Rare disease

Middle cranial fossa cystic schwannoma

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The authors present the unusual case of a 37-year-old man, with a history of hepatitis C, presenting with a 3 year history of progressive trigeminal, facial and vestibular nerve defects. Intracranial imaging demonstrated a cystic middle and posterior fossa lesion, that was ultimately diagnosed as a cystic trigeminal schwannoma. Due to the uncertainties of diagnosis he was managed in two stages with an open biopsy and then a subsequent partial resection. Surveillance of the tumour remnant is planned. The rare nature of this diagnosis is discussed, along with the more common findings in intracranial schwannoma, and the wide differential to be considered. Briefly, the authors describe the surgical option for a complex multi-fossa schwannoma and the importance of the multi-disciplinary team in the diagnosis and management of such patients.

BACKGROUND

While trigeminal schwannoma is the second commonest intracranial schwannoma, they remain rare, and thus both the presentation and management options are imperfectly understood. A minority exhibit calcification or cystic changes, with even fewer an intracystic fluid level. This case illustrates the complex presentation, wide differential to be considered and initial surgical management of a large partly calcified, cystic middle fossa trigeminal schwannoma.

CASE PRESENTATION

A 37-year-old man with hepatitis C as a consequence of previous intravenous drug misuse, presented with a 3 year history of symptoms, of progressive hearing loss, worsening left-sided facial spasms and facial numbness in the left ophthalmic region of the trigeminal nerve. The symptoms commenced 3 years previously with progressive hearing loss, and thereafter a shorter history of hemi-facial spasm and finally facial numbness. He had a previous history of facial trauma and felt that this underpinned his symptoms and therefore he did not seek medical attention until late on. His medical history was otherwise non-contributory and his only regular medication was methadone. There was no other significant social or family history to note.

Neurological examination revealed hypoaesthesia in the distribution of the left ophthalmic branch of the trigeminal nerve with loss of corneal reflex, a House-Brackmann grade 2 left lower motor neuron facial nerve palsy, and intermittent left sided hemi-facial spasms. There were no symptoms or signs of raised intracranial pressure or vestibulopathy. He was afebrile and there were no systemic findings suggestive of concurrent infection.

INVESTIGATIONS

Routine bloods revealed a white cell count of 11.4, with a C reactive protein <5.

A formal audiogram revealed sensorineural hearing loss in the left ear, other auditory and vestibular testing was not undertaken.

MR CT and MRI of the brain revealed a large, left-sided space demanding cystic lesion in the middle cranial fossa extending posteriorly to the tentorial hiatus (figure 1).

- ▶ Axial T2 MR demonstrates a fluid level in the cystic mass which is indenting the left hemi pons and expands Meckel's cave. The rim is partly hypodense, reflecting its calcified nature.
- ▶ Bone windowed CT demonstrates that the lesion remodels and replaces the left petrous apex, expands the

Figure 1 Three different views of the cystic lesion in the middle cranial fossa

Figure 2 An intraoperative view with frontal and temporal lobes retracted, demonstrating the initial fenestration of the tumour cyst.

greater sphenoid wing and erodes the posterior margin of the supraorbital fissure, indicating its chronicity.

- Parasagittal T1 postcontrast MR reconstruction demonstrates the expansive lesion in the course of the trigeminal nerve which is partly effacing the left cerebellum.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for this lesion included temporal lobe abscess, a trigeminal or vestibular nerve schwannoma, metastatic disease, chronic inflammatory processes, arachnoid cyst or rarely haemangioblastoma.

TREATMENT

Based on the patient's progressive neurological deficits, the late presentation, and the need for histological diagnosis, urgent surgical intervention and decompression of the cystic component was deemed necessary, rather than a more conservative medical approach. The patient underwent a pterional craniotomy, fenestration and biopsy of the cyst wall and marsupialisation of the cyst after having his case reviewed and discussed in a regional multidisciplinary team (MDT) meeting. Following biopsy the patient consented to undergo surgical debulking. At surgery, the tumour was adherent to the trigeminal ganglion, the cavernous sinus and the petrous apex. A partial resection was therefore performed. Surveillance imaging with the use of stereotactic radiosurgery is planned for any tumour recurrence. Postoperatively the patient had numbness in all three divisions of the trigeminal nerve. Facial nerve function remained at House-Brackmann Grade 2. Unfortunately the patient had an ipsilateral oculomotor palsy following surgery (figure 2).

OUTCOME AND FOLLOW-UP

Formal histology demonstrated sections a tumour composed of elongate, spindle shaped cells, showing some nuclear clustering, with a fibrous background. Mitotic figures were not noted. Following immunocytochemistry the tumour was strongly positive with an antibody to S100 protein and negative with an antibody to glial fibrillar acidic protein. A fascicle of peripheral nerve was passing through

Figure 3 Histological image of tumour showing bundles of elongate cells with a fibrous background (H&E stain, x100 magnification).

the edge of the tumour and scattered collections of perivascular lymphocytes and foamy macrophages and chronic inflammatory cells, including macrophages, were dispersed throughout the lesion. The morphological appearances and immunophenotype indicate a schwannoma.

Based on the interpretation of the results from imaging and histopathology combined, a diagnosis of a trigeminal nerve schwannoma was made (figure 3).

DISCUSSION

A schwannoma is a benign tumour that originates from the neural sheath of schwann cells. Schwannoma of cranial nerves are common intracranial tumours, constituting up to 8% of intracranial neoplasms. Trigeminal schwannomas although rare, are the second commonest intracranial schwannoma, with the vast majority occurring on the vestibular nerve.^{1 2} Most patients present in the fourth decade of life, with the duration of symptoms ranging from 1 month to 13 years.

Intracranial cystic neurogenic tumours constitute an uncommon subset of tumours with a distinct clinico-biological behaviour. The presence of fluid-fluid levels within the tumours, although rare, confirms the cystic nature of the neoplasms. Barring the acoustic schwannomas, cystic cranial nerve schwannomas are exceptionally uncommon.³ Cystic components in vestibular schwannomas are associated with a less favourable surgical outcome, probably due to the rapid tumour growth and symptoms caused by compression of the posterior fossa structures.⁴

Definitive surgical management of a multi-compartment trigeminal schwannoma is controversial. Potential surgical management options for tumours involving both middle and posterior fossa include the conventional approaches (subtemporal and transsylvian approaches for the middle fossa, retrosigmoid for posterior fossa) and skull base approaches. The latter includes anterior petrosal approach advocated by Yoshida,⁵ zygomatic middle fossa approach with access to the posterior fossa component through Meckel's cave,⁶ or a presigmoid approach.⁷ Current microsurgical options allow for complete or near-total removal in greater than 70% of patients.⁸

Recurrence rates are low, between 0 and 17%, and most commonly occur at the cavernous sinus or Meckel's cave⁹ and there is a clear role for stereotactic radiosurgery (SRS), for adjunctive treatment of postsurgical tumour recurrence or residuum.¹⁰ Furthermore, some have advocated SRS as a primary treatment modality for tumours less than 15 cm.³

Learning points

- ▶ Patients presenting with neurological symptoms or signs should be offered prompt intracranial imaging.
- ▶ Trigeminal schwannoma is uncommon causes of middle fossa space demanding lesions, and are associated with multiple cranial nerve palsies.
- ▶ The differential diagnosis of cystic intracranial lesions is wide, and some (cerebral abscess) mandate urgent surgical exploration.
- ▶ Definitive diagnosis and management options should be co-ordinated via a skilled MDT team.

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REFERENCES

1. **Garen PD**, Harper CG, Teo C, *et al*. Cystic schwannoma of the trochlear nerve mimicking a brain-stem tumor. Case report. *J Neurosurg* 1987;**67**:928–30.
2. **Colreavy MP**, Lacy PD, Hughes J, *et al*. Head and neck schwannomas—a 10 year review. *J Laryngol Otol* 2000;**114**:119–24.
3. **Hasegawa T**, Kida Y, Yoshimoto M, *et al*. Trigeminal schwannomas: results of gamma knife surgery in 37 cases. *J Neurosurg* 2007;**106**:18–23.
4. **Fundová P**, Charabi S, Tos M, *et al*. Cystic vestibular schwannoma: surgical outcome. *J Laryngol Otol* 2000;**114**:935–9.
5. **Yoshida K**, Kawase T. Trigeminal neurinomas extending into multiple fossae: surgical methods and review of the literature. *J Neurosurg* 1999;**91**:202–11.
6. **Al-Mefty O**, Ayoubi S, Gaber E. Trigeminal schwannomas: removal of dumbbell-shaped tumors through the expanded Meckel cave and outcomes of cranial nerve function. *J Neurosurg* 2002;**96**:453–63.
7. **Prasad S**, Gupta S, Patankar T, *et al*. Fluid-fluid levels in intracranial schwannomas. *Australas Radiol* 1999;**43**:529–31.
8. **Ramina R**, Mattei TA, Sória MG, *et al*. Surgical management of trigeminal schwannomas. *Neurosurg Focus* 2008;**25**:E6; discussion E6.
9. **MacNally SP**, Rutherford SA, Ramsden RT, *et al*. Trigeminal schwannomas. *Br J Neurosurg* 2008;**22**:729–38.
10. **Kano H**, Niranjan A, Kondziolka D, *et al*. Stereotactic radiosurgery for trigeminal schwannoma: tumor control and functional preservation Clinical article. *J Neurosurg* 2009;**110**:553–8.

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