Cervical dumbbell ganglioneuroma producing spinal cord compression

Sir,

A 39-year old man presented with complains of progressive weakness and numbness of all four limbs for six months. There was moderate spastic tetraparesis that was more marked on the left side, and hypoesthesia below the C5 dermatome.

Magnetic resonance imaging (MRI) showed a large extramedullary dumbbell mass at the C4-C5 level. The tumor was hypointense on the T1 and hyperintense on T2 images. The spinal cord was severely compressed [Figure 1 A and B].

A two-staged operation was performed to resect the tumor. First, the patient was operated through a posterior cervical approach. Wide lumbectomy of C4 and C5 was done. The mass was solid, well capsulated, elastic, moderately vascularised, purely extradural and ventrolaterally located to the spinal cord. The lesion originated from cervical nerve C5, which was resected with tumor [Figure 2A]. resection of intraforaminal mass was performed through the foramen which had been already enlarged by tumor growth. After four weeks the patient underwent second operation. The paraspinal extradural component of tumor was excised through the left lateral cervical approach [Figure 2B]. The vertebral artery was dissected off the surface of encapsulated tumor.

At a three-year follow-up the patient had regained the motor strength in all four limbs. There was no radiographic signs of recurrence.

Histological examination of both tumor masses confirmed that the lesion was a ganglioneuroma [Figure 3].

Discussion

Kyoshima et al.\[1\] surveyed the literature on the subject and identified a total of only five pathologically confirmed cases of cervical spine ganglioneuromas. One patient was an 18-month old child and rest of the patients were young adults. Von Recklinghausen’s disease was present in two patients. The symptoms spinal cord compression were present in all reported cases. Two patients had bilateral tumors. The origin of tumors was sensory root ganglion or cervical nerve. In all the reported cases, the tumor growth was in dumbbell pattern. Intraspinal extradural growth was observed in three patients, while in-
tradural extension was seen in two patients.

On MRI about 75% of ganglioneuromas are isointense and 25% are hypointense on T1 images. Most of them are hyperintense on T2 images. The non-homogeneous appearance corresponds to areas of cystic degeneration, hemorrhage or necrotic degeneration.\[4,5\]

Ganglioneuromas are well encapsulated tumors and can be completely excised. Even when they are intradural, the tumor could be removed without cord injury because they are not adherent to the spinal cord.\[1\] This and previously reported cases indicate that spinal ganglioneuromas could be completely removed and cured.

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References

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Multicentric glioma presenting as man-in-the-barrel syndrome

Sir,

Primary motor cortex is somatotopically organized, and the motor representation in the precentral gyrus forms a motor homunculus – the leg and perineum is represented over the medial aspect of the motor strip, and the arm and the hand over the convexity. It is well known that precise and circumscribed weakness may affect one limb only if the appropriate area of the motor cortex or its projection pathway is selectively damaged.\[1\]

Bilateral upper limb weakness with relative sparing of lower limbs is usually seen in lesions involving the medullary decussation of the pyramidal tracts, or cervical spinal cord. Such a clinical syndrome due to lesions occurring bilaterally in the motor cortex is a rare event. These bilateral cortical lesions producing brachial diplegia are usually infarcts secondary to cerebral hypoperfusion following shock or aortic surgery. Cerebral tumor causing such paralysis is extremely rare.

A 37-year-old Nepalese national was admitted with seven-week history of gradually progressive worsening weakness of both arms. Weakness involved predominantly the shoulders, elbows and to a lesser extent, the wrist movements. Hands were relatively spared. He was unable to raise his arms or flex his elbows. He had remained ambulatory, continent, seizure-free, with no visual or gait disturbances. For two days prior to admission, he had complained of dull, generalized headache accompanied by one episode of vomiting.

Clinical examination revealed well-built and nourished normotensive male, with no abnormality of higher mental functions. Funduscopy revealed early bilateral papilledema. There was no nystagmus or involvement of facial or of lower cranial nerves. Motor system examination revealed power in both deltoids to be grade 0/5, that in elbow flexors 1/5 with wasting of deltoids. Tone was increased in both upper limbs with brisk biceps and triceps jerks. No fasciculations were observed. There was no sensory impairment. MRI brain showed bilateral frontal convexity space occupying lesions with surrounding edema [Figure 1].

After initial treatment with cerebral decongestants and dexamethasone, the right sided tumor was excised by craniotomy, while the left sided tumor was biopsied stereotactically at a later date. Histopathology of the excised specimen confirmed both the tumors to be glioblastoma multiforme.

The syndrome of disproportionate weakness of the upper

Figure 1: MRI brain showing bilateral frontal convexity space occupying lesions with surrounding edema