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Case Report

Diagnostic and Therapeutic Doubts in Retrobulbar Neuritis in Children

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A B S T R A C T

Retrobulbar neuritis is often very complicated clinical entity. The most common cause of retrobulbar neuritis is demyelinating disease of CNS. This report is to express some other uncommon causes of it. Three children, age 8 to 12 with sudden and severe visual loss are presented. The diagnosis of retrobulbar neuritis is made by complete ophtalmological examination in consultation with neuropediatrics and neuroradiologist. Different ethiological causes of retrobulbar neuritis are found: pranasal sinusitis, functional visual loss and pseudotumor cerebri. In first two children complete therapeutically effort was as expected, and by child with pseudotumor cerebri there was no improvement of visual acuity, even after 6 months. In this presentation the authors want to emphasise some uncommon causes of retrobulbar neuritis.

Key words: *retrobulbar neuritis, children, diagnoses, therapy*

Introduction

Optic neuritis is a clinical entity of unknown etiology. There is corelation between idiopathic optic neuritis and multiple scelrosis exspecially in cases of reccurent attacs of neuritis[1–3]. In childhood optic neurits is very rare an isolated process1. Usually is related with neurological and systemic diseases like acute meningitis or some systemic viral infections with encephalitis. It is also related with demyelinating diseases like Devic's neuromyelitis optica or Schilder s disease. It s also possible to become optic neuritis after high doses of antibiotics (chloramphenicol)[4,5].

The condition is characterized by acute, often progressive visual loss associated with retrobulbar pain or pain on eye movement. It usually affects patients 15 to 45 years of age. Generally the condition is unilateral, and there is loss of visual acuity, decreased colour vision and the central or paracentral scotoma in the affected eye. Occasionally arcuate field defect occur. Brightness sense is profoundly diminished. An afferent pupillary defect almost occurs in the acute phase, and the visual evoked potential usually shows a prolonged latency.

Optic disc appears normal. Phosphenes or photopsias induced by eye movements or sound occasionally occur. The clinical course is characterized by a rapid deterioration of vision followed by a steady recovery. In most patients vision recovers gradually over the next several weeks, but incomplete[2]. Uhthoff s symptom (transient decrease in vision after exercise or elevation of body temperature) occurs in at least one-half patients after recovery[6]. Evaluation of optic neuritis should include complete eye examination, including visual field testing of both eyes,testing optic nerve function[7].The history should include questions about recent viral illness, family history of visual loos and past episodes of neurological dysfunction. Laboratory evaluation, CT or MRI scan of the head and orbit[6,7].

Steroid therapy has been controversial. Patients treated with oral prednisolone did not have a better outcome than those on placebo. High dose intravenous methylprednisolone leads to a faster recovery of visual acuity and better visual outcome. Patients with visual loss of 50% or better may do very well without any treatment[8].

Differential diagnosis of optic neuritis includes retinal disorders (central serous retinopathy), anterior ischaemic optic neuropathy, syphilis, postviral optic neuritis, Leber's optic neuropathy, nutritional reasons, toxic, malignant and other compressive lesions[9–12].

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Patients and Methods

Case 1

An 8-years old boy was hospitalised in our Clinic with sudden and severe bilateral visual loss in the period of two months (VOD:0.1; VOS:0.1). The complete ophthalmologic evaluation was normal with exception of visual fields testing that showed generalised constriction of isopters with homonymous qudrantanopias. VER was with normal amplitudes and prolonged latency bilateral. In LHON analyses no mutation was found.

MRI scan of the head and orbits showed signs of sinusitis maxillaris and inflammation of both mastoid sinuses.

During the neuropediatric evaluation lumbal puncture was done and CSF was syringing from the needle (ICP was 470mmHg!). CSF analysis showed all signs for infective, toxic or demyelinating process negative.

Our patient had three of four general conditions for diagnose of pseudotumor cerebri. In consultation with paediatricians we were decided for that diagnose and corticosteroid therapy combined with diuretics was given according to the diagnose.

Case 2

A 12-years old boy was addressed to our Clinic because of sudden and severe unilateral visual loss associated with pain on eye movements in the period of the one month (VOD:0.7). The complete ophthalmologic evaluation was normal with exception of visual fields testing that showed paracentral and arcuate scotoms.

MRI scan of the brain and viscerocranium showed inflammatory findings in all paranasal sinuses, so the therapy was peroral antibiotics in 14 days with the diagnose of retrobulbar neuritis.

Case 3

A 12 years old girl came with sudden unilateral visual loss associated with discrete pain on eye movement in the period of 24 hours (VOS: counting fingers on 0.5 m).

This girl has interesting anamnesis with frequent respiratory infections, gastrointestinaly disturbances and fever evaluated in Children's hospital. Also has done psychologists testing and was characterized as extremely conscientious and careful. Four years ago was ophtalmologicaly evaluated because of visual disturbances and double vision, but all findings were normal.

In our case, all ophthalmologic, neuropediatric and radiological evaluation were normal, so we were concerned for the diagnosis of functional visual loss.

Results

We used complete ophthalmologic, neuropediatric and neuroradiologic evaluation in our cases to find three different etiology of retrobulbar neuritis by children

In the first case we made a doubt on pseudotumor cerebri as a cause of visual loss. Therapy according to the diagnose has been lowering ICP, but results with visual loss were disappointing. Even after six months visual acuity was as at the beginning.

The second case was retrobulbar neuritis caused by sinusitis, and with antibiotic therapy there was complete restitution of visual acuity and whole status of the child.

In the third case we confirm our diagnose next day without any therapy.

Discussion

With our review we wanted to show some different and unexpected etiologi1es of retrobulbar neuritis and there treatment. Always when we have diagnose of retrobulbar neuritis we must exclude demyelinating process with all available methods, but that doesn't mean that MS is causative factor of visual loss. MS is characterized by relapses and remissions of neurological disturbance, very often by optic neuritis, increases in severity over a week or two and after approximately one month begins to remit[6]. In the early stages, clinical recovery is virtually complete, though persistent abnormalities of conduction can be detected by VER and structural abnormalities can be detected by MRI. Cerebrospinal fluid examination for oligoclonal IgG provide supporting evidence for the diagnose[6,13].

In our first case we were deciding on diagnose of pseudotumor cerebri because our patient had three (elevated ICP, normal cerebral anatomy, normal CSF) of four general conditions for this diagnose[14,15]. Pseudotumor cerebri is an idiopathic disorder characterized by papilledema and elevated intracranial pressure without a mass lesion. Our patient did not have signs and symptoms of increased ICP except disturbance in visual acuity. In differential diagnoses we must think about Leber's hereditary optic neuropathy associated with mitochondria (mt)DNA point mutations. This mutations raised the possibility to genetic susceptibility to MS[16,17]. Treatment of patients with moderate visual loss because of pseudotumor cerebri is with acetazolamid as a first choice for decreasing CSF production in combination with corticosteroids[14,15]. In our case results with the therapy were disappointing in improvement visual acuity, even after six months.

In the second case therapy was just aimed to the cause of retrobulbar neuritis and in a few days the success was complete, visual acuity and visual field were as before the illness. This case was just one of the common described in the literature[2,5,8]. The clinical course, etiology, success of therapy according to etiology was just like in literature with complete visual recovery[8,18].

In the third case we did not need any treatment because of complete recovery of visual acuity and all symptoms in very short period. In this case we also have done the whole evaluation according to the diagnose, but all findings were in normal rates, so in consultation with other specialists and literature we concluded that was

subjective visual loss not attributable to an organic cause, called functional visual loss[19,20].

In diagnoses of retrobulbar neuritis by children we have to exclude demyelinated process but already think that etiology is sometimes unusually with very simple therapy and unexpected results. Optic neuritis presents differently in children than in adults. Visual prognosis is poorer in children than adults, the development of MS is less common in children. Children who present with unilateral involvement have a better visual prognosis, however, they also develop MS at a greater frequency than children with bilateral involvement[3,7].

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DIJAGNOSTIČKE I TERAPIJSKE DVOJBE KOD RETROBULBARNOG NEURITISA DJECE

SAŽETAK

Retrobulbarni neuritis je složen i često teško razjašnjiv klinički entitet. Najčešće je klinički povezan s demijelinizirajućim bolestima CNS-a. Ovim radom želimo upozoriti na još nekoliko etioloških uzroka retrobulbarbog neuritisa. Na Klinici je obrađeno troje djece u dobi od 8 do 12 godina sa naglim i značajnim padom vidne oštrine pod radnom dijagnozom retrobulbarnog neuritisa. Učinjena je kompletna oftalmološka obrada u suradnji sa neuropedijatrima i neuroradiolozima. Kompletnom oftalmološkom, neuropedijatrijskom i neuroradiološkom obradom ustanove se različite etiologije bolesti: paranazalni sinusitis, funkcionalni gubitak vidne oštrine, dok se kod trećeg djeteta na osnovu dijagnostičke obrade posumnja na pseudotumor cerebri. U prva dva slučaja terapija i ishod bili su u skladu s našim očekivanjima, dok kod pseudotumora cerebri na ordiniranu terapiju nije došlo do očekivanog poboljšanja vidne oštrine (čak niti nakon 6 mjeseci). Ovim radom želimo upozoriti da kod razriješavanja slučajeva retrobulbarbog neuritisa treba posumnjati i na neuobičajene etiološke momente.