSF flow and viscosity.[11] It therefore seems more likely that the high CSF protein interferes with the passage of CSF into the venous system at the arachnoid granulations, perhaps through receptor saturation. However, while most people with vestibular schwannoma have an elevated CSF protein, only a minority develop hydrocephalus, raising the possibility that while hyperproteinorrachia is necessary for this pathology, it is not sufficient alone and other factors contribute.

The location and size of vestibular schwannoma may influence the subsequent development of communicating hydrocephalus. Of the cases described, all extended into the cerebellopontine angle with no purely intracanalicular tumours (Grade 1 on the Koos grading scale) presenting with communicating hydrocephalus. However, wholly intracanalicular tumours are by definition smaller than schwannomas extending into the cerebellopontine angle raising the possibility that size rather than location influences hydrocephalus development. Furthermore, tumours in this location have limited exposure to CSF when compared with tumours in the cerebellopontine angle which may impact on tumour protein access to CSF.[12] A univariate analysis has shown that while tumour size and CSF protein relate the presence of hydrocephalus, only protein concentration was significant on multiple logistic regression analysis.[5] Therefore, tumour size and location may significantly impact hydrocephalus development only by their impact on CSF protein concentration.

Despite mechanistic uncertainty, there is an established and growing body of documented cases with vestibular schwannoma and communicating hydrocephalus. Neurological practice should reflect this with careful assessment of gait, cognitive or urinary disturbance in people with known vestibular schwannoma or unilateral hearing loss. Patients developing a NPH-like syndrome often respond well to CSF shunting and may not require primary tumour resection.

CONCLUSIONS

Communicating hydrocephalus causing a NPH syndrome is a rare but increasingly recognised association of vestibular schwannoma. These patients should be considered for CSF shunting given the high rate of clinical improvement and the possibility of avoiding tumour resection. MR imaging should be performed in all patients presenting with Hakim's triad, both for Radscale score calculation and for detecting an underlying cause such as vestibular schwannoma.

Contributors AH drafted the original manuscript and produced videos 1 and 2. CC reviewed the manuscript from a neurological perspective. AG reviewed the manuscript from a radiological perspective and produced figures 1 and 2. MW reviewed the manuscript from a neurosurgical perspective. All authors reviewed and approved the final manuscript. CC takes overall responsibility for the manuscript.

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Competing interests AH, MW and AG have no disclosures. CC holds an MRC-CARP Grant to investigate the cause of shunt-responsiveness in normal pressure hydrocephalus (NPH), is the chair of the ABN NPH Specialist Interest Group and holds a 1% share in the HCA-Chiswick Medical Centre.

Key points

- Vestibular schwannoma may rarely be associated with communicating hydrocephalus, probably from elevated cerebrospinal fluid (CSF) protein interfering with CSF dynamics.
- Such patients may present with Hakim's triad of gait apraxia, cognitive impairment and urinary incontinence.
- Temporary CSF diversion by large volume lumbar puncture and/or lumbar drain can identify those patients most likely to improve with shunt surgery.
- Shunt surgery can lead to long-term improvement and preclude the requirement for primary resection of the schwannoma.

Patient consent for publication Consent obtained directly from patient(s).

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Data availability statement Data are available upon reasonable request. No data are available.

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