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Case Series

Neurofibroma of C2 Root: A Series of Cases

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Abstract

Intra and extradural neurofibromas of C2 root is a challenging pathology, usually with indolent evolution, that is why the onset symptoms are neurologic deficit like upper cervical myelopathy symptoms and explains the late diagnosis of giant tumors with severe spinal cord compression with involvement and distortion of great vessels anatomy. These facts turn the surgeries more difficult, a need of dural reconstruction was necessary in all cases and the follow-up must be very close, because of possibility of CSF fistula. It is a series of three cases of neurofibromas, two of them are giant dumbbell-shaped C2 neurofibromas and the third one is a case of neurofibromatosis type 1 of C2 roots bilaterally, C3 root unilateral with unusual postoperative course.

Keywords: C2 root; Dumbbell-shaped tumor; Neurofibroma

Abbreviations: NF1: Neurofibromatosis Type 1; WHO: World Health Organization; VA: Vertebral Artery; CSF: Cerebral Spinal Fluid; ACDF: Anterior Cervical Decompression And Fusion.

Introduction

Neurofibromas arise mainly from the C2 root [1,2]. C2 neurofibromas can grow to a large size while remaining asymptomatic and extend via the intervertebral foramina to achieve a dumbbell shape [1,2]. C2 neurofibroma has specific characteristics, such as multiplicity, a dumbbell shape, a relationship with the vertebral artery and particular associated surgical problems [1-4]. Peripheral nerve sheath tumors arising from the second cervical nerve root are relatively common and constitute approximately 15% of all spinal peripheral nerve sheath tumors [1]. Neurofibromas account for about 2-5% of all primary spinal tumors [5-7], including neurofibromatosis type 1 (NF1, Von Recklinghausen's disease) [8]. Neurofibromas have a high prevalence at the C2 level in patients with and without NF1 [1,9-12].

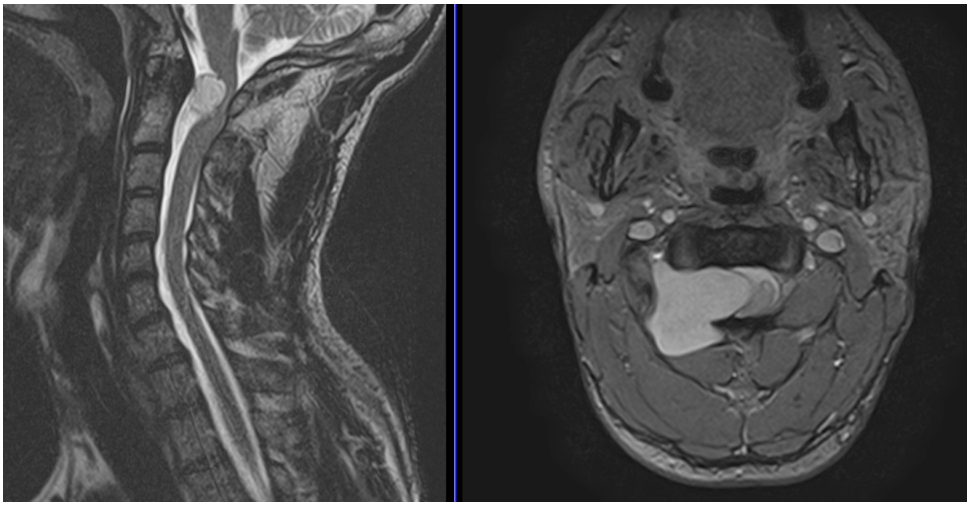
Surgical Procedure

The posterior approach was selected in all patients, using a midline incision, exposing occiput, C1, C2, C3 posterior elements. A C1 laminectomy and C2 hemilaminectomy was performed and extended laterally to expose the tumor and the dural tube. The posterior dural wall of the tumor was coagulated and incised, and intra tumoral debulking was performed. The dura was cut in a T shape, longitudinal and the perpendicular incision to the tumor side. The involved C2 root was resected together with the tumor, avoiding vascular structures. The venous bleeding was the main sources of hemorrhage during surgery: careful coagulation and the use of hemostatic foam associated with gentle compression reduces the bleeding. Reconstruction of dural defect was performed if needed. Fixations and fusions were not necessary. According to Atul Goel, C2 neurinomas arise in the region of the C2 ganglion, and despite the fact that some achieve a large size, they remain confined within the dura. Radical tumor resection can be achieved by working within the layers of the dural cover. Bone removal and opening of spinal dura for tumor exposure and resection can be avoided [13]. Unfortunately, in our cases it was not possible, we considered dural opening safer and we recommend the nuchal

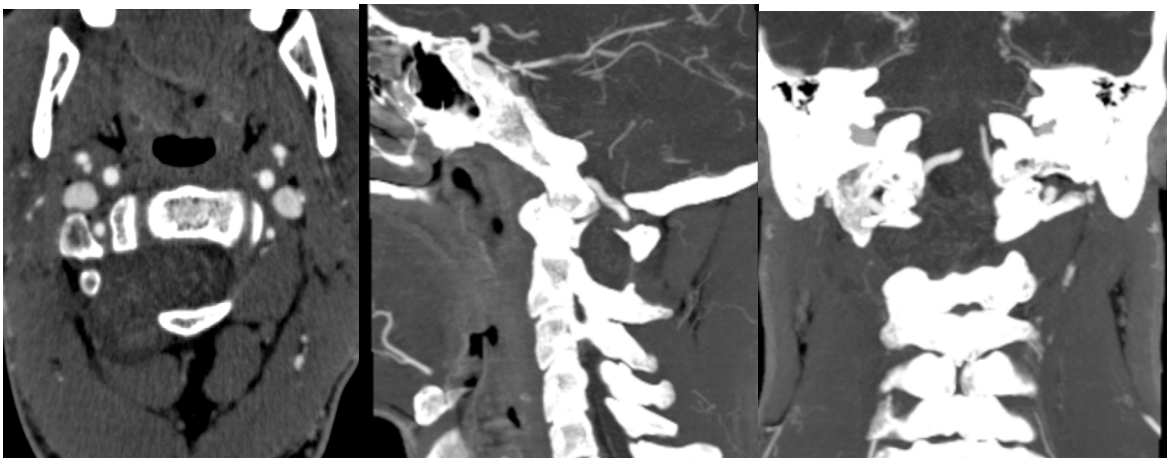
ligament as good plastic material for dural reconstruction. Each patient has sufficient size of ligament, it is easy to choose a good layer, and it is strong and resistant, there are no difficulty to suture it.

Case 1

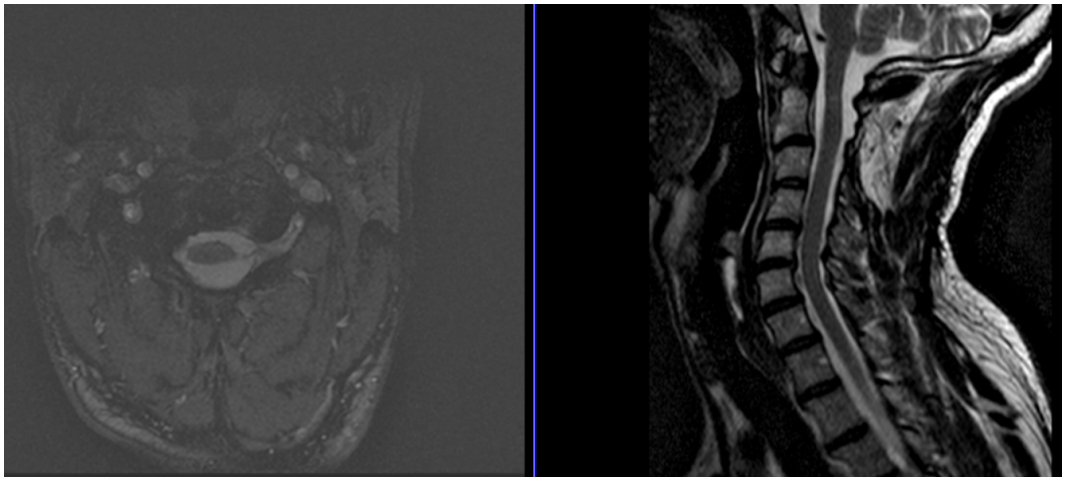
42 years old man, presented with spastic tetra paresis at the admission time, but still maintaining autonomy (Karnofsky 80). First sign was right hand weakness, rapidly progressing to other limbs. No history of previous diseases, no smoking. MRI revealed severe spinal cord compression by dumbbell-shaped extramedullary, intra, extradural tumor (Figure 1A). For preoperative planning and in order to study the vascular anatomy, contrast enhanced CT scan was performed, that showed impingement of VA upwards (Figure 1B). The patient was selected for removal of the tumor through median posterior approach, as described previously. At the end of the surgery, the reconstruction of the dural defect was performed. As material for reconstruction was used the nuchal ligament, previously harvested during the approach. Postoperative period was uneventful, with excellent neurological and functional recovery. Pathologic anatomy was neurofibroma grade I (WHO), Three-year postoperative MRI confirmed no sign of residual tumor, right C2 root amputation, slight intramedullary T2 hypersignal (Figure 1C).



A: Preoperative MRI revealing severe spinal cord compression by dumbbell-shaped extramedullary, intra, extradural tumor.



B: Contrast enhanced CT scan, revealing right C1-C2 foramina enlargement and relationship with vertebral artery.



C: Postoperative MRI, identifying good spine cord decompression, right C2 root amputation, slight intramedullary hypersignal.

Figure 1: Right sided dumbbell-shaped neurofibroma of the right C2 root.

Case 2

31 years old African man with a two-week history of paresthesia of inferior limbs with rapid progression to tetra paresis and subsequent inferior paraplegia (Karnofsky 50). CT scan revealed right-sided, dumbbell-shaped C2 root tumor with severe spine cord compression (Figure 2A) and C1 arch erosion (Figure 2B). He was operated on, the tumor was removed totally through posterior medial approach as well, the tumor was growing from C2 root, at the end of the surgery also a dural reconstruction was needed (Figure 2C). The patient's postoperative recovery was good, the only sequel remained is right hand proximal weakness, still recovering. Postoperative CT scan excluded local complication.

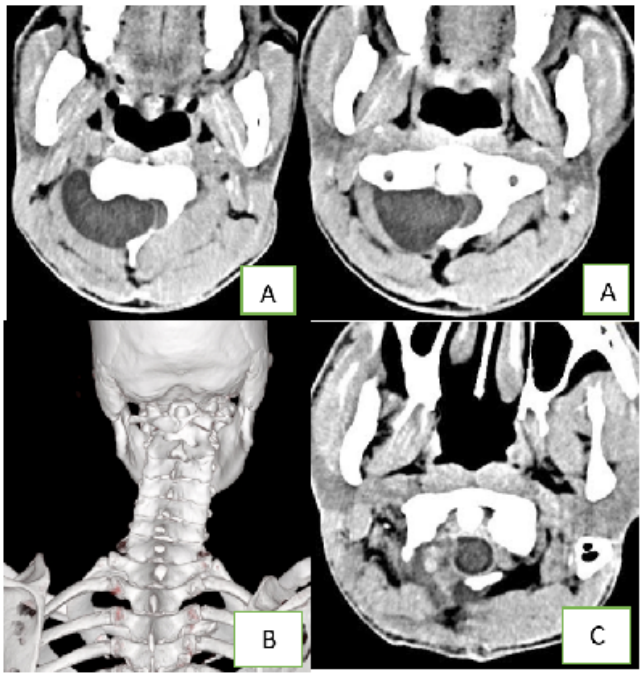


Figure 2: A. Preoperative CT scan; B. CT scan reconstruction revealing C1 and C2 lamina erosion C. Postoperative CT scan

Case 3

34 years old lady, with NF1 diagnosed since the age of 18 years, admitted because of unstable gait (Karnofsky 80). MRI revealed bilateral C2 roots tumors with severe bilateral compression of the spinal cord, spinal cord is squeezed in between C2 bilateral tumors and C3 root left sided tumor (Figure 3A). She was operated, it was performed removal of tumors with extra, intradural growth through posterior approach through C2 laminectomia with subsequent dural reconstruction also with nuchal ligament harvested during the approach. In the postoperative period she developed a huge, tense, subaponeurotic CSF collection (Figure 2B), solved only after inserting de ventriculoperitoneal shunt with the use of navigation, because of small ventricles (Figure 3C). Two years after the neurofibroma removal she was bothered by neck pain and fixed position in anterior flexion of the head. X-Ray, MRI diagnosed post laminectomy regional kyphosis C2-C3 with anterior luxation. She was operated: circumferential arthrodesis (ACDF C3-C4, C4-C5, posterior mass lateral fixation C2-C6 on the right side, C2, C3, C5, C6 on the left side) with good recovery and pain relief (Figure 3D). Three years after she started to be bothered by upper limb paresthesia and MRI revealed initial cervical syringomyelia. One year later her gait was progressively more unstable and MRI showed significant syringomyelia progression (Figure 3E) and next surgery was performed: siringopleural shunt insertion trough upper thoracic laminectomy Th1, Th2 and unilateral intralaminar fixation C7-Th3 was performed at the end of the surgery (Figure 3F). The decision to fix C7-Th3 with intralaminar unilateral screws was dictated by previous experience with cervical instability and weak muscles of the patient. The postoperative period was uneventful, the gait improved significantly.

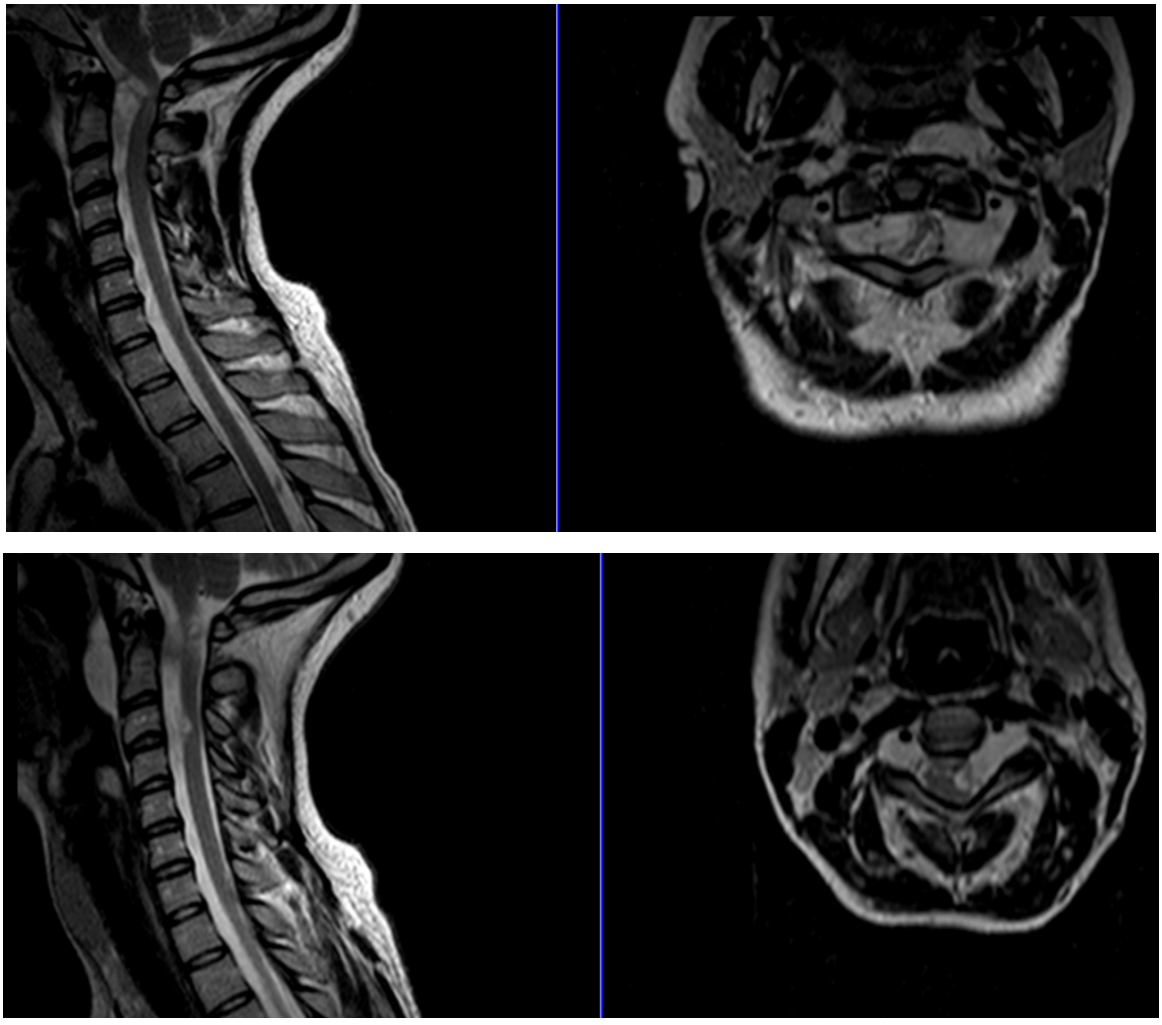


Figure 3A: MRI revealing bilateral C2 root tumor with severe bilateral spine cord compression and C3 unilateral tumor.

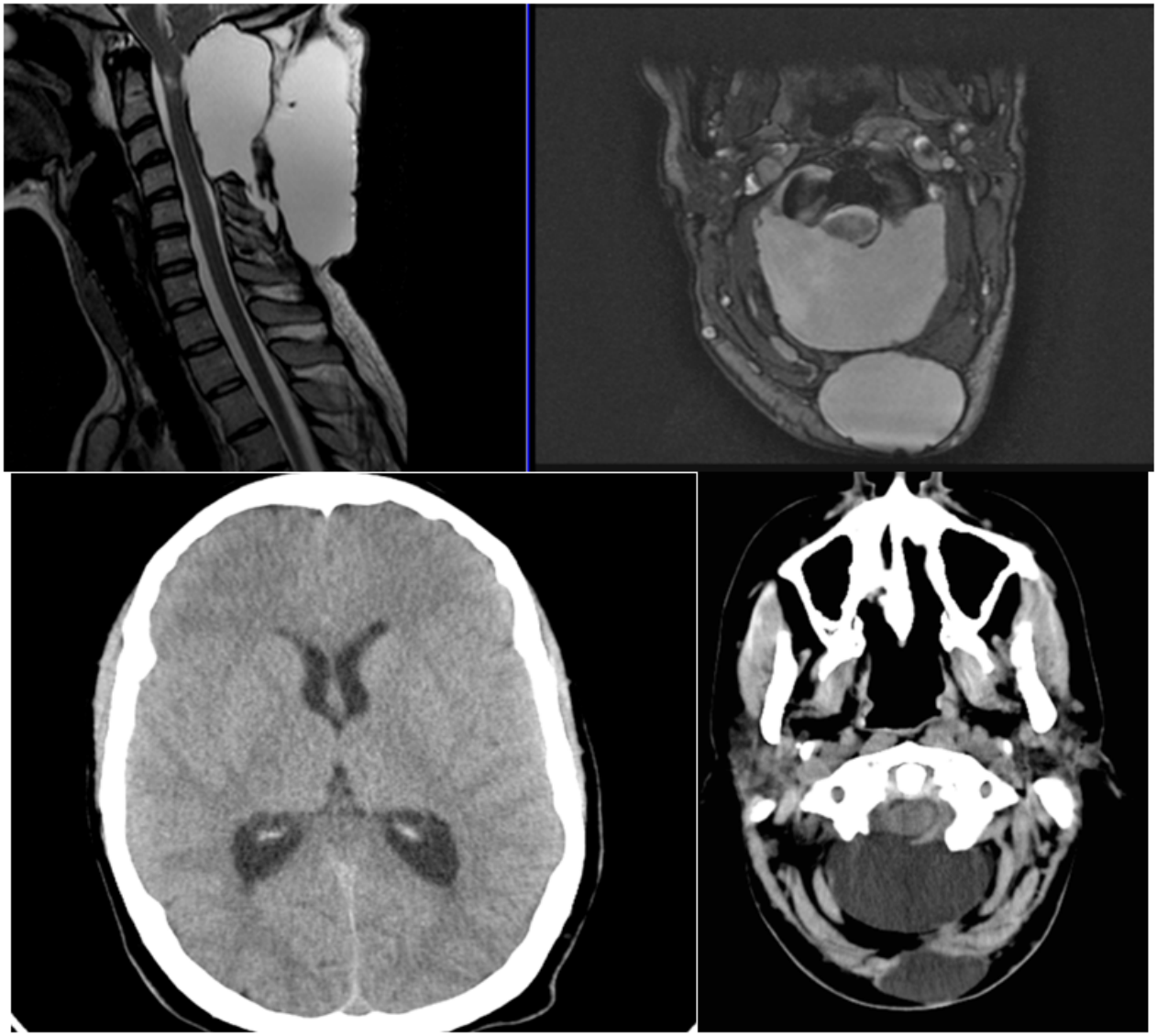


Figure 3B: MRI and CT scan showing huge, postoperative pseudomeningocele, no signs of hydrocephalus, no signs of residual tumors.

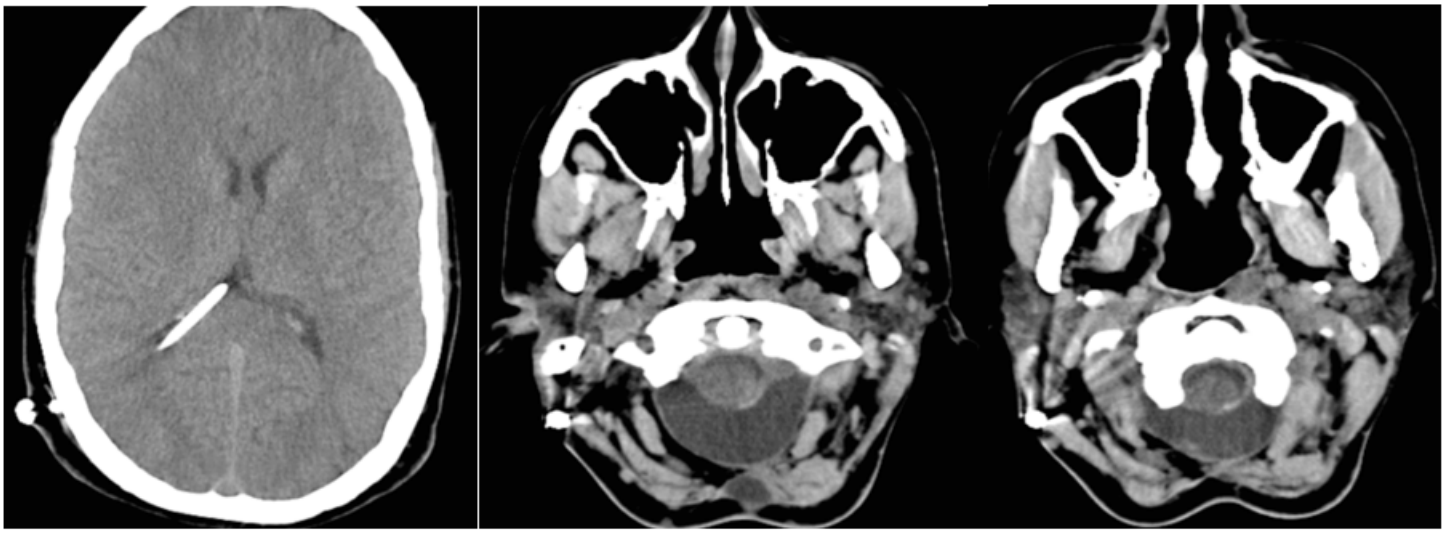


Figure 3C: Serial CT scan after ventriculoperitoneal shunting, meningocele diminished on serial CT scan.

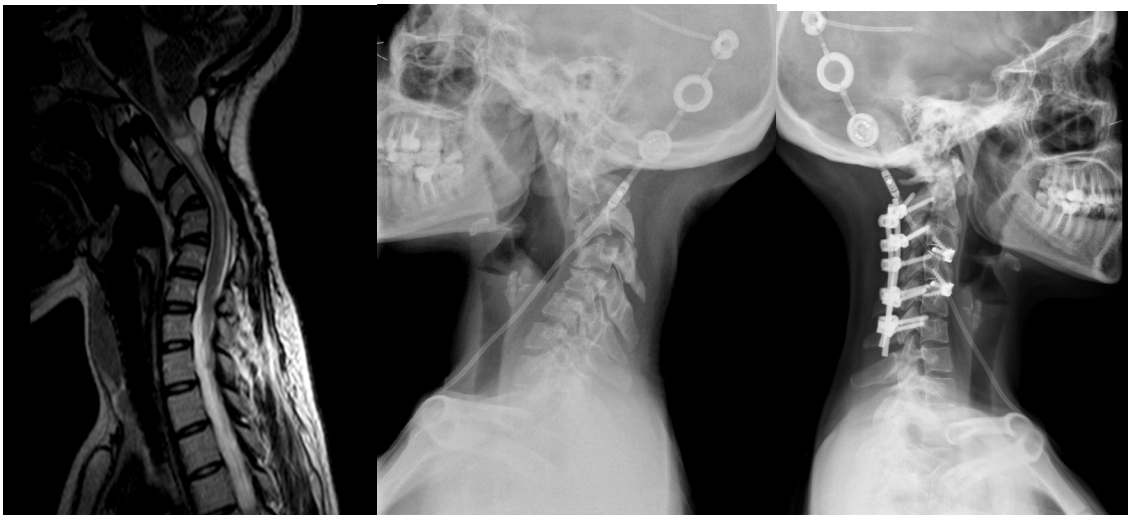


Figure 3D: MRI revealing upper cervical post laminectomy regional kyphosis, syringomyelia, no signs of meningocele. X-Ray before and after 360-degree arthrodesis.

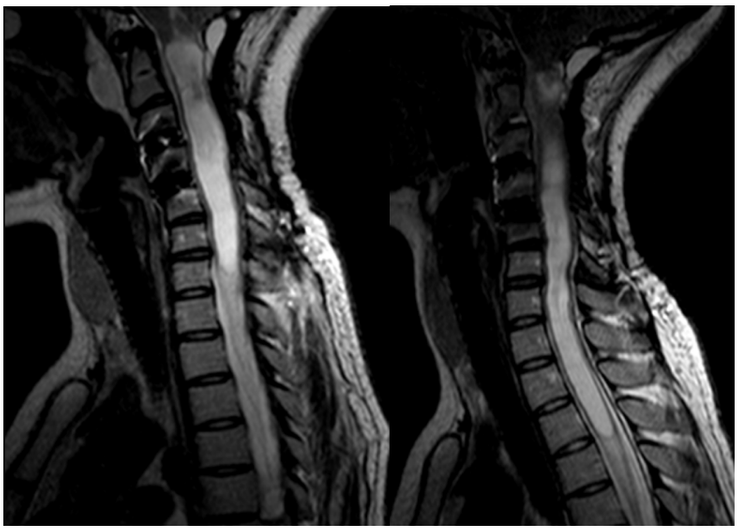


Figure 3E: Serial MRI with 1-year time difference, showing syringomyelia progression to upper thoracic spine cord.

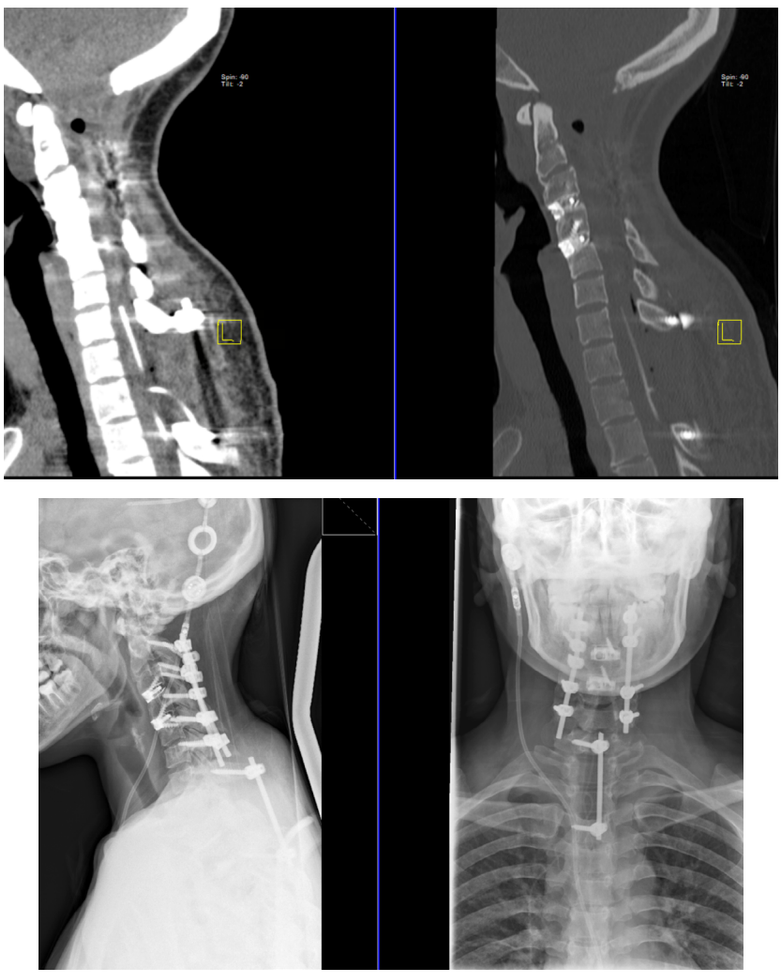


Figure 3F: CT scan, X-Ray after syringopleural shunting.

Conclusion

The patients in this series were very symptomatic with good recovery. The surgical procedures were challenging, total tumor removal was possible, the reconstruction of dural defects was needed in all cases. In the case of NF1, the postoperative period was practically just the follow up of complications and addressing them. This fact stresses the importance of meticulous preoperative planning and close follow up. Postoperative quality of life was better than preoperative in all the cases. The purpose of the surgical treatment is the decompression of the spinal cord, tumor removal and prophylaxis of CSF fistula by closing the dural defects.

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