New onset strabismus in association with ear pain

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CASE HISTORY
A 7-year-old previously well boy presented to the emergency department (ED) with diplopia and pain in his right eye for 2 days. The previous week, while on holiday abroad, he developed right ear pain and vomiting. He was seen by the local physician, diagnosed with right otitis media and was treated with ear-drops. On returning to the UK, his general practitioner started him on oral amoxicillin, but his parents brought him to the ED due to worsening symptoms. There was no history of trauma, headache, vomiting or fever. He was fully immunised, was not on any regular medications and had no known allergies.

On examination, he was distressed due to photophobia in his right eye and diplopia. There was crusted discharge in his right ear canal with a perforated tympanic membrane. Cranial nerve examination revealed a loss of abduction of the right eye, but the rest of the neurological and systemic examination was normal.

QUESTIONS
1. What abnormalities are seen on the brain MRI? (figure 1)
2. What is the likely diagnosis?
3. Based on the clinical presentation, what other important differential diagnoses should be considered?

Answers can be found on page 268.
1. There is fluid in the right middle ear cavity and mastoid air cells, and an area of high T2 signal in the right petrous apex, which represents petrous apicitis or petrous apex abscess. Enhancement of the adjacent dura and the right trigeminal nerve can also be seen.

2. The clinical and MRI findings are consistent with Gradenigo’s syndrome—a triad of (i) suppurative otitis media, (ii) paralysis of the external rectus muscle and (iii) pain in the ipsilateral orbit.12 Symptoms develop as the infection spreads to the petrous apex of the temporal bone, where the abducens nerve and trigeminal ganglion are in close proximity, separated only by the dura mater. The abducens nerve is affected by surrounding inflammation as it passes through Dorello’s canal, the anatomical region between petrous apex and petrosphenoidal ligament (figure 2).3

MRI is the optimal imaging modality—acute petrous apicitis with abscess formation is appreciated as a hyperintense lesion on T2 and low intensity lesion on T1 weighted images that may enhance post gadolinium. Most cases are due to aerobic organisms but anaerobic infections have been reported, warranting anaerobic antibiotic cover.4 Intravenous antibiotics are recommended for at least 2–3 weeks, although patients with an associated osteomyelitis may require up to 6 weeks of medical treatment.

Surgery is only required in those who do not respond to antibiotic therapy, or have an abscess in the petrous apex, surrounding dura or brain tissue. Debridement or myringotomy is a less invasive procedure in patients with a bulging tympanic membrane who fail to respond to antibiotics.5

3. Intracranial abscess

A brain abscess usually presents with an indolent and protracted prodrome of nonspecific symptoms, including low-grade fever, headache, mental state changes and lethargy. As the associated inflammation and cerebral oedema progress, they usually manifest with symptoms of a space-occupying lesion, including vomiting, severe headache, seizures, papilloedema, focal neurology and coma.1

Meningitis

- While otitis media can be complicated by meningitis, the presenting features tend to include headache, generalised photophobia, fever and meningeal irritation, which may progress rapidly. Our patient had an isolated sixth nerve palsy and right-sided photophobia with no other features of meningitis.

Sinus venous thrombosis

- Lateral sinus venous thrombosis is a rare but potentially fatal intracranial complication of acute otitis media. Patients present with headache, fever, otorrhoea, and signs of increased intracranial pressure, which result from occlusion of the sinus lumen and the consequent interruption of cortical venous circulation. Tenderness and oedema over the mastoid (the Griesinger sign) are often present.

PATIENT OUTCOME

Our patient was managed conservatively and received antibiotics for a total of 6 weeks. The sixth nerve palsy resolved within a few days and he made a full recovery within 3 weeks. The follow-up MRI at 6 weeks showed complete resolution of the petrous apicitis and surrounding inflammation.

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Figure 2 Rotated three-dimensional computer graphic views of the Dorello canal as the space between the petrous apex and superolateral portion of the clivus, bounded superiorly by the petrosphenoidal ligament of Gruber (asterisk, left). Also depicted are cranial nerves III (white arrowhead), IV (white arrow), V (black arrow) and VI (black arrowhead). Reproduced with permission.5
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