Unusual Location of Intracranial Vagus Neurinoma

—Case Report—

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Abstract

A 55-year-old male presented with hearing disturbance and tinnitus in the left ear. Computed tomography (CT) and magnetic resonance imaging demonstrated a well-defined, homogeneously enhanced mass in the left cerebellomedullary cistern without extension close to the jugular foramen. A three-dimensional image reconstructed from thin-slice CT scans demonstrated that the mass was clearly separated from the jugular foramen. The mass lesion was totally removed surgically. At surgery the tumor was found to originate from one rootlet of the vagal nerve just after its exit from the medulla oblongata. The histological diagnosis was neurinoma. Intracranial neurinomas of the glossopharyngeal, vagal, or accessory nerve usually originate within or close to the jugular foramen. This unusual location made it difficult to achieve a correct preoperative diagnosis.

Key words: vagus neurinoma, jugular foramen, cerebellomedullary cistern, magnetic resonance imaging, three-dimensional image, surgery

Introduction

Intracranial neurinomas originating from the glossopharyngeal, vagal, or accessory nerve are relatively rare. The majority of the reported cases with detailed descriptions of the tumor location arose from the glossopharyngeal, vagal, or accessory nerve within or close to the jugular foramen,^{1,3–7,11,14–16,18,19} except for four cases of spinal accessory neurinomas located in the cisterna magna.^{2,8,12}

We describe an unusual case of intracranial neurinoma arising from the vagal nerve just after its exit from the medulla oblongata with no tumoral extension close to the jugular foramen.

Case Report

A 55-year-old male presented with a 5-year history of progressive hearing disturbance and tinnitus in the left ear. He reported no difficulty with swallowing or change in his voice. Neurological examination showed left sensorineural hearing disturbance and ataxic gait, but no dysphagia, hoarseness, or impaired

pharyngeal movement. Preoperative computed tomography (CT) revealed a well-enhanced lesion in the left cerebellomedullary cistern just above the jugular tuberculum, but neither the internal auditory canal nor the jugular foramen was dilated (Fig. 1). A three-dimensional image reconstructed from thin-slice CT scans demonstrated that the mass was clearly separated from the jugular foramen (Fig. 2). Magnetic resonance (MR) imaging showed the mass as a well-defined low intensity area on the T₁-weighted images and a high intensity area on the T₂-weighted images, with homogeneous enhancement by gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA) (Fig. 3). Cerebral angiography showed no tumor blush. The preoperative diagnosis was jugular tuberculum meningioma.

Left far lateral suboccipital craniectomy with C-1 hemilaminectomy was performed with the patient in the park-bench position. The glossopharyngeal and vagal nerves were flattened, elongated, and displaced caudally by the tumor. The proximal parts of these nerves were firmly attached to the tumor, but the distal parts could be easily peeled off the tumor. We confirmed that the tumor originated from one rootlet of the vagal nerve just after its exit from the medulla oblongata. The facial and vestibulocochlear nerves

Fig. 1 Coronal postcontrast CT scan demonstrating a well-enhanced mass in the left cerebello-medullary cistern just above the jugular tuberculum without extension close to the jugular foramen.

Fig. 2 Three-dimensional image reconstructed from thin-slice CT scans demonstrating that the mass was clearly separated from the jugular foramen (arrow).

were not involved, and the tumor was not attached to the dura. There was no tumoral extension close to the jugular foramen. The tumor was totally removed with its origin (Fig. 4).

Histological examination of the tumor specimen showed dense and loose-meshed tissues. The dense area contained interwoven bundles of long bipolar cells with elongated nuclei aligned in a palisading fashion (Fig. 5). The histological diagnosis was neurinoma.

The postoperative course was uneventful, except for mild swallowing difficulties and hoarseness.

Fig. 3 Axial T₁-weighted MR image with Gd-DTPA demonstrating a well-defined, homogeneously enhanced mass in the left cerebellomedullary cistern compressing the medulla oblongata medially.

Discussion

Neurinomas originating from the glossopharyngeal, vagal, or accessory nerve are often called jugular foramen neurinomas without identifying the specific nerve of origin. Kaye *et al.*¹⁰ reported 13 cases of jugular foramen neurinomas classified into the following three types: type A, tumors primarily intracranial in location, with only a small extension into bone; type B, tumors with the main mass within bone, with or without an intracranial component; and type C, primarily extracranial tumors with only minor extension into bone or into the posterior fossa. They postulated that the location of the tumor depends on the site of origin from the nerves passing through the pars nervosa of the jugular foramen. The more proximal lesions will enlarge into the posterior fossa, but the more distal lesions will expand extracranially. Tumors arising in the mid region will tend to expand primarily into the bone. According to this classification, all three types of tumors are located in or close to the jugular foramen, and this is often the most valuable diagnostic feature. Two cases of intracranial vagus neurinoma similar to our present case have been reported by Sawamura and de Tribolet¹³ and Kawamura and Sze.⁹ These neurinomas were not located within the jugular foramen, but the detailed relationship between the tumor location and the jugular foramen was not described.

The majority of acoustic neurinomas originate from the vestibulocochlear nerves within the internal auditory canal. Tos *et al.*¹⁷ reported a rare type of

Fig. 4 upper: Operative photograph showing that the proximal parts of the glossopharyngeal and vagal nerves are firmly attached to the tumor (Tu). lower: Operative photograph after the tumor was totally removed. Arrowheads indicate the rootlet of the vagal nerve from which the tumor originated. Notice that the tumor origin is close to the medulla oblongata (Md), and that the distal parts of the glossopharyngeal (IX) and vagal nerves are intact.

Fig. 5 Photomicrograph of the tumor specimen showing interwoven bundles of long bipolar cells with elongated nuclei aligned in a palisading fashion. HE stain, $\times 400$.

acoustic neurinoma which they called the medial tumor, defined as an extrameatal tumor without tumor mass in the internal auditory canal. Such tumors probably originate from the vestibulocochlear nerves outside the porus in the cerebellopontine angle. Our present case seems to correspond to this "medial tumor," and should be differentiated from the type A tumors as defined by Kaye *et al.*

The intracranial neurinoma in this patient originated from the vagal nerve just after its exit from the medulla oblongata with no tumoral extension close to the jugular foramen. This unusual location made it difficult to achieve a correct preoperative diagnosis.

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