Clinical Note

Brain stem glioma, a rare cause of hearing loss and dysequilibrium

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The complaints of hearing loss and imbalance or vertigo are commonly encountered in otolaryngology practice. Typically, main causative disorders are endolymphatic hydrops, acoustic neuroma or cerebellopontine angle tumors and other inner ear diseases. Much less frequently, these symptoms may be the first symptom of rare intracranial pathologies. Brain stem gliomas comprise approximately 15% of all childhood and adolescent brain tumors.[1] However, they are rare in the adult population. The presenting symptoms of gliomas are varied, and they are among the rare etiopathologic causes reported in association with hearing disturbance. In this article, we present a case of brain stem glioma associated with progressive sensorineural hearing loss and disequilibrium and discuss its pathological, clinical, radiological, and therapeutic features.

A 40-year-old man was admitted to our department with the complaints of unsteadiness, unilateral right-sided hearing loss, and dysarthria for 3 months. Routine ENT examination was normal. A spontaneous nystagmus toward the left side was detected. Also, a generalized ataxia was seen during the gait test, and a falling tendency to the right side was observed during Romberg test. Pure tone audiometry revealed a right-sided moderate sensorineural hearing loss. Speech discrimination score was 10% on the right, and 90% on the left. Magnetic resonance imaging demonstrated an iso/hypointense, diffuse brainstem lesion (**Figure 1**). Radiological findings were consistent with diffuse brainstem glioma. He was consulted with the neurosurgery and radiology departments, and referred to the radiation oncology department. Unfortunately, he died during the radiation therapy.

Brainstem gliomas are rare in the adult population. These tumors can be divided mostly into 2 groups: the classic diffusely infiltrative pontine glioma and focal gliomas outside the ventral pons. Diffuse pontine gliomas are infiltrative astrocytomas of varying grade, whereas the focal gliomas are frequently pilocytic astrocytomas.[1] The MRI has certainly been more sensitive for the detection of these neoplasms. The overall MRI appearance of diffuse pontine glioma is variable, with regard to signal intensity and enhancement with intravenous contrast.[1] Intratumoral heterogeneity is occasionally present, but cysts are not common. There is usually obvious anatomic distortion of the pons morphologically. When gross enlargement of the brainstem is present, the traditional indirect signs of pontine glioma are also seen on MR imaging, including invagination or engulfment of the basilar artery and effacement of the prepontine cistern. An undulating ventral border of the brainstem on the sagittal image may be the initial finding of a mass lesion and when present, significantly limits the differential diagnosis.[1] Contrast enhancement has been reported in approximately one-half of cases and is often focal and nodular. Pilocytic brainstem gliomas are often better circumscribed and sometimes markedly exophytic. Dorsally exophytic lesions often enhance intensely and are grossly multicystic. In our case, MRI findings were matched closely with the diagnosis of diffuse pontine glioma.

Clinically, the patient with diffuse pontine glioma presents with a triad of cranial nerve deficits, long tract signs, and ataxia over just several months or less.[1] In the present case, progressive sensorineural hearing loss, and disequilibrium were the first symptoms. In the English literature, there have been very few reported brain stem glioma cases leading to otologic, neuro-otologic, or both, symptoms and signs. Licht et al[2] described a temporal lobe glioblastoma case causing hearing disturbance and vertigo. Schade et at[3] reported a brain stem glioma case presenting with the symptoms of unilateral tinnitus and incomplete facial paresis. Also, Elgamal and Coakham[4] presented an adult patient with hemifacial spasm due to fibrillary pontine glioma. These symptoms and signs are probably due to mass compressive effect of the tumor.

Histopathological investigation of the specimen or biopsy material gives the certain diagnosis. However, as in our case, the radiological diagnosis is important where the lesion is located in a complex neuro-anatomic region. Diffuse adult gliomas are unique among tumors in human beings for the scale to which their imaging features relate to clinical behavior and histopathological characteristics.[5] Because of their typical infiltrative nature and location, diffuse brainstem gliomas are usually not surgically resectable, so the mainstay of therapy has been

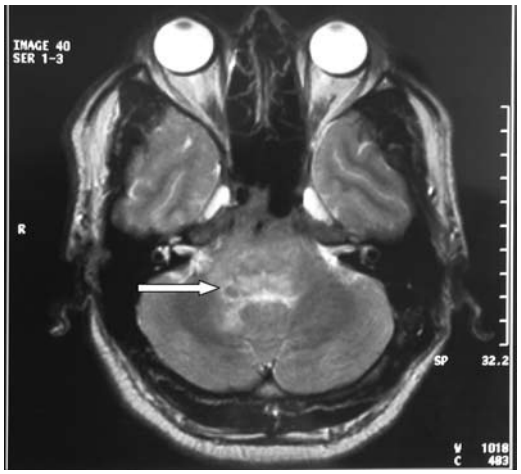

Figure 1 - An magnetic resonance imaging demonstrated an iso/hypointense, diffuse brainstem lesion. The arrow indicates the diffuse brain stem glioma.

irradiation.[1] The prognosis for patients with infiltrative brainstem astrocytoma is dismal; 5-year survival rates are less than a few percent. Other potential causes of abnormal signal intensity in the brainstem of a child or young adult include encephalitis, demyelinating disease, cavernous angioma, or arteriovenous malformation, capillary telangiectasia, and rarely infraction.[1] Although hearing loss and imbalance or vertigo are common complaints in otolaryngology practice, we believe that patients who have unilateral and progressive symptoms should be investigated by MRI.

Received 3rd June 2007. Accepted 22nd July 2007.

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