Unusual Case of Cerebellopontine Angle Epidermoid Causing Cerebellar Atrophy

Sir,

A 33-year-old male patient presented with swaying to the right side while walking, inability to perform coordinated movements using his right hand and slurring of speech of 1-year duration. Clinical examination showed right cerebellar signs-gaze evoked horizontal nystagmus, hypotonia of right upper limb and lower limb, dysdiadochokinesia, impaired finger nose, and heel-knee test. Examination of cranial nerves, rest of motor and sensory systems was unremarkable. His magnetic resonance imaging (MRI) [Figure 1a-d] showed 4.6 cm × 4.5 cm × 4.8 cm well-defined extraaxial lesion in right cerebellopontine (CP) angle cistern, extending into the right quadrigeminal cistern with significant compression of the right cerebellar hemisphere, right middle cerebellar peduncle and pons. The lesion was T2 hyperintense, heterogeneously T1 hypointense, fluid-attenuated inversion recovery (FLAIR) heterogeneously hypointense without contrast enhancement, suspicious of epidermoid cyst. The right cerebellar hemisphere was showing volume loss with T2/FLAIR hyperintensity suggestive of atrophy [Figure 1c and d, short arrow] as compared to the normal left cerebellar hemisphere [Figure 1d, long arrow]. No cerebellar infarct was seen. He underwent right suboccipital retrosigmoid craniotomy and excision of lesion. Intraoperatively, the lesion contained flakes of pearly white fragmented tissue characteristic of epidermoid [Figure 1e]. Histopathology confirmed the diagnosis. The lesion was adherent to V-VIII cranial nerves, cerebellum, and brain stem. Postoperatively, he had partial improvement in symptoms when reviewed after 1-month in gait and fine movements using right hand.

Intracranial epidermoid cysts account for 0.2–1.4% of all intracranial tumors, constitute 40% CP angle lesions, and are the third most tumor of the region.[1,2] The onset of symptoms occurs between the second and fifth decades of life. Common presentations include a long history of tinnitus and hearing loss, diplopia, visual impairment, apathy, dizziness, hypoacusia, gait ataxia, symptoms of trigeminal neuralgia, hemifacial spasm, occasionally with vestibular symptoms, headache, or rarely rupture with meningeal features and symptoms of hydrocephalus.[3,4] The patient presented exclusively with cerebellar features even though the size was enormous and was causing compression on the brain stem and cranial nerves both radiologically and intraoperatively. Though there are reported cases of CP angle epidermoid presenting as cranial nerve features alone, there are seldom reports with isolated cerebellar features. As histological study of the cerebellum was not done, atrophy of right cerebellar hemisphere is

Figure 1: Magnetic resonance imaging of brain showing large right cerebellopontine angle lesion. (a) Hypointense on T1-weight. (b) Hyperintense on T2-weight. (c) Heterogeneously hypointense on fluid-attenuated inversion recovery. (d) Right cerebellar hemisphere gliosis seen (short arrow). (e) Pearly white tissue seen intraoperatively
solely an MRI finding. Exclusive cerebellar presentation in our case was attributed to cerebellar atrophy rather than direct mass effect. The cerebellar atrophy in our case is an unusual feature which we postulate it to be due to excessive focal leakage of cyst content because atrophy from direct compression has not been reported in the literature. Though leakage of cyst content of epidermoid can cause mild chemical inflammation causing arachnoid adhesions with adjacent structures (specifically cranial nerves), we describe unusual case of cerebellar atrophy secondary to epidermoid.

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