Accessory nerve schwannoma

Lower cranial nerve schwannomas are rare and when present are located in the jugular foramen most commonly arise from the glossopharyngeal or vagal nerves with accessory nerve involvement being least common. The other locations in the accessory nerve course where schwannomas arise are extracranial beyond the jugular foramen in the cervical region and cisternal proximal to the jugular foramen. Both of these are very rare locations [1].

Epidemiology

Till 2013, a total of 28 cases of spinal accessory nerve schwannomas have been reported, among which four were reported in the craniocervical region, which extended from C1 or C2 to the vermis, with only three being located in the cervical region [2].

Classification

see Accessory nerve schwannoma classification.

Clinical features

Julow presented two cases of accessory nerve schwannomas and suggested that they could be divided into two groups, namely, intrajugular schwannomas that grow into the cisterna magna and intracisternal accessory nerve schwannoma that grow into the jugular foramen [3].

The symptoms of intrajugular schwannomas consist of various combinations of 5th to 12th cranial nerve palsy, cerebellar dysfunction, and myelopathy, depending on the degree of their extension [4].

Patients with intrajugular schwannomas usually present with a jugular foramen syndrome because the tumor is confined entirely within the foramen. Radiological diagnosis of jugular foramen tumors using CT or conventional x-rays is often difficult [5].

Patients with intracisternal accessory nerve schwannoma usually have an accessory nerve palsy, cerebellar syndrome, and/or myelopathy. Deficits of glossopharyngeal or vagal function have not been reported [6].

Diagnosis

Imaging studies help in the accurate diagnosis of schwannomas, with a reasonably good accuracy.

In some cases enlargement of the jugular foramen is evident on CT and this is a valuable diagnostic feature. MR has made diagnosis much easier even though it is still not possible to identify the nerve of origin [7] [8].

They usually have regular contours and a round or oval shape, lack edema, and enhance homogenously with contrast. Cystic degeneration is generally seen in large tumors and this may alter the otherwise uniform texture of the mass. [9].

These tumors are generally found against the posterior border of the foramen occipitalis, close to the midline [10] [11].

MRI

MRI scans are very helpful in identifying the tumours and their correlations with the surrounding vascular structures, muscles, and nerves [12].

This imaging provides information on the tumour size, location, extent and the surrounding anatomy, and it enables the surgical planning [13].

Treatment

A gross total resection remains the treatment of choice for these tumours, because they are radio resistant.

The other treatment option is sterotactic radiosurgery (SRS). SRS is gaining popularity, but it depends on many factors, which include the size and the location of the Schwanoma and the age of the patient. Whenever it is possible, SRS should be avoided in young patients Jayaraman M, Smirniotopoulos JG, Davis LM, Patel MR, Krasny RM. Imaging in cranial nerve schwannoma. Available at url. http://emedicine.medscape.com/article/336141-overview (Accessed on 4 April, 2019)

Outcome

Postoperative accessory nerve deficits also seem to depend on the degree of damage to the nerve. In the case reported by Caputi and colleagues, dissection was performed along the arachnoid interface, exposing the tumor attachment to the spinal accessory nerve, from which it was removed en bloc with no apparent damage caused to the nerve [14].

Their patient recovered completely without any impairment of neck movement. However, in another case in which the fibers of the accessory nerve entered the tumor and were coagulated and cut, the patient experienced shoulder muscle weakness and impairment of neck motion postoperatively [15].

So, in cases where the mass originates from a spinal accessory rootlet, the damage to the rootlet does not lead to the development of specific accessory nerve signs [16].

In the case of Juang et al., a spinal accessory rootlet near the spinal cord was the place of origin of the tumor and resulted in a lack of postoperative symptoms and signs [17].

Total removal of these tumors is recommended as recurrence is probably unavoidable if removal is incomplete [18].

Case reports

A 61-year-old man who presented with a 3-month history of dysmetria, ataxic gait, and frequent falls. Magnetic resonance imaging revealed a giant rim-enhancing cystic lesion at the right cerebellomedullary cistern, which markedly displaced the brainstem and caused a critical compression on surrounding structures and mild hydrocephalus. Even though the nature of this lesion was not clear, it received a radiological diagnosis of meningioma as first option. Surgery was performed through an extended far lateral retrosigmoid approach with C1 hemilaminectomy, with intraoperative neurophysiological monitoring. A near-total resection was achieved due to the adhesion of the lesion to the brainstem and to the cranial nerves VII, VIII, IX, X, XI, and XII. Intraoperatively, the tumor was found to arise from the accessory nerve. The histopathological analysis concluded with a final diagnosis of ancient schwannoma, a rare histological subtype characterized by degenerative changes, typical from long-standing tumors.

Very few cases of intracranial ancient schwannomas have been described. To the best of our knowledge, this is the first report of this extremely rare histological variant arising from the intracisternal component of the XI nerve. The rarity of this disease at this location may lead to preoperative misdiagnosis [19].

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