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Brain & Development 27 (2005) 155–159

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Case Report

Acute onset of abducens nerve palsy in a child with prior history of otitis media: a misleading sign of Gradenigo syndrome

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Received 14 July 2003; received in revised form 2 February 2004; accepted 16 February 2004

Abstract

Gradenigo syndrome in children during otitis media is a very rare complication since the use of antibiotics: nevertheless, it must be taken into account in case of otitis media, abducens nerve paralysis and facial pain in regions innervated by the first and second division of trigeminal nerve. We report the case of a 4-year-old boy whose neurological signs and clinical history prompted us to entertain a diagnosis of Gradenigo syndrome, which was not confirmed by CT and MRI findings, revealing otomastoiditis and sinus thrombosis.

Keywords: Gradenigo syndrome; Abducens nerve palsy; Otitis media; Cerebral venous sinus thrombosis; MR angiography; Falsely localising sign

1. Introduction

Gradenigo syndrome consists of the association of otitis media, facial pain in the regions innervated by the first and second division of trigeminal nerve and abducens nerve paralysis [1]. Cranial nerve dysfunction is caused by osteitis and local leptomeningitis near the apex of the petrous bone, where the trigeminal nerve ganglion and the abducens nerve lie closely together, separated from the petrous apex only by the dura mater [2]. Both nerves can be easily affected by a local inflammatory process.

Cerebral venous sinus thrombosis can be a sequel to local inflammations, coagulation disorders (activated protein C resistance, factor V Leiden mutation) and systemic inflammatory diseases. It can present with a wide spectrum of signs and symptoms. Headache is the presenting symptom in 70–90% of cases and most commonly evolves over a period of days to weeks [3]. It is most frequently associated with nausea, vomiting and vision changes. Seizures, cranial nerve syndromes, neurologic deficits and confusion are also common presentations [4–7]. Headache, nausea and pyramidal signs can present with a rapid decline of consciousness to eventual coma followed by death, if the deep cerebral veins are involved. The classic triad of dural sinus thrombosis is headache, papilledema and high opening cerebrospinal fluid pressure. This makes diagnosis difficult to distinguish because more benign entities such as pseudotumor cerebri present in a similar fashion [3–5,8].

2. Case report

A 4-year-old boy with a 3-day-history of fever, abdominal pain and one episode of vomiting was referred to the emergency department of our institution. His past medical history was unremarkable except for an episode of otitis media requiring oral antibiotics (amoxicilline + clavulanate), prescribed by his general practitioner a fortnight before the onset of these symptoms.

On admission, the child was afebrile, heart rate was 120/min, blood pressure 95/60 mm Hg, and respirations 35/min. The chest was clear, the abdomen was not distended, and no visceromegaly or masses were palpable. His height was 102 cm, weight 17 kg (both at the 50th percentile): mild dehydratation (weight loss $<5\%$) was present. On otolaryngological examination the oropharynx was clear, but both tympanic membranes were slightly bulging and bilateral cervical lymphadenopathy was evident. The rest of the examination was unremarkable, including neurological signs. White blood count was

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doi:10.1016/j.braindev.2004.02.003

12,290/mm^3 with 72% segmental neutrophils, 15.8% lymphocytes, 9.1% monocytes, 0.2% eosinophils, 0.25% basophils, and 2.2% band form; red blood count was $4{,}48 \times 10^6$/mm^3, hemoglobin level 10,9 g/dl, hematocrit 32,1%, and platelet count 438×10^3/mm^3. C-reactive protein was 109 mg/l. Serum chemistries, including electrolytes, and BUN were within normal ranges. A chest radiograph showed mild, bilateral infiltrates and a normal heart size.

The child was discharged on antibiotics (6-day course of ceftriaxone). On follow-up visit one week later, he looked comfortable. Fever had promptly disappeared with antibiotics administration. White blood count was 9410/mm^3 with 54.5% segmental neutrophils, 34.4% lymphocytes, 8.6% monocytes, 0.2% eosinophils, 0.3% basophils, and 2% band form; red blood count was $5{,}18 \times 10^6$/mm^3, hemoglobin level 13,1 g/dl, hematocrit 37,3%, and platelet count 535×10^3/mm^3. C-reactive protein was below 4.6 mg/l, electrolytes were within normal ranges.

Four days later he was readmitted to our emergency department because of onset of right eye strabismus: he was afebrile, heart rate was 76/min, blood pressure 105/65 mm Hg, and respirations 32/min. His weigth was 16,830 kg-a decrease of 170 grammes since his 1st admission. On physical examination, right abducens nerve palsy was evident. Owing to the young age, presence of ipsilateral diplopia could not be ascertained. Facial motricity was normal. There were neither fever nor meningeal signs, the child was alert and orientated. On account of the prior history of episodes of otitis media, a diagnosis of Gradenigo syndrome was considered. However, when questioned whether any facial areas were painful, the child was unable to give a definitive answer. The ophtalmologic examination revealed bilateral papilledema, more evident on the right side. Visual acuity was not ascertained due to lack of collaboration. The otolaryngological examination was not conclusive because of obstructing cerumen. There was no mastoid tenderness. White blood count was 9.390/mm^3 with 47.4% segmental neutrophils, 43.9% lymphocytes, 4.9% monocytes, 0.4% eosinophils, 0.3% basophils, and 3% band form; red blood count was 5.39×10^6/mm^3, hemoglobin level 13.3 g/dl, hematocrit 38.8%; the platelet count was high at 760×10^3/mm^3. C-reactive protein was below 4,6 mg/l. Fibrinogen, antithrombin-III, and D-dimer (a fibrin degradation product) levels came back slightly positive (415 mg/100 ml, 142%, and 135 μg/l, respectively). Serum chemistries, including electrolytes, and BUN were within normal ranges. Blood cultures were negative, as well as those from nasal and pharyngeal swabs. Anticardiolipin antibodies and a coagulation profile were performed and turned out to be normal.

Contrast enhanced computerised tomography (CT) scan did not show abnormal density areas in the brain. However, it did demonstrate a triangular area of enhancement with a relatively low-attenuating center on transverse images obtained in the region of the left sigmoid sinus (Fig. 1).

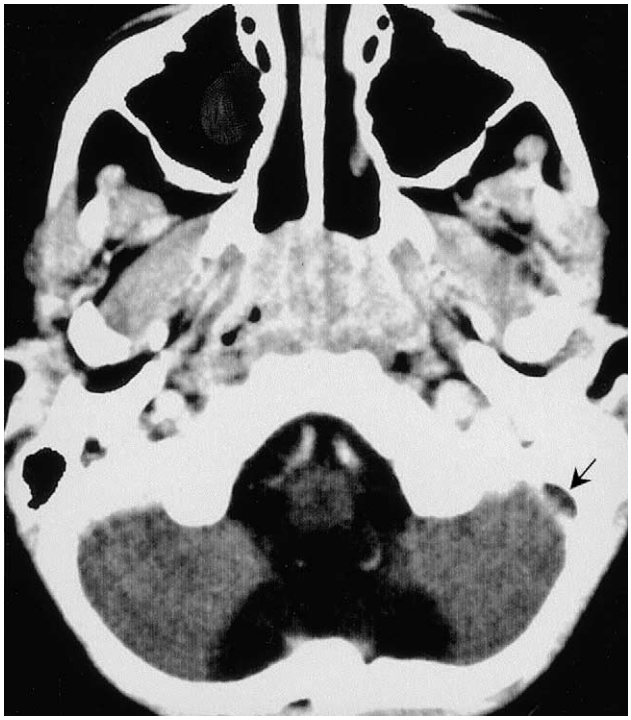

Fig. 1. Contrast-enhanced axial CT scan shows a filling defect at level of left sigmoid sinus (arrow), consistent with thrombosis.

Owing to these findings, the child was then referred for magnetic resonance imaging (MRI) (Fig. 2). Two-dimensional time-of-flight (TOF) technique was used for venous MR angiography; it showed absent canalisation of the left sigmoid sinus, right transverse sinus, and of the proximal portion of the left transverse sinus. There was collateral canalisation through the occipital sinus draining into the right jugular bulb. Other dural sinuses, and especially the cavernous sinuses, were canalised. There were neither morphological nor signal abnormalities involving the nervous tissues both before and after intravenous paramagnetic contrast medium administration. No detectable pathology involved the sixth cranial nerves. The ventricular system was slightly enlarged. Inflammatory debris were detected in the left mastoid.

EEG showed a diffusely, mildly slowed background. Intravenous antibiotics (ceftriaxone, metronidazole and teicoplanin) were started. The treatment included antithrombotic therapy with subcutaneous low-molecular-weight heparin (enoxaparin) at the dose of 150 U/kg/day, the management of intracranial hypertension with IV corticosteroids and seizure prevention with phenobarbital.

Three days later, follow-up MRI demonstrated complete recanalisation of the occluded left transverse sinus and partial recanalisation with residual stenosis of the left sigmoid and right transverse sinuses (Fig. 3).

After having 10 days of IV antibiotics, the patient was discharged home on ceftriaxone for another 5 days. Treatment with subcutaneous low-molecular-weight