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Jugular Foramen Neurilemmoma Mimicking an Intra-axial Brainstem Tumor

- A Case Report -

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Jugular foramen neurilemmoma is frequently manifested as a jugular foramen syndrome or extraaxial mass. Neurilemmoma arising from the cranial nerves of the foramen, although rare, may be manifestated as an intracranial or extracranial mass lesion. When the tumor is located only in the intracranium, it is often misdiagnosed as an acoustic neurinoma or a brainstem tumor because of their similarity in clinical or radiological findings. We present a rare case of jugular foramen neurilemmoma with only intracranial extension with clinical and radiologic features of an intra-axial brainstem tumor.

Key Words : *Jugular foramen, Neurilemmoma, Schwannoma*

INTRODUCTION

Neurilemmoma is an encapsulated nerve sheath tumor arising from Schwann cells or neurilemmal cells(Slager, 1972). Its incidence accounts for about 8% of all primary intracranial tumors, and it constitutes the great majority of primary intracranial extracerebral neoplasms (Christoferson et al., 1959). The most common form of neurilemmomas is the acoustic neurinoma arising from the vestibular branch of the vestibulocochlear nerve. Other than the acoustic nerve, cranial nerves rarely constitute sites of origin for neurilemmomas. Only those tumors of the acoustic or trigeminal nerves and jugular foramen are numerically significant. Neurilemomas arising from the cranial nerves of jugular foramen are rare and represent only 2.9% of all the intracranial neurilemmomas (Bordi et al., 1989 ; Hiscott et al., 1982). We encountered a case of jugular foramen neurilemoma, with only intracranial extension, mimicking an intra-axial brainstem tumor on clinical and radiological findings.

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CASE REPORT

A 46-year-old woman suffered from chronic headache and slow progressive speech disturbance for five years. On neurologic examination, the patient was alert and well oriented. Down beat nystagmus, scanning speech, ataxic gait, and long tract signs suggesting an intra-axial posterior fossa lesion were noted. Signs involving the lower cranial nerves or other abnormal neurologic findings indicating an extra-axial brainstem lesion were not detected. Plain skull X-rays failed to show any abnormalities. Brain MR images demonstrated a large posterior fossa mass on the right cerebellopontine angle, which was interpreted as an intra-axial mass because of an unclear interface between the mass

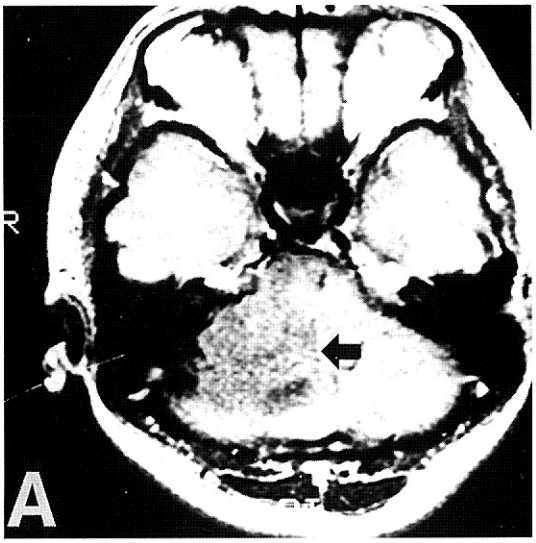

Fig. 1A. Inhomogenous low signal intensity mass lesion (arrow) is on the right cerebellopontine angle and cerebellum. Interface between the mass lesion and the cerebellum and brainstem is unclear (T1-weighted image).

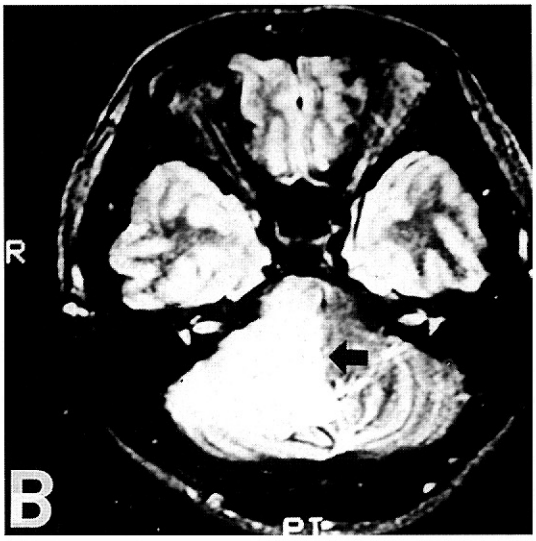

Fig. 1B. The mass lesion(arrow) shows high signal intensity (T2-weighted image).

and the cerebellum and brainstem. The mass showed an inhomogenous low signal intensity on T1-weighted MR image and high signal intensity on T2-weighted MRI with strong enhancement after intravenous administration of gadolinium (Fig. 1A,1B).

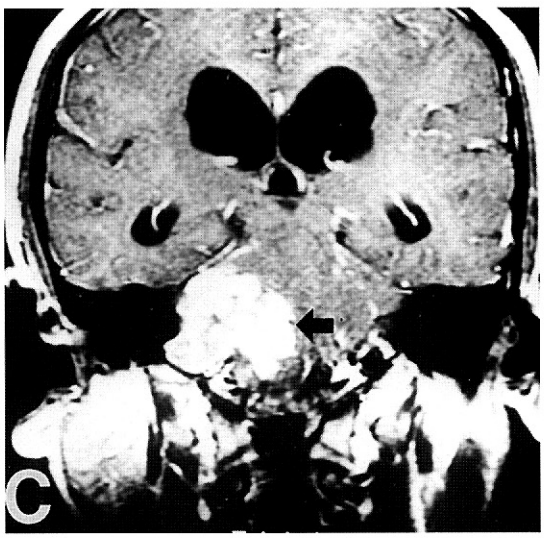

Fig. 1C. The irregularly enhanced mass(arrow) lesion compresses the brain stem on right cerebellopontine angle. Brainstem is shifted to the left side (T1-weighted enhanced image).

The brainstem was shifted to the left side (Fig. 1C). In audiometric test, the speech response threshold was 30dB in each side. Absolute latency of I, III, V waves and I-III, III-V, I-V interpeak latency were within normal limits in right and left brainstem auditory evoked potential. With an impression of an intra- axial posterior fossa tumor, surgical removal was performed. The mass was firmly attached to the lower cranial nerves at the level of the jugular foramen. Histologically, the mass was a cellular spindle cell tumor, composed of numerous characteristic Verocay bodies(Fig. 2). The origin of the tumor was a cranial nerve at the jugular foramen, but the definite cranial nerve was not identified. She died of aspiration pnuemonia after operation.

DISCUSSION

Neurilemmoma originating from the intracranial segments of the glossopharyngeal, vagus, and accessory nerves called neurilemmoma of the jugular foramen nerves because of their proximity (Henschen, 1955 ; Hakuba et al., 1979). Anatomically, the jugular foramen is divided into the anteromedial and posterolateral compartments by a fibrous or bony septum. The medial compartment transmits the 9th, 10th, and 11th cranial nerves and the posterior

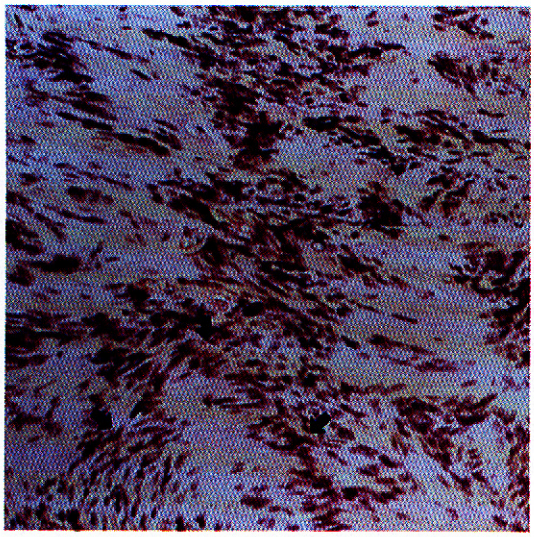

Fig. 2. This figure shows cellular spindle cell tumor, composed of numerous characteristic Verocay bodies (arrows) (hematoxylin-eosin stain × 100).

compartment transmits the internal jugular vein and the posterior meningeal artery (Arenberg and McCreary, 1971). It has been postulated that the position of the tumor depends on its point of origin from the nerves passing through the pars nervosa of the jugular foramen(Kinney et al., 1982). The most useful and detailed investigation that will reveal the extent of any intracranial component is MR image, especially with gadolinium enhancement (Tan et al., 19 90). According to the radiological and surgical features, the tumors were classified into four types : type A, a tumor primarily at the cerebellopontine angle with minimal enlargement of the jugular foramen; type B, a tumor primarily at the jugular foramen with intracranial extension; type C, a primarily extracranial tumor with extension into the jugular foramen; and type D, a dumbbell-shaped tumor with both intra and extracranial components (Samii et al., 1995). In patients with type A tumors, various degree of extra-axial symptoms, such as deafness, vertigo, and ataxia were usually present. Although this case was type A, she had no signs or symptoms of extra-axial mass lesion. These clinical findings could be explained by the innervation overlap of the lower cranial nerves (Waxman and Degroot, 1995). In other word, the signs of one lower cranial nerve palsy can be masked by the other lower cranial nerves on the same side due to innervation overlap. According to the clinical and MRI findings, we considered the lesion was an intra-axial posterior fossa mass. We therefore conclude that extra-axial lesions should be included in the differential diagnosis, even though a patient manifests only symptoms and signs of intra-axial lesions.

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