

Hypoglossal Schwannoma Presenting as Hemi-Atrophy of the Tongue

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Abstract- Schwannoma of the hypoglossal nerve is extremely rare. We report the clinical manifestations of a patient with Schwannoma of the hypoglossal nerve with hemi-atrophy of the tongue and numbness in the lip. Magnetic resonance image study of the brain showed a lobulated mass at the right posterior fossa with an extension to the right upper neck. Surgical intervention was performed with right occipital craniotomy and a partial resection of C1 and occipital condyle. Pathological studies confirmed a Schwannoma with hemorrhages and necrosis.

Key Words: Schwannoma, Hypoglossal nerve, Tongue atrophy

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INTRODUCTION

Schwannomas of the hypoglossal nerve are exceedingly infrequent^(1,2). To our knowledge, there has been only one reported case of hypoglossal Schwannoma in Taiwan and our case marks the 70th reported case of hypoglossal neurinoma in the Medline^(1-7,11,13-17). Because of their scarcity, early diagnosis of these lesions is intricate⁽⁴⁾. We here report a pathologically proven case of the left hypoglossal nerve Schwannoma in a 66-year-old woman who presented with progressive left tongue atrophy. The brain magnetic resonance imaging showed a lobulated lesion at the right posterior fossa with an extension to the right upper neck was reported.

CASE REPORT

A 66-year-old woman presented with hoarseness and impairment of the lingual mobility. She first noticed these symptoms seven months previously, together with tinnitus, slurred speech and numbness in the right part of the lip. During the ensuing weeks, she experienced headache with intermittent interruption of sleep and numbness in the right half of the face. Hence, she was admitted to the hospital. On neurological examinations, the patient was alert and oriented. There was obvious dysarthria but without dysphasia. The cranial nerve functions were almost intact except for a tongue deviation to the right side with hemi-atrophy of the tongue

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(Fig. 1). The muscle strength and muscle tone in four extremities were normal. There was no loss of systemic vibration sensation and the remaining sensory modalities were also preserved. The tendon reflexes were brisk. The plantar responses were flexor. The gait was steady and the Romberg's test was normal.

Laboratory data including routine biochemical and hematological examinations were normal. Brain magnetic resonance imaging (MRI) showed a right jugular foramen mass and a glomus jugulare tumor was impressed. On a gadolinium enhancement study of the brain, there



Figure 1. Deviation of the tongue on protruding with hemiatrophy of the tongue on the right side.

was a lobulated tubular lesion in the right posterior fossa and the mass extended to the right upper neck (Fig. 2).

Right subcortical craniotomy was performed with a partial resection of the C1 lateral mass and occipital condyle. There was a soft tumor within the enlarged hypoglossal foramen with a compression of medulla oblongata noted during the operation. The pathological examination confirmed a Schwannoma with Antoni Type A and Antoni Type B cells with hemorrhages and necrosis (Fig. 3).

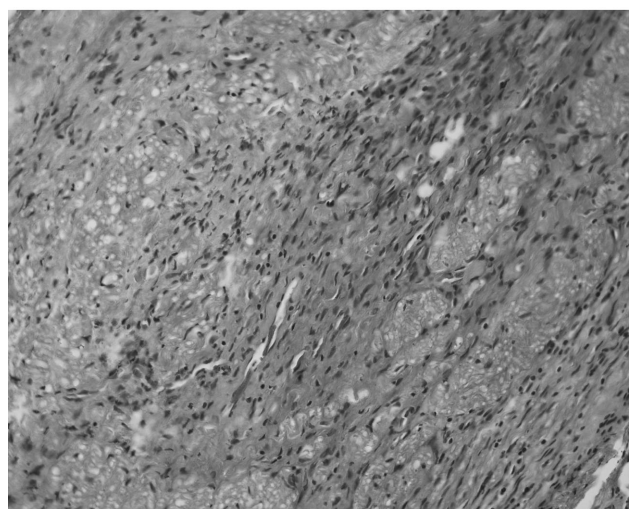


Figure 3. Pathological studies indicates a characteristic picture of neurinoma with Antoni A and Antoni B type tissues (H&E; ×100)

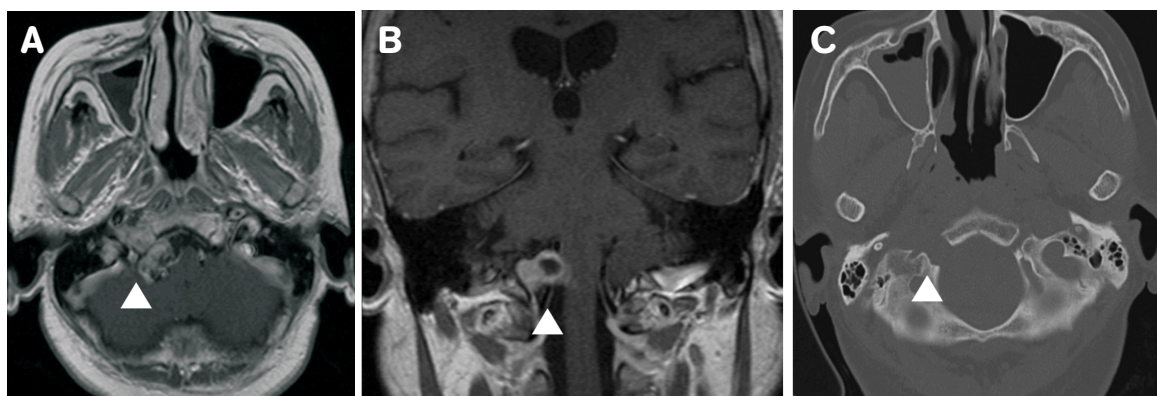


Figure 2. (A) Brain T1- weighted MR image with enhancement reveals a mass extending from the extracranial part and protruding to the intracranial part with a mass effect on the left pontomedulla region (white arrow head). (B) The coronal section of contrast-enhanced MR image shows a heterogeneously enhanced mass (white arrowhead). (C) Axial brain CT scan reveals the enlarged hypoglossal canal on the right side (white arrowhead).

DISCUSSION

Schwannomas account for 8.5% of all intracranial tumors and more than 90% of the tumors originate from the 8th cranial nerve⁽⁸⁾. The first case of hypoglossal neurinoma was noted in 1933⁽⁹⁾. Schwannoma is the second most common intracranial extra-axial neoplasm after meningioma. The leading symptoms of the hypoglossal neurinoma include headache and dizziness^(10,12). High resolution CT scan of the posterior fossa with bony details of the foramen passing through by cranial nerve is the neurodiagnostic procedure of choice⁽¹⁰⁾. Histologically, Antoni type A neurilemoma has elongated spindle cells in irregular streams and is compact in nature, and type B neurilemoma has a looser organization, often with cystic spaces. The cystic spaces can result in high signal intensity on T2-weighted MRIs^(7,18).

The hypoglossal nerve arises from the hypoglossal nucleus and then passes through the hypoglossal canal, and provides motor fibers innervating the muscles of the tongue⁽¹⁹⁾. Most of the hypoglossal neurinomas have a dumbbell shape and involve both intracranial and extracranial segment of the hypoglossal nerve⁽¹⁶⁾. However, intracranial hypoglossal neurinomas are unusual⁽¹⁾. The most distinguishing clinical findings of patients with hypoglossal nerve Schwannoma are unilateral tongue atrophy and fasciculations^(3,8). The differential diagnosis of tumor involving hypoglossal canal includes chemodectoma, chordoma, meningioma, lymphoma and metastatic tumors⁽²⁰⁾.

Our patient had slurred speech initially and then numbness in the right half of the face. Tinnitus is a rare complaint in the previous reported cases of hypoglossal Schwannoma. This may be related to the mass effect involving the cochlear fiber at the level of medulla portion. Initial brain MRI showed a right posterior fossa tumor compressing the pontomedulla junction and a jugular foramen tumor was suspected at first. However, high resolution computed tomography revealed an enlarged right hypoglossal canal (Fig. 2C) and thus a hypoglossal tumor was impressed. Pathological studies revealed a Schwannoma with of Antoni A and Antoni B types. The numbness of the right half of the face was

most likely due to the compression of the right spinotrigeminal tract. The symptom of numbness in the lip improved after the operation.

Hypoglossal nerve neuropathy is frequently associated adjacent cranial nerve involvement. Isolated hypoglossal neuropathy is rare. Tumor, infection, trauma of skull base, radiation and vascular insult may attribute the isolated hypoglossal nerve palsy⁽²¹⁾.

In conclusion, patients with hypoglossal Schwannomas can present with muscle atrophy of the tongue, but they can also be asymptomatic initially⁽¹¹⁾. Early diagnosis can be achieved after a detailed neurological examination.

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