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Caso clínico

Pediatric Otitic Hydrocephalus associated with Otogenic Lateral Sinus Thrombosis in Acute Mastoiditis

Mastoidite Aguda Complicada de Trombose do Seio Lateral e Hi- drocefalia Otfica em Idade Pediátrica

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Abstract

Introduction: Acute mastoiditis may be associated with intracranial complications such as lateral sinus thrombosis. As a consequence, children may, exceptionally, develop otitic hydrocephalus, presented by symptoms and signs of elevated intracranial pressure. A high clinical suspicion is essential for an early diagnosis, in order to achieve the best clinical outcome.

Case Report: A seven-year-old female patient, previously diagnosed with acute otitis media in the right ear, presented with a progressively increasing frontal headache, photophobia and diplopia. A head computed tomography revealed a right ear mastoiditis with lateral sinus

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thrombosis. Magnetic resonance angiography confirmed an ipsilateral transverse sinus, sigmoid sinus and internal jugular vein thrombosis. Neuro-ophthalmologic examination confirmed bilateral abducens nerve palsy and severe bilateral papilledema, following intracranial hypertension. The patient underwent prompt surgical treatment with myringotomy with transtympanic tube placement and mastoidectomy with perisinus empyema drainage, having started intravenous antibiotic therapy and anticoagulation. After diuretic therapy, systemic steroids and two lumbar punctures the patient had a complete recovery.

Discussion: Otitic hydrocephalus is a rare but potentially fatal complication of otitis media. It is associated with cerebral venous thrombosis, therefore a multidisciplinary assessment, involving specialties such as neurology and ophthalmology is essential to evaluate ocular and neurologic symptoms. In the presence of intracranial hypertension, early treatment is critical to improve headache and prevent permanent vision loss.

Keywords: Acute mastoiditis; Intracranial complications; Otitic hydrocephalus

Resumo

Introdução: A mastoidite aguda pode estar associada a complicações intracranianas como é o caso da trombose do seio lateral. Na sequência desta, excepcionalmente, pode desenvolver-se um quadro de hidrocefalia otítica, manifestando-se por sinais e sintomas de hipertensão intracraniana. Um elevado grau de suspeição clínica é essencial para um diagnóstico precoce, de forma a melhorar o prognóstico destes doentes.

Caso Clínico: Criança do sexo feminino, de 7 anos de idade, previamente diagnosticada com otite média aguda à direita e que inicia quadro de cefaleias frontais intensas, fotofobia e diplopia. A tomografia computadorizada (TC) cerebral revelou otomastoidite com suspeita de trombose do seio lateral. Realizou Angio-Ressonância Magnética (Angio-RM) cerebral que confirmou a presença de trombose da porção distal do seio transversal, seio sigmóide e veia jugular interna. A observação por Oftalmologia e Neurologia Pediátrica confirmou paralisia bilateral do VI par craniano e papiledema bilateral, na sequência de hipertensão intracraniana. Foi submetida a miringotomia com colocação de tubo de ventilação transtimpânico e mastoidectomia simples com drenagem de empiema perisinusal, tendo iniciado antibioterapia endovenosa e hipocoagulação. Após terapêutica diurética, corticoterapia sistémica e duas punções lombares evacuadoras verificou-se uma boa evolução clínica, com recuperação completa.

Discussão: A hidrocefalia otítica é uma complicação rara mas potencialmente fatal da otite média. Na presença de trombose venosa cerebral, é essencial uma avaliação multidisciplinar, envolvendo especialidades como Neurologia e Oftalmologia, de forma a avaliar a presença de sintomas oculares e/ou neurológicos. Na presença de hipertensão intracraniana, o tratamento precoce é fundamental para melhorar a cefaleia e prevenir perda de visão permanente.

Palavras-chave: Mastoidite aguda; Complicações intracranianas; Hidrocefalia otítica

Introduction

Acute otitis media (AOM) may lead to intracranial or extracranial complications. In the pre-antibiotic era, acute mastoiditis was a widespread and feared complication of otitis media, presenting high rates of morbidity and mortality. The use of antibiotics has led to a decrease in the incidence of complications from approximately 17% to 1%.^{1,2}

Otitic hydrocephalus (OH) is a rare intracranial complication of middle ear infection, consisting of increased intracranial pressure without focal neurologic abnormalities other than those due to the elevated pressure.

Taylor, in 1890, first described patients with increased intracranial pressure with a normal composition of CSF, in the absence of a tumor. However, Quincke, in 1897, was the first who published a comprehensive report and referred the condition as “serous meningitis” and Symonds first suggested the term “otitic hydrocephalus” in the 1930s.^{3,4} Elevated opening pressure during lumbar puncture with normal cerebrospinal fluid (CSF) composition and radiological evidence of sinus thrombosis is the mainstay of diagnosis.

Clinically, the manifestations of OH are common to all other causes of elevated intracranial pressure which include headache, nausea and vomiting, blurred vision, papilledema, and palsy of VI nerve with diplopia.^{5,6} It can be presented weeks after an AOM or many years after the start of chronic middle ear disease.¹

Case report

A 7-year-old female patient previously diagnosed with AOM in the right ear and treated with oral antibiotics (amoxicillin and clavulanic acid 50mg/kg/day) for 7 days, presented to the emergency department with a progressively increasing frontal headache, photophobia and diplopia. Right tympanic membrane was erythematous, thick and bulged. A contrast head CT-scan revealed a right ear mastoiditis with lateral sinus thrombosis. Magnetic resonance angiography confirmed an ipsilateral transverse sinus, sigmoid sinus and internal jugular vein thrombosis (Fig. 1). Neuro-ophthalmologic examination revealed bilateral abducens nerve palsy and severe papilledema (Fig. 2), following intracranial hypertension. The patient underwent prompt surgical treatment with myringotomy with transtympanic tube placement and mastoidectomy with perisinus empyema drainage, having started intravenous antibiotic therapy (ceftriaxone + vancomycin), antioedematous drugs (acetazolamide, mannitol and high-dose steroid) and anticoagulation drugs. Two lumbar punctures were performed and demonstrated an elevated opening pressure with normal CSF composition. Cultures of blood and mastoid were negative. Antibiotic therapy was continued for 3 weeks after surgery. The patient subsequently improved and was discharged from hospital five weeks after surgery in general good condition, still medicated with a corticosteroid tapering regimen and anticoagulation. Two months after discharge the abducens palsy recovered completely and fundoscopic examination results were normal (Fig. 3).

Figure 1: MRA with thrombus of right transverse sinus, sigmoid sinus, and internal jugular vein.



Figure 2: Limited abduction of both eyes (A and B) and fundoscopic images shows bilateral papilledema (C and D).

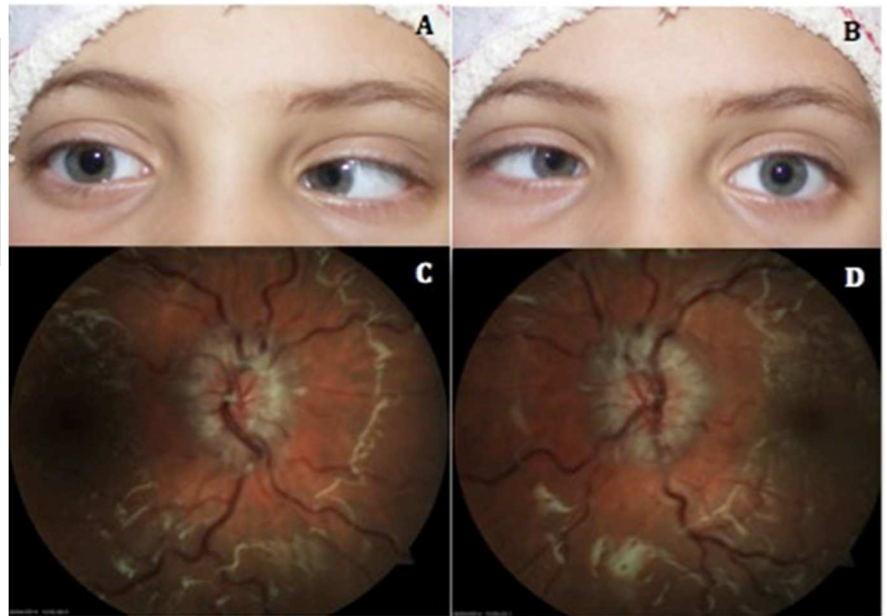


Figure 3: Normal bilateral ocular mobility (A and B) and fundoscopic examination (C and D).



Discussion

The risk of intratemporal or intracranial complications of suppurative ear disease is potentially high due to spread of infection through bony or vascular dissemination.⁷ The combination of widespread antibiotics and prompt surgical intervention has markedly decreased intracranial complications of middle ear infection.^{1,2}

However, complaints of headache and fever in patients with otitis media should raise the clinical index of suspicion for an early diagnosis of intracranial complications.

Otitic hydrocephalus may be a result of acute, as well as chronic inflammation in mastoid. The clarification of its complex *pathogenesis* still remains unclear. It is largely accepted that otitic hydrocephalus is a syndrome of reduced CSF absorption in the setting of sinus thrombosis.^{6,8} Although thrombosis of the lateral venous sinus is almost always a constant component of this disease, debate remains about the role of superior sagittal sinus thrombosis.⁸ According to Fishman, increased pressure in the superior sagittal sinus leads to reduced CSF absorption by the arachnoid villi and a consequent increase in CSF pressure.⁹

Manifestations of elevated intracranial pressure (headache, nausea and vomiting, papilledema and diplopia), in patients with a history of ear disease and an elevated opening pressure during lumbar puncture with normal CSF composition are necessary to make the diagnosis of otitic hydrocephalus and exclude meningitis.⁹

Magnetic resonance venography is considered the diagnostic imaging exam of choice because it allows an enhanced evaluation of the venous sinuses.^{10,11}

The aim of the treatment is to eradicate ear disease and lower the elevated intracranial pressure.

Management of otitic hydrocephalus includes surgical procedures, appropriate antimicrobial therapy, medical treatment of raised intracranial pressure and anticoagulants.

The surgical approach includes cleaning the middle ear and mastoid disease and draining perisinus abscesses if present. Mastoidectomy eradicates perisinus infection and removes granulation tissue thereby reducing the thrombophilic nidus and promoting recanalization of the sinus without clot evacuation.^{10,11}

Medical treatment comprises systemic antibiotics and the use of corticosteroids, diuretics and hyperosmolar dehydrating agents to reduce intracranial pressure. Multiple lumbar punctures, once very popular, are not free of risk in the presence of raised intracranial pressure and have been reported to be ineffective.^{6,8}

The role of routine anticoagulation in the management of lateral sinus thrombosis is still doubtful. The weight of potential benefits in preventing thrombus propagation, recanalization, and improved clinical outcome must be assessed against the risk of hemorrhagic complications, evidence of recanalization without anticoagulants, and reports of clinical recovery despite persistent thrombus. However, anticoagulant therapy has been shown to be generally safe in young children with proper monitoring.¹⁰

In conclusion, otitic hydrocephalus is a rare but potentially fatal complication of otitis media, therefore a prompt multidisciplinary assessment, involving specialties such as neurology and ophthalmology is essential to evaluate ocular and neurologic symptoms. In the presence of intracranial hypertension, early

treatment is critical to improve the clinical condition and prevent permanent vision loss.

Conflicts of interest: No conflicts of interest were declared by the authors.

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