SURGICAL CORRECTION OF SUBAXIAL KYPHOSIS IN A CHILD WITH TYPE I NEUROFIBROMATOSIS: RARE CLINICAL CASE AND LITERATURE REVIEW

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The paper presents a case of rapidly developing severe cervical kyphosis in a 5-year-old child which was the first clinical manifestation of type I neurofibromatosis (NF1). Surgical correction was carried out in two stages with preliminary hardware halo-traction and subsequent reconstruction of the spine using titanium mesh cage with bone autograft and posterior instrumentation. The deformity was corrected from the magnitude of 79° to the restoration of physiological lordosis. Long-term results were followed-up for 2.5 years with full maintenance of the achieved deformity correction. A 15-year literature review on the cervical spine lesions in NF1 is presented.
Key Words: cervical spine, children, surgical treatment, cervical kyphosis, type I neurofibromatosis, spine deformity.

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Neurofibromatosis type I (Recklinghausen's disease, NF1) is a hereditary disease associated with a mutation in the NF1 gene located in chromosome 17q [9, 22]. Typical spinal manifestations of NF1 include combined spinal deformities (kyphoscoliosis, lordoscoliosis) that are most often located in the thoracic and lumbar spine [3]. The pathology less often affects the cervical and cervicothoracic spine, manifesting as severe kyphosis that may be complicated by neurological disorders [3, 17]. Most publications have reported single clinical cases or small groups of patients, with approaches to surgical treatment being quite different. The presented case may be interesting as one of the treatment options.

Parents of a 5-year-old girl M. first presented to doctors in February 2015 about a "nodus" on her dorsal neck, which had developed about 1 month before. An examination at a local clinic revealed restricted motion and deformity in the cervical spine. A radiographic examination revealed changes in the C3–C6 vertebrae, which were assessed as a potential developmental anomaly or a destructive process complicated by spinal canal stenosis and spinal cord compression on the site of a gross kyphotic deformity (Fig. 1). In this case, there were no neurological disorders (clinical manifestations of myelopathy) in the child. There was a concomitant disease, West syndrome (age-dependent epileptic syndrome), in remission. Laboratory studies (complete blood count, blood chemistry, and clinical urine analysis) and chest radiography revealed no pathology.

At admission to the clinic (15.04.2015), the child's condition was quite satisfactory; there were no subjective complaints. Of particular interest were multiple, variably sized, and rather pale cafe-au-lait pigment spots on the girl's body. Palpation of the spinous processes and paravertebrally at the C2–C5 level revealed a moderately painful dense nodule; the skin was normal. Motion in the cervical spine was constricted; the patient constantly wore a Philadelphia collar; the gait was normal. There was a slight scoliotic deformity of the thoracic spine; the length of the limbs was D = S; there was a full range of motion in the joints, painless. There were no neurological disorders (Frankel E).

X-ray and CT studies demonstrated a pronounced cervical kyphosis with the apex at C3. The C3 and C4 bodies were wedge shaped, with the C3 body lacking its anterior half (Fig. 1); there was a scoliotic thoracic curve.

According to the MRI data (Fig. 2), in the setting of a sharp-angle deformity of the spinal canal, the residual C3 body was displaced posteriorly into the vertebral canal; the anteroposterior size of the spinal cord was reduced at this level, but no radiological signs of myelopathy were detected. Paravertebral tissues were intact; there were no microcirculatory changes in the vertebral bodies.

Given instability of the cervical spine, rapidly progressing kyphotic deformity, and a high risk of neurological disorders, surgical treatment was planned. In order to minimize risks associated with a sharp change in the anatomical relationships in the neck, slow traction was performed after application of a halo-cast

(21.04.2015) for 14 days, which reduced the kyphosis from 79 to 51° (Fig. 3).

After removing one of the anterior bars (05.05.2015), the patient underwent reconstruction of the C2–C5 segment with decompression of the spinal canal and correction of the deformity by means of anterior spinal fusion using a titanium mesh cage filled with an iliac crest fragment (surgery was performed using neuromonitoring, NIM Eclipse, Medtronic). In surgery, an approach along the anterior edge of the right *m. sternocleidomastoideus* was used; soft tissues were not affected; the C3 and C4 bodies were completely removed; the spinal canal contents were exposed throughout the reconstruction area. The vertebrae were removed by fragmentation and treatment with a high-speed drill; no visual signs of inflammation or destruction were noted. Surgery was completed by restoring the stability of an external fixation device.

After 14 days, the final surgical stage was performed – posterior laminar fixation of C2–C6 and removal of the halo apparatus.

A histological examination of the surgical material revealed dystrophic changes.

All inter- and postoperative periods proceeded without complications; wounds healed primarily. By the time of discharge, there were no subjective complaints. For some time, the patient wore a Philadelphia collar during long trips; the collar was discontinued after 6 months. There was no pain syndrome or signs of neurological deficiency.

After surgery, the patient was followed-up at the place of residence and distantly, via a distance consultation system. According to control CT at 6 and 30 months after surgery (Fig. 4), a solid bone block formed inside the mesh cage; the posterior joints were ankylosed. There was no deformity progression and subjective complaints.

Discussion

The characteristic clinical, radiologic, and structural and functional (non-dystrophic and dystrophic) features of spinal deformities in type I neurofibromatosis, as well as the experience of their surgical treatment (primarily instrumental correction), are in detail described in the domestic medical literature [1–4]. It is noteworthy that these publications are devoted to deformities of the thoracic and lumbar spine, while the pathology of the cervical spine is described in detail without indicating possible treatment options.

On the contrary, foreign authors have described various ways of surgical correction of cervical kyphosis in children with NF1, indicating the risk of spinal cord compression due to both vertebral body displacement and the intracanal component of a tumor (neurofibroma) [15]. The largest sample included data on 22 children [10], and the earliest age of surgery was 21 months [20].

A surgical approach usually includes decompression of the spinal canal contents using corpectomy at the apex of kyphosis and isolated anterior or posterior fixation or their combination. In some cases, after decompression, surgery is limited only to anterior fixation using replacement of the defect with a bone autograft (fibula or iliac crest) [12], sometimes supplementing it with the anterior plate [7, 18]. It should be noted that in these studies, the maximum kyphotic deformity was 52°, and the correction was 25°. The authors explained their refusal of posterior fixation by the adequacy of anterior stabilization for bone block formation and by a risk of injury to neurovascular structures during transpedicular fixation.

Some authors have used combined techniques with posterior instrumental fixation/deformity correction by transpedicular devices [8, 10, 16, 19–22] or, in cases of pronounced dystrophic changes in the vertebrae (scalloped vertebral body edges, spinal canal widening, thinning of pedicles), translaminar screws.

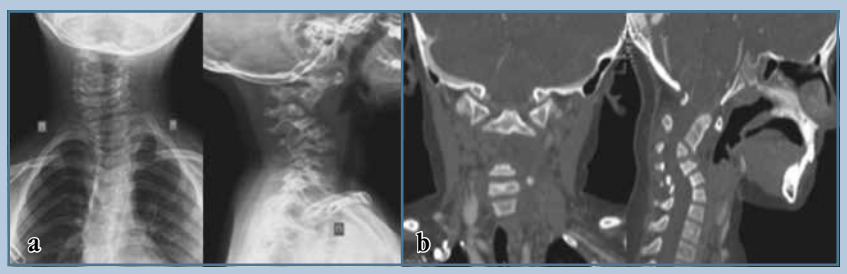

Fig. 1
Radiographs **(a)** and CT scans **(b)** of the cervical spine of the 5-year-old patient M. in two standard projections (see explanations in the text)

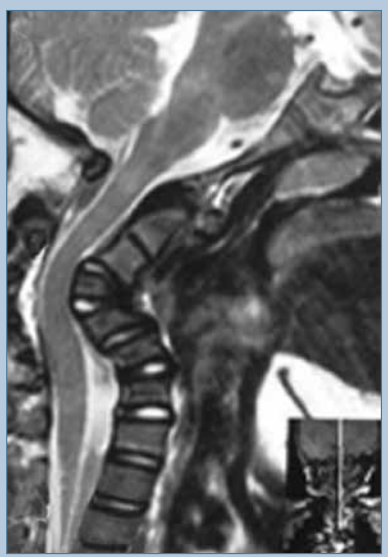

Fig. 2
MRI of the cervical spine of the 5-year-old patient M. in the sagittal projection: stenosis of the spinal canal and angulation and compression of the spinal cord are seen

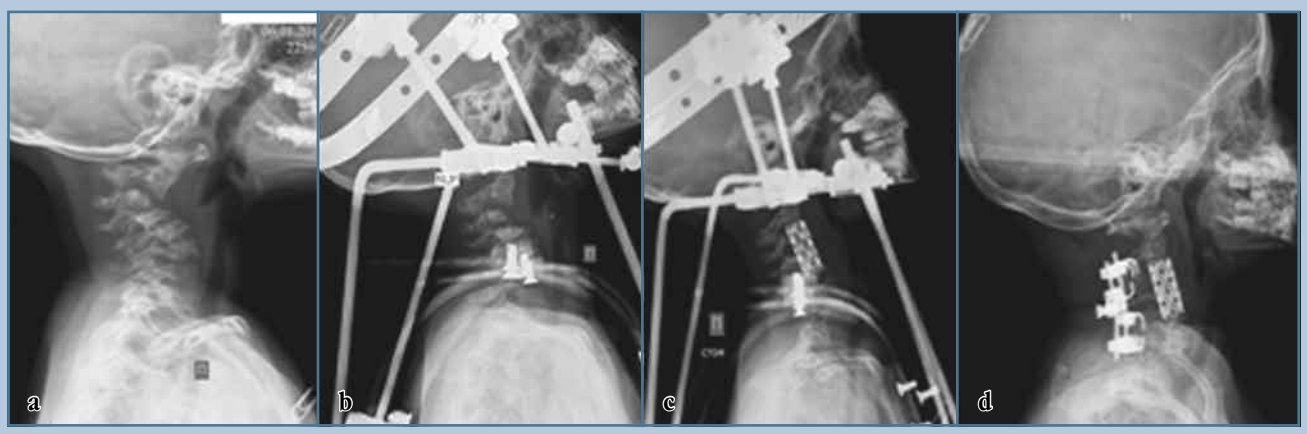

Fig. 3
Sagittal radiographs of the cervical spine of the 5-year-old patient M. at the correction stages: **a** – initial kyphosis of 79°; **b** – by the time of halo-traction completion, the kyphosis was 51°; **c** – after anterior reconstruction, the kyphosis was completely corrected; **d** – the result of correction was retained after posterior fixation and removal of an external fixation device

Gardner et al. [8] successfully corrected cervicothoracic kyphosis from 90 to 20°, despite compression of the spinal cord, in a 17-year-old girl without neurological complications using 2-stage treatment: first, anterior reconstruction was performed using a fibular graft and a LCP plate; subsequently, at a 7 day interval, scoliosis was corrected using lateral mass screw fixation in the cervical spine and translaminar screw fixation, one per each level, in the thoracic spine to avoid a fracture at the junction between the spinous process and the lamina. On a follow-up examination at 18 months, bone block formation was observed. Sublaminar fixation was not used because of a risk of injury to a deformed dura mater. Kawabata et al. [13] described three neurofibromatosis cases, including a 10-year-old girl with subaxial kyphosis that was corrected from 81 to 55° after one month of halo-traction. A fibular graft was fixed with a cortical screw to the C7, and posterior fixation was performed using laminar screws. Within 1 month after surgery, the patient wore a halo cast. Correction of the kyphosis to 15° and bone block formation were achieved after 36 months. The authors indicated risks of complications during correction of cervical kyphosis in NF1: the risk of spinal cord injury due to its previous compression, presence of spinal cord tumors and neurofibromas, risk of surgical bleeding requiring embolization, and technical difficulties of posterior fixation in the case of dystrophic vertebral changes. In addition, the authors recommended MR angiography or contrast-enhanced CT before performing halo-traction to prevent vertebral artery aneurysm rupture.

Among the options used to fix the cervical spine in a small child with NF1, an anterior resorbable biopolymer plate was also reported [14].

There are cases when posterior instrumentation alone in a patient with the dural ectasia provided bone block formation after 2 years [11, 15]. In critical situations (spastic tetraparesis), palliative interventions, such as decompression laminectomy with partial tumor resection, may also improve the neurological status [17].

Interestingly, despite various ways of correcting kyphotic deformity, all authors have noticed good formation of a bone block without deformity progression in the long-term postoperative period, although a high rate of pseudoarthrosis, loss of correction, and worsening of deformity during growth are characteristic of children with thoracolumbar scoliosis in NF1.

Our clinical case differs from the data presented in the review by the following features:

– the cervical spine deformity was the first manifesting symptom of NF1 that had earlier manifested only with minimal diagnostic signs: multiple cafe-au-lait spots; other signs of the disease, especially its vertebral syndrome [6], were revealed on examination at the clinic;

– the primary diagnosis of cervical spine malformation seems to be quite typical: vertebral changes were considered as malformations because there were no structural disturbances and changes in the size of the spinal canal and intracanal structures, except the shape of the cervical vertebrae and thinning of vertebral ends of the ribs (Fig. 1a). However, it should be clarified that congenital cervical kyphosis is an oddity: this spine region is characterized by different anomalies in the form of lateral or combined deformities; the dystrophic changes detected by a morphological study once again confirm the absence of a relationship between the pathology and the congenital anomaly;

– the surgical approach was chosen due to, first, the risk of one-step surgery and the desire to safely perform both a preparatory traction stage of correction and anterior decompression, the

complexity of which increases as the magnitude of kyphosis is growing; the second cause was successful experience of using combined anterior reconstructions (titanium mesh cage + bone autograft) at various levels of the spine in children [5]; third, in our opinion, in the case of normal laminae and a priori expected dystrophic changes in cancellous bones in NF1, the use of anchor supports for posterior instrumentation may have advantages over screws, regardless of the procedure used for their fixation: transpedicular, laminar, or to the cervical lateral masses. In our opinion, the proposed treatment option may be included, as another effective and sufficiently safe option, in the arsenal of spinal surgeons engaged in the treatment of gross kyphotic deformities of the cervical spine in children.

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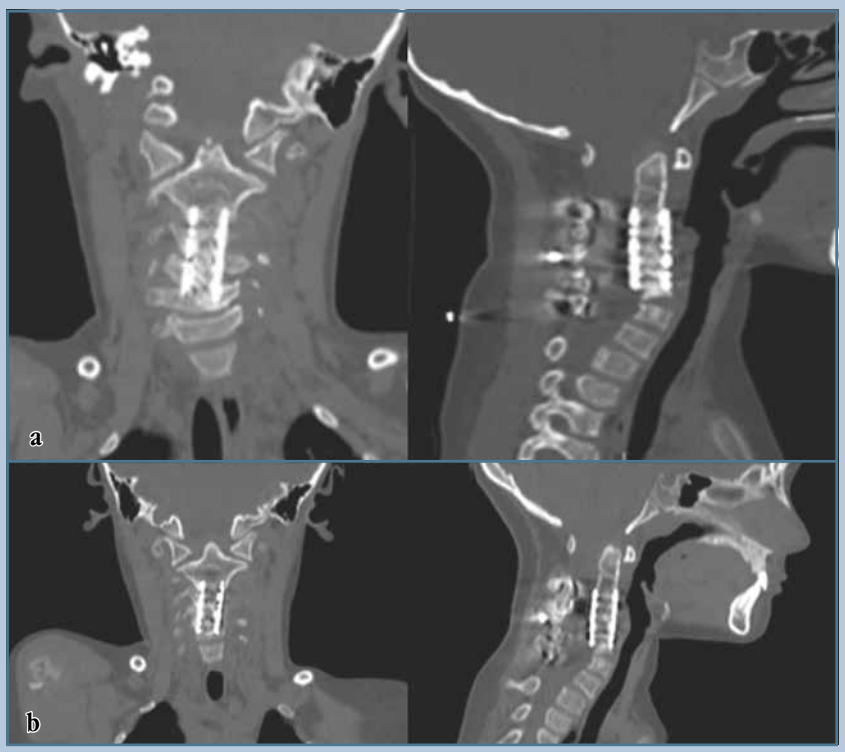

Fig. 4
CT scans of the cervical spine of the patient M. at 6 **(a)** and 30 **(b)** months after surgery

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