

## Tic Convulsif Caused by Cerebellopontine Angle Schwannoma

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A case is presented of painful tic convulsif caused by schwannoma in the cerebellopontine angle (CPA), with right trigeminal neuralgia and ipsilateral hemifacial spasm. Magnetic resonance images showed a 4 cm round mass displacing the 4th ventricle and distorting the brain stem in the right CPA. The schwannoma, which compressed the fifth and seventh cranial nerves directly, was subtotally removed by a suboccipital craniectomy. Postoperatively, the patient had a complete relief from the hemifacial spasm and marked improvement from trigeminal neuralgia. The painful tic convulsif in this case was probably produced by the tumor compressing and displacing the anterior cerebellar artery directly.

**Key Words:** Tic convulsif, schwannoma, cerebellopontine tumor

### INTRODUCTION

“Painful tic convulsif” was a term coined by Cushing<sup>1</sup> to describe the coexistence of trigeminal neuralgia and ipsilateral hemifacial spasm. Such cases have only rarely been reported, and the majority of reported cases of painful tic convulsif have involved vascular abnormalities around the fifth and seventh cranial nerves. Posterior fossa tumors are possible causes of tic convulsif, but epidermoid tumors are most common, followed by meningioma.<sup>2</sup> Schwannoma in the cerebellopontine angle (CPA) has not been reported, and we report here upon a case of painful tic convulsif caused by a schwannoma.

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### CASE REPORT

The patient, a 55-year-old woman, was referred to our department for neurological assessment in February 1997. She had a 9-month history of right hemifacial spasm and severe episodic pain on the right mandibular, maxillary and ophthalmic region apparently induced by tooth brushing and eating.

### Examination

Her physical examination was unremarkable and hematologic and biochemical studies normal. Neurological examination revealed a right-sided hemifacial spasm and severe neuralgia in the region of all branches of the right trigeminal nerve. A trigger point for touch was in the anterior region of the tragus. Neuro-otologic assessment indicated right-sided high frequency sensorineural hearing loss. Brain magnetic resonance images (MRI) revealed a 4cm round mass in the right CPA displacing the fourth ventricle and distorting the brain stem. The tumor enhanced heterogenously after gadolinium-diethylenetriamine penta-acetic acid (GdPA) administration (Fig. 1). A vertebral angiogram showed no tumor staining.

### Operation

The patient underwent a right suboccipital craniectomy in February 1998. A large tumor was attached to the pons, and directly compressed the 5th and 7th cranial nerves. The tumor was

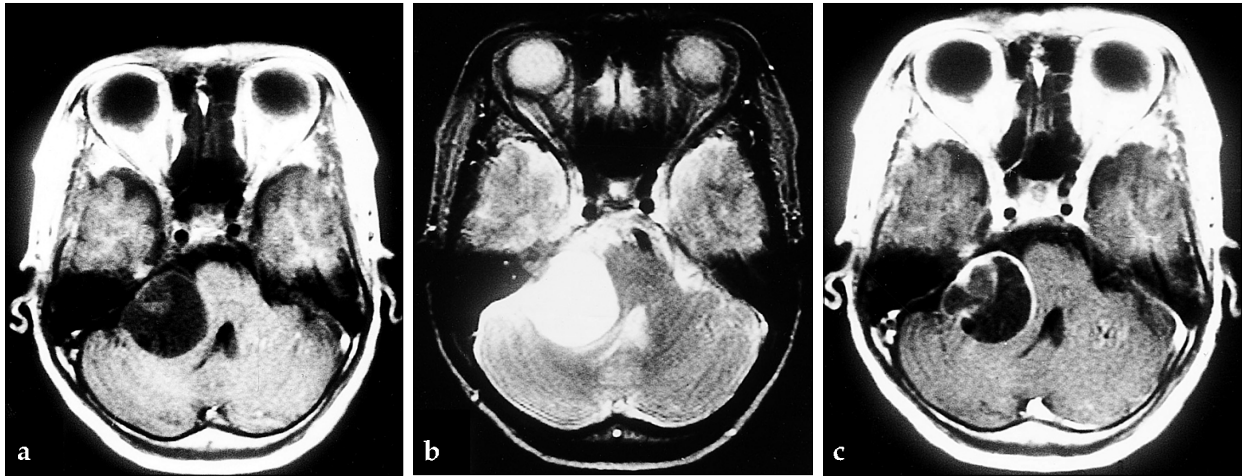


Fig. 1. Pre-operative brain MRI showing the tumor. It was attached to the right CPA, and showed heterogenous, ring-shaped enhancement with GdPA. (a. T1 weighted image, b. T2 weighted image, c. T1-GdPA image).

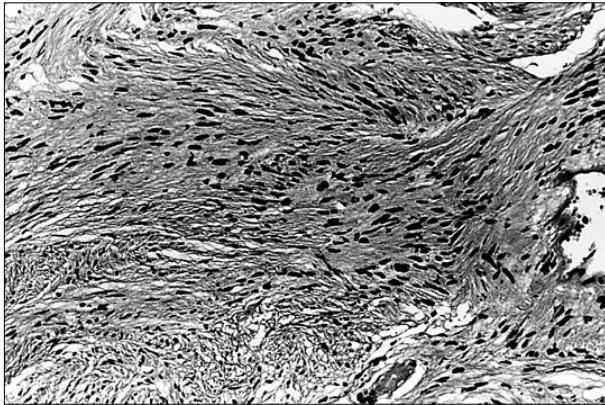


Fig. 2. Microscopic examination of the tumor showing elongated cells with cytoplasmic processes arranged in fascicles and vague nuclear palisading (H&E,  $\times 200$ ).

subtotally removed by operation. On microscopic examination, the tumor was found to be composed of spindle cells with cytoplasmic processes arranged in fascicles. The tumor cells had elongated bipolar nuclei without mitotic activity. There were "nuclear free zones" of processes that lay between regions of vague nuclear palisading, termed Verocay bodies (Fig. 2). The immunohistochemical stains were positive for S-100 protein and negative for EMA.

#### These findings are compatible with schwannoma

Postoperative course. Both the hemifacial spasm and trigeminal neuralgia gradually improved after operation. Radiotherapy was undertaken (5,400

cGy/6 weeks). Nine months later, a follow up brain MRI showed a small enhancing lesion in the right CPA. The patient's right-sided hemifacial spasm had resolved completely and her trigeminal neuralgia was improved markedly.

#### DISCUSSION

Trigeminal neuralgia is commonly caused by compression of the trigeminal nerve at its entry zone, and the same mechanism is thought to be responsible for hemifacial spasm.<sup>3,4</sup> The coexistence of trigeminal neuralgia and ipsilateral hemifacial spasm is uncommon. Since Cushing's described this condition as "painful tic convulsif", a small number of cases have been reported.<sup>1</sup> The majority of reported cases of painful tic convulsif have had vascular abnormalities around the fifth and seventh cranial nerves, due to, ectatic vertebral or basilar arteries, arteriovenous malformations and angiomas.<sup>5</sup> Tumors causing painful tic convulsif have been reported in 9 cases. All tumors, except tentorial meningioma, also reported to cause painful tic convulsif, have been located at the CPA. Epidermoid cyst was the most common tumor causing painful tic convulsif. In the cases of epidermoid cysts,<sup>6</sup> the fifth and seventh cranial nerves were surrounded or compressed by the tumor. The capsule and contents of the tumor irritate the fifth and seventh nerves, and might cause trigeminal neuralgia and

hemifacial spasm.<sup>2</sup>

When painful tic convulsif is caused by a tumor, two different (symptomatic?) mechanisms might be proposed. In the case of a tumor in the CPA, the tumor may directly compress the fifth and/or seventh nerve. In the case of tumor located outside the CPA, the tumor may compress and displace the brain stem, causing secondary neurovascular compression of the fifth and seventh nerves (the so-called "remote effect").<sup>2</sup> Therefore, decompression of the fifth and seventh nerves by removing the tumor might be the first choice for painful tic convulsif caused by posterior fossa tumor located outside the CPA. Cook and Jannetta<sup>1</sup> have reported a meningioma compressing the anterior inferior cerebellar artery at the root entry zone of the seventh nerve. In a case like this, additional microvascular decompression of the root entry zone of the nerves might be necessary if facial pain and/or hemifacial spasm remain despite tumor removal.

In our case, the CPA schwannoma caused compression, distortion, and rotation of the brain stem and vessels and directly compressed the root entry zone of the fifth and seventh nerve. Decompression of the root entry zone of nerve after (by?) removal of the mass caused an improvement of clinical symptoms. Because of residual schwannoma surrounding the brain stem, radiation

therapy was performed.

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