Unusual presentation of more common disease/injury

Embryonic rhabdomyosarcoma presenting as Gradenigo's syndrome

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The authors report a previously undocumented case of parapharyngeal rhabdomyosarcoma presenting as Gradenigo's syndrome and review the history and aetiology of Gradenigo's syndrome. With sensitive diagnostic acumen this rare cause of Gradenigo's like syndrome was identified and correct treatment initiated.

BACKGROUND

Gradenigo's syndrome is a rare but still clinically important disease with potentially life-threatening complications, regardless of its aetiology. Despite classically being an otolaryngology diagnosis this case and the literature review demonstrate the range of pathologies that can mimic true Gradenigo's syndrome. This case seeks to highlight and refresh the reader's knowledge of Gradenigo's syndrome and to demonstrate with all 'classical' syndromes and diagnoses it is important to retain diagnostic acumen and consider different and unexpected causes for seemingly well-known illnesses. This is becoming more and more pertinent in our increasingly specialised medical world with the decline of the true generalist doctors.

CASE PRESENTATION

A previously fit and healthy 5-year-old boy presented to his general practitioner with left retro-orbital pain. He was diagnosed with tonsillitis and treated with oral antibiotics. A week later he was reviewed and found to have no improvement of his symptoms. Otological examination revealed a bulging left tympanic membrane. He was referred to the ENT department at Worcester Royal Hospital and underwent unilateral grommet insertion for presumed glue ear. Myringotomy revealed a serosanguinous effusion. Unfortunately, the grommet did nothing to relieve his symptoms and he went on to develop diplopia and a left abducens nerve palsy. MRI to investigate the soft tissues of the neck revealed a large left parapharyngeal mass, there was evidence of erosion through the skull base and compression of the VI and VII cranial nerves and trigeminal ganglion.

A biopsy was performed and revealed the diagnosis of embryonal rhabdomyosarcoma. Due to the site and size of the tumour it was deemed unsuitable for surgical reduction or resection.

Triple agent chemotherapy of ifosfamide, vincristine and actinomycin was therefore commenced. Eventually nine cycles of chemotherapy were undertaken with good result. The tumour responded well as did the symptoms of

pain and diplopia. Currently the patient remains in remission with no residual nervous symptoms.

INVESTIGATIONS

MRI revealed a large left parapharyngeal mass, there was evidence of erosion through the skull base and compression of the VI and VII cranial nerves and trigeminal ganglion (figure 1).

Biopsy revealed the histological diagnosis of embryonal rhabdomyosarcoma.

TREATMENT

Triple agent chemotherapy of ifosfamide, vincristine and actinomycin.

OUTCOME AND FOLLOW-UP

The tumour responded well with complete resolution of the symptoms of pain and diplopia. Currently the patient remains in remission with no residual nervous symptoms.

DISCUSSION

Guiseppe Gradenigo first described his triad of suppurative otitis media, trigeminal neuralgia and abducens nerve palsy in 1904.¹ Before the advent of widespread antimicrobial use it was a well-described complication of otitis media,² however, in the later half of the twentieth century it became a rare diagnosis.³ It is caused by the spread of infection from the mastoid air cells to the petrous apex of the temporal bone. This may cause a localised meningitis affecting the trigeminal ganglion and the adjacent abducens nerve causing the classic triad.

Spread can be through the pneumatized air cell tracts of the mastoid, direct invasion via fascial planes or via the vascular network.⁴

If left untreated the infection can cause a more widespread meningitis, cerebral abscess, Vernet's syndrome and parapharyngeal abscess.⁵ Traditional management of Gradenigo's syndrome included both surgery to remove the suppurative material from the mastoid coupled with

Figure 1 Coronal MRI image shows a mass in the left parapharyngeal space.

intensive antibiotic therapy. However, modern antibiotics and improved imaging techniques have enabled successful conservative management with antibiotics alone.⁶

The classical description of Gradenigo's syndrome implicates a suppurative otitis media as the cause. The most common pathogens include *Streptococcus*, *Staphylococcus aureus* and *Pseudomonas aeruginosa*. A review of the literature has revealed several other interesting, non-infective, causes.

Dorn *et al*⁷ describe a case of right-sided facial pain and headache with ipsilateral abducens nerve palsy secondary to fresh thrombosis of the right transverse and sigmoid sinuses. This was treated with intravenous antibiotics and heparin followed by mastoidectomy. Capanna⁸ reports a 19-year-old who developed facial pain and abducens palsy following a gunshot wound to the right temporo-parietal area. Angiography of his carotid trees revealed an enlarging aneurysm at the carotid siphon on the right. This was treated with ligation of the affected common carotid which unfortunately made no difference to the symptoms. Bourne and Maclaren⁹ report a case of a 45-year-old man who presented with horizontal diplopia, left facial pain and left otalgia following a vigorous episode of blowing his nose. His symptoms rapidly deteriorated to include palsy of the V, VI, VIII, X and XII cranial nerves. MRI revealed the presence of an extramedullary plasmocytoma arising from the left middle cranial fossa eroding the petrous apex and involvement of the affected nerves.

In our case, it is a rhabdomyosarcoma of the parapharyngeal space compressing the trigeminal ganglion and abducens nerve creating the symptoms of Gradenigo's syndrome.

Rhabdomyosarcoma is a relatively uncommon paediatric malignancy accounting for only 3.5% of all childhood cancers with tumours arising in the head and neck account for a quarter of those. Other sites commonly affected are the genitourinary tract (22%) and limbs (18%).

They are a member of the family of small round blue cell tumours that include non-Hodgkin's lymphoma, Ewing's sarcoma and neuroblastoma. If found early and treated before metastasis, curative rates of up to 70% can be achieved.¹⁰

Gradenigo's syndrome is a rare but still clinically important disease with potentially life-threatening complications regardless of its aetiology. With all 'classical' syndromes and diagnoses it is important to retain diagnostic acumen and consider different and unexpected causes for seemingly well-known illnesses. This case and the others described illustrate the range of pathologies capable of producing a similar clinical picture and fooling the unwary clinician. With appropriate imaging, malignant causes of Gradenigo's syndrome can be differentiated from others and in the case of embryonal rhabdomyosarcoma, treated with triple agent chemotherapy.

Learning points

- ▶ Gradenigo's syndrome is the triad of otitis media, trigeminal neuralgia and abducens nerve palsy.
- ▶ When dealing with classical disease descriptions it is important to maintain diagnostic acumen and pathological inquisitiveness to avoid missing unlikely diagnoses.
- ▶ Careful consideration of imaging is needed to avoid unnecessary investigation.
- ▶ Knowledge of cranial nerve anatomy will often help identify the origin of the pathology in multiple cranial nerve palsies.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Ricks RG, Hollis L. Embryonic rhabdomyosarcoma presenting as Gradenigo's syndrome. *BMJ Case Reports* 2012;10.1136/bcr.08.2011.4723, Published XXX

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