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CASE REPORT

Opsoclonus-ataxia syndrome in an adolescent: an acute otitis media complication[☆]



Síndrome de opsoclonus-ataxia em adolescente: uma complicação de otite média aguda

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Introduction

Opsoclonus is defined as non-rhythmic, involuntary, spontaneous, multidirectional, and hyperkinetic eye movements, with a large amplitude and high frequency (10–15 Hz). These movements are present during visual fixation and convergence, and remain active during sleep and when the eyelids are closed. They should not be confused with an acquired nystagmus, which is usually unidirectional, with fast and slow components. In addition, opsoclonus is different from eye flutter, as this movement is restricted to the horizontal plane.

The opsoclonus-myoclonus-ataxia syndrome (OMAS) is a rare condition characterized by opsoclonus associated with compensatory head movements, myoclonus, and cerebellar

ataxia most likely secondary to an autoimmune mechanism. The etiologies include parainfectious, paraneoplastic, toxic, and metabolic causes. Among the paraneoplastic causes, neuroblastoma is prominent in childhood and small-cell lung carcinoma stands out in adults.^{1,2}

The objective of this article was to present a case of OMAS secondary to a CNS infection resulting from an unusual course of an acute otitis media (AOM) complicated by otomastoiditis in a previously healthy adolescent.

Case report

NCO, an eleven-year-old girl, experienced an episode of bacterial pharyngitis that was treated with benzylpenicillin. Fever persisted, and she developed recurring vomiting, after which AOM was found; amoxicillin was initiated and cefuroxime was added. Approximately 15 days after the onset of symptoms, she complained of dizziness and unstable gait, and her mother noticed non-rhythmic eye movements in both eyes.

She was admitted to the University Hospital. On the general physical examination, the patient was in a normal

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Figure 1 Mastoid computed tomography, cross-sectional view. Within the clear circles, a bilateral partial clouding of mastoid cells is observed.

state, exhibiting irritation; her skin was pale, the tympanic membranes were bulging and hyperemic bilaterally, and no discharge was observed from the ear canal. No other changes, such as enlarged lymph nodes, enlarged liver and spleen, or a change in heart or lung sounds were observed. During otoneurological examination, the patient was conscious and had rapid, frequent, involuntary, multidirectional (both horizontal and vertical), unpredictable, and disordered eye movements, in addition to axial cerebellar ataxia (stable Romberg, Fukuda with broad base, and no dysmetria or dysdiadochokinesia). Rhinoscopy was normal. No myoclonus, meningeal signs, or cranial nerve impairments were observed.

The complete blood count showed increased white blood cells (40,700 leucocytes), with neutrophils predominating (89%) and no left shift. Cranial tomography revealed a bilateral partial clouding of mastoid cells (**Fig. 1**). Lumbar puncture showed pleocytosis (18 cells, predominance of lymphocytes and monocytes) with no other findings. Audiometry showed mixed (sensorineural and conductive) hearing loss. Cranial MRI was normal, and the entire metabolic investigation and infectious agents screening, including HIV, hepatitis, rubella, toxoplasmosis, cytomegalovirus, herpes, and Epstein-Barr virus, were negative. Following a 28-day course of parenteral antibiotics (ceftriaxone and oxacillin), there was complete regression of all signs and symptoms.

Discussion

The present patient had an AOM complicated by bilateral mastoiditis, progressing to encephalitis over a three-week period. This course was unusual not only for the early diagnosis in this patient, but also due to the antimicrobial agents currently available.^{3,4}

In addition, when the patient was admitted to the University Hospital, she had no earache or retroauricular pain, but her gait was unstable and she had associated eye movements, thus meeting the criteria for the opsoclonus-ataxia syndrome.^{1,2} These symptoms could only be linked to an AOM after otomastoiditis complicated by encephalitis was found, which explained the findings. A number of patients may have neurological symptoms as a complication of otomastoiditis, but it is unusual to find an OMAS.⁴ The richness of semiological data for differential diagnosis of the eye movements and the rareness of this condition, usually present in younger children, make this case unique.²

Since there was clinical and laboratorial evidence of bacterial infection associated with a good response to antibiotics, the use of anti-inflammatory agents and immunosuppressing drugs usually used in OMAS management was not required. The findings demonstrate that there is no definitive rule in treating these cases and, by considering the several etiologies, management should be chosen in accordance with the uniqueness of each patient.^{5,6}

The prompt diagnosis in this case particularly favored the good clinical course and avoided additional unnecessary and costly methods usually employed to establish a diagnosis in patients with OMAS. At the 30-day and the 90-day follow-up, the patient was asymptomatic and her neurological examination was completely normal.

Final remarks

The rare character of this OMAS case, occurring after an acute otitis media, and the favorable course highlight the importance of a thorough history and a careful clinical examination to guide the investigation and therapeutic strategies.

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Sahu JK, Prasad K. The opsoclonus-myoclonus syndrome. *Pract Neurol*. 2011;11:160–6.
2. Desai J, Mitchell WG. Acute cerebellar ataxia, acute cerebellitis and opsoclonus-myoclonus syndrome. *J Child Neurol*. 2012;27:1482–8.
3. Johnson NC, Holger JS. Pediatric acute otitis media: the case for delayed antibiotic treatment. *J Emerg Med*. 2007;32:279–84.
4. Osborn AJ, Blaser S, Papsin BC. Decisions regarding intracranial complications from acute mastoiditis in children. *Curr Opin Otolaryngol Head Neck Surg*. 2011;19:478–85.
5. Glatz K, Meinck H-M, Wildemann B. Parainfectious opsoclonus-myoclonus syndrome: high dose intravenous immunoglobulins are effective. *J Neurol Neurosurg Psychiatry*. 2003;74:277–82.
6. Brunklaus A, Pohl K, Zuberi SM. Outcome and prognostic features in opsoclonus-myoclonus syndrome from infancy to adult life. *Pediatrics*. 2011;128:e388–94.