Clinical cases

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Visual function abnormalities in Arnold-Chiari syndrome: Analysis of two cases

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Purpose: To analyze clinical manifestations of visual function abnormalities in Arnold-Chiari syndrome.
Material and Methods: Two patients with Arnold-Chiari syndrome (ACS) were under our observation at SI "The Filatov Institute of Eye Diseases and Tissue Therapy of the National Academy of Medical Sciences of Ukraine" and Eye Microsurgery Center, Multipurpose Medical Center, Odesa National Medical University. They underwent general clinical eye examinations.
Results: We reviewed the cases of two patients with ACS. Major manifestations of the disease and the potential for treatment for preserving visual functions were determined.
Conclusion: Taking a comprehensive approach to medical issues (especially those related to comorbid conditions) was always important. There are several ophthalmological issues that require consultations with neuropathologists and neurosurgeons for making a correct diagnosis and starting a subsequent treatment early. The cases presented showed that possible ACS manifestations may include e.g., strabismus and papilledema. Knowing these manifestations is a prerequisite for solving the diagnostic and treatment issues of the disease. There is no specific pathognomonic clinical sign or group of symptoms for the clinical diagnosis of ACS, and the disease can be diagnosed only by neuroimaging. Early identification of these ocular abnormalities allows suspecting and detecting this malformation early. Comprehensive treatment over many years can stabilize visual functions in patients with ACS.

Key words:
Arnold-Chiari syndrome, head MRI, optic nerve, neuritis, treatment

Introduction

Arnold-Chiari malformation (ACM) can develop both in children and adults in the presence of fast growth of the brain and relatively slow growth of skull bones. An ACM is usually considered a congenital abnormality.

Basilar invagination (BI) and Chiari I malformation (CM1) are the most common adult craniovertebral junction malformations, they are frequently associated with each other and present synchronously [1]. Additionally, they are very important anomalies that introduce instability and compression in the occipitocervical transition region and have complex clinical characteristics [1, 2]. The incidence for a combination of BI with CMI is estimated between 2.4/100,000 in children and 9.6 to 19.7/100,000 in adults [3].

Arnold-Chiari syndrome (ACS) is a disorder manifested by the displacement of the cerebellum and medulla oblongata through the foramen magnum inside the spinal canal due to the discrepancy between the size of the posterior fossa and its contents [4, 5].

Cleland was the first to describe Chiari II or ACM in 1883. The Austrian pathologist Chiari and the German professor of pathology Arnorld described the syndrome in 1891 and 1894, respectively. The incidence of the disease ranges from 3.3 to 8.2 per 100,000. The pathogenesis has not been elucidated yet. Most frequently, the involvement of congenital osteoneuropathy, traumatic injury to the sphenoid and occipital bones of the clivus due to birth trauma, and cerebrospinal fluid (CSF)-induced hydrodynamic shock at the walls of the central spinal canal have been considered. Most researchers, however, believe that the disease has a genetic basis. The status of the structures located in the posterior fossa may undergo changes in patients with this syndrome [6, 7].

The ACM is a condition in which there is a downward displacement of cerebellar structures (and sometimes the medulla oblongata and pons) through the foramen mag-

num, leading to their compression. The upper neck segments of the spinal cord, lower cranial nerves and cerebellar arteries also undergo compression. The CSF outflow from the fourth ventricle becomes inadequate, causing CSF circulation disorder [4], which leads to CSF accumulation and increased intracranial pressure (ICP) with resultant CSF hypertension syndrome and hydrocephalus. Hydrocephalus is a serious condition characterized by enlarged ventricles, due to excess of CSF. This may potentially cause increased ICP, leading to brain tissue damage and other serious complications. Early diagnosis and treatment of hydrocephalus is very important for improved prognosis and quality of life of patients. Visual abnormalities in hydrocephalus are associated with excess accumulation of CSF in the ventricles, which causes increased ICP, potentially leading to brain tissue damage and affecting visual functions and functional changes. The ocular manifestations include strabismus, papilledema, optic atrophy and neuritis. A diagnosis of one of these conditions should raise the suspicion for ACM [8].

Chiari malformation (CM) is a cluster of related developmental anomalies of the posterior fossa ranging from asymptomatic to fatal. Cranial and spinal decompression can help alleviate symptoms of increased CSF pressure and correct spinal deformity [9].

Several theories have been proposed to explain the development of ACM [10]:

1. Molecular genetic theory; in this theory, the disease develops due to primary defects in the genes responsible for hindbrain segmentation and the development of the occipital bone and related structures. However, the vast majority of Chiari malformations do not have a hereditary background and are known to be sporadic malformations. Therefore, if we accept this theory as one of the underlying causes of Chiari malformations, the occurrence of new mutations spontaneously or as the result of fetal exposure to teratogen agents is the more probable explanation [11].

2. Hydrodynamic Theory. In this theory, the primary development of hydrocephalus poses a downward pressure upon the posterior fossa contents and result in the herniation of the cerebellum and brainstem through the foramen magnum [12].

3. Compression theory; it this theory, an inadequate growth of the posterior fossa causes a disproportion between size of the posterior fossa and its neural contents, leading to neural tissue compression and displacement through the foramen magnum [13].

4. CSF insufficiency; in this theory, a neural tube closure defect during early fetal development causes CSF loss, with the CSF volume available being insufficient to provide an appropriate stretching of the ventricular system, which leads to a decrease in the size of the posterior fossa [14].

There are four types of ACM. Arnold-Chiari type I malformations (ACM-I) are characterized by abnormally shaped cerebellar tonsils displaced below the level of the foramen magnum. Hydrocephalus is rare in CM-1 [11]. Usually, ACM-I is a disorder of mesodermal origin, but neuroectodermal and acquired forms have also been reported. This is a group of congenital conditions involving the downward displacement of either the posterior cerebellum alone or the posterior cerebellum and lower medulla oblongata through the foramen magnum into the cervical canal. The disorder is often accompanied by increased ICP and the development of visual abnormalities. Cerebellar symptoms of ACM-I include ataxia and nystagmus. Other symptoms include hoarseness, vocal cord paralysis, dysarthria, palatal dysfunction, pharyngeal achalasia, chanting speech, nystagmus, oscillopsia, central and obstructive apneas, headache, syncope, sensorineural deafness, sinus bradycardia, hiccups, general weakness, hyperreflexia, Babinski sign, and sensory and motor neurological deficit caused by syringomyelia which frequently accompanies the disease [15].

ACM-II is the most common type of ACM and involves more severe changes (the downward displacement of the cerebellum through the foramen magnum with the dislocation of the brainstem). This variant is usually associated with spina bifida and other developmental abnormalities of the brain, spinal cord and meninges. Hydrocephalus is present in 70% of cases and is obstructive in nature. It is in ACM-II that fundus changes, low visual acuity, visual field loss, diplopia, strabismus, upward gaze paralysis, downward globe displacement and headache are most common [16, 17].

Arnold-Chiari type III malformations (ACM-III) are the rarest. These malformations combine a small posterior fossa with a high cervical or occipital encephalocele. ACM-III is often associated with displacement of cerebellar structures into the encephalocele with inferior displacement of the brain stem into the spinal canal. Hydrocephalus is present in 50% of cases and is obstructive in nature due to stenosis of the aqueduct of Sylvius or the presence of Danfy-Walker malformation [18]. ACM-III is associated with a high rate of newborn mortality and survivors suffer from neurologic deficits (epilepsy, limb paresis or paralysis, cranial nerve lesions, cerebellar disorders), mental retardation, and visual acuity and ocular motility abnormalities [19].

Arnold-Chiari type IV malformations (ACM-IV) are characterized by cerebellar hypoplasia unrelated to other ACM [20].

Unfortunately, as there is no specific pathognomonic clinical sign for ACM, the diagnosis of ACM can be only confirmed by neuroimaging evidence. Sometimes years are required to establish the correct diagnosis.

Case descriptions

Case 1

A six-year-old boy (medical record ID, 595698) was born by cesarean delivery at 37 weeks (birth weight, 2550 g; birth height, 49 cm). He was referred to magnetic resonance imaging (MRI) due to the presence of swallowing difficulty, excessive saliva, irritability during feeding, vomiting, and strabismus, and was diagnosed with ACS. At two

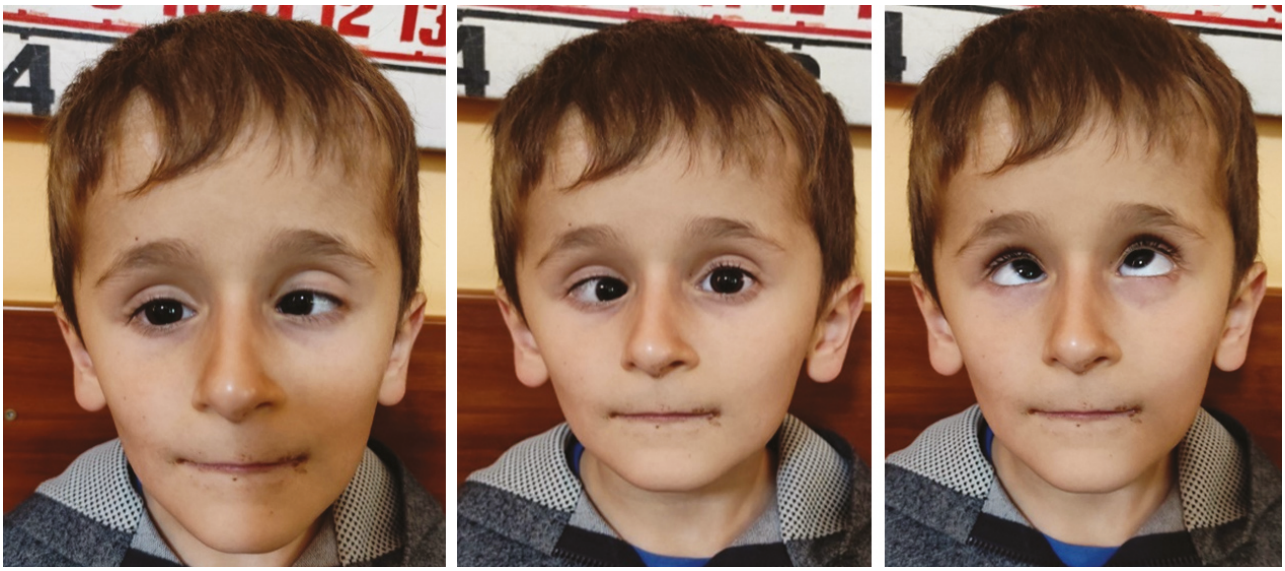

Fig. 1. Photographs of eye movements in a six-year-old patient

weeks after birth, the child underwent ventriculoperitoneal shunting for hydrocephalus. Subsequently, his parents took him to an ophthalmologist due to strabismus and diplopia. The patient was meticulously examined by both the ophthalmologist and a neuropathologist who confirmed the diagnosis of ACM-I. The neurosurgical examination found no motility deficit, no neurological deficit caused by syringomyelia, and no specific symptoms of ACM (Fig. 1).

The ophthalmological diagnosis was anisotropy OU, mixed astigmatism OU, comitant esotropia OU, severe strabismic amblyopia OS and optic neuropathy OS.

A neurological diagnosis of ACM-I was established based on the symptoms (headache, strabismus, and MRI evidence of a disproportion between the size of the posterior fossa and its neural contents).

The visual acuity was 1.0 with a correction of +1.0D sph OD and 0.04 with a correction of +3.0 D sph 2.0D cyl ax 168° OS. His sensorimotor examination demonstrated a comitant esotropia of 10 degrees (Fig. 1). The color test showed monocular color vision. The patient showed foveal fixation in the right eye and macular fixation in the left eye.

The axial length as assessed by ultrasound A-scan biometry was 23.15 mm OD and 22.25 mm OS. Pachymetry measurement was 0.565 mm OD and 0.565 mm OS. Phosphene current threshold was 77 μA at 45 Hz OD and 80 μA at 45 Hz OS. Maculotester was used for assessing the function of the macula lutea. The minimum illuminance at which the patient could see Haidinger's brushes was 5.0 lux OD and 9.0 lux OS. The right eye could determine the shape, color and direction of rotation of rotated Haidinger's brushes. It was not, however, able to discriminate the stopped Haidinger's brushes. The left eye was able to determine the color and direction of rotation, but not the shape of Haidinger's brushes. The patient had a peripheral visual field loss of 15–200 in the right eye and nasal visual field depression in the left eye. The retina was attached in both eyes on B-scan ultrasonography.

On examination, both eyes showed a clear cornea, a moderately deep anterior chamber, and a round pupil (about 3 mm in diameter, with a normal pupil light reflex). The lens and vitreous were clear. The fundi showed some pallor in the temporal halves of discs, the discs had clear boundaries, and there were optic nerve drusen in the right eye. The arteries were constricted and veins were tense, but not congested. In both eyes, 7-mm rectus muscle recession was performed for cosmesis and recovery of binocular vision. After surgery, the Hirschberg test showed no deviation. Surgery was conducted because the patient was unresponsive to other treatments (prismatic treatment and pleoptic treatment). It strengthened the oculomotor muscles and restored eye alignment in the patient. Subsequently, the patient was administered pleoptic treatment and a balanced vitamin and mineral supplement containing carotenoids and anthocyanins, and was allowed to do swimming. He is still under observation, his health status is stable, and the binocular vision has recovered.

On the completion of treatment, the visual acuity was 1.0 with a correction of +1.0D sph OD, and 0.08 with a correction of +3.0D sph 2.0D cyl ax 168° OS.

Discussion of case 1

Unrecognized intraocular hypertension may cause the displacement of the brainstem through the foramen magnum, leading to fatal consequences. The decision for selecting the anesthetic technique for this neurosurgical patient was made by both the anesthesiologist and the neurosurgeon. The decision for performing ventriculoperitoneal shunting at two weeks after birth was grounded, and this surgery saved the patient's life. Strabismus surgery aims to achieve precise ocular alignment not only to improve cosmetic appearance, but also to restore the binocular vision. In this case, surgery for esotropia with hyperfunction of the inferior oblique was performed for cosmesis, and to improve visual acuity and restore binocular vision. Visual acuity improved due to additional pleoptic treatment.

Given that ACM is a rare disease and the literature is scant on ACM treatment data, a long-term observation by the ophthalmologist and neurosurgeon is required. Performing surgical decompression with dura mater plasty before puberty may prevent scoliosis progression in children with ACM-I. During surgical decompression, the neurosurgeon removes a small bone fragment by cutting the dura mater and inserts autofascia within the latter to increase the space for the brain and decrease the ICP.

Extending the fornix may be recommended in a child with ACM associated with craniosynostosis. Children with ACM-I may do sports due to a low risk of increased disease severity. Early diagnosis and adequate surgical decompression with dura mater plasty before puberty are critically important for the amelioration of the effect of the disease on the child's well-being [21].

Cavernous angioma of the vertebral body with basilar invagination and/or syringomyelia has been reported in patients with ACM but was not observed in this case. The pathogenesis of cavernous angioma of the vertebral body with or without basilar invagination and/or syringomyelia is supposed to be associated primarily with atlantoaxial instability. Surgical treatment in such cases should be aimed at atlantoaxial stabilization and segmental arthrodesis. With the exception of atlantal assimilation, the involvement of the occipital bone is not indicated and does not provide optimal stability [22, 23, 24].

Case 2

A male patient was under our observation for 15 years. He presented to us in 2009 when he was 38 years old. After he had suffered a viral infection, he noted metamorphopsia and decreased visual acuity in both eyes.

On examination at presentation, the best-corrected visual acuity (BCVA) was 0.3 in the right eye and 0.2 in the left eye. The cornea was transparent and spherical. Isolated vitreous floaters were seen. The optic disc was edematous and hyperemic. There were individual hemorrhages in the periphery and around the optic disc due to venous congestion and compression and abnormal blood supply from capillaries. The optic disc projected within 3.5-4.0 D into the vitreous above the level of the retina, disc margins and vacular funnel were blurred due to edema that extended to the posterior pole and macular region exhibiting pathological reflexes, plasmorrhagia in the form of whitish-yellow spots and transudation foci in the edematous area. The retina showed dilated and tortuous veins of irregular caliber and narrowed arteries (Figs. 2A, 2B).

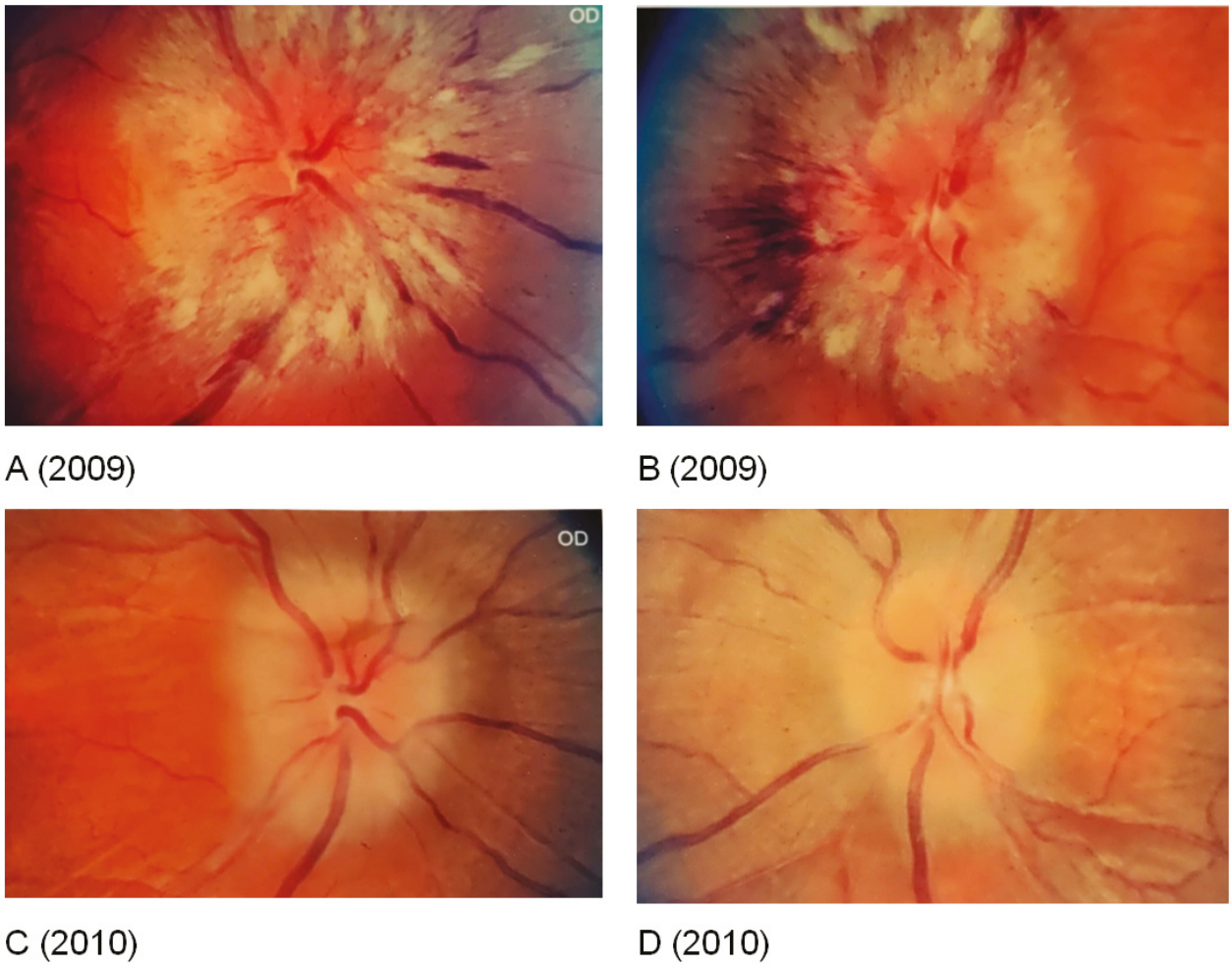

Fig. 2. Fundus photographs for a patient born in 1971: right and left fundus photographs at presentation (A and B, respectively) and at one year after the initiation of treatment (C and D, respectively).

Perimetry revealed a 3-4-cm-enlarged blind spot in both eyes. Phosphene current threshold was 120 μA OD and 110 μA OS. A diagnosis of optic neuritis OU was presumed. Brain MRI was performed to differentiate between optic neuritis and papilledema, and found an increased foramen magnum. There was no MRI evidence of direct optic nerve lesion.

The neuropathologist was consulted and made a diagnosis of decompensated ACM.

On the basis of clinical findings and results of investigation, the patient was finally diagnosed with papilledema OU.

The patient had no MRI with contrast because the above changes were revealed with standard MRI.

Corticosteroids were prescribed according to the scheme. Antiviral therapy was administered because an acute viral infection preceded the eye disease. Inosin pranobex 1000 mg was administered thrice daily for 15 days. Endonasal electrophoresis of non-steroidal anti-inflammatory drugs combined with corticosteroids were administered to relieve optic nerve edema and for anti-inflammatory and neurotrophic purposes. Antihistamines and phonophoresis of resorptives were administered because an autoallergic component is always present in optic nerve inflammation (Fig. 2).

The neuropathologist was consulted repeatedly and agreed with our prescriptions.

At one year, in the presence of the treatment, the BCVA improved to 0.6 OD and 0.5 OS. The optic disc projected within 2.0-2.5 D into the vitreous above the level of the retina.

Perimetry revealed a 1.5-2-cm-enlarged blind spot in both eyes. Phosphene current threshold was 100 μA OD and 96 μA OS.

There was a reduction in edema near the optic nerve and some relief of plasmorrhagia and hemorrhages (Figs. 2C, 2D).

Over the last fifteen years, the patient received diode-laser optic disc stimulation [25], citicoline 1000 mg daily (administered as oral solution) for a month, and meldonium 500 mg daily (administered as oral solution) for a month for neuroprotection.

In 2024, on examination, the BCVA was 0.4 OU. The status of the fundus stabilized. Optic disc pallor developed, the disc margin was clear cut, and the vessels were narrowed (Fig. 3A).

Phosphene current threshold was 100 μA OD and 96 μA OS.

There was OCT evidence of some bilateral thinning of the retinal ganglion cell complex (GCL+IPL) (96 and 97 nm, in the right eye and left eye, respectively) (Fig. 3B). Additionally, there were reductions in the thickness of almost all optic disc-related parameters, but the parameters of peripapillary retinal nerve fiber layer (RNFL) were normal (Fig. 3C).

The patient was diagnosed with partial optic atrophy and ACM.

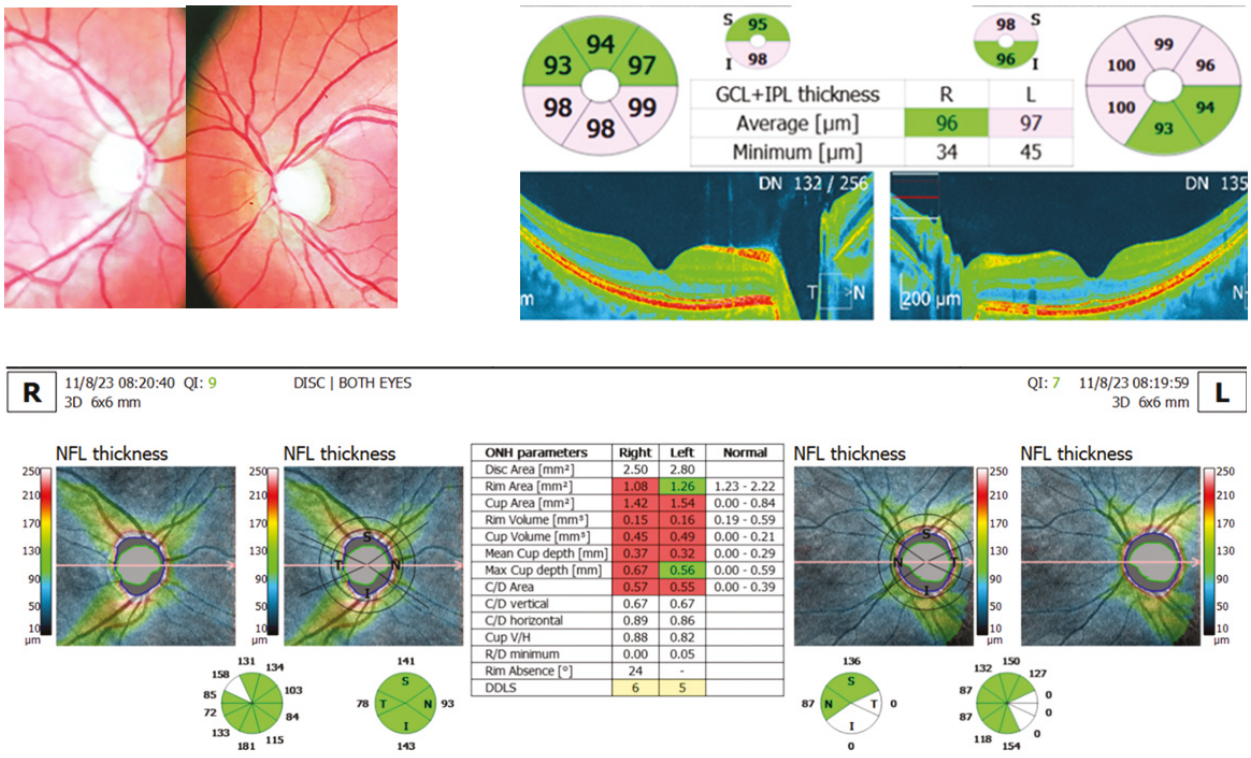

Fig. 3. Data for the 2024 examination in a patient born in 1971: right and left fundus photographs at presentation at 15 years after the initiation of treatment (A); retinal ganglion cell complex (GCL) and inner plexiform layer (IPL) optical coherent tomography of the right and left eyes at 15 years after the initiation of treatment (B); retinal nerve fiber layer (RNFL) of the right and left eyes at 15 years after the initiation of treatment (C).

Discussion of case 2

Hydrocephalus is an ACS complication caused by the displacement of cerebellar structures inside the spinal canal, which prevents normal CSF outflow, leading to CSF accumulation in the brain. Cerebellar structures are displaced through the foramen magnum, preventing normal CSF flow between the intracranial space and the spinal space, which may cause increased ICP [26, 27]. The diagnosis of ACM-I is based on brain MRI evidence of cerebellar tonsils displaced below the level of the foramen magnum, which reflects the overflow of the underdeveloped posterior fossa. However, the displacement of the cerebellar tonsils may be seen also in some patients with a normal-sized posterior fossa or even in some patients with a small posterior fossa. The history and course of the disease in patients presented suppose that clinical manifestations of the syndrome were triggered by a viral infection. It is most likely that a viral lesion was complicated by arachnoiditis localized mostly in the optic chiasmal region, resulting in the development of intracranial hypertension. In the presence of intracranial hypertension, due to the displacement of cerebellar structures, a viral lesion complicated by arachnoiditis most likely contributed to ACS manifestations. Therefore, chiasmal arachnoiditis and the displacement of brain structures contributed to the development of intracranial hypertension (manifesting in the eye as papilledema). Intensive steroid, anti-inflammatory and resorptive therapy contributed to disease regression, with the arrest of chiasmal arachnoiditis and a reduction in intracranial hypertension. A reduction in papilledema and improvement in visual acuity were observed with the treatment. The status of the patient and early therapy allowed avoiding surgery [28].

Conclusion

First, success in the diagnosis requires the efforts of a multidisciplinary team including ophthalmologists, roentgenologists, and neurologists to perform new tasks (e.g., those related to machine learning models) that have never been done before.

Second, the cases presented showed that possible ACM-1 manifestations may include e.g., strabismus and papilledema. Knowing the manifestations is a prerequisite for solving the diagnostic and treatment issues of the disease.

Third, there is no specific pathognomonic clinical sign or group of symptoms for the clinical diagnosis of ACM, and the disease can be diagnosed only by neuroimaging. The development of papilledema or inflammation or atrophy of the optic nerve in a patient should alert the ophthalmologist to the possibility of ACM, and he or she must initially refer the patient for MRI or computed tomography. Early identification of these ocular abnormalities allows suspecting and detecting this malformation early.

Finally, comprehensive treatment over many years can stabilize visual functions in patients with ACS.

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Disclosures

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Ethical Statement: *This study involved human subjects, was approved by the Bioethics Committee of SI "The Filatov Institute of Eye Diseases and Tissue Therapy of the National Academy of Medical Sciences of Ukraine" (Approval Date: 14.09.2024), and followed ethical standards as outlined in the Declaration of Helsinki of the World Medical Association and the European Convention on Human Rights and Biomedicine, and relevant laws of Ukraine.*

Informed Consent: *Consent to publish data has been obtained from the patients' parents.*

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Abbreviations: *ACM, Arnold-Chiari malformation; CM1, Chiari I malformation; MRI, magnetic resonance imaging*