Medical
Research
Journal

CLINICAL MANIFESTATIONS, DIAGNOSIS, AND CONSERVATIVE MANAGEMENT OF KIMMERLE ANOMALY IN NEUROLOGICAL PRACTICE

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Abstract. Kimmerle anomaly (KA) is one of the most common osseous anomalies of the craniovertebral junction and may lead to vertebrobasilar insufficiency and neurovascular complications. The aim of this study was to analyze the incidence of KA in a neurological hospital, to identify the main clinical syndromes, and to substantiate rational conservative treatment approaches. The study included a screening examination of 8,436 patients hospitalized in a neurological clinic over a six-year period, using clinical neurological assessment, radiography, CT, MRI, and ultrasound of cerebral and cervical vessels. KA was identified in 68 patients aged 18–67 years. Three major clinical syndromes were distinguished: pain syndrome, vascular insufficiency syndrome, and autonomic dysfunction syndrome, as well as three degrees of disease severity. Conservative therapy consisting of non-steroidal anti-inflammatory drugs, muscle relaxants, vitamin complexes, physical therapy, and massage proved effective in symptom control and achievement of long-term remission. The findings support the effectiveness of pathogenetically oriented conservative management of KA, particularly in early and moderate stages of the disease.

Key words: Kimmerle anomaly; craniovertebral junction; vertebrobasilar insufficiency; vertebral artery syndrome; conservative treatment.

КЛИНИЧЕСКИЕ ПРОЯВЛЕНИЯ, ДИАГНОСТИКА И КОНСЕРВАТИВНОЕ ЛЕЧЕНИЕ АНОМАЛИИ КИММЕРЛЕ В НЕВРОЛОГИЧЕСКОЙ ПРАКТИКЕ

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Аннотация. Аномалия Киммерле (АК) представляет собой одну из наиболее распространённых костных аномалий краниовертебрального перехода, потенциально приводящую к развитию вертебробазилярной недостаточности и нейроваскулярных осложнений. Целью исследования явился анализ частоты выявления АК в условиях неврологического стационара, уточнение клинических синдромов, а также обоснование рациональной консервативной тактики лечения. Материалы и методы включали скрининговое обследование 8436 пациентов, госпитализированных в неврологическую клинику в течение шести лет, с применением клинико-неврологического, рентгенологического, нейровизуализационного и ультразвукового методов исследования. АК была выявлена у 68 пациентов в возрасте от 18 до 67 лет. В результате анализа выделены три ведущих клинических синдрома: болевой, синдром сосудистой недостаточности и вегетативной дисфункции, а также три степени тяжести заболевания. Показано, что консервативная терапия, включающая нестероидные противовоспалительные препараты, миорелаксанты, витаминотерапию и немедикаментозные методы, позволяет эффективно купировать симптомы и достичь длительной ремиссии. Полученные данные подтверждают

целесообразность патогенетически обоснованного консервативного лечения АК на ранних и средних стадиях заболевания.
Ключевые слова: аномалия Киммерле; краниовертебральный переход; вертебробазилярная недостаточность; синдром позвоночной артерии; консервативное лечение.

KIMMERLE ANOMALIYASINING KLINIK KO‘RINISHLARI, DIAGNOSTIKASI VA KONSERVATIV DAVOLASH USULLARI

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Annotatsiya. Kimmerle anomaliyasi (KA) kraniovertebral sohaning eng ko‘p uchraydigan suyak anomaliyalaridan biri bo‘lib, vertebrobazilyar yetishmovchilik va neyrovaskulyar asoratlarning rivojlanishiga olib kelishi mumkin. Tadqiqotning maqsadi nevrologik statsionar sharoitida KA uchrashish chastotasini baholash, asosiy klinik sindromlarni aniqlash va konservativ davolashning oqilona usullarini asoslashdan iborat. Tadqiqotda 6 yil davomida nevrologik klinikaga yotqizilgan 8436 bemor skrining tekshiruvdan o‘tkazildi. Tekshiruvlar klinik-nevrologik, rentgenologik, KT, MRT va ultratovush usullarini o‘z ichiga oldi. KA 18–67 yoshdagi 68 bemorda aniqlandi. Uchta asosiy klinik sindrom: og‘riq sindromi, tomir yetishmovchiligi sindromi va vegetativ disfunksiya sindromi, shuningdek kasallikning uch darajali og‘irlik shakllari ajratib ko‘rsatildi. Konservativ davolash (NSAIDlar, miorelaksantlar, vitaminoterapiya va reabilitatsion usullar) klinik belgilarni samarali kamaytirish va uzoq muddatli remissiyaga erishish imkonini berdi. Olingan natijalar KA ning erta va o‘rta bosqichlarida konservativ yondashuvning samaradorligini tasdiqlaydi.
Kalit so’zlar: Kimmerle anomaliyasi; kraniovertebral soha; vertebrobazilyar yetishmovchilik; umurtqa arteriyasi sindromi; konservativ davolash.

Introduction. The craniovertebral region (CVR), a mobile structure of the cervical spine, is also its most vulnerable area. Clinical manifestations of CVR damage are varied. The vertebral artery and its accompanying nerve structures are most involved in the pathological process, forming the primary pathogenetic core of the disease.

One of the most common anomalies of the atlas is the bony bridge, described in 1923 by H. Hayek [1, 2]. It is located between the posterior edge of the articular process of the atlas and the posterior border of its arch and forms an opening through which the vertebral artery (VA) and occipital nerve pass. This pathology was described in more detail in 1930 by the Hungarian physician A. Kimmerle, who noted that this change could lead to cerebrovascular disorders, and subsequently received his name – Kimmerle anomaly (KA).

The development of this bone pathology is based on a mismatch between the neural elements of the spinal cord and vertebral tissues. These include occipitalization of the atlas or remnants of the proto-atlas with progressive growth, gradual calcification of the atlanto-occipital membrane due to microdamage or microbleeds sustained during CVO injuries [8]. As a result of these changes, the atlanto-occipital ligament (AOL) passes not through the groove of the atlas arch, but through a foramen formed on one side by the groove and on the other by the ossified atlanto-occipital ligament, which forms a kind of bony bridge. This bridge limits the free movement of the AOL and occipital nerve during head movements, especially during head turns. A.A. Lutsik [7] attaches primary importance to cicatricial degeneration of the arterial wall and periarterial tissue due to prolonged trauma to the VA in the AC region, which leads to the development of intimal damage, early formation of atherosclerotic plaques narrowing the lumen of the vessel, and/or dissections [2, 3, 8, 10]. As a result, sudden head movements can provoke a process of destabilization of the hemodynamics of the affected artery and turn into a source of blood supply disturbances in the distal parts of the VBS via the Bow Hunter Stroke mechanism [7, 9, 11].

Thus, the pathogenesis of hemodynamic disorders in the VBS with AC is etiologically determined by the development of a complex compression-stenotic irritative effect, as a result of which a decrease in the volumetric blood flow through the PA and the development of circulatory failure to varying degrees in the VBS are observed. During anatomical studies, various variants of AC are detected in approximately 30% of the population [5, 7]. In patients with clinical manifestations of vertebrobasilar dysfunction (cochleovestibular disorders, ataxia, visual impairment, etc.) or with myofascial pain similar to Tonzik's syndrome, AC is detected in 7.6% of cases [2, 11]. The bridge may be incomplete, unilateral or bilateral, therefore, different degrees of this anomaly are distinguished. It should be noted that the thickness of the bridge and the size of the opening also vary significantly [7]. The clinical presentation of the initial manifestations of AK is characterized by a variety of symptoms (dizziness, autonomic dysfunction, headaches, transient visual changes, neck pain), which creates difficulties in determining the prevalence of the initial manifestations of AK. This is due to the fact that patients, and often even physicians, do not consider the recurring headaches in the initial stages of the disease to be a serious illness. Consequently, they are observed for a long time with various diagnoses, such as migraine, cephalgia, tension headache, vegetative-vascular dystonia, etc., by different specialists and receive disparate examinations and treatments, which ultimately prevents the formation of the necessary statistical base.

In the advanced stage, which develops in 20% of all cases of AC, the clinical picture of the disease is dominated by symptoms of ischemia of the structures supplied by the VBS, such as initial manifestations of cerebral circulatory failure (IMCSF) and/or paroxysmal cerebrovascular accidents (PACE) [4, 8, 11]. IMCSF manifests as dizziness, hearing impairment, noise, tinnitus, headache in the occipital region, sensation of "dots or sand" before the eyes, and sometimes color photopsies. In the interictal period, patients complain of a veil before the eyes, pressure in the external auditory canal, noise, hyperacusis, fatigue, and sleep disturbances. Focal neurological symptoms are absent. The dependence of each of the symptoms on body movement in space is clearly visible. NMCDs are characterized by more pronounced symptoms: paroxysmal non-systemic dizziness, often with nausea and vomiting (when extending and turning the head); hearing impairment (noise, ringing in the ears); headaches (mainly in the occipital region); visual impairment (blurred vision, photopsia, visual field defects); pathological pyramidal signs; sensory disturbances; cerebellar and bulbar symptoms; mono-, para-, or tetraparesis; sudden falls without loss of consciousness (drop attacks); sudden falls with loss of consciousness (Unterharnscheidt syncopal syndrome). All symptoms of NMCD should persist for no more than 24 hours [6, 10].

The most pronounced complication of circulatory disorders in the cerebrovascular system is ischemic stroke in the cerebrovascular system, in which small foci of softening are formed in the region of the cerebellum and medulla oblongata, which is clinically manifested by persistent neurological insufficiency in the form of general cerebral and focal neurological symptoms, damage to the mid-brainstem and cerebellar structures, which persist in patients for more than 24 hours [3]. Study Objective. The aim of our study was to analyze the incidence of AK in a neurological hospital, develop diagnostic criteria, and develop rational treatment methods.

Characteristics of materials and methods. Our study used the results of a screening examination of patients hospitalized in the neurology clinic of the Voronezh State Medical University over a six-year period (a total of 8,436 patients). Exclusion criteria included the presence of verified traumatic brain injury, cervical spine injury, vascular anomalies, and tumors of this region. In addition to the clinical neurological examination, the complex of examinations included X-rays of the spine and skull, CT and MRI of the brain and spinal cord, ultrasound of the neck and brain vessels (ultrasound, dopplerography, and electroencephalography), and electroencephalography (EEG). All examinations were performed over a three-year period, with a frequency of two to six times per year. The study identified 68 patients with AK aged 18 to 67 years, including 42 (61.8%) women and 26 (38.2%) men. In most cases (74.6%), clinical manifestations of neurological impairment stabilized by the age of 20–25 years. However, anamnesis studies revealed that 43.8% of those examined had previously experienced specific complaints indicative of CVO pathology, which were assessed by specialists as spasmodic torticollis, cervicobrachialgia, syncope, hemocerebrospinal fluid flow disturbances, and others.

Results. The clinical picture identified in patients with AK was characterized by a variety of symptoms, but, taking into account the leading clinical sign, 3 main neurological syndromes were identified.

1. Pain syndrome characterized by a half-headache localized predominantly in the occipital region (91.6% of observations) with irradiation to the eye or ear canal, accompanied in 43.9% of cases by photopsies, visual field loss (34.6%) or metamorphopsia (21.5%).
2. Vascular insufficiency syndrome, manifested in 42.6% of cases as dizziness (79.4%), unsteadiness when walking, and intermittent impairments of coordination tests (61.7%), accompanied by nausea and vomiting in 54.4% of cases. Less frequently, patients reported the appearance of noise illusions in the form of buzzing, rustling, squeaking, and whistling, localized either in the ear or in the head (45.8%). A characteristic feature of this syndrome was its association with head rotation, manifested by its intensification with movement, which forced patients to assume a certain posture (so-called head positioning).
3. Vegetative syndrome (panic attack syndrome) was recorded in 41.1% of those examined. It was characterized by sudden onset, accompanied by a feeling of "hot flush" in the head or neck, accompanied by unmotivated fear, anxiety, a feeling of suffocation, and pronounced dysfunction of the autonomic nervous system (chill-like tremors, numbness in the hands, blood pressure instability, etc.). After the attack, patients experienced polyuria and panasthenia. Their duration ranged from tens of minutes to several hours.

Observation of patients over time allowed us to identify three degrees of disease severity:

I mild – characterized only by the presence of pain syndrome in combination with hemoliquorodynamic disturbances;

II moderate – manifestations of the first degree were accompanied by permanent and paroxysmal autonomic dysfunctions with a frequency of paroxysms of 3–4 times a year;

III severe – in this case the frequency of the main syndromes varied from monthly to weekly. The follow-up analysis showed a direct dependence of the severity of the course on the duration of the disease and the possibility of a gradual and progressive transformation of the clinical forms of the disease from mild through moderate (68.7±0.3%, $p<0.05$) to severe (31.3±0.2%, $p<0.05$).

Treatment of AK. The choice of treatment strategy for the clinical manifestations of AK was based on the pathogenetic principle, as etiologic treatment requires surgical intervention not only on the atlas ring itself but also reconstructive angioplasty of the vertebral artery, which is not always advisable in the early stages of the disease, which represent the majority of clinical cases. Therefore, preference in choosing a treatment regimen was given to therapeutic methods.

The primary pathogenetic model was based on the principles of a "vicious circle," which centers on irritation of the occipital nerve, leading to pain and protective tension in the neck and scalp muscles. In turn, the developing myotonia disrupts normal biomechanical relationships, leading to further irritation of the occipital nerve and a corresponding progression of pain, accompanied by autonomic dysfunction due to irritation of the Frank nerve and the plexuses accompanying the vertebral artery, as well as cerebrovascular insufficiency syndromes in later stages. The therapeutic impact on this pathogenetic model was based on methods that allow for the relief of pain, reduction of myotonic symptoms, and improvement of the trophism of vascular and nerve structures. The basic medications used were fast-acting non-steroidal anti-inflammatory drugs (lornoxicam, etc.), peripheral muscle relaxants (tolperisone), and complex vitamin-containing drugs with trophic and anabolic action. Additionally, the course of treatment included therapeutic exercise and massage.

Table 1 presents the main clinical effects observed in patients of various age groups receiving lornoxicam for pain relief.

Age (years)	Number of patients	Pain syndrome relief, %	Cardiac dysfunction, %	Gastrointestinal disorders, %
18–20	4	98	0	0
21–30	13	94	0	5
31–40	10	87	4	11
41–50	25	79	9	19

51–60	9	73	17	23
61–67	7	61	28	32

As can be seen from the data presented, lornoxicam, prescribed for pain relief, was effective in all patients. Cardiac dysfunction and gastrointestinal disorders were more common in older patients. Thus, based on the data obtained, the NSAID lornoxicam has proven to be a highly effective analgesic in patients of all age groups; however, due to the rapid development of side effects, the drug is best prescribed once for emergency pain relief.

Discussion and Conclusions. Ossification of the atlantooccipital ligament is a common anomaly of the atlantooccipital ligament structure. This anomaly occurs in virtually all age groups, with a peak prevalence in those 25 years and older. Three main syndromes predominate in the clinical presentation: pain, vascular insufficiency, and autonomic dysfunction. The severity of clinical manifestations corresponds to three degrees, reflecting different types of progression – mild, moderate and severe, characterized by the frequency of exacerbations and depends on the severity of the anatomical defect. The close arrangement of various elements, as well as the combination of various pathological processes developing in the cervical spine, in most cases complicates active surgical interventions, which requires the development of conservative treatment methods aimed at relieving pain, reducing myotonic tension in the muscles of the neck and scalp, and activating reparative processes in the affected area.
Oxicam-type painkillers, particularly lornoxicam, are highly effective in patients with AK as an emergency treatment for the development of pain after a single dose. If longer-term use of this medication is necessary, protective measures should be taken to prevent damage to the gastrointestinal tract. The drug should also be prescribed with caution to older patients due to the risk of developing cardiac pathology. The use of muscle relaxant and reparative (vitamin) drugs in the treatment regimen allows for the effective removal of the main clinical manifestations of AK and the introduction of the disease into a state of remission for a period of up to 9 months.

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