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SPINAL SURGERY COMPLICATED WITH SOME CONGENITAL AND CHILDHOOD NEURAL DISEASES

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SUMMARY

Objective: This study was a retrospective clinical research based on challenging cases with clinical and radiological data analysis, and the review of literature. The aim of this study was to determine the factors affecting spinal deformity treatment in patients with diseases of the central nervous system following neurosurgical management.

Material and Methods: Three sample cases presented and the relevant literature were interpreted by the lights of these analyses. Patients are consisted of 20 and 44 years old females and 9 years old boy with childhood neural diseases. These accompanying diseases were detected during evaluation for spinal surgery by neurologic examination and imaging modalities.

Results: All patients have been doing neurologically well with almost no deficits and manageable pain problems. A 44-year-old women who had CSF diversion operation previously, was resolved her arachnoiditis by extending the cervical stenosis operation. Neurofibromas that complicated kyphosis operation of a 9-year-old boy were removed. And tethered cord of a 20-year-old girl was untethered during scoliosis operation. All patient's neurological status is well after operations and follow-ups.

Conclusions: Surgical treatment of patients with spinal diseases may become a quite challenging case when they have congenital anomalies or CNS related problems. Staging surgical approach should be kept in mind for these kinds of patients. Besides, growing of spine and spinal cord at different rates shouldn't be forgotten for children. The whole spinal axis must be kept under lifetime control by some regular intervals for spine patients with additional congenital or CNS anomalies.

Key Words: Congenital spinal diseases, Congenital neurologic diseases, Deformity, Degenerative spinal diseases, Intraspinal anomalies, Neurofibromatosis-I, Spinal operations

Level of Evidence: Retrospective clinical trial based on a case study, level IV

INTRODUCTION

Spinal surgery has been doing in huge amount all around the world. Some diseases of the brain and spinal column are seen frequently at younger ages, children or early childhood. Congenital anomalies and diseases of the craniospinal system are expected to affect these groups of patients more than those of the older patients, and may also be associated with an increased risk for subsequent neurological compromise during spinal surgeries. Those pediatric patients are already vulnerable to any kind of surgery; associated anomalies of those patients may increase the risk of surgery. At the same time, it may be encounter some technical difficulties during surgery in older age group of patients that have some associated congenital anomalies [(1)].

The aim of the study is to reveal that how congenital spinal anomalies effect the outcomes of spinal surgery, to discuss decision making process of three sample cases. Besides, a review of the literature was executed using PubMed database. The objective of this research was to find any publication about that how congenital spinal anomalies influence the decision, and prognosis of spine surgery.

MATERIALS AND METHODS

The authors presented three of their patients who needed excessive evaluation and investigation with a lot

of preoperative tests and studies before making a decision of patients' treatment plans. Three of our patients are discussed in this manuscript. Twenty and 44 years old females and a 9-year-old boy with some spinal disorders complicated with childhood neurologic diseases are evaluated and discussed. Neurological diseases of the patients need extra surgical effort, longer surgery time or extension of operation. These associated diseases are explained in sample cases. This study is a retrospectively case analysis, so it needn't any ethics committee approval.

Sample Cases:

Case 1:

The patient was a 44-year-old woman admitted to our clinic in 2007. Her complaints were headache and severe, increasing pain in her right arm for almost 4 years. When she was 22 years old, she had a ventriculoperitoneal (V/P) shunt operation in another clinic with the diagnosis of hydrocephalus. At the night of operation a left hemiplegia developed with loss of consciousness. She had been hospitalized more than 2 months with one month in intensive care unit (ICU). In a year, her hemiplegia had been resolved by an intense physical therapy and rehabilitation. Since then she has some complaints such as moderate to severe headache, pain in her neck and especially right arms. She connected with many doctors and she had a lot of cranial magnetic resonance imagings (MRIs) including cerebrospinal fluid (CSF)-flow sequences. She was done lumbar puncture to measure the CSF pressure in another hospital few years ago, and she was hospitalized due to severe headache, and vomiting. Her last MRI revealed a right parieto-occipital calcified epidural/subdural hematoma, moderate enlargement of lateral and third ventricles with ventricular catheter on the right ventricle (Figure-1), and CSF flow in the aqueductus Silvius was in normal ranges and patterns. Her neurological examination disclosed that monoparesis of the right arm, hypoesthesia on C4,5,6,7 dermatomes. Deep tendon reflexes of all extremities were hyperactive. Hoffmann reflex was bilaterally positive, Achilles clonus and, Babinsky reflex were positive on the right side. Right shoulder impingement was also detected during abduction and external rotation. A cervical MRI was performed showing C3-4, C4-5 and C5-6 severe spondylosis with C4-T1 arachnoid cysts compressing the spinal cord especially on the right side (Figure-2). Further evaluation and tests were performed such as electromyoneurography (EMG), somatosensory evoked potentials (SSEP), and motor evoked potentials (MEP) showing myelopathy and multilevel radiculopathies. A roentgenogram showed distal catheter breakage of V/P shunt (Figure-3). Ophthalmological examination revealed that bilaterally mild optic atrophy. V/P shunt reservoir tapping was done. The CSF pressure was low, but spontaneous CSF flow through the needle was seen. Because of the possibility of partially obstruction of ventricular catheter, it couldn't be a reliable index of measuring CSF pressure for this patient. So, we decided to explore the arachnoid cyst first, because her complaints seemed mainly related with cervical compression. We explained to the patient and her family underlining that there was a risk of herniation during laminectomy and cervical dural opening. A written consent was taken for that the patient may have needed exploration of her shunt, and/or insertion of an external ventricular drainage system. C4 and C5 total laminectomy and C6 superior hemilaminectomy were performed. During the operation it was seen that there was no a single cyst, it was like arachnoiditis, with a lot of membranes firmly attached to the spinal cord and the dura forming honeycomb appearance. It was thought that arachnoiditis was caused by hemorrhages from V/P shunt insertion 22 years ago. After operation, most of her complaints were resolved mostly due to surgical decompression and duroplasty enlarging medullary canal.

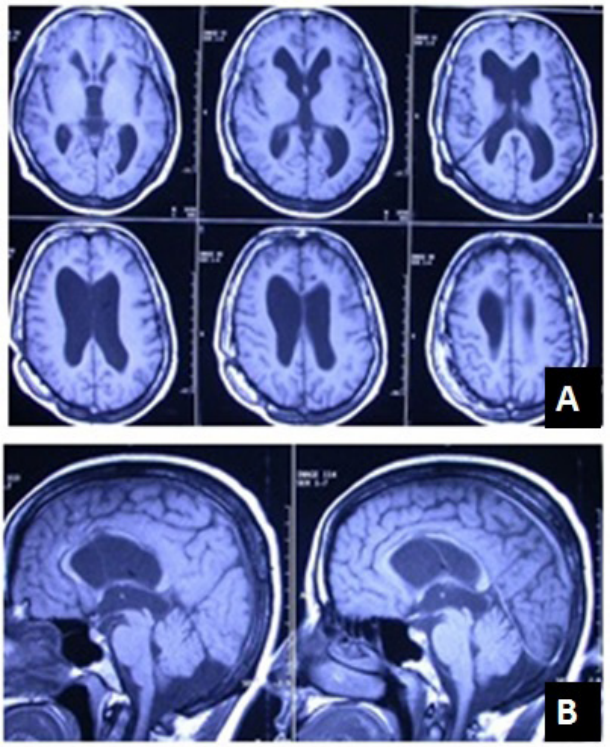

Figure-1. T1 weighted axial **(A)** and sagittal **(B)** MR images showing right parieto-occipital calcified epidural/subdural hematoma, moderate enlargement of lateral and third ventricles with ventricular catheter on the right ventricle, and normal-sized fourth ventricle.

Since the first operation she has been under follow-up routinely. Almost for 5 years she has been quite well. In 2013 she hospitalized again with the similar complaints. Her neurological examination was revealed that right-sided hemihypoesthesia, and hemiparesis (both symptoms heavy on the right arm), hyperactive deep tendon reflexes, bilateral positive Hoffman

reflex, bilateral Achilles clonus, and Babinsky reflex. There was still impingement syndrome of the right shoulder. Cranial and cervical MRIs were performed again. There was no new lesion in the cranial MRI; and the cervical MRI showed that the spondylosis and the cystic enlargement continued (Figure-4). 3D CSF flow MRI (1) was performed and showed CSF flow through the aqueductus Sylvius, Foramen of Magendie, and spinal subarachnoid spaces (Figure-5). The patient underwent operation again. Under intraoperative neuromonitoring (IONM), three level anterior cervical discectomies, corpectomies (C3-4,C4-5,C5-6), interbody cage and bony fusion with DBM and instrumentation with anterior cervical plate were performed. Then the patient was turned to prone position. Bilateral C4,5,6 lateral mass screws were inserted, C3,6,7 total laminectomies, and T1 superior hemilaminectomy were done. Dura was opened, it seemed that the arachnoiditic appearance was resolved, and there were fewer adhesions. The thick arachnoid band at superior T1 level (Figure-6) was opened and the cyst marsupialization was done. After dural closure, the posterior instrumentation and bony fusion were completed (Figure-7). At the 6th month of follow-up with physical therapy and rehabilitation program, her complaints resolved very well. She had no hemiparesis, and decreasing hemihypoesthesia. Babinsky, and Achilles clonus were disappeared on her left side. She is still under follow-up.

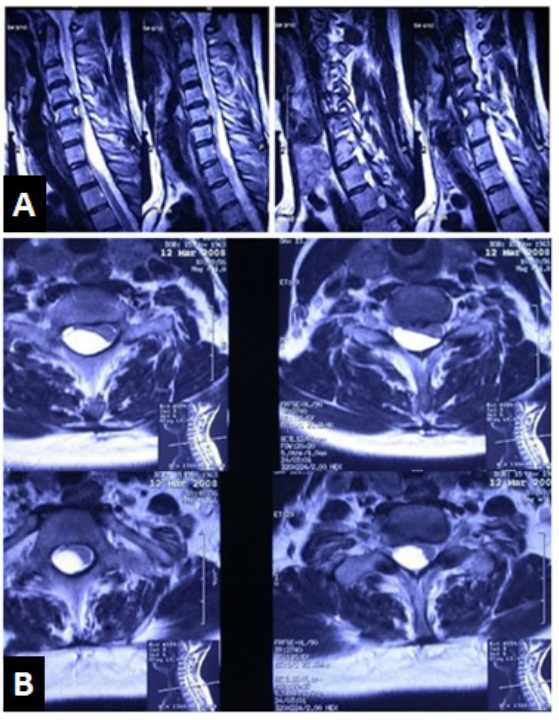

Figure-2. T2 weighted sagittal **(A)** and axial **(B)** cervical MRI showing C3-4, C4-5 and C5-6 severe spondylosis with osteophytic supur formation causing foraminal narrowing, and C4-T1 arachnoid cysts compressing the spinal cord especially on the right side.

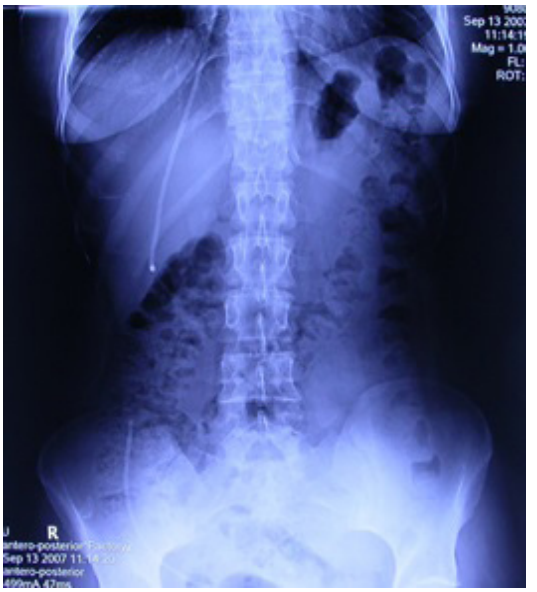

Figure-3. PA abdominal x-ray showed distal catheter breakage of V/P shunt

Case 2:

The patient was a 9 year-old boy applied to our clinic in 2011. He was diagnosed as Neurofibromatosis-1 (NF-1) three years ago.

Cranial and spinal MRIs for routine control showed left C4-5 intradural neurofibroma, and it was doubled in size during 9 months (Fig-8).

He had also congenital cervical kyphosis 20 degree (Fig-9). Neurological examination revealed that left arm abduction was 4/5 in muscle strength. He had huge (10x15 cm) subcutaneous plexiform neurofibroma at the left side anterior cervical area. He underwent operation.

Left C4 hemilaminectomy and C5 sup hemilaminectomy were done. Midline structures and facet joint were kept intact.

Gross total tumor excision was achieved with control cervical MRI showing no residual tumor. Philadelphia type semirigid collar was recommended in order to reduce pain for short period (3-5 days). Early isometric cervical exercises were thought to him, and his mother. At 6th month he came for control. His mother said that he has kept cervical collar 3-4 weeks, he didn't do exercises. His complaint was neck pain.

His neurological examination was intact. His cervical X-ray showed that his cervical kyphosis increased to 68 degree (Fig-10). He underwent intensive physical therapy and rehabilitation program. 3 months later his pain was resolved but the degree of kyphosis didn't change.

His control cervical MRI showed that no residual or recurrent intradural tumor with cord compression at C4 level.

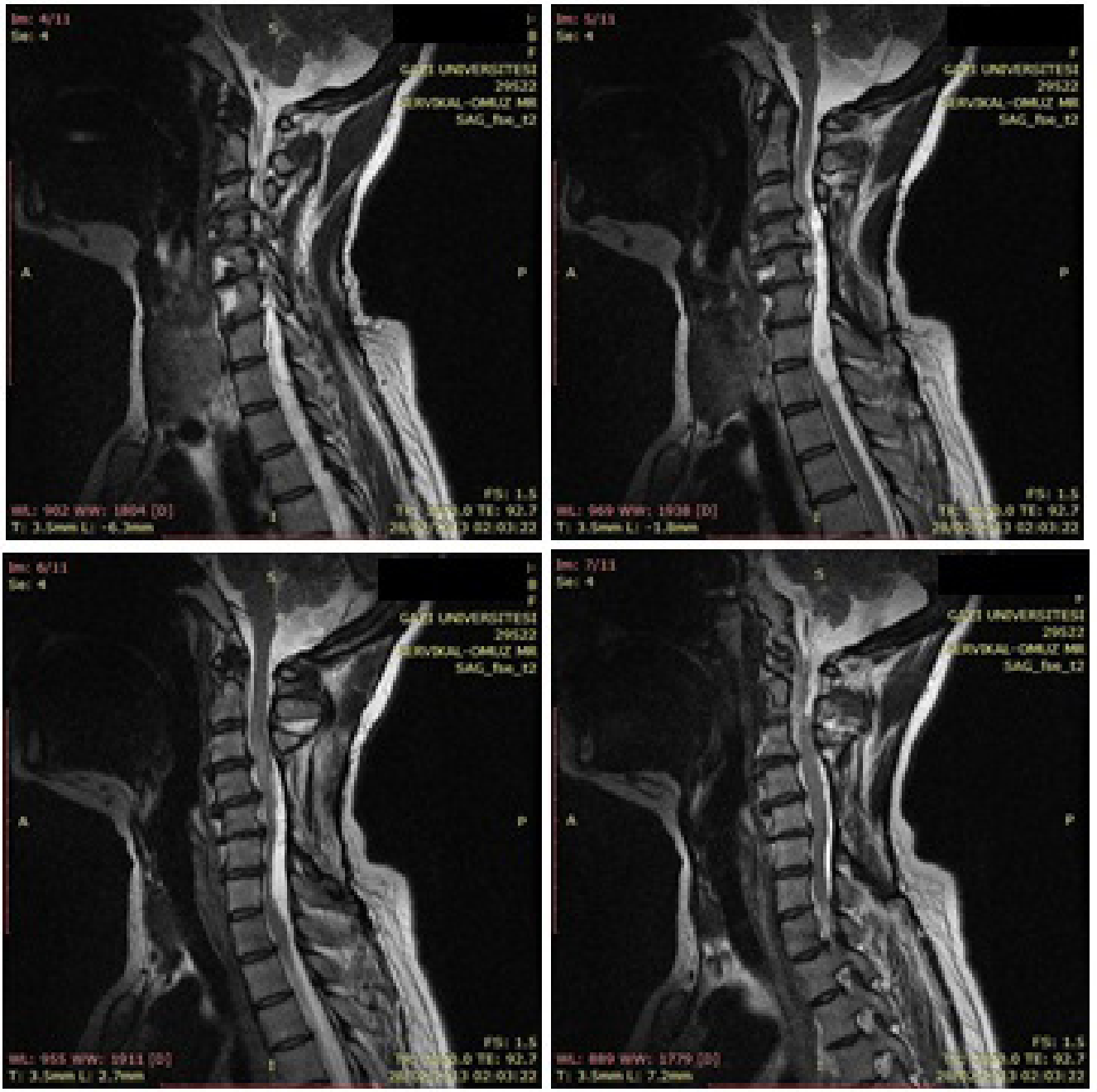

Figure-4. T2 weighted sagittal cervical MRI showing C3-4, C4-5 and C5-6 severe spondylosis with osteophytic supur formation causing foraminal narrowing, and C4-T1 cystic enlargement continued. C4 and C5 total laminectomy and C6 superior hemilaminectomy areas visualized.

The family was informed widely, emphasizing that he has NF-1, and he may have other neurofibromas at the cervical area ever, and he was a child and his spinal column would lengthen. But his kyphosis must have been corrected because it was shown that when the Gore angle was between 21 and 78 degree, the intramedullary pressure was increasing more than 50 mmHg (2). Thus he underwent operation with intraoperative neuromonitoring, C4 corpectomy, insertion of cage and anterior fusion with C3-5 anterior cervical plating, then C3,4,5 lateral mass fusion with screws were performed (Fig-11). Due to the reasons mentioned above the craniocervical instrumentation and fusion were not recommended at this stage. The patient was under follow-up for 2 years without any pain and neurological deficit.

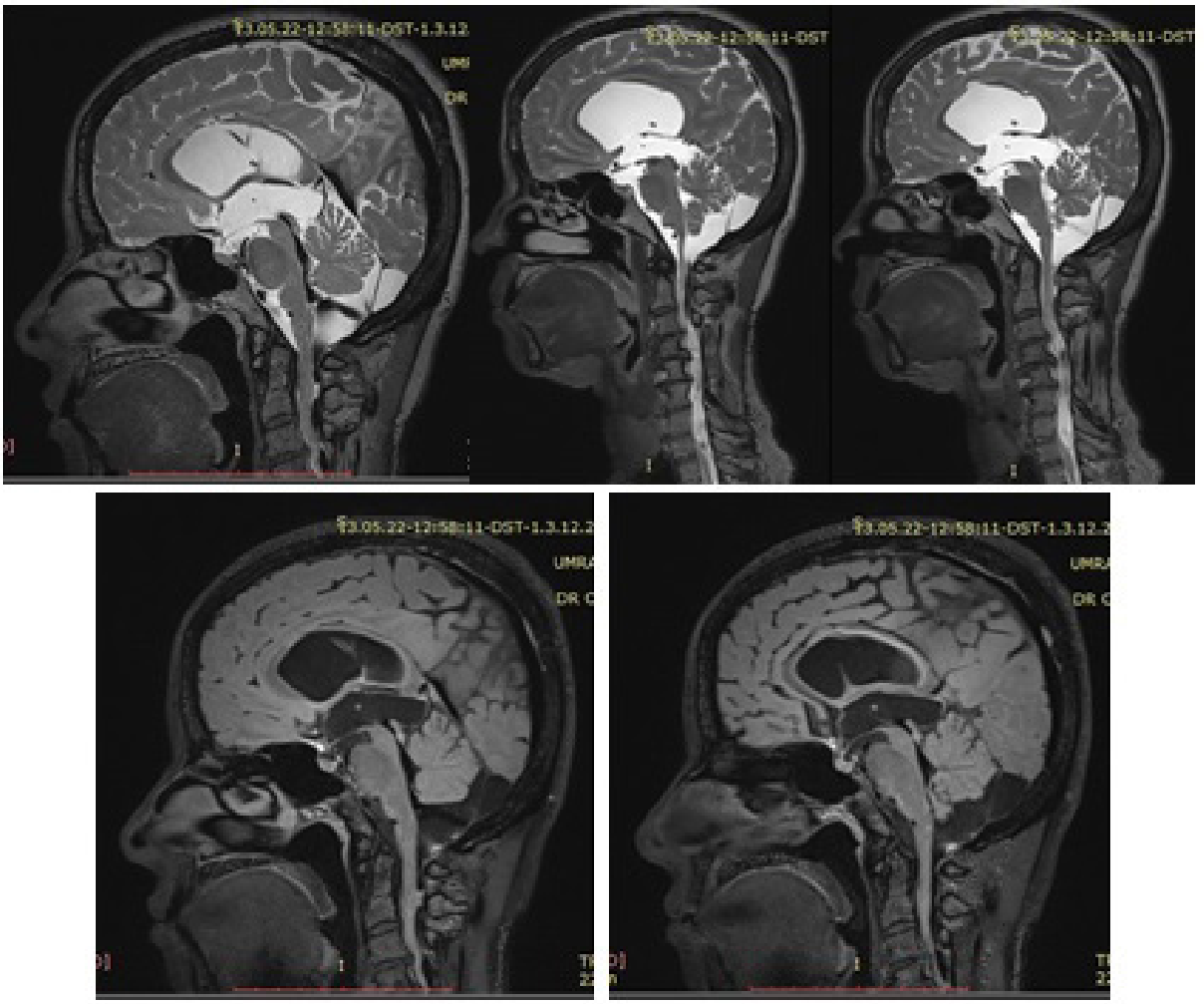

Figure-5. 3D CSF flow MRI was performed and showed CSF flow through the aqueductus Sylvius, Foramen of Magendie, and spinal subarachnoid spaces

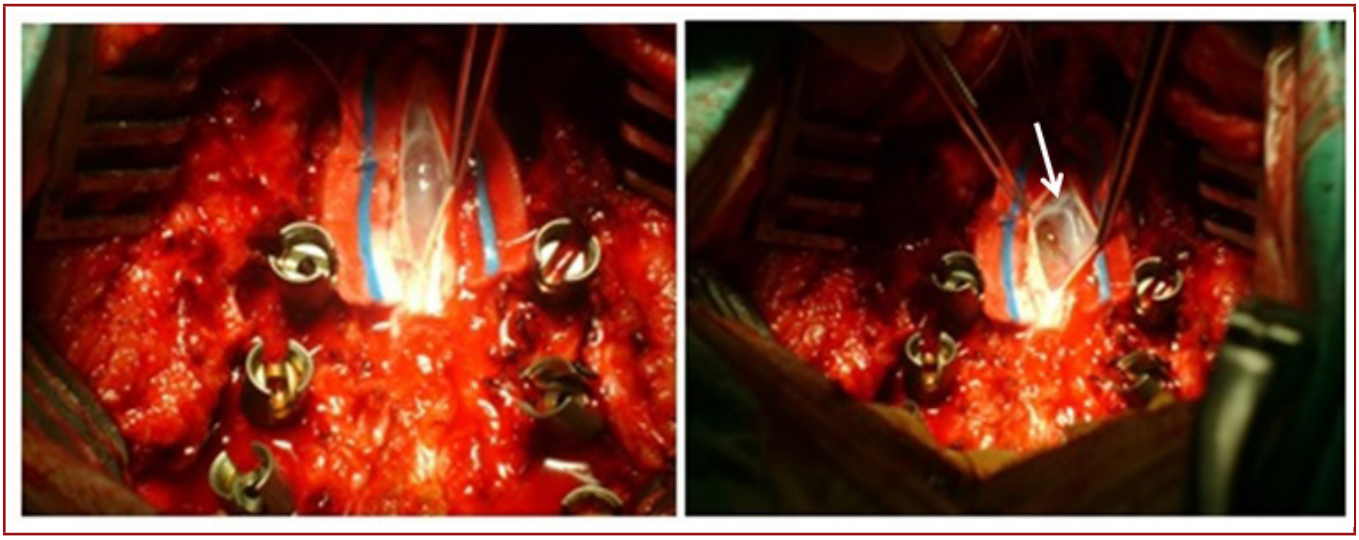

Figure-6. Intraoperative picture showing the thick arachnoid band (arrow) at superior T1 level.

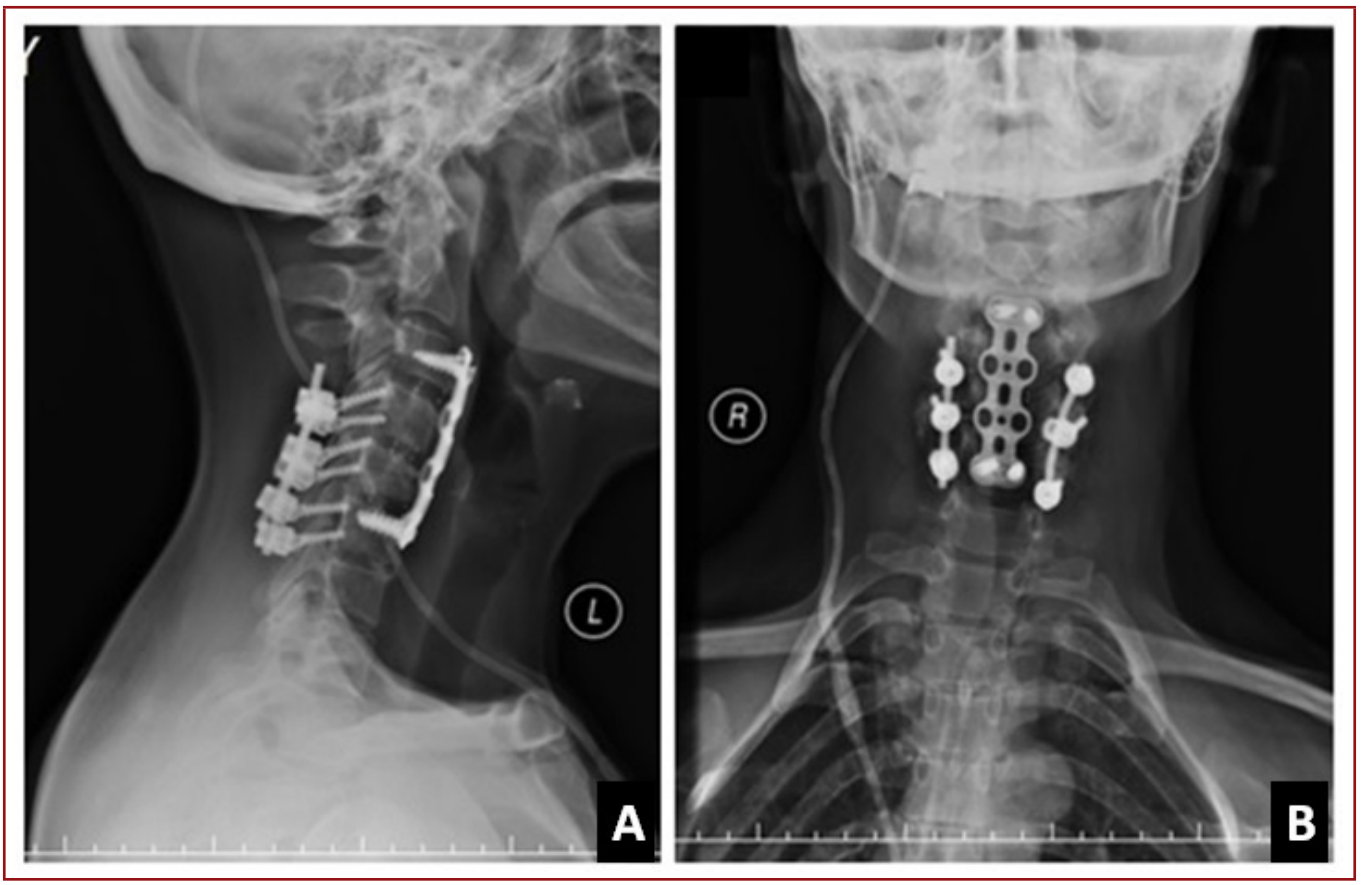

Figure-7. Lateral (A) & PA (B) cervical x-rays showed 360 degree enstrumentation of the cervical area

Case 3:

The patient was a 20 year-old girl applied to our clinic in 2012. She was operated on for tethered cord and scoliosis when she was 11 year-old. Thoracolumbar pedicle screw fixation and bone fusion which was not known if it was allograft bone matrix or synthetic bone graft substitutes were done. After operation her complaints (back and leg pain) did not resolve. 2 years later her instrumentation was removed because of back pain. She has had physical therapy and rehabilitation intermittently ended with increase of back and leg pain, and hypoesthesia. For the last 1 year, her complaints increased especially on the left leg with difficulty in walking. When she walked, she felt pain on her back, and pulling on the left leg with a severe pain on the posterior cervical and occipital area. Her neurological examination was revealed monoparesis and hypoesthesia on the left leg, heavy on the distal part. Cranial and whole spinal MRIs showed that she had no Chiari malformation, but she had scoliosis with vertebrae anomalies (L2 hemivertebrae, L1 butterfly vertebrae) (Fig-12), and tethered cord (Fig-13). She underwent operation, and after paravertebral muscle dissection it was seen that her lumbar area had 3,5 cm-thick, and 7 cm-wide intensive bony fusion (Fig-14). Using high-speed drill the spinal dura was found on the thoracolumbar area. The first dural opening was at the L4-5 level for untethering from the first operation. The dura was opened until S2 level, and it was seen that tethering continued until that level. The cord was released, and dura was closed as watertight fashion. The procedure was ended at this stage because the bony fusion was so intensive and there was no reason trying to crush it for correcting the scoliosis. After operation she underwent intensive physical therapy and rehabilitation. Her numbness and weakness of the left leg resolved significantly. Her back and neck pain ended.

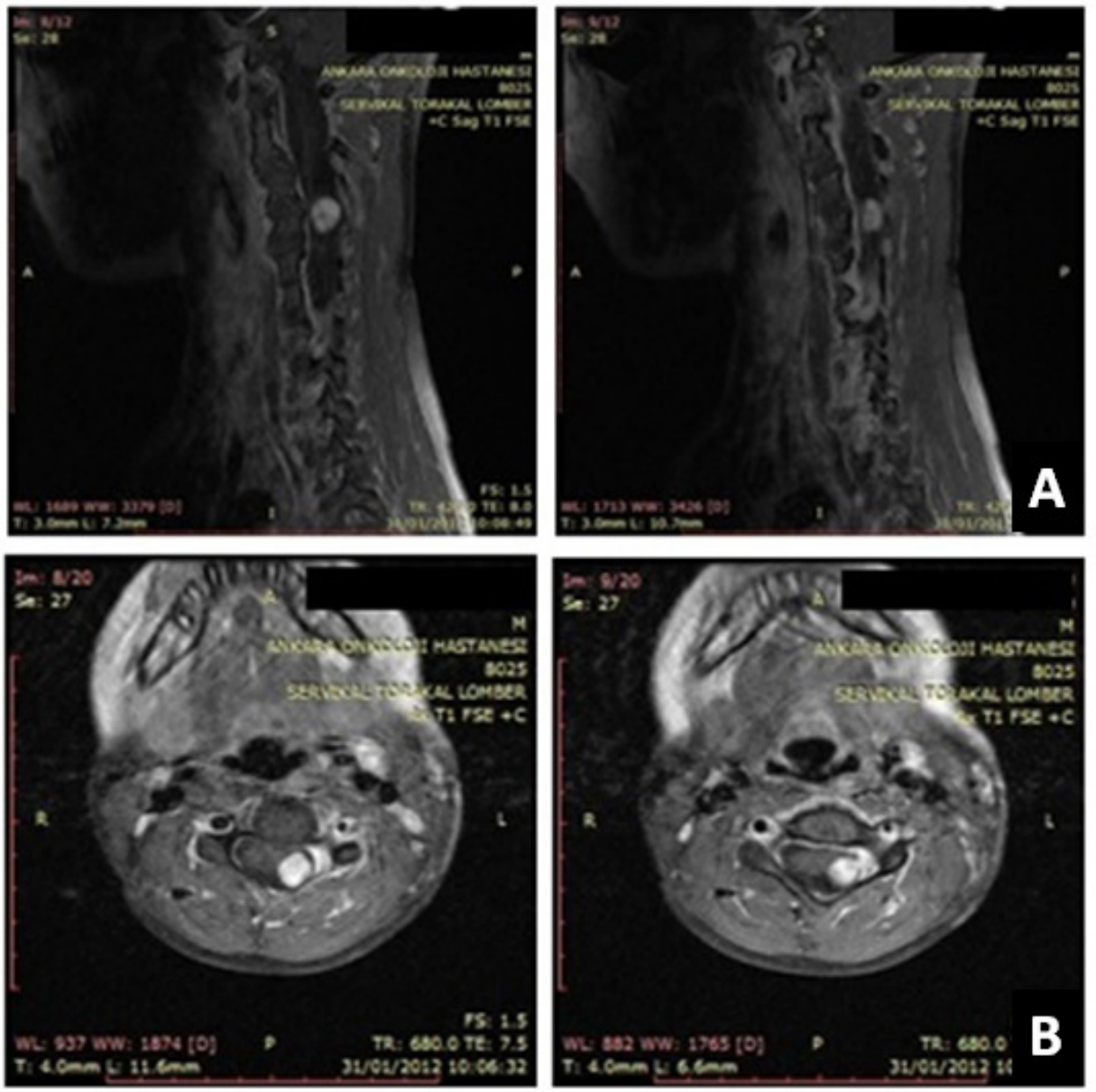

Figure-8. T1 weighted sagittal (A) and axial (B) MR images with contrast showing intradural neurofibroma at C4-5 level, located on the left side.

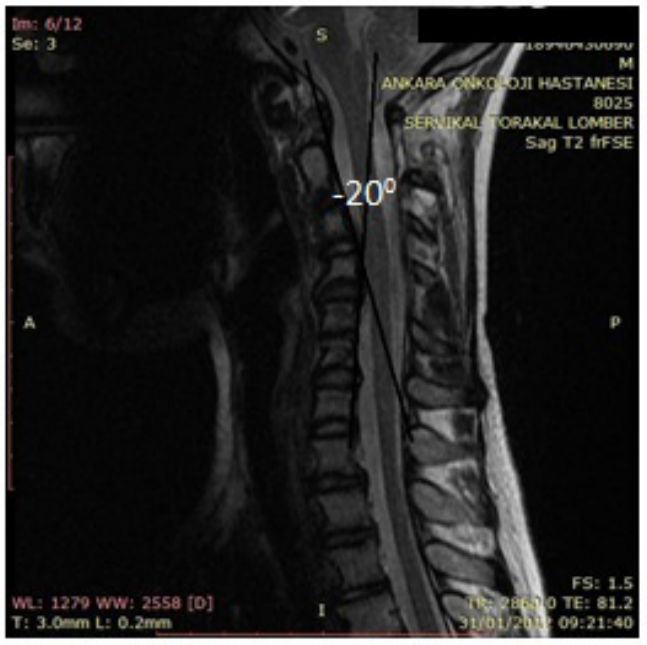

Figure-9. T2 weighted sagittal cervical MRI showing the pre-operative cervical kyphosis angle measured by posterior tangent method. It was 20 degree.

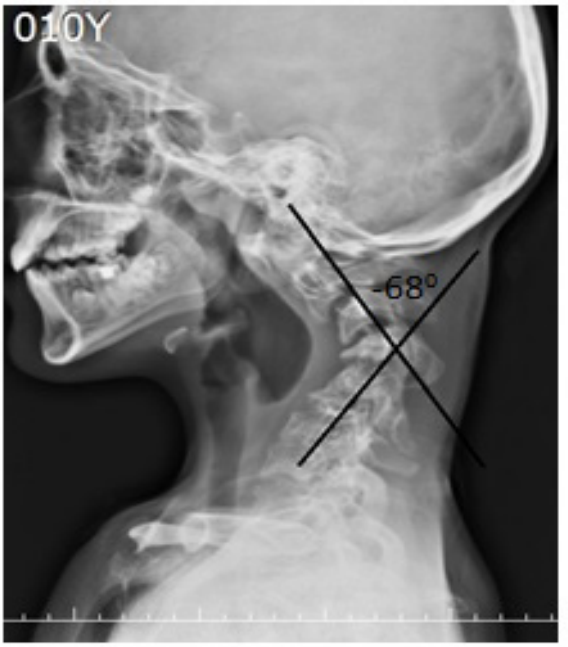

Figure-10. Lateral cervical x-ray showing cervical kyphosis angle measured by posterior tangent method at post-operative 6th month. It was 68 degree.

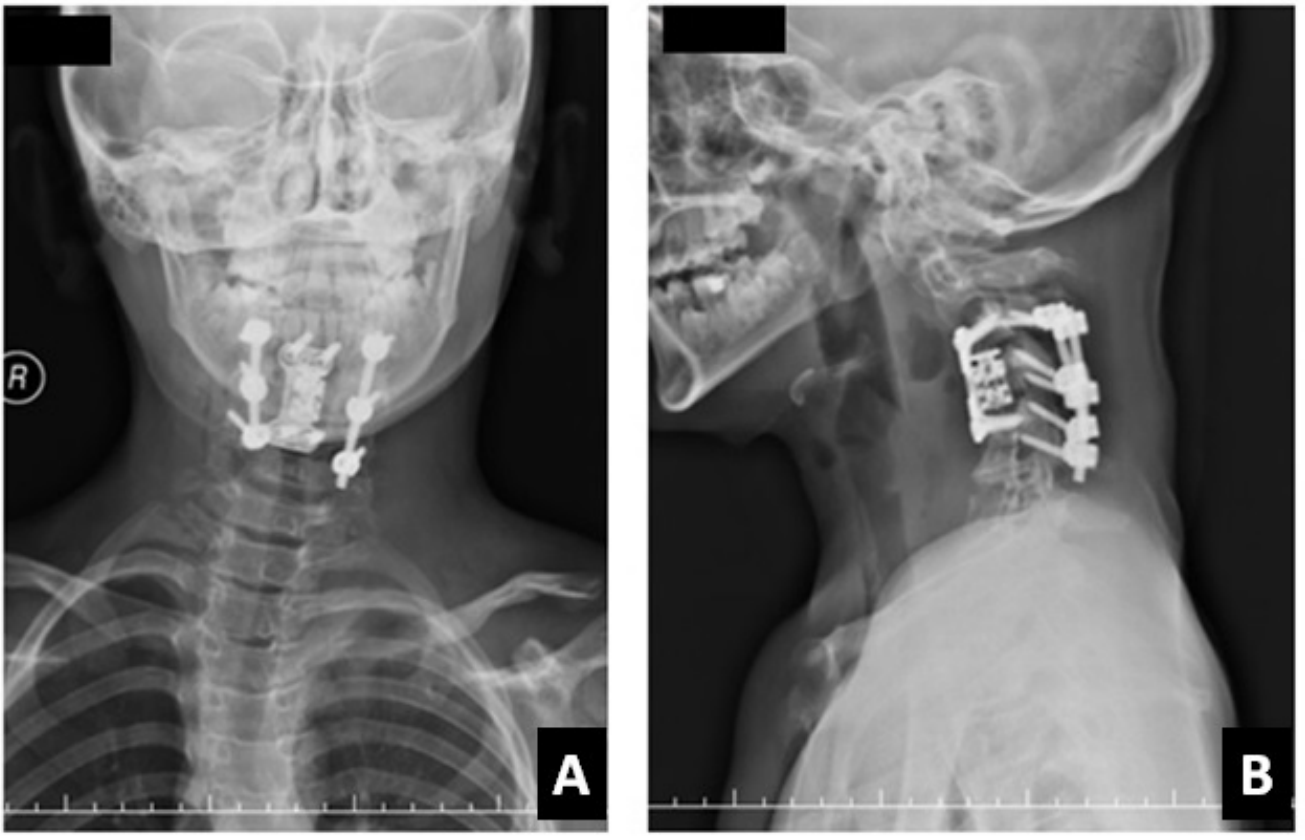

Figure-11. PA **(A)** & Lateral **(B)** cervical x-rays showed 360 degree instrumentation and fusion of the cervical area.

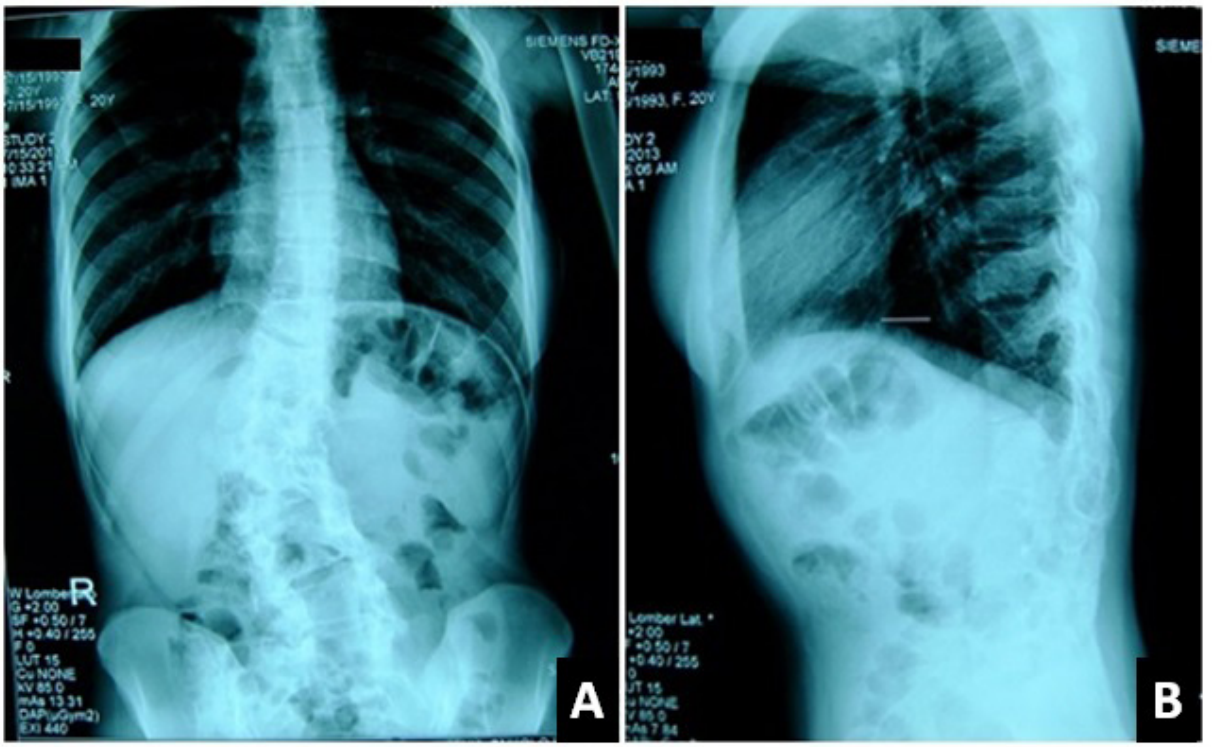

Figure-12. PA **(A)** & Lateral **(B)** cervical x-rays of Case-3 showed scoliosis, and previous L4, L5, S1 laminectomy areas.

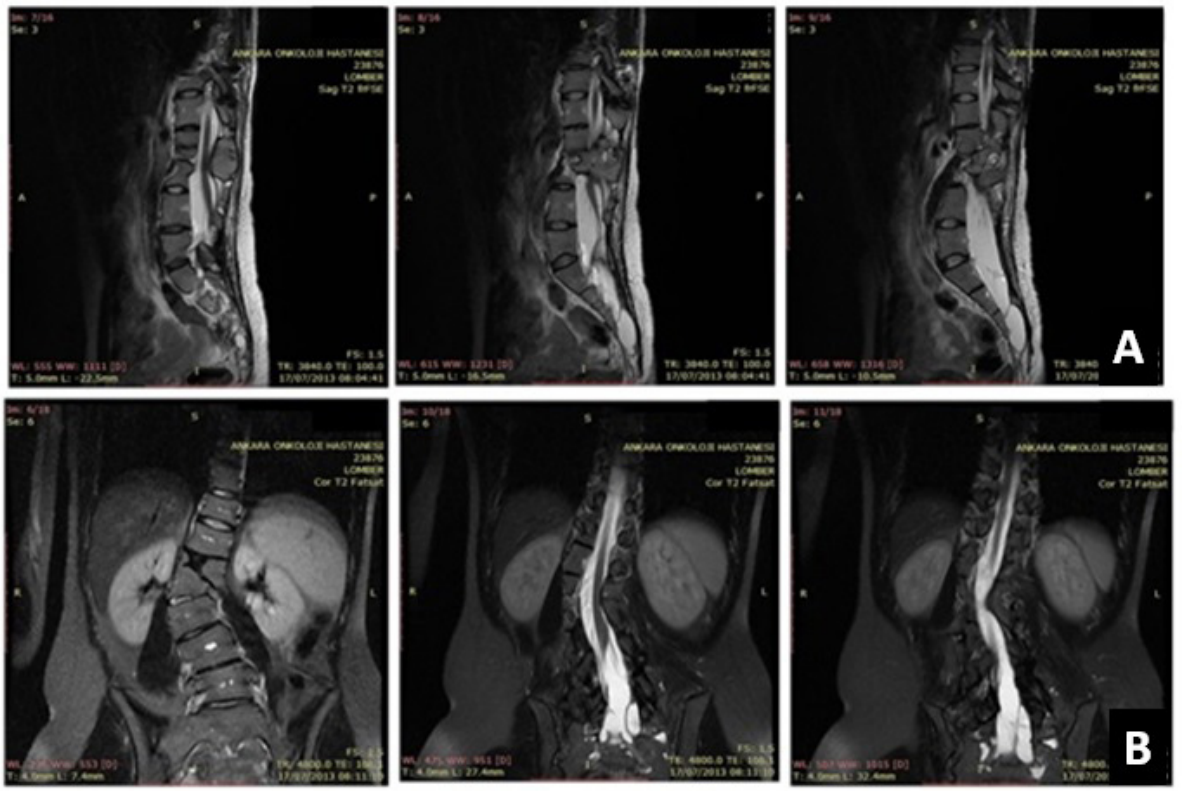

Figure-13. T2 weighted sagittal **(A)** and coronal **(B)** lumbar MR images showing scoliosis with L2 hemivertebrae, L1 butterfly vertebrae, conus ending almost at L5-S1 level with intradural septations resembling tethered cord.

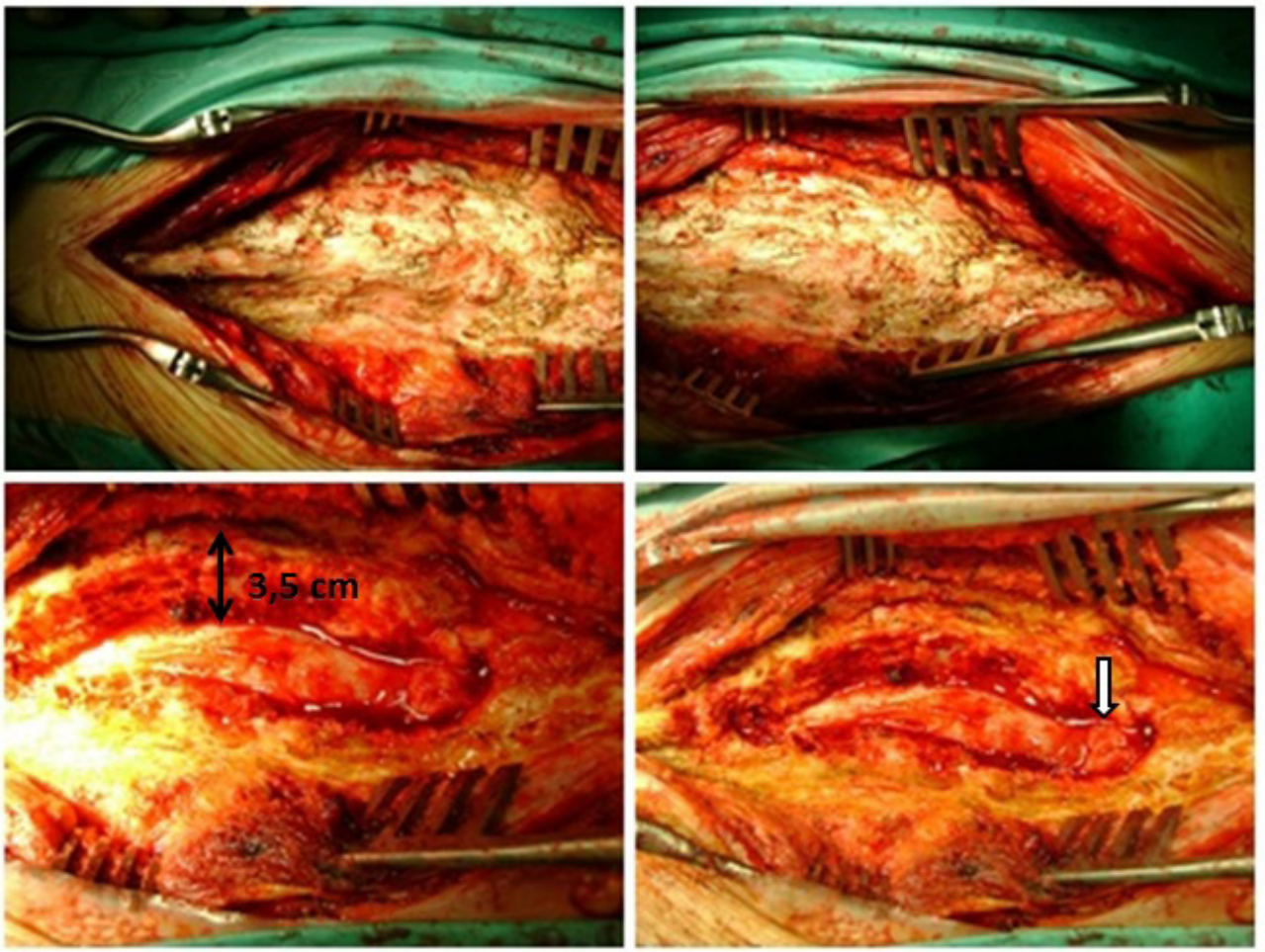

Figure-14. Intraoperative picture of the Case-3 showed 3,5 cm-thick bony fusion (black arrow) of the whole lumbar area, and previous dural opening was at the L4-5 level (white arrow)

Complications

Case-3 had CSF fistula after operation, which was resolved in 3 days with bed rest and dressing the wound.

RESULTS

All patients have some associated childhood neurologic disorders that complicated or prolonged surgery. Patient 1's previous CSF diversion yielded high risk of cerebellar herniation. Removing arachnoiditis needed extra laminectomy and dural opening. Difficulty of case 1's treatment was performing laminectomy with avoiding cerebellar herniation. Neurofibromas of the patient 2 required excision of these lesions. For this case, necessary of intradural and extradural tumor resection made difficult and prolonged the kyphosis correction. For untethering during scoliosis correction of the patient 3's tethered cord craved time consuming extra procedures, neuromonitoring, dural opening and microtechnique. Associated neural pathologies requires additional preoperative evaluation, using extra imaging modalities and excessive surgical planning. At the same time, surgical complication rates are getting higher with every associated disease. Fortunately, we saw only one serious complication after these 3 patient's operations. After untethering of the patient 3's tethered cord, a CSF fistula developed. Fistula closed after a 3-day bed resting and wound care. This is the only serious complication of these three patients. It needed no reoperation for dural repair. These procedures were challenging and time consuming.

DISCUSSION

For the Patient-1, it was difficult to decide from where to start. If the patient was shunt dependent, the cervical laminectomies, and dural opening would have the risk of cerebellar herniation. So it would have been necessary to fix the shunt first. But she may have also had arrested hydrocephalus and posterior cervical decompression, anterior discectomies, and spondylectomies in one session would have been enough for resolving her

complaints. Shunt reservoir tap was performed but it had been not a suitable method to measure the CSF pressure from a 22 year-old shunt, because of the ventricular catheter may have been hemi-obstructed, too. The history of severe headache and vomiting after LP also suggested hydrocephalus but this was not supported by other clinical tests such as ophthalmic evaluation for papilledema. During the first exploration of the cervical cyst, it was thought that arachnoiditis was caused by hemorrhages from V/P shunt insertion 22 years ago as an only possible reason. There is no literature in PubMed related to V/P shunt complication, subdural/epidural hematoma and cervical arachnoiditis and/or arachnoid cysts. So we decided to make staged surgery not to harm, and over treat the patient. At every step, the patient evaluated, examined again and again. So we took the risk and started from the cervical area first. Her shunt is still untouched but under follow-up with ophthalmological and radiological tests. Regular follow-up for such a patient by the same neurosurgical team and early intervention of the cervical cyst may have prevented the compression of the cervical cord and excessive degeneration of the cervical column. V/P shunt dysfunction may have been realized earlier, and the shunt pump may have been changed to programmable one with the replacement of distal and if needed proximal catheters. Ultimately, continuing question mark about the shunt dysfunction may have been extinct.

For Patient-2, previous studies of asymptomatic volunteers have revealed that the greatest variation in regional sagittal neutral upright spinal alignment occurs in the cervical spine with "normal" alignment ranging up to +15 to +20° kyphosis. Minor intramedullary pressure (IMP) increases of 2-5 mmHg were observed when the Gore angle was <+21°. Gore angles ranging from +21° to +78° resulted in statistically significant increases in IMP ranging to >50 mmHg [2].

Cervical kyphosis is relatively common and can develop due to infection, tumors, or surgery; however congenital cervical kyphosis is a rare clinical condition. It is defined as kyphosis induced by an abnormal vertebral body, including congenital failure of formation (type-1), congenital failure of segmentation (type-2), and mixed failure (type-3). Cervical kyphosis may occur with NF1 and is often associated with vertebral dysplasia. Outcomes of cervical spinal fusion in patients with NF1 are not well described because of the rarity of the condition. There are only two references in the literature (3-4). Most of the patients underwent anteroposterior cervical fusion [3-4]. Kawabata et al. [4] suggested dystrophic changes in the vertebrae make surgical correction and fusion of the deformity extremely difficult. The kyphosis angles improved after surgery but the patients had still some degree of kyphosis. The partial dislocation of the distal fibula graft after removing the halo west is the surgical complication. Helenius et al. recommended anteroposterior surgery provided better correction of cervical kyphosis than posterior fusion in children with NF-1. Thirteen patients of 22 patients had complications and nine needed revision surgery [3]. In light of the foregoing findings, cervical kyphosis in NF-1 patients is difficult to treat, may need consecutive operations, and should be followed with close attention.

Patient-3 has congenital scoliosis which is the presence of abnormal coronal plane curvature in spine secondary to a failure of formation, segmentation, or a combination of the two arising from abnormal vertebral development. Congenital scoliosis occurs 1 in 1000 newborns [5-6]. As a developmental anomaly, it is associated with various organ system abnormalities including neural axis. The close relationship of embryonic development of vertebrae and spinal cord causes co-existence of neural and vertebral malformations such as tethering of the cord, diastematomyelia, lipomas, syringomyelia, and lipomeningocele [7-10]. The MRI studies showed the intraspinal anomalies range from 24,5 % to 47 % [10-12]. Hemivertebrae was the most common anomaly. The incidence of intraspinal anomaly in patients with failures of segmentation and mixed defects were significantly higher than patients with failures of formation [9-10,13]. Tethered cord was the most common intraspinal abnormality in congenital scoliosis patients.

The options for conservative management for congenital scoliosis are less effective when compared with idiopathic scoliosis [14]. An MRI examination of the entire spine is recommended before any surgical intervention [15-16]. Neurosurgical evaluation and operation are recommended for any anomaly which tethers the cord before attempting surgical correction of the deformity. Surgical treatment is indicated for deformities that are increasing in severity, or an anomaly that is predicted to have a high risk for progression [17]. In situ fusion and convex hemiepiphysiodesis have been described for congenital scoliosis with minimal deformity over a short section [18]. But both procedures have been reported to have limited success with minimal deformity correction [19], and the development of crankshaft phenomenon [20]. Earlier hemivertebrae excision and short-segment posterior spinal fusion have been advocated to prevent future curve progression of the deformity and/or the development of large compensatory curves [21]. This patient underwent her first operation almost 14 years ago. Untethering of the cord and correction of the spinal deformity were planned in the same session. Untethering of the cord could have not been achieved, so the scoliosis was fixed without correction. When the reason for tethered cord is not a thick/fatty filum terminale, the untethering operation can be long and complicated procedure. For Patient-3, it could have been better just untethering the cord first, and then following the progression of scoliosis. Then, a detailed plan for the second operation could have been prepared for the favorable correction of the scoliosis. But inadequate planning and insistence on going on the operation abolished the chance of any other operation for scoliosis.

Surgical treatment of patients with spinal diseases may become a quite challenging case when they have congenital anomalies or CNS related problems. Expectations of only one surgery that solve all the problems makes surgeon trail to desperation and pessimism. In very first planning, staging surgical approach should be kept in mind for these kinds of patients. Besides, growing of spine and spinal cord at different rates shouldn't be forgotten for children. The whole spinal axis must be kept under lifetime control by some regular intervals for spine patients with additional congenital or CNS anomalies.

That this series were consisted of only three patient and varieties of associated neurological disorders are the main limitation of this study. However, the thing that emphasize in this manuscript, spinal surgeons should be awake for associated neurologic disorders in spinal deformity or degenerative spine cases. These associations are time consuming and needs extra effort. Complication rates are higher than solely spinal surgeries. We need larger series for statistical analysis. Besides, classification of associated neurological disorders is the requirement of this kind of studies. Making any classifications also needs larger patient population.

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