Giant craniocervical junction schwannomas. Report of two clinical cases and literature review.

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Introduction

Schwannomas of the first cervical nerve root are extremely rare. These lesions constitute 2.9%–4% of all intracranial schwannomas[1]. Preserving and restoring the function of the lower CNs becomes the main goal when managing schwannomas in the craniocervical junction (CCJ)[3].

Various surgical approaches for dumbbell cervical tumors, such as a single posterolateral, anterolateral or lateral approach and combined posterior and anterior or anterolateral approaches in two stages, have been described, together with their respective advantages and disadvantages, as well as limitations of the exposed area[2]. The approach to these lesions is difficult because of the close proximity of the medulla and cervical spinal cord, lower cranial nerves, and the vertebral artery[7].

The clinical presentation is variable, depending on location, and can be associated with neurofibromatosis[2]. It is estimated that in approximately 2% of patients with NF1, symptomatic spinal nerve root tumors develop[4].

In this report we describe our surgery-related experience with two cases of giant dumbbell schwannoma in both non-NF1 and NF1 patients, treated via the lateral approach.

Case reports

Case 1: A 43-year-old male presented with a 3-month history of experiencing numbness and weakness of the left upper limb, he reported of objects falling off his left hand, progressive weakness of the lower limb. Neurological examination reviled spastic tetraparesis, Grade 4/5 on his upper extremity. Hoffman (+) MRC 46; CCI 2; KPS 90 He had decreased sensation on the left upper limb to light touch and pinprick. Reflexes were normoactive. On a head Magnetic Resonance Image (MRI), a dumbbell-shaped tumor was observed at the C1 - C2 level on the left side (Figure 1).

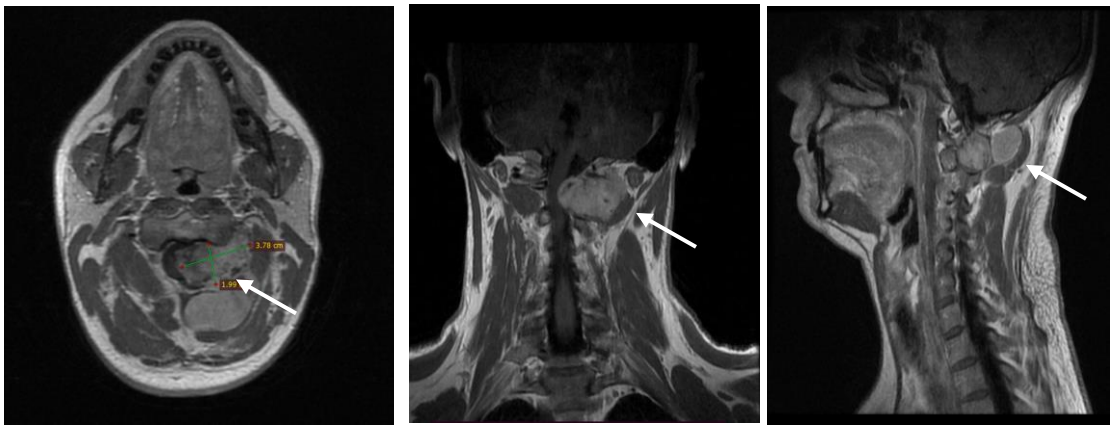

Figure 1: Huge Left Antero-lateral C1-C2 schwannoma

As part of the vascular evaluation, selective Digital Subtraction Angiography was performed, and dominance of right Vertebral Artery (VA) was noted, as well as blush tumor from the third segment of left VA. No neurological changes were observed during Balloon Occlusion test (BOT) (Figure 2).

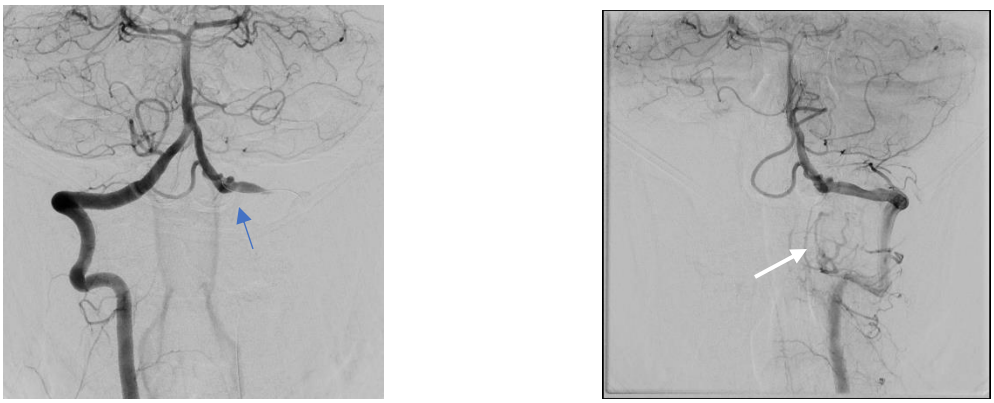

Figure 2: Diagnostic DSA. Black arrow shows BOT; White arrow shows tumor flushing

Surgery was performed in Concord position, and through a midline incision and a left C1 laminectomy, the extradural encapsulated tumor was found to be arising from the left C2 dorsal root. Total gross total resection (GTR) was achieved through decompression with cusa. (Figure 3,4) Histological findings confirmed the diagnosis of the schwannoma. Postoperative neurological evaluation showed a significant improvement of motor force and fine touch sensibility.

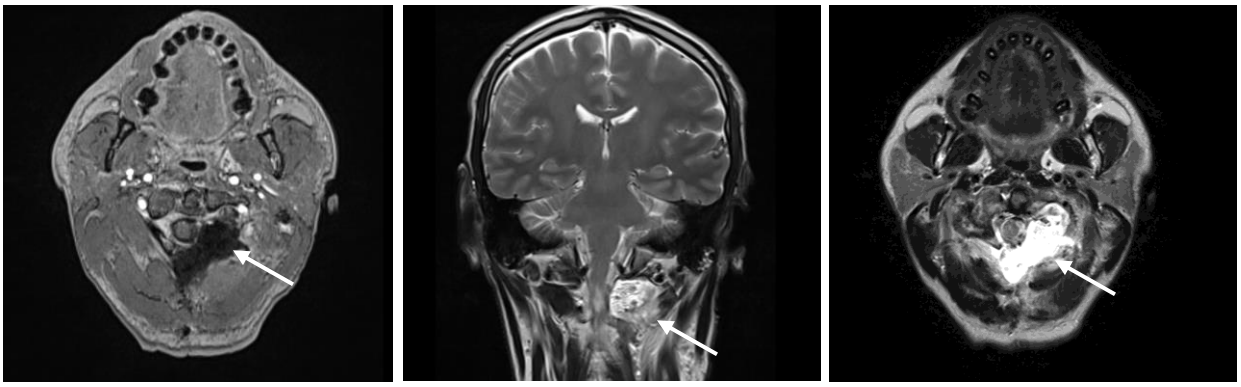

Figure 3: Immediate Post operative MRI confirmed GTR.

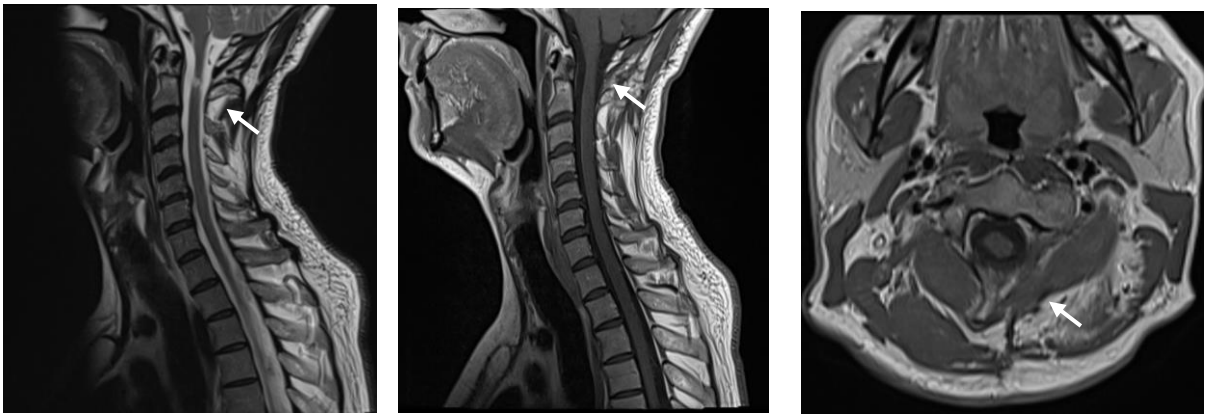

Figure 4: Nine months follow up MRI, GTR was confirmed

Case 2: A 26-year-old male, with NF1, was referred to our department for spastic tetraparesis. Ten years before, he underwent surgery for a brainstem lesion; Pilocytic Astrocytoma (PA), after was treated with RT and was considered cured. The clinical and radiological follow-up for PA five years after, identified an asymptomatic spinal lesion in the level of C1 with extension in the cranio-cervical junction, and yearly follow up clinical and MRI was recommended and showed increased tumor size, with tetraparesis.(Figure 5).

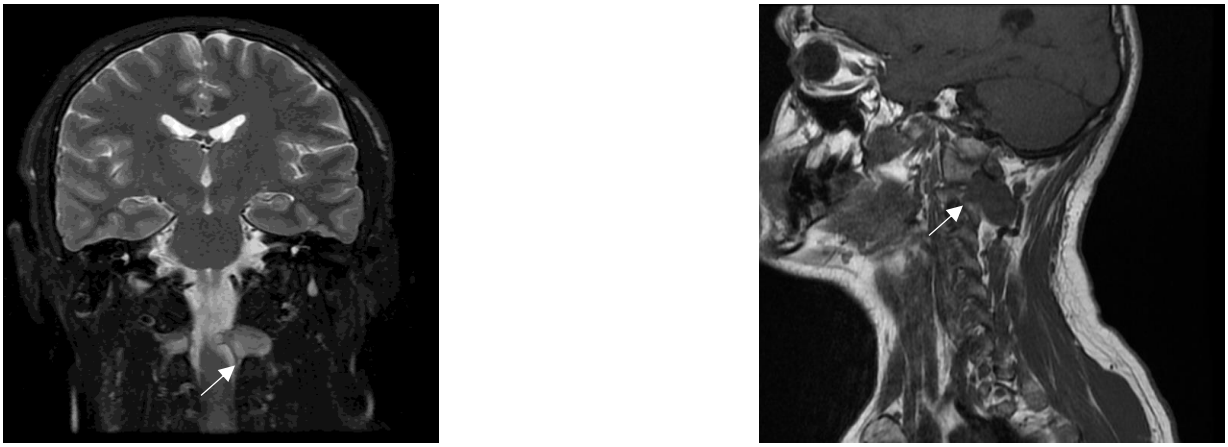

Figure 5: White arrows shows the spinal lesion in the level of C1 with extension in the CCJ, MRI performed at age 21

He began to experience progressive weakness of his right upper limb, 6 months before being admitted in our clinic (Figure 6). A Brain MRI showed an increase in tumor size.

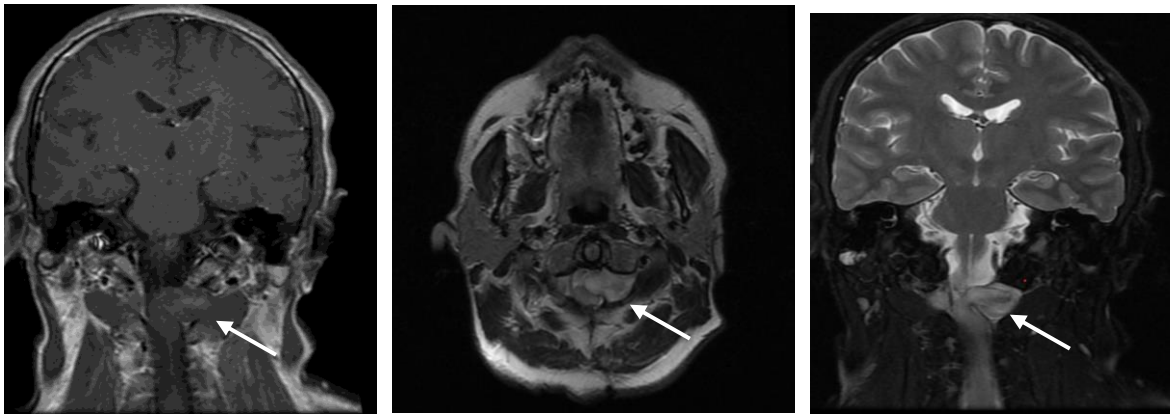

Figure 6: Brain MRI performed on admission before surgery

Neurological examination revealed residual right peripheral facial nerve palsy, right lingual hemi atrophy, trapezoid muscle atrophy and gait ataxia were due to the previous PA considered cured. There was no evident swallowing difficulty. Recent severe spastic tetraparesis; MRC54; CCI 5; KPS 85.

The patient underwent surgery on March 8th, 2023 on sitting position. Through a midline incision, previous C1 laminectomy and extended C2 hemilaminectomy the extra dural lesion was seen in the left C1 nerve root with anterior craniocervical junction extension, gray, encapsulated, hard even with cusa, hemorrhagic and non-aspirable. GTR was archived (Figure 7). HP confirmed Neurofibroma. Post-operative period was uneventful, with a important improvement of motor force and his tetraparesis

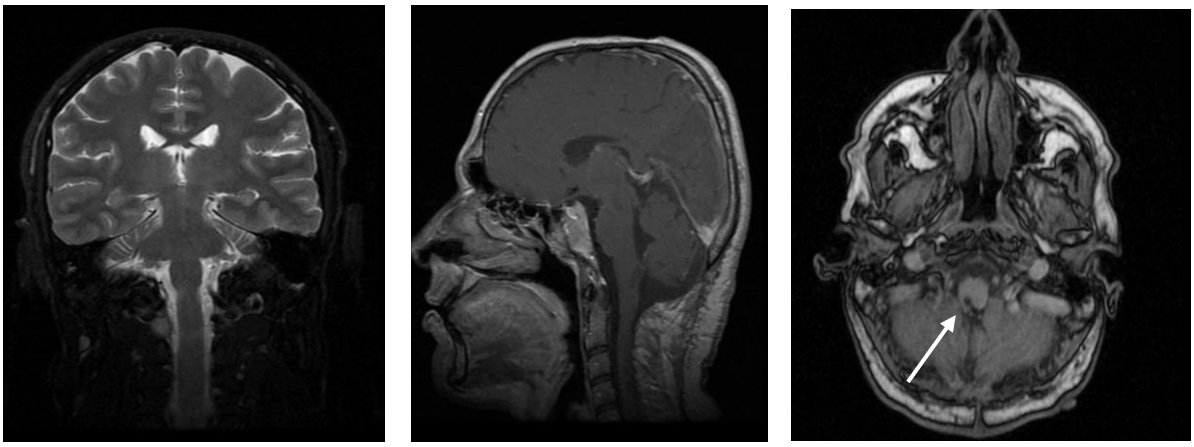

Figure 7: Follow up MRI. White arrow shows post-operative encephalomalacia from the first PA intervention.

Discussion:

Schwannomas in the CCJ are rare skull base neoplasms and are usually described separately in series on surgical approaches, series on tumors involving the jugular foramen and lower cranial nerves series [1,3].

In the largest series of 42 patients with C-1 and C-2 schwannomas analyzed by George and Lot[5], 83% had motor deficits and 60% had sensory impairments.

Schwannomas arising from C1 are very rare. C1 controls the function of the rectus capitis lateralis and, historically, was not thought to have a sensory component. In our cases the patients were both presented with spastic tetraparesis. In 1988, Guidetti and Spallone[4] reported three cases of C-1 tumors, one of which was "hourglass" shaped and is the first and only clearly distinguished intradural/extradural C1 schwannoma reported.

A solitary case report of an extradural C1 schwannoma resulting in intermittent vertebral artery compression with symptoms was published by Kalavakonda et al.[6] in 2000. Three of 15

cases were C1 schwannomas, according to a 2003 study by Parlato et al.[7] on the treatment of benign craniovertebral junction tumors; however, the site of the tumors was not specified.

According to George and Lot [5] the treatment for craniocervical schwannomas depends on the tumor's extension. Several surgical techniques are available for the CCJ. For lesions inside or extending to the anterolateral CCJ, the far-lateral or posterolateral approach, the extreme lateral or anterolateral approach, and its variations offer superior surgical access. When it comes to C-1 and C-2 neurinomas, these methods allow access to any extension of the tumor, particularly the extradural component near the vertebral artery. As was done in the present cases, a gross total resection of schwannomas is the goal.

Recently, Neurosurgery in its operative editions showed a surgical video of a C1-C2 cervical dumbbell schwannoma with ventral extension and dorsal spinal cord compression[8].

Conclusion: We present two clinical cases of giant CCJ schwannomas, where GTR was achieved, which is the target goal, with significant neurological improvements. The approach to craniocervical schwannomas must be chosen based on the extension of the tumor and its relationship to the dura mater and neurovascular structures. High cervical dumbbell schwannoma should be radically resected while preserving and improving preoperative neurological function.

Keywords: Schwannoma, craniocervical junction CCJ, C1 first cervical nerve root, neurofibromatosis, dumbbell shape

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