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LESION OF THE CRANIOVERTEBRAL JUNCTION IN ANKYLOSING SPONDYLITIS: RARE CLINICAL OBSERVATIONS

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Two cases of surgical treatment for lesions of the craniovertebral junction in patients with ankylosing spondylitis accompanied by severe neurological symptoms are described. Patients underwent one-step combined interventions. The first stage included occipitospondylodesis using screw instrumentation, and the second — transoral removal of the C2 odontoid process. Surgical treatment resulted in elimination of the spinal cord compression, regression of neurological deficit existed in patients before surgery, and achievement of stable fixation in the intervention area.

Key Words: ankylosing spondylitis, Bechterew's disease, craniovertebral junction, transoral decompression, occipitospondylodesis.

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Ankylosing spondylitis (Bechterew-Marie-Strumpell disease) is a chronic non-infectious inflammatory disease that affects joints of the axial skeleton and belongs to the seronegative spondyloarthritis group. Its incidence rate is 23.8 cases per 10,000 population in Europe, 16.7 cases in Asia, 31.9 cases in America, and 7.4 cases in Africa. The prevalence of disease in males is 3 to 4 fold higher. Ankylosing spondylitis manifests at an age of 20–30 years. The disease is characterized by hereditary predisposition whose genetic marker is the HLA-B27 antigen which is present in more than 90 % of patients [1].

There are five disease forms that differ in involvement of certain skeleton joints as well as internal organs (heart, aorta, kidney). However, spinal lesions are typical of all forms of disease. In ankylosing spondylitis, chronic inflammation of the joints and ligaments leads to ossification and ankylosing of the spine. The usual outcome of the disease is the loss of mobility and the development of kyphotic spinal deformity [2].

Among all non-infectious inflammatory diseases, craniovertebral junction lesions are most often associated with rheumatoid arthritis. There is little data about this pathology associated with ankylosing spondylitis, despite the prevalence of the disease. According to the available data [3–5], the rate of craniovertebral junction lesions in ankylosing spondylitis varies from 0.5 to 35.0 %, which may be explained only by a small number of reported cases. The largest series of cases reported in the current literature included 8 patients operated on in 1990–2008 [2]. As in rheumatoid arthritis, ankylosing spondylitis of the craniovertebral junction is characterized by dislocations and inflammatory changes causing compression of the spinal cord and medulla oblongata [5].

Clinical case 1. A 57-year-old patient *Ts.* suffered from a rhizomelic form of Bechterew's disease for 27 years. Earlier, he underwent surgery for C3–C4 disc herniation. Clinically, the craniovertebral junction lesion manifested as numbness, paresthesias, and weakness in the upper and lower extremities one year before hospitalization. Despite the treatment, neurological disorders progressed, up to the development of spastic tetraparesis. According to an additional outpatient examination, the patient was diagnosed with a craniovertebral junction lesion. At admission to the hospital, he could not move independently; in the neurological status, there were primarily sensory pathway disorders with strength decreased to 3 points in the left upper and lower extremities and to 1–2 points in the right upper and lower extremities. The patient was graded with functional class IIIB according to the Ranawat scale, which was proposed for comprehensive assessment of neurological disorders in rheumatoid lesions of the craniovertebral junction. Blood test: ESR, 25 mm/h; C-reactive protein (–); rheumatoid factor (–); ACCP, 5.4 U/mL; HLA-B27 (+). A CT examination revealed pronounced destruction of the lateral atlantoaxial joints, vertical dislocation of a deformed odontoid process, an additional round-shaped soft-tissue lesion (along the posterior contour of the odontoid process) with ossification sites, which narrowed the spinal canal up to 12 mm in the sagittal plane, and the atlas assimilation. An MRI examination identified a soft-tissue lesion of an inhomogeneous structure at the apex of the C2 odontoid process, which compressed the spinal cord. The substance of the cervical spinal cord contained a focus of myelomalacia. Functional spinal radiography revealed no atlantoaxial instability (Fig. 1).

The patient underwent combined single-stage surgery. Anesthesia was performed using nasotracheal intubation. At the first step, with the patient in the prone position and with his head fixed in the Mayfield frame, occipitospondylodesis was performed using screw instrumentation. Because of destruction of the atlantoaxial joints and anatomical fea-

tures (thin C2 pedicles), polyaxial screws were inserted into the C3 and C4 lateral masses. A plate was fixed to the occipital squama by cortical screws. After modeling, rods were fixed to screw heads and to the occipital plate, and bone autograft was placed along the rods. At the second step, after turning the patient, we performed transoral resection of the anterior atlas arch, C2 odontoid process, and anterior margin of the foramen magnum, removal of inflammatory granuloma, and decompression of the spinal cord and medulla oblongata.

The intervention provided restoration of the anatomical relationships in the craniovertebral region, decompression of the spinal cord and medulla oblongata, and stable fixation (Fig. 2). In the early postoperative period, there was partial regression of neurological disorders in the form of increased strength in the left upper and lower extremities, up to 3–4 points, and in the right upper and lower extremities, up to 2–3 points.

A histopathological examination of tissues removed from the craniovertebral region revealed bone tissue fragments with resorption and formation of bone trabeculae, areas of a markedly thickened fibrotic synovial membrane, multiple histiocytic collagen granulomas, and inclusions of small bone sequesters and osteoid areas.

Clinical case 2. A 71-year-old male patient M. suffered from a central form of Bechterew's disease for more than 30 years. The craniovertebral junction lesion manifested as the development of motor and sensory pathway disorders one year before hospitalization. An additional outpatient examination revealed a pathological process in the craniovertebral junction, with an unknown origin. At the time of hospitalization, spastic tetraparesis with decreased strength in the upper and lower extremities, up to 3 points, prevailed in the neurological status. The patient was graded with functional class II according to the Ranawat scale. Blood test: ESR, 30 mm/h; C-reactive protein, 100.98 mg/mL; rheumatoid factor (–); ACCP, 5.8 U/mL; HLA-B27 (+). A CT examination revealed erosion and vertical dislocation of the odontoid process with pronounced ossification of the ligamentous apparatus and the presence of an additional soft-tissue lesion along the posterior contour of the process; the lesion narrowed the spinal canal to 14 mm. An MRI examination revealed a soft-tissue lesion of an inhomogeneous structure located along the posterior contour of the C2 odontoid process, which compressed the spinal cord. A focus of myelomalacia was found in the spinal cord at the C1 level. According to functional spinal radiography, the deformity was rigid (Fig. 3).

The patient underwent surgery similar to that described in Case 1. At the first step, occipitospondylodesis was performed using screw instrumentation. In this case, given the condition of bone structures, the lower fixation points were the C1 and C2 vertebrae (screws were inserted into the atlas lateral masses and transpedicularly into the C2 body). At the second step, transoral resection of the C2 odontoid process, removal of inflammatory granuloma, and decompression of the spinal cord were performed.

The intervention resulted in decompression of the spinal cord and medulla oblongata and stable fixation of the craniovertebral junction (Fig. 4). In the early postoperative period, there was regression of the neurological deficit in the form of increased strength in the upper and lower extremities, up to 4 points.

The results of a histopathological examination were similar to those of biopsy in Case 1.

Despite the similarity with rheumatoid arthritis, the pathological changes in the craniovertebral junction in Bechterew's disease in the presented cases were characterized by specific features. In addition to the specific signs of spinal column pathology, X-ray and tomographic studies revealed more pronounced destruction of the lateral atlantoaxial joints, erosion and destruction of the odontoid process, and heterotopic ossification of the ligamentous apparatus. The question whether atlas assimilation in the first clinical case was an independent nosology or resulted from concres-

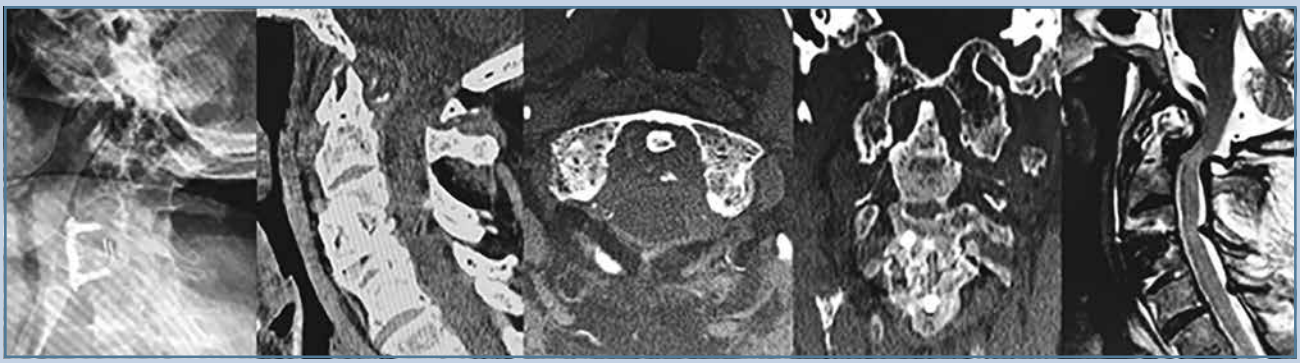

Fig. 1
Preoperative radiographs and X-ray CT and MRI scans of a 57-year-old patient Ts. with a craniovertebral junction lesion in the setting of ankylosing spondylitis

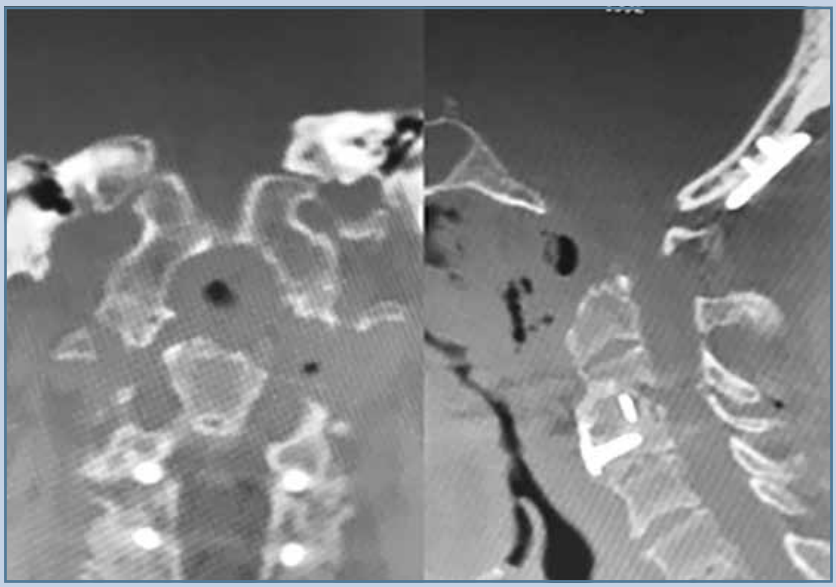

Fig. 2
Postoperative X-ray CT scans of a 57-year-old patient Ts. with a craniovertebral junction lesion in the setting of ankylosing spondylitis

cence of the atlas lateral masses with the occipital condyles in the setting of ankylosing spondylitis may not be answered; however, there are reports in the literature about the acquired nature of these changes [2, 6]. Thus, we may suppose that the cause for the development of inflammatory granulomas was a stress load on the most mobile atlantoaxial junction due to lost mobility of the subaxial spinal segments.

Transoral decompression involves resection of the osteoligamentous structures controlling craniovertebral junction stability. Despite the rigid nature of deformities, we used craniocervical fixation through the posterior approach at the first step. There are various techniques for stabilization of the craniovertebral junction, with most of them being of historical interest. Screw instrumentation available at present at the medical market provides the most stable fixation of the craniovertebral junction, being the optimal method for occipitospondylodesis. Certain technical difficulties occurred only at the transoral step of surgery and were due to the tilted head positioning characteristic of ankylosing spondylitis. Positioning of the patient on the operating table and the use of a special mouth gag, an operating microscope, and long tools enabled solving this problem. It should be noted that the soft palate was not dissected in either of the cases.

We decided to present these clinical cases not only to exemplify a rare localization of spinal lesions in Bechterew's disease but also to illustrate the features of pathomorphology and diagnostic criteria of ankylosing spondylitis of the craniovertebral junction as well as the efficacy of the chosen surgical treatment.

The study did not have sponsorship. The authors declare no conflict of interest.

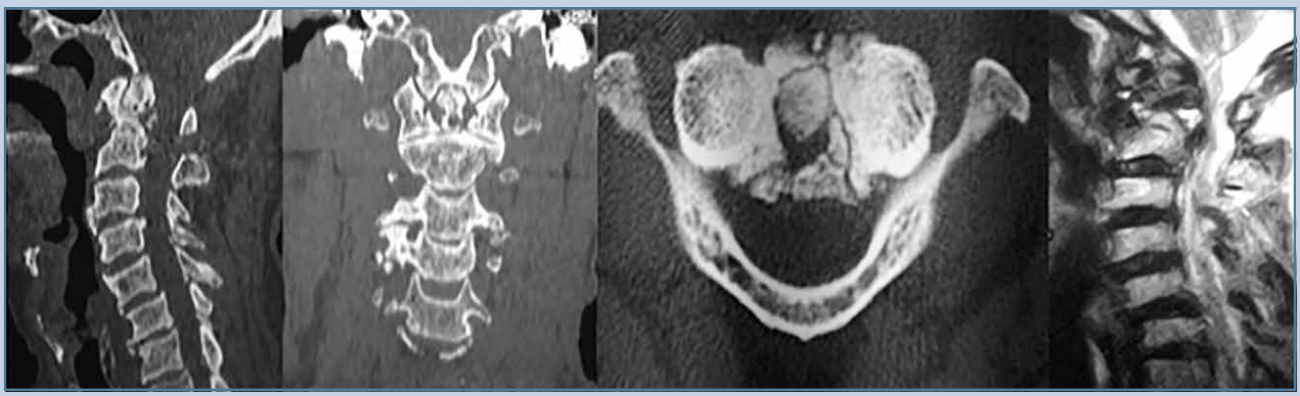

Fig. 3
Preoperative X-ray CT and MRI scans of a 71-year-old patient M. with a craniovertebral junction lesion in the setting of ankylosing spondylitis

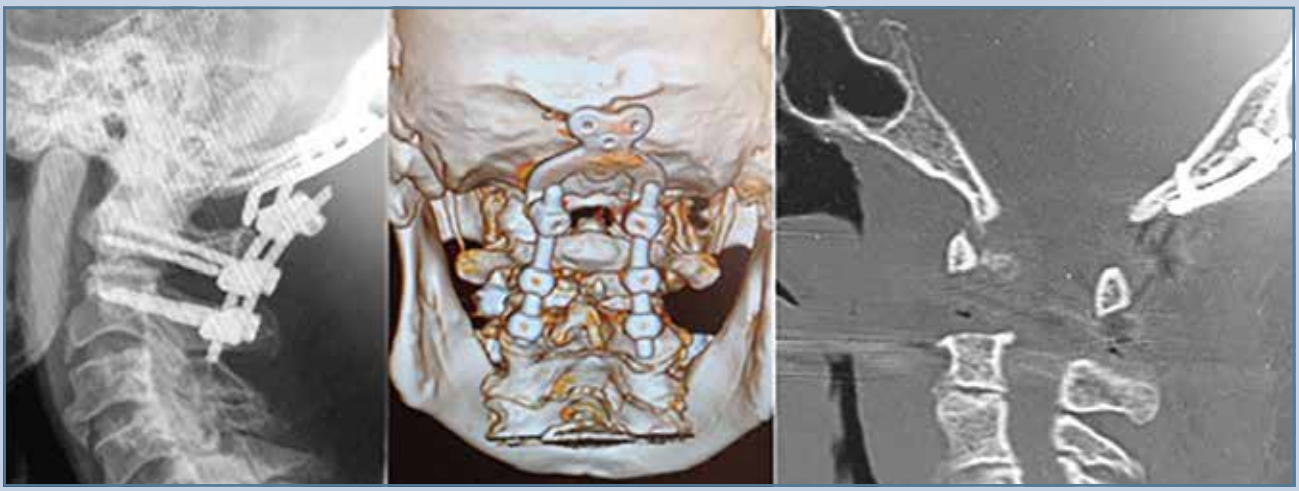

Fig. 4
Postoperative X-ray CT scans of a 71-year-old patient M. with a craniovertebral junction lesion in the setting of ankylosing spondylitis

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