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Rare disease

Conservative management of Gradenigo's syndrome in a child

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Summary

Gradenigo's syndrome, the triad of suppurative otitis media, abducens nerve palsy and pain in the ophthalmic division of the trigeminal nerve, remains a rare complication of otitis media. A case in a paediatric patient is described, successfully managed conservatively. There is little evidence to support increased use of antibiotics in acute otitis media to prevent this complication.

BACKGROUND

With the advent of antibiotic therapy, intracranial complications of otitis media have become rare. In the pre-antibiotic era, the incidence of intracranial complications of ear disease has been quoted at 2.3–6.4%, with a reduction to 0.04–0.15% due to the introduction of widespread use of antibiotics.[1] These complications include meningitis, cerebral abscess formation, as well as several distinct syndromes.[2]

First described in 1904, Gradenigo's triad of suppurative otitis media, pain in the ophthalmic branch of the trigeminal nerve and ipsilateral abducens palsy was unusual even within his own case series.[3] There have been few cases published in the paediatric literature. Traditionally management was surgical, with mastoidectomy occasionally proceeding to decompression of the petrous apex. In recent years, conservative management has been suggested. We present a case of successful outcome from medical management alone.

CASE PRESENTATION

A previously fit and well 11-year-old New Zealand Maori girl presented acutely to the emergency department. She had been suffering from severe headaches for 2 days, and complained of a deep pain around the right eye associated with nausea, vomiting and fevers. Her vital signs were unremarkable with a temperature of 37.5°C. At presentation, her neurological examination was normal, and a full systems examination revealed no pathology, although the right eardrum could not be visualised. There was no mastoid or sinus tenderness. Inflammatory markers were raised with a white cell count of 11.7×10^9/l, predominantly neutrophils, and an elevated C reactive protein of 94 mg/l. She was admitted overnight for observation.

The following morning, the patient complained of double vision on right lateral gaze, and over the course of several hours, she developed a complete failure of abduction of the right eye. Re-examination demonstrated loss of the corneal reflex on the right eye, suggesting impairment of the afferent pathway in the ophthalmic branch of the trigeminal nerve.

INVESTIGATIONS

An urgent MRI was requested, but prior to imaging there was copious purulent discharge from the right ear. The MRI demonstrated extensive mastoiditis on the right temporal bone, oedema at the petrous apex and overlying inflammation of the dura (figure 1). This supported the clinical diagnosis of Gradenigo's syndrome with suppurative otitis media, pain in the ophthalmic branch of the trigeminal nerve and an ipsilateral abducens nerve palsy.

TREATMENT

Conservative management was commenced, with intravenous ceftriaxone at intracranial sepsis dose. High-dose dexamethasone was administered, resulting in a rapid improvement in pain. However, on terminating the dexamethasone, this pain re-occurred and a longer tapering course was required. Inflammatory markers normalised within 1 week. No growth was obtained either on swabs from the purulent discharge or blood cultures. A magnetic resonance venography was performed to exclude sinus thrombosis.

OUTCOME AND FOLLOW-UP

Fortnightly MRIs showed improvement of the mastoiditis and gradual resolution of oedema at the apex. The patient's facial pain improved rapidly, but improvement of the sixth nerve palsy did not occur until 6 weeks of treatment, when some lateral rectus activity became apparent. Antibiotics were discontinued at 8 weeks, and the lateral rectus palsy continued to improve.

DISCUSSION

Gradenigo's syndrome results from infection or inflammation in the area of the petrous apex. Classically, this is

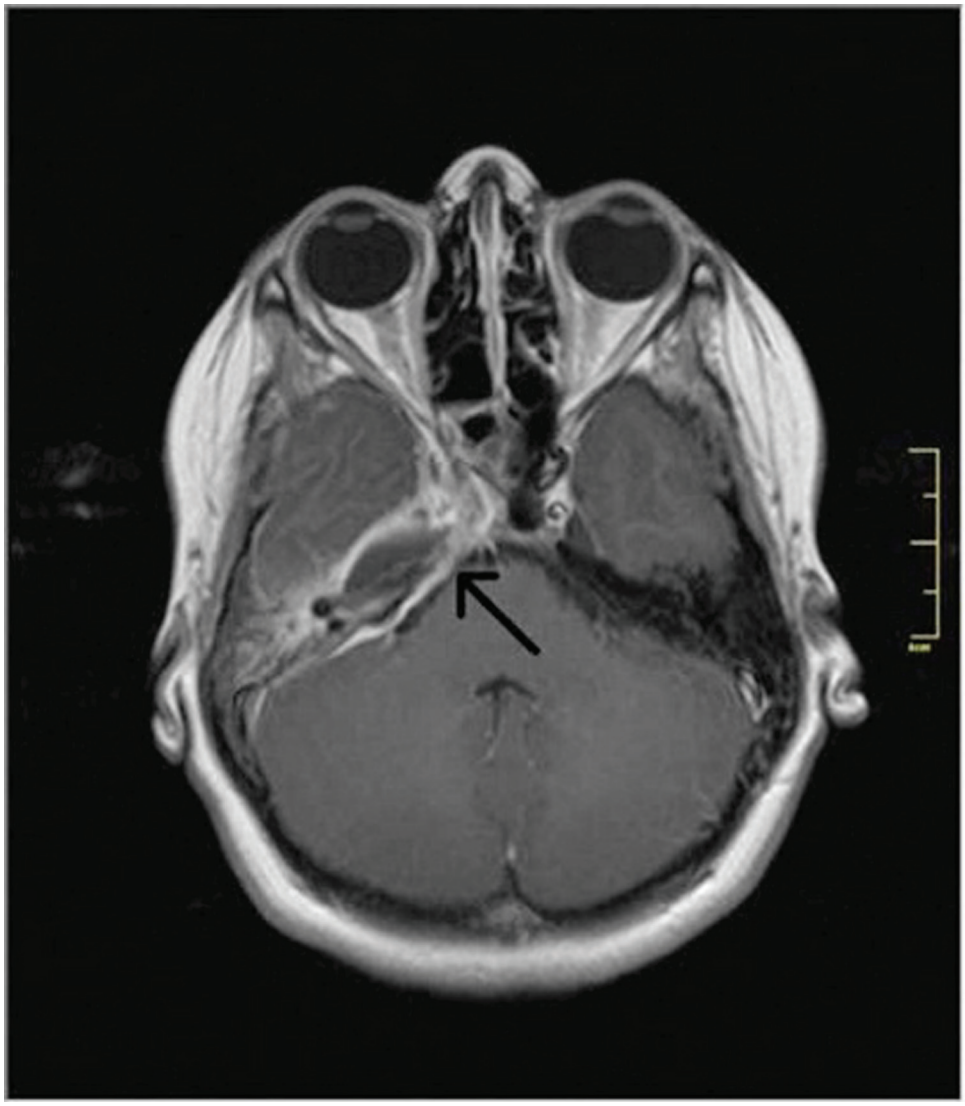

Figure 1 MRI demonstrating oedema in the area of the apex of the petrous bone.

a result of middle ear infection spreading to the mastoid bone. Subsequent spread to the apex can be direct through the petrous bone via air cells or as a true osteomyelitis, or via vascular channels.[4]

In conjunction with inflammation of the petrous apex, inflammation of the overlying dura is often seen, and may contribute to the neurology. The abducens nerve is particularly vulnerable in Dorello's canal, an inflexible channel roofed by Gruebber's ligament. Distal to the Gasserian ganglion, at this point, the ophthalmic branch of the trigeminal nerve is closely related to the abducens nerve. The lateral rectus palsy, pain and absence of a corneal reflex are most easily explained by a lesion at this point, selectively causing irritation of the ophthalmic branch.

Advances in imaging have assisted diagnosis and monitoring of this condition. CT will demonstrate bony abnormalities well, but MRI may provide diagnostic information relating to petrous apex lesions not provided by CT.[5 6] In this case, MRI was chosen as the first line of imaging as it was immediately available, likely to yield diagnostic information without using further modalities, and avoided unnecessary irradiation.

Historically, the management of Gradenigo's syndrome has been surgical, with drainage of the mastoid and decompression of the petrous apex.[7] Duration until resolution of the sixth nerve palsy has been variable, from almost instantaneous to up to 6 weeks. This is usually the last symptom to resolve. There has been a shift away from surgical management in favour of conservative management of Gradenigo's syndrome. It has now been proposed that in the context of acute infection, surgical treatment may be reserved for those not responding to conservative management.[7] Reports of conservative management have described resolution times of the lateral rectus palsy from 4 days to up to 3 months.[3 8] Surgical intervention may still be indicated, particularly in the context of chronic infection where bony destruction may result in cystic collections resistant to conservative management.[3]

It is common to have difficulty isolating an organism, and empirical treatment is often necessary. This may be due to antecedent use of antibiotics or inadequate culture

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techniques.[3] One report isolated *Pseudomonas*.[9] In this case, *Pseudomonas* was not specifically covered for, due to the prompt clinical response to antibiotics. Empiric antibiotic choice is usually based on providing cover for organisms most likely to cause otitis media. It is not yet clear as to how long intravenous antibiotic therapy should be administered, but in this case, treatment duration was based on clinical grounds.

Cases reported in the literature include paediatric and adult patients, and a review of these cases shows no discrimination with regard to age. Several case reports of this syndrome in the immunocompromised population would support investigation for any predisposing factors.[2] This patient had normal immunoglobulins, and diabetes mellitus and HIV were excluded.

Potential intracranial complications of Gradenigo's syndrome include meningitis, intracranial abscess, Vernet's syndrome, cavernous sinus thrombosis and hydrocephalus.[2] For this reason, an awareness of the anatomy of the petrous apex and the varied presentation of pathology is useful in order to initiate investigation and treatment promptly. If complications are avoided, the prognosis remains excellent.

The literature demonstrates a decline in mastoiditis and intracranial complications with the widespread use of antibiotic for otitis media. A recent study shows that antibiotic use halves the progression to mastoiditis.[10] This remains such a rare complication that the number needed to treat to prevent one case of mastoiditis is estimated at 4831.[10] There are no figures available to estimate the proportion of mastoiditis that progresses to intracranial complications, but this is rare, and as such despite the severity of the sequelae, this syndrome is unlikely to contribute to the argument regarding treatment of acute otitis media. Over the recent decades, with increased antibiotic resistance, there has been a move to decrease antibiotic use in otitis media, although this continues to be debated.[11] Ongoing surveillance of mastoiditis and diseases such as Gradenigo's syndrome is required to monitor for disease increase with more selective antibiotic use for treatment of otitis media being current practise.

Learning points

- In conclusion, this case, while unusual, provides a reminder that intracranial complications still occur, and require early recognition and treatment to avoid serious and potentially fatal sequelae.
- Furthermore, timely intervention may allow conservative management to succeed, avoiding the need for surgery.
- This case demonstrates good resolution of symptoms despite no clinical improvement in the lateral rectus palsy until 6 weeks. We would support persistence in conservative management, when surgical management is not otherwise indicated.

Competing interests None.

Patient consent Obtained.

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