Isolated spinal accessory neuropathy and intracisternal schwannomas of the spinal accessory nerve

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Abstract

We report a 40-year-old female patient presenting with isolated left spinal accessory neuropathy that developed insidiously over 6 years. She complained of ill-defined deep neck and shoulder pain. On examination, prominent sternocleidomastoid and trapezius muscle weakness and atrophy, shoulder instability, and lateral scapular winging were observed. MRI identified a small mass of the cisternal portion of the spinal accessory nerve. Its appearance was typical of schwannoma. Surgical treatment was not offered because of the small tumor size, lack of mass effect and the questionable functional recovery in the presence of muscular atrophy.

Isolated spinal accessory neuropathy (SAN) is not uncommon after iatrogenic surgical injury of the accessory nerve (AN) during various procedures in the posterior triangle of the neck [1,2]. Less frequent aetiologies include traction injury, blunt or penetrating trauma, neurololgic amyotrophy, infection, or tumors involving the AN along its complicated course (intraaxially; intradurally in the high cervical segment of the spinal canal and at the foramen magnum; extradurally but intracranially, at the skull base/jugular foramen; extracranially, distal to the jugular foramen in the neck) [1,2]. Schwannomas of the AN are very rare and occur extracranially [3] or intracranially. The intracranial ones are further divided by location into intrajugular and intracisternal. Intrajugular masses result in jugular foramen syndrome and only occasionally present as isolated SAN [4].

Intracisternal AN schwannomas are exceedingly rare and usually manifest by the mass effects they produce [3]. The patient we report is – to the best of our knowledge – the first with this type of neoplasm to present with isolated SAN.

A 40-year-old female patient presented with poorly localized deep pain in the neck and left shoulder of 6 years duration. Her complaints were stationary over the last years, and she denied any other symptoms. There was no history of surgery or trauma to the neck. Examination showed prominent atrophy of the left trapezius and sternocleidomastoid (SCM) muscles (Fig. 1), drooping of the left shoulder and lateral scapular winging, accentuated by shoulder abduction.

Muscle power testing (MRC scale) was 4/5 for the trapezius and 3/5 for the SCM. Abduction of the left arm was limited to 90 degrees. The rest of her neurological examination was normal. In particular, there was no evidence of tongue atrophy or asymmetry, the soft palate and uvula were symmetrical with preserved gag reflexes. Direct laryngoscopy was normal. Formal gustometry was not performed, but the patient denied any changes in taste sensation and testing for sour of the posterior third of the tongue (citric acid strip 16.5%) revealed no loss of taste. Clinical impression was of an isolated left spinal accessory neuropathy (SAN).

Nerve conduction studies revealed normal latencies but markedly reduced compound muscle action potential amplitude of the left trapezius muscle (0.4 mV versus 6 mV on the right) (Fig. 2).

Needle EMG examination of trapezius (upper part) and SCM on the left was difficult due to atrophy. There was no spontaneous activity. Chronic neurogenic remodeling of motor units action potentials and severely reduced recruitment were evident.

MRI of the brain and cervical spine (Fig. 3) revealed a small (5×8 mm), oblong mass within the subarachnoid space lateral to left side of medulla oblongata, in direct relation with the cisternal portion of the spinal accessory nerve (AN). The mass was isointense to brain parenchyma on axial T1 and hyperintense – on axial T2 sequences. It enhanced on T1 after gadolinium administration, revealing central hypointensity. It thus exhibited the typical radiological features of schwannoma. Mass effect was not seen.

Surgical removal of the tumor was decided against after consideration of the expected risks versus benefits. The patient was referred for physiotherapy and observation.
Intracisternal AN schwannomas are exceedingly rare. About 20 cases are reported or cited in current literature, mostly arising from the spinal root of the nerve (summary in Agrawal et al. [3]) [6,8,9]. These tumors tend to expand from the pontocerebellar angle towards the midline, about the posterior edge of the foramen magnum. They may grow large and exert mass effect. On MRI they appear as oval-shaped masses with regular contours, hypo- or isointense in T1, slightly hyperintense in T2, and enhancing homogeneously with contrast. Cystic degeneration may occur in larger tumors [6,8,9]. Patients present with unspecific combinations of symptoms such as neck pain [10], cerebellar signs, myelopathy or foramen magnum syndrome mostly resulting from the mass effect of the tumor; SAN features may be present or absent altogether [7,9]. Some cases manifest their onset by intracranial hypertension [10], mimic normal pressure hydrocephalus or fourth ventricle tumor [5,6]. Our case is the only one described presenting as isolated SAN, as the tumor was small, not exerting mass effect.

Operative strategy in intracisternal schwannomas is determined by the effects of the mass versus expected deficits. Tumors compromising the cerebellum, medulla, CSF circulation are resected even if the nerve has to be sacrificed. In other cases en bloc removal is feasible without damage to the nerve.

In our patient, surgical intervention was not offered as the small size of the lesion without mass effect after a long clinical course made a major neurosurgical procedure unwarranted. Besides, the patient had advanced muscle atrophy and the nerve appeared encased in the tumor, making an atraumatic resection unlikely and the chance for effective reinnervation minimal. Radiosurgery was discussed but its delayed results (complete neuropathy) are expected to be worse than an attempt for microsurgical resection. The patient remains under follow-up with clinical assessment and MRI scheduled over one year periods.

We do not have tissue diagnosis of the tumor. However, the long-standing and indolent clinical course, as well as the typical radiological appearance strongly supports the diagnosis of SA nerve schwannoma. The rare intracranial neurofibromas have a fusiform appearance and would be accompanied by the other stigmata of neurofibromatosis type 1, that were not observed in our patient. The origin of the tumour from the accessory nerve is in fact quite noticeable on the coronal postcontrast MRI as it follows and nicely “shapes” the course of the spinal accessory nerve (Fig. 3C). Besides, the absence of “foramen jugulare syndrome” features like tongue wasting, soft palate weakness, ageusia for the posterior third of the tongue also supports the origin of the mass from the cisternal part of the SA nerve.

Spinal accessory nerve schwannomas are extremely rare but should be considered in patients with isolated SAN of unclear etiology.

We have no conflict of interests regarding this study.

References


