SCHWANNOMA OF THE CRANIOCERVICAL JUNCTION

Surgical approach of two cases

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ABSTRACT - We report two cases of craniocervical junction schwannomas with a special focus on the surgical approach. The patients underwent a far-lateral approach in the sitting position that facilitated the lesion removal. This report is meant to improve the understanding of this surgical technique as well as improve awareness of its usefulness for similar cases.

KEY WORDS: craniocervical junction, far-lateral approach, schwannoma.

The craniocervical junction is the possible site of several pathological processes, such as degenerative disease, tumors, aneurysms, vascular malformations and traumatic lesions¹². Salas et al., 1999³ reported 51 patients with tumors of this region. There were 24 meningiomas, 10 chordomas, 7 paragangliomas, 3 schwannomas, 2 chondrosarcomas, 2 neurofibromas, and one each of hemangiopericytoma, osteochondroma, and metastatic carcinoma. The schwannomas are benign tumors of the spinal and cranial nerves⁴⁵ and represent about 8% of all intracranial tumors⁶. Schwanommas located at the craniocervical region are rare³. The clinical presentation is variable, depending on its location and can be associated with neurofibromatosis⁴. The structures that must be considered when planning the surgical approach to the craniocervical junction are the medulla and the spinal cord, the lower cranial and upper spinal nerves, the vertebral artery and its branches, the veins and dural sinuses, and the ligaments and muscles uniting the atlas, axis and occipital bone²⁸.

A number of surgical access routes to this region are currently in use, including the transoral, transcervical, bifrontal transbasal, infratemporal, transpetrosal, retromastoid, the far-lateral approach and its transcondylar, supracondylar, and paracondylar extensions and combined supra- and infratentorial approaches¹⁸⁹. Lateral, far-lateral or extreme lateral approaches are most widely used to remove tumors anterior or lateral to the brain stem. Bony resection is important to gain free exposure of the area and to limit manipulation of the vascular and neural structures⁸¹⁰. Samii et al., 1996¹⁰ pointed out that the far-lateral or extreme lateral approaches are a variant of the lateral suboccipital approach with opening of the foramen magnum. They explained that the angle of approach and the working space for the surgeon are sufficient when a standard lateral suboccipital craniotomy is combined with drilling the posterior third of the occipital condyle.

We report the sequential steps of the far-lateral approach and the details of the microsurgical technique used in two cases of schwannoma of the craniocervical junction.
CASES

Case 1. A 43-year-old man developed a four-month history of progressive quadriparesis. There was a more recent history of headache. When he was admitted, he had been walking with difficulty and had weakness and dysesthesias of the upper extremities. He was submitted to a Gadolinium-enhanced, T1-weighted magnetic resonance imaging (MRI) which revealed a 2.2 X 3.8 cm lesion, located at the craniocervical junction, extending from the anterior portion of the foramen magnum to the right side up to the limit of the posterior arc and lateral mass of C1. There was no signal alteration in the brain stem, which would suggest anatomical lesion.

Case 2. A 59-year-old woman developed a six-month history of progressive quadriparesis and increasing difficulty in breathing and swallowing. When she was admitted she was no longer able to walk and had an episode of aspiration pneumonia thought to be secondary to a IX and X cranial nerves paresis. She was submitted to a Gadolinium-enhanced, T1-weighted MRI that showed a 3.5 X 1.7 cm lesion, located at the craniocervical junction, extending from the anterior portion of the foramen magnum to the right side over to the vertebral artery (Fig 1-A and B). The lesion was leading to compression of the upper cervical cord and lower medulla.

Both patients were submitted to the same procedure described here. The illustrations shown below were recorded from Case 2. Under general anesthesia the head was fixed with a Mayfield device. In the sitting position, using a hockey stick incision extending from the mastoid process and through the superior nuchal line as far as the midline, continuing inferiorly down to C4 vertebral level, a far-lateral approach was performed (Fig 1-C). The myocutaneous flap was elevated to expose the posterior craniocervical junction, deviating laterally on the subocciput toward the right side. Ipsilateral posterior arc of the C1 vertebrae was removed (Fig 1-D). The lesion originating from the right C1 root was totally removed (Fig 2-A). The C1 root ipsilateral to the tumor was cut in both cases because of the great extension of the lesion originated from that root.

The patients had a prompt recovery upon total removal of the tumor with marked improvement of their neurological status. Post-operatively they were submitted to rehabilitation and at a six-month follow up visit both patients were able to walk, talk, swallow and breathe without any difficulties. Pathology analysis reported the tumors to be schwannoma (Fig 2-B) in both cases. Postoperative Gadolinium-enhanced, T1-weighted MRI showed no residual tumor (Figs 2-C and 2-D).

DISCUSSION

This report describes a surgical approach to an anterolateral placed schwannoma of the craniocervical junction. In our two cases the patients were operated on from the sitting position.

Some authors prefer to place the patient in the supine position with the body rotated 45° to the opposite side. Others prefer the sitting position,
with the head flexed ventrally and rotated approximately 30° ipsilaterally and tilted slightly to the contralateral site of the surgical approach. The extreme-lateral craniocervical technique developed progressively as a more lateral approach to lesions in the midline on the anterior aspects of the foramen magnum.

The surgical anatomy determines the access gained through lateral approaches and the intradural structures are visualized through windows between 9th and 12th cranial nerves and C2. Lateral, far-lateral or extreme lateral approaches are most widely used to remove tumors anterior or lateral to the brainstem. Bony resection is important to gain free exposure of the area and to limit manipulation of the vascular and neural structures.

Samii et al., 1996, reported that the far-lateral or extreme lateral approach is a variant of the lateral suboccipital approach with opening of the foramen magnum. They demonstrated that the angle of approach and the working space for the surgeon are sufficient once a standard lateral suboccipital craniotomy is combined with drilling the posterior third of the occipital condyle.

Regarding the neural structures, the C1 root can be cut without any apparent adverse effects, as in these case reports.

The far-lateral approach with the patients in the sitting position was markedly important and facilitated the total removal of the schwannoma. This report is meant to increase awareness of the usefulness of this surgical technique when approaching these cases.

REFERENCES