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Ganglioglioma of the trigeminal nerve: MRI

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Abstract Ganglioglioma of the cranial nerves is extremely rare; only a few cases involving the optic nerves have been reported. We present a case of ganglioglioma of the trigeminal nerve, which was isointense with the brain stem on all MRI sequences and showed no contrast enhancement.

Key words Ganglioglioma · Nerve trigeminal, magnetic resonance imaging

Introduction

Gangliogliomas of the brain, spinal cord, optic nerve, and optic chiasm have been described [1]. We report a ganglioglioma of the trigeminal nerve, not previously described in the medical literature.

Case report

Approximately 1 year prior to admission, a 34-year-old man was near a 55-gallon drum which exploded. Two weeks later, while getting out of bed, he fell due to a lack of balance. He continued to have episodic dizziness and light-headedness; Episodes occurred twice a day and as frequently as every 4–5 days. They lasted from 20 min to 4 h.

Examination was normal apart from mild hyperacusis in the left ear; he showed nystagmus thought to be physiologic.

On MRI a small, round mass on the cisternal portion of the left fifth cranial nerve was identified (Fig. 1 a, b); it showed no significant contrast enhancement (Fig. 1 c). MRI 6 months later showed no change.

A left suboccipital craniotomy was undertaken. The left vestibular nerve was divided to treat the clinical post-traumatic vestibular dysfunction. The mass was resected from the fifth cranial nerve. Histological examination demonstrated a benign tumor typical of a ganglioglioma (glial neuronal hamartoma) [2], composed of large, haphazardly arranged neurones containing abundant, eosinophilic cytoplasm with prominent nuclei and nucleoli, in a matrix of delicate fibrillar neural parenchyma containing scattered benign astrocytes (Fig. 2). Postoperatively the patient has not fallen or had facial paresthesiae. Facial and trigeminal function is normal, as is hearing.

Discussion

Gangliogliomas can occur almost anywhere in the central nervous system. With decreasing frequency, they have been identified in the cerebral hemispheres, cerebellum and spinal cord [1], and rarely in the brain stem, hypothalamus, third and fourth ventricles, pineal region and optic nerve [3].

They constitute 0.4–0.9 % of central nervous system tumors [4].

On MRI this tumor gave nonspecific homogeneous signal indistinguishable from the adjacent brain stem with all imaging sequences (Fig. 1 a, b) and showed no contrast enhancement [5]. The reason for the latter are

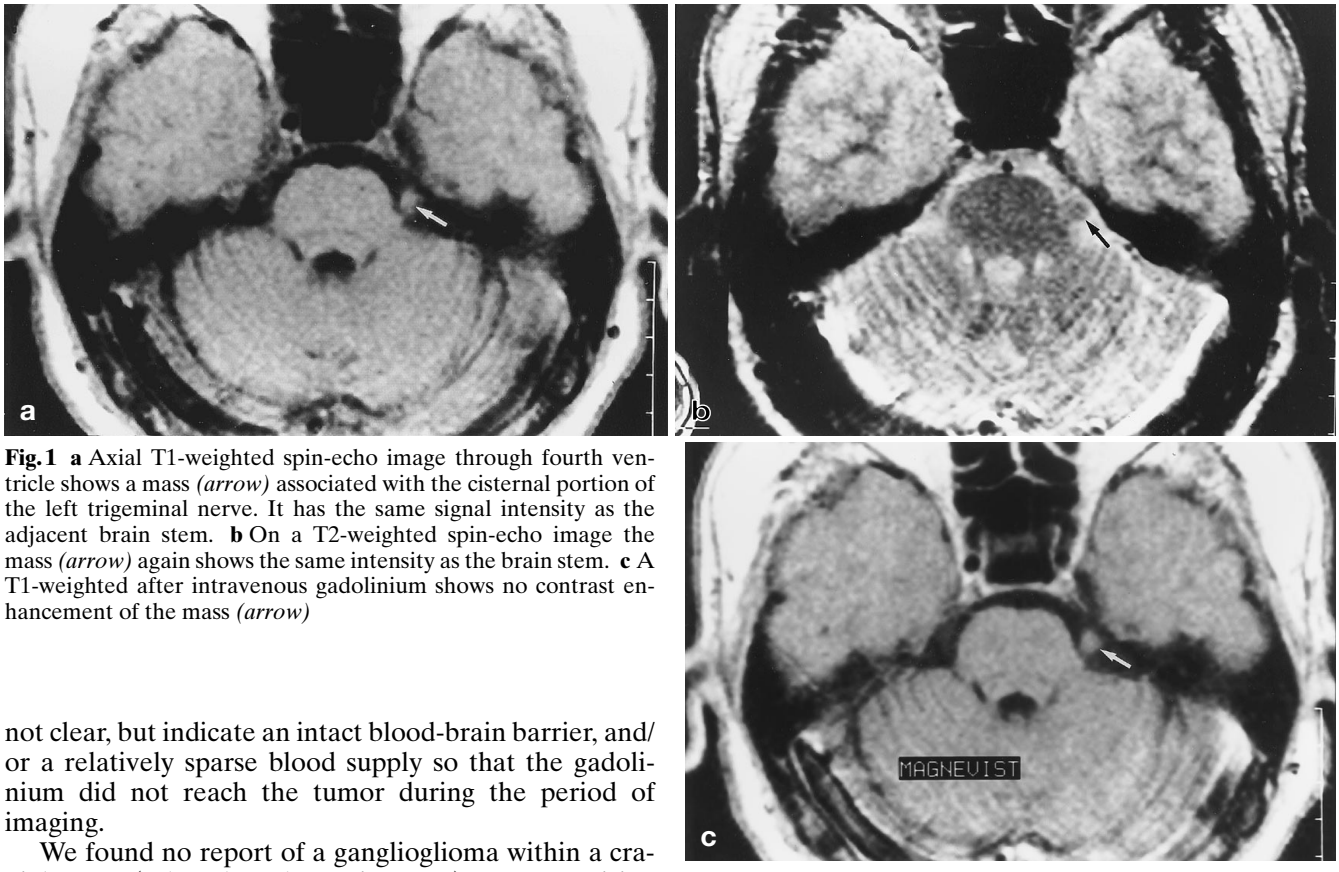

Fig. 1 **a** Axial T1-weighted spin-echo image through fourth ventricle shows a mass *(arrow)* associated with the cisternal portion of the left trigeminal nerve. It has the same signal intensity as the adjacent brain stem. **b** On a T2-weighted spin-echo image the mass *(arrow)* again shows the same intensity as the brain stem. **c** A T1-weighted after intravenous gadolinium shows no contrast enhancement of the mass *(arrow)*

not clear, but indicate an intact blood-brain barrier, and/or a relatively sparse blood supply so that the gadolinium did not reach the tumor during the period of imaging.

We found no report of a ganglioglioma within a cranial nerve (other than the optic nerve). Tumors arising from the trigeminal nerve are commonly schwannomas or neurofibromas. The presence of a ganglioglioma in the fifth cranial nerve is difficult to explain. It has a corollary in the occurrence of neuroblastomas along sympathetic ganglia and, rarely, in areas where ganglion cells do not have cell bodies. Neuroblastomas, malignant tumors of primitive neuroectodermal origin, may mature or differentiate to become gangliogliomas, which are less malignant. Differentiation to a different

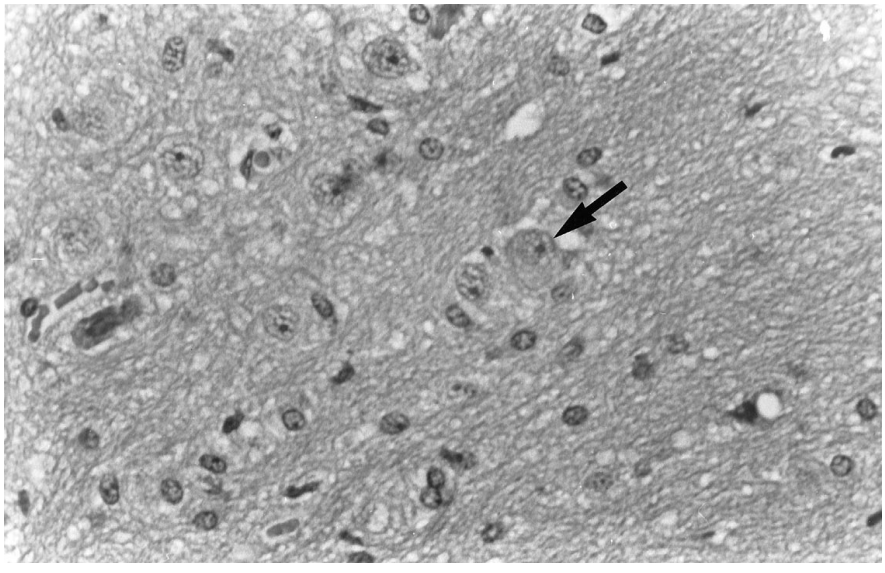

Fig. 2 Light microscope (hematoxylin and eosin) shows large, haphazardly arranged ganglion (neuron) cells *(arrow)* containing abundant, eosinophilic cytoplasm with prominent nuclei and nucleoli. These are arranged in a matrix of delicately fibrillar neural parenchyma containing scattered, benign astrocytes (original magnification 200)

benign form of tumor (ganglioneuroma or ganglioglioma) is hypothesized in this case. The ganglioneuroma is primarily composed of ganglion cells with Schwann-cell elements, whereas gangliogliomas contain glial cells in addition to the ganglion cells. Gangliogliomas contain a gliomatous component; clinical evidence of neurofibromatosis should be sought [2]. Our patient was examined for evidence of this disease, but none was found.

This tumor might also have arisen from an embryonic rest of central nervous system tissue (glial and ganglion cells) that became displaced into the fifth cranial nerve from the adjacent brain stem during embryogenesis.

The differential diagnosis would include schwannoma or neurofibroma and metastasis. However, most of these would be expected to enhance. Inflammatory processes were also a possibility, but again would be expected to enhance. Since the signal was typical of a hamartomatous lesion, following the signal of the brain stem on all sequences, this was a diagnostic possibility.

Although it may not be possible in all cases to follow such lesions conservatively, gangliogliomas are well known for their slow growth, and malignant transformation is uncommon [6]. Despite its rarity, ganglioglioma should be considered when a lesion associated with a cranial nerve closely parallels the brain stem tissue in signal and does not enhance.

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