ATLANTOAXIAL DISLOCATION IN AN ADOLESCENT WITH JUVENILE SPONDYLOARTHRITIS

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Atlantoaxial dislocation with spinal canal stenosis not associated with craniovertebral junction malformations and traumatic injury is a rare pathology in children and adolescents. A clinical case of the diagnosis and surgical treatment of atlantoaxial dislocation of non-traumatic genesis combined with spinal canal stenosis in an adolescent with juvenile idiopathic arthritis is presented. A minimal instrumental fixation of the upper cervical spine allowed eliminating stenosis of the spinal canal, repairing instability of the upper cervical vertebrae, and jugulating neurological disorders.

Key Words: spinal canal stenosis, atlantoaxial dislocation, children, juvenile idiopathic arthritis, surgical treatment.

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Atlantoaxial dislocation accompanied by a posterior displacement of the odontoid process with the development of spinal stenosis and spinal cord compression is most often caused by traumatic injuries [1] and some congenital pathologies of the craniovertebral junction, such as developmental anomalies of the skull base and upper cervical vertebrae (os odontoideum) and vertebral formation and segmentation defects [2]. A number of genetic syndromes and systemic diseases are accompanied by weakness of the ligamentous apparatus of the atlantoaxial joints and can cause spinal stenosis in children. These include Down and Morquio syndromes, mucopolysaccharidosis type I, II, and IV, spondyloepiphyseal dysplasias, osteogenesis imperfecta, and neurofibromatosis [3, 4]. Acquired pathology that causes atlantoaxial dislocation can manifest as complications of upper respiratory tract infection particularly in retropharyngeal abscess– Grisel syndrome [5].

Diseases constituting a group of spondyloarthritides, which include adult rheumatoid arthritis, juvenile idiopathic arthritis, and HLA-B27-associated arthritis (ankylosing spondylitis, psoriatic arthritis, Reiter's syndrome), can also cause instability of the cervical spine, leading to spinal stenosis [2, 6–8]. Juvenile idiopathic arthritis is a heterogeneous group of inflammatory joint diseases developing in children under 16 years of age, with a chronic (more than 6 weeks) progressive course. In juvenile arthritis, atlantoaxial joint with the synovial membrane can undergo inflammatory changes similar to lesions of peripheral joints: excessive production of synovial fluid; hypertrophic changes in the synovial membrane with formation of the pannus causing erosion of the cartilage and subchondral bone [7, 9]. In this case, inflammatory changes can cause degeneration of the cervical vertebral ligaments, which leads to joint instability in 17–85 % of cases [2, 8, 10].

Regardless of cause of spinal canal stenosis at the craniovertebral junction, various surgical techniques are used to treat these patients. In recent years atlantoaxial complex in most cases has been stabilized by screws at the C1–C2 level, both in adult and in pediatric patients [3, 4, 9]. However despite the fact that atlantoaxial subluxation is the most common type of cervical spine instability in juvenile arthritis [6], neurological complications of this process rarely develop in childhood or adolescence as an onset of the disease [1, 2, 10].

The aim of this study was to analyze the course and treatment of atlantoaxial instability in adolescents with juvenile spondylitis.

A 16-year-old male patient O. presented to the Department of Spinal Pathology and Neurosurgery at the Turner Scientific Research Institute for Children's Orthopedics with atlantoaxial dislocation. He did not have any significant co-morbidities and previously had never been consulted with any specialists (neurologist, orthopedist, rheumatologist).

The patient consider himself sick for the past year, during which there was a periodic cervical spine pain not associated with physical or static load. The pain was temporary and resolved spontaneously. He underwent MRI of the cervical spine at the place of residence (data not available; based on the medical records), which revealed atlantoaxial subluxation without spinal stenosis. He received several sessions of osteopathy; after latter session (6 months after the onset of cervical spine pain) patient developed left-sided hemiparesis. Repeated MRI of the cervical spine revealed progression of atlantoaxial dislocation and development of spinal stenosis at the level of C1–C2. The patient was hospitalized to the neurosurgical department; there he underwent traction using Glisson's loop with 4.0 kg of weight, which led to a slight improvement in strength of the left

upper and lower extremities. Then patient was transferred to a specialized center for surgical treatment.

At the admission: patient's body type was normosthenic. His neurological status showed central tetraparesis, more significant in his left upper and lower extremities (with a decrease in muscle strength to 3 points) and asymmetric hyperreflexia (more significant on the left side) with foot clonuses in lower extremities. MRI revealed atlantoaxial dislocation with spinal stenosis and spinal cord compression at this level in the form of a hyperintensity in T2 of the spinal cord at the odontoid process level (myelopathic signal). There were no reliable signs of paradental pannus formation (synovial hypertrophy); the spinal cord was compressed between the odontoid process and the posterior arch of the atlas more on the left (Fig. 1).

A CT examination didn't reveal any cervical or skull base bony malformations; the anterior atlantodental interval was increased to 13.9 mm due to posterior dislocation of the odontoid process with narrowing of the retrodental distance to 3.0 mm, with an asymmetric decrease of the anteroposterior dimension of the spinal canal at the C1–C2 level, up to 3.7 mm on the left and up to 6.0 mm on the right. An uneven contour of the anterior surface of the odontoid process was noted, which may be a sign of the erosive process associated with arthritis (Fig. 2).

Given the time from the disease onset, prolonged conservative treatment, pronounced neurological disorders, and the lack of etiopathogenic therapy, a halo apparatus was applied under general anesthesia as the first stage of treatment for stabilization of the craniovertebral region and gradual, controlled closed reposition of the atlas. A control CT examination showed that gradual traction and reposition with the apparatus reduced the amount of dislocation with decrease in the atlantodental interval to 8.8 mm and increase in the anteroposterior dimension of the spinal canal at this level up to 8.6 mm (Fig. 3).

During the process of treatment with halo apparatus, the patient developed signs of right knee joint arthritis (pain, swelling, and restricted movements) on the 3rd day after surgery. These changes were observed for the first time. Ultrasound of the joint revealed signs of exudative synovitis and an increase in the superior recess size to 45×19 mm due to fluid collection.

On day 7 after surgery, the patient complained of pain in the area of right temporomandibular joint during chewing. On examination, rheumatologist suspected chronic arthritis in the setting of juvenile rheumatoid arthritis. Complete blood count showed an accelerated ESR up to 27 mm/h, the rheumatoid factor was 21 IU/ml, and the C-reactive protein was 12 mg/L. PCR typing identified the HLA-B27 gene. The combination of clinical polyarthritis signs and laboratory parameters enabled the diagnosis of juvenile spondylitis. After bicillin prophylaxis and a course of non-steroidal anti-inflammatory therapy for 7 days the articular syndrome (in the knee and temporomandibular joints) was completely reversed. The patient repeatedly consulted a rheumatologist and pediatrician; no contraindications for surgical intervention were revealed.

On day 10 after the first surgery, the patient underwent the main stage of surgical treatment involving elimination of spinal stenosis, posterior open reposition, and transpedicular fixation of the C1 and C2 vertebrae using Harms technique (screws were inserted through the atlas lateral masses and translaminarly into the axis) followed by removal of the halo apparatus.

Control CT examination of the cervical spine revealed complete restoration of the anatomical relationships at the C1–C2 level and elimination of spinal stenosis: anterior atlantoaxial interval was 2.8 mm, and the anteroposterior dimension of the spinal canal was 18.5 mm. Screws were positioned correctly (Fig. 4).

Immediately after the main stage of surgery and removal of the halo apparatus, the patient underwent MRI of the knee and temporomandibular joints. There was effusion in the superior recess of the right knee joint (Fig. 5) and trabecular edema of the subchondral region of the mandibular condyle in the temporomandibular joint on the right (Fig. 6), which indicated an inflammatory lesion.

After the intervention, the patient immediately noted an increase in strength in his left upper extremity; he was verticalized on the 2nd postoperative day. A cervical collar was recommended in the postoperative period. He was discharged from the hospital in satisfactory condition on the 10th day. There was complete regression of neurological symptoms.

The postoperative follow-up period was 2.5 years. Patient was followed-up by a rheumatologist; during the follow-up period, the patient had no complaints of joint

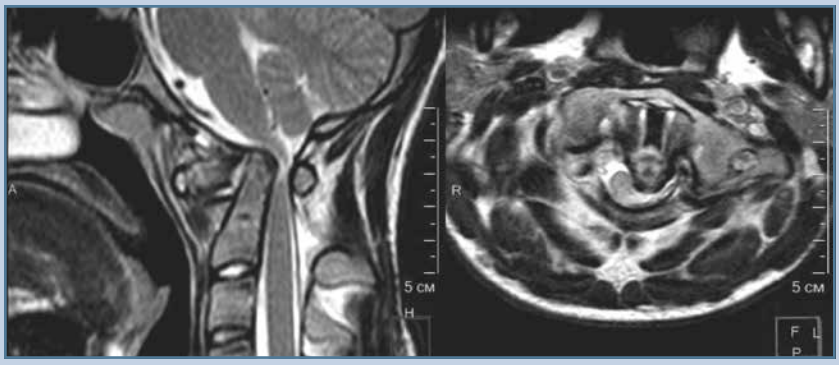

Fig. 1
MRI scans of the cervical spine of the 16-year-old male patient O.: T2-weighted images in sagittal and axial projections; atlantoaxial dislocation with spinal stenosis and spinal cord compression, (myelopathic/myeloischemic) focus with a hyperintense MRI signal at the C1–C2 level, more pronounced on the left

pathology and cervical spine pain; there were no neurological symptoms. A follow-up CT examination 2 years after surgery revealed no changes in the relationships between the atlas and axis and no destabilization of the instrumentation; there were signs for the formation of a bone block in between C1 and C2 (Fig. 7).

In our opinion the feature of this case is an extremely rare primary manifestation of the quite frequent pediatric disease – rheumatoid arthritis with signs of atlantoaxial dislocation and spinal stenosis. Osteopathic treatment which led to deterioration of the neurological condition, and traction with the Glisson loop, which is absolutely contraindicated in instability of the cervical spine, were absolutely erroneous in this patient.

Another feature of this case is a full clinical picture of polyarthritis manifested with clinical radiological (knee and temporomandibular joint lesions) and laboratory (inflammation markers) signs in the early period after the first stage of surgical treatment – application of the halo apparatus. Standard background therapy of rheumatoid arthritis was effective for stopping its clinical manifestations, which is supported by the results of patient follow-up by a rheumatologist for 2.5 years.

In contrast to atlantoaxial dislocations in adult patients with ankylosing spondylitis, which are characterized by pronounced rigidity of this segment and require resection of structures in the craniovertebral junction for complete decompression of the spinal canal contents, all deformity components in the presented case were eliminated due to mobility of the affected segment [10].

Involvement of the cervical spine joints as a first sign of rheumatoid arthritis is rare, but cervical spine pain, especially in combination with neurological manifestations, should alert doctors of any specialty: inflammatory changes in the cervical spine of patients with systemic, polyarticular and enthesitis-associated forms of the disease are typical manifestations of juvenile idiopathic arthritis [7, 8], and a chronic inflammatory process can lead to severe anatomical changes manifesting in atlantoaxial instability with potential risk of spinal cord compression [3] and patient disability.

Conclusion

Non-traumatic atlantoaxial dislocation in children and adolescents should be the reason for comprehensive examination, including testing for juvenile idiopathic arthritis. A quick and correct diagnosis may provide timely etiopathogenic therapy to prevent the development of life-threatening complications.

Indirect spinal cord decompression and reposition of C1–C2 dislocation in the halo apparatus followed by posterior stabilization using Harms technique enabled restoration of the correct relationships in the spinal motion segment and complete reversal of neurological deficit, with the minimal length of fixation. The sequential and staged intervention enabled gradual return of the spinal motion segment to the physiologically correct position, elimination of spinal canal stenosis and neurological deficit, restoration of the segment stability and preservation of the achieved result in the long-term period.

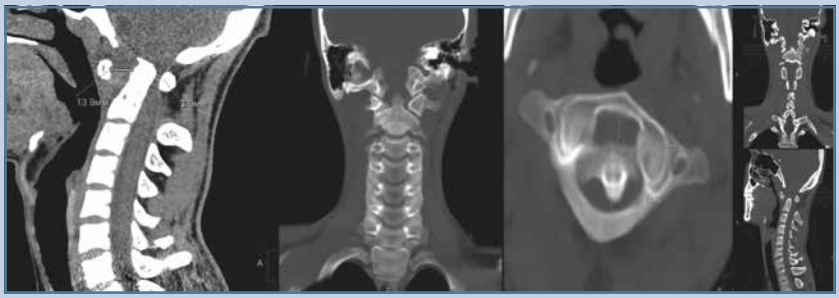

Fig. 2
MSCT scans of the cervical spine of the 16-year-old male patient O. with atlantoaxial dislocation: a significant increase in the anterior atlantodental interval, spinal canal stenosis at the atlas level and erosion of the anterior surface of the odontoid process

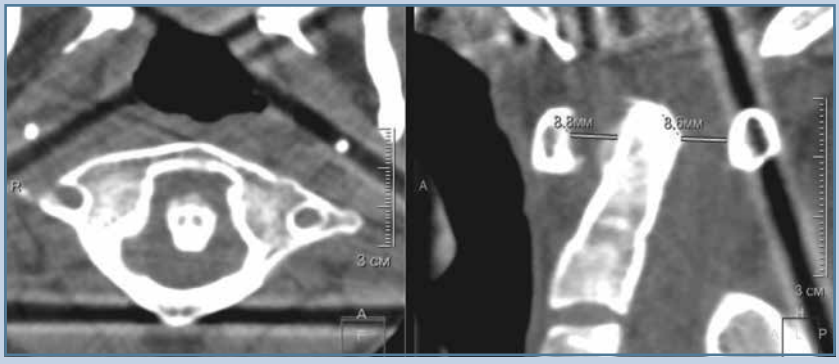

Fig. 3
MSCT scans ofthe 16-year-old male patient O. with atlantoaxial dislocation during reposition in the halo apparatus on the 5th day

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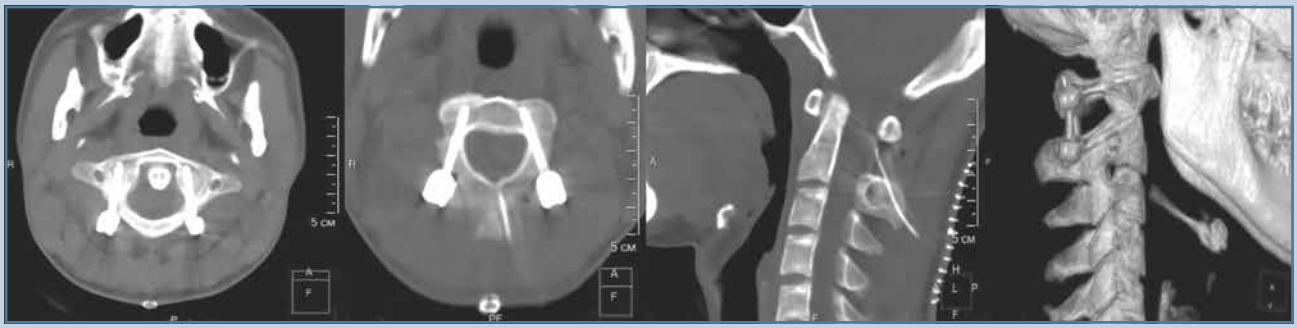

Fig. 4
MSCT scans of the cervical spine of the 16-year-old male patient O. after surgical treatment of atlantoaxial dislocation: there is no spinal stenosis; correct screw position in the C1 and C2 vertebrae

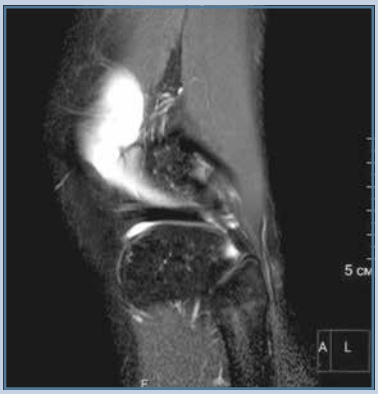

Fig. 5
MRI of the right knee joint of the 16-year-old male patient O.: effusion in the superior recess

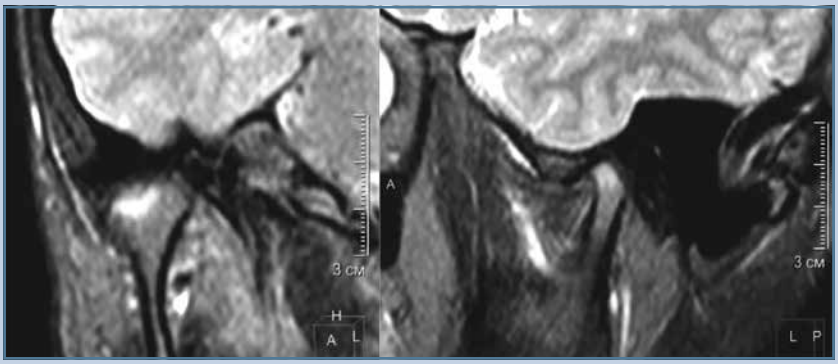

Fig. 6
T2-weighted MRI scans of the right temporomandibular joint of the 16-year-old male patient O. in the frontal and sagittal projections: local area of a hyperintense MRI signal of the head of mandible on the right (signs of bone edema in the subchondral region)

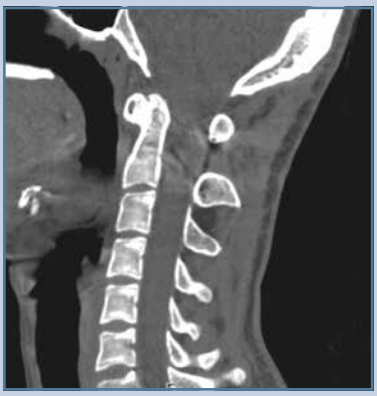

Fig. 7
MSCT of the cervical spine of the patient O. 24 months after surgery: there is no spinal canal stenosis; signs of complete bone ankylosis formation between C1–C2

References

1. **Lvov IS, Grin' AA, Nekrasov MA, Kordonskiy AYu, Sytnik AV.** Ventral decompression techniques in patients with traumatic and non-traumatic atlanto-axial dislocations. Zh Vopr Neirokhir Im N N Burdenko. 2018;82(1):33–40. In Russian.
2. **Brockmeyer DL, Sivakumar W, Mazur D, Sayama CM, Goldstein HE, Lew SM, Hankinson TC, Anderson RCE, Jea A, Aldana PR, Proctor M, Hedequist D, Riva-Cambrin JK.** Identifying factors predictive of atlantoaxial fusion failure in pediatric patients: lessons learned from a retrospective pediatric craniocervical society study. Spine. 2018;43:754–760. DOI: 10.1097/BRS.0000000000002495.
3. **Roonprapunt C, Kokoszka MA, Ulkatan S, Ghatan S.** Value of neurophysiologic monitoring in confirming indirect decompression of severe pediatric non-traumatic C1–C2 subluxation. A case report. Interdiscip Neurosurg. 2018;13:95–99. DOI:10.1016/j.inat.2018.04.005.
4. **Martinez-Del-Campo E, Turner JD, Rangel-Castilla L, Soriano-Baron H, Kalb S, Theodore N.** Pediatric occipitocervical fixation: radiographic criteria, surgical technique, and clinical outcomes based on experience of a single surgeon. J Neurosurg Pediatr. 2016;18:452–462. DOI:10.3171/2016.2.PEDS15544.
5. **Fath L, Cebula H, Santin MN, Coca A, Debry C, Proust F.** The Grisel's syndrome: a non-traumatic subluxation of the atlantoaxial joint. Neurochirurgie. 2018;64:327–330. DOI:10.1016/j.neuchi.2018.02.001.
6. **Bodakci E, Uskudar Cansu D, Erturk A, Calisir C, Korkmaz C.** Can neck pain be an initial symptom of rheumatoid arthritis? A case report and literature review. Rheumatol Int. 2018;38:925–931. DOI:10.1007/s00296-018-4019-z.
7. **Dai Y, Jian FZ, Huang JY.** Research progress of the pathogenesis of atlantoaxial dislocation caused by rheumatoid arthritis. Chin J Contemp Neurol Neurosurg. 2018;185:366–371. DOI: 10.3969/j.issn.1672-6731.2018.05.012.
8. **Pinto JRR, Pedron IG, Utumi ER, Miranda ME, Pinto ECP, Nucci LP.** Temporomandibular joint disorders as the only manifestation of juvenile idiopathic arthritis: a case report. Einstein (Sao Paulo). 2018;16:eRC4003. DOI: 10.1590/S1679-45082018RC4003.
9. **Lisitskiy IYu, Kiselev AM, Kiselev SE.** Rheumatoid atlanto-axial dislocation: a surgical approach. Zh Vopr Neirokhir Im NN Burdenko. 2018;82(1):41–47. In Russian.
10. **Burtsev AV, Gubin AV, Ryabykh SO, Kotel'nikov AO, Pavlova OM.** Syndromic approach in assessing the surgical pathology of the cervical spine. Genii Ortopedii. 2018;24(2):216–220. In Russian.

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