

Scedosporium and Cutibacterium skull base osteomyelitis complicated by blindness from fulminant papilloedema

James Corbett, Nigel Raymond, Rebecca Garland, Andrew Parker, Jesse Gale

A 48-year-old Pacific man with a background of diabetes developed chronic right otitis with mastoiditis in 2016 and underwent a modified radical mastoidectomy in 2018. Post-operatively he developed skull base osteomyelitis (SBO) and dural venous sinus thrombosis diagnosed with computed tomography (CT) with venogram. In 2020 his vision deteriorated with severe papilloedema and he was referred to our centre.

On review he had thick, green ear discharge and warm, non-tender swelling behind the right ear. Visual acuity was hand movements in right eye and no light perception in left eye. Ocular examination showed bilateral end-stage pallid optic disc swelling and bilateral retinal central retinal vein occlusions (Figure 1), without retinal ischaemia.

Full blood count showed mild neutrophilia. Glycated haemoglobin was 53mmol/mol (reference range <41mmol/mol) and C-reactive protein was 26mg/L (reference range <6mg/L). Superficial swabs from the right ear canal grew two yeasts identified as belonging to the *Candida parapsilosis* complex, thought to be commensal flora.

A follow-up CT with venogram showed the right mastoidectomy, mastoid air cell opacification, sclerotic and erosive changes affecting the right petrous temporal bone, sphenoid bone and clivus, and soft tissue inflammation including subgaleal collection. Right transverse and sigmoid sinus thrombosis was demonstrated.

Sterile subgaleal collection aspirate showed leukocytes but no organisms on Gram stain (no additional stains used), and grew environmental fungus *Scedosporium apiospermum*, sensitive to voriconazole with minimum inhibitory concentration 0.5mg/L.

Magnetic resonance imaging showed extra-axial enhancing soft tissue along the inner aspects of right occipital and temporal bones, extending into the posterior and middle

cranial fossae. Soft tissues in the scalp and neck were oedematous and enhancing (Figure 2). Lumbar puncture opening pressure was >38cmH₂O. Cerebrospinal fluid analysis showed 48x10⁶/L white cells (97% mononuclear) with raised protein and normal glucose. No organisms were isolated following extended incubation.

A multidisciplinary team including infectious disease, otorhinolaryngology, ophthalmology and neurosurgical specialists managed this case. Oral voriconazole 200mg BD and terbinafine 250mg daily were commenced with monitoring of serum levels and liver function. Oral acetazolamide 250mg TDS continued for 8 months to reduce intracranial pressure. Ventriculoperitoneal shunt was deemed inappropriate due to active extra-axial infection. Right optic nerve sheath fenestration was conducted to protect remaining vision: optic disc oedema resolved quickly but remaining light perception in the right eye was gradually lost.

Neuroimaging over the following year showed gradual improvement. Terbinafine was discontinued after 12 months due to increasing cholestatic liver enzymes. Subsequently, right-sided headaches recurred with neutrophilia and increased C-reactive protein (39mg/L). A CT demonstrated increased temporoparietal lobe oedema and extracranial soft tissue inflammation. Another sterile deep tissue biopsy was taken, including deep fascia, occipital bone and extradural tissue. *Cutibacterium acnes* was grown from bone and fascia. Oral doxycycline 100mg BD was initiated for possible bacterial coinfection given excellent bony penetrance, with gradual clinical improvement. He received 26 months of antifungal treatment and 12 months of antibacterial treatment.

Skull base osteomyelitis is a rare and dangerous infection spreading from otological or sinus infections, typically caused by *Pseudomonas aeruginosa* with malignant otitis externa.^{1,2,3} *S. apiospermum* is a ubiquitous fungus that most

Figure 1: a) Colour fundus photography at presentation showing pale, swollen optic discs, and bilateral central retinal vein occlusions with collateral vessel formation. b) Colour fundus photography 3 years later showing optic atrophy with gliosis. Some minor pigment irregularities in the mid-periphery represent panretinal photocoagulation treatment, which was not completed.

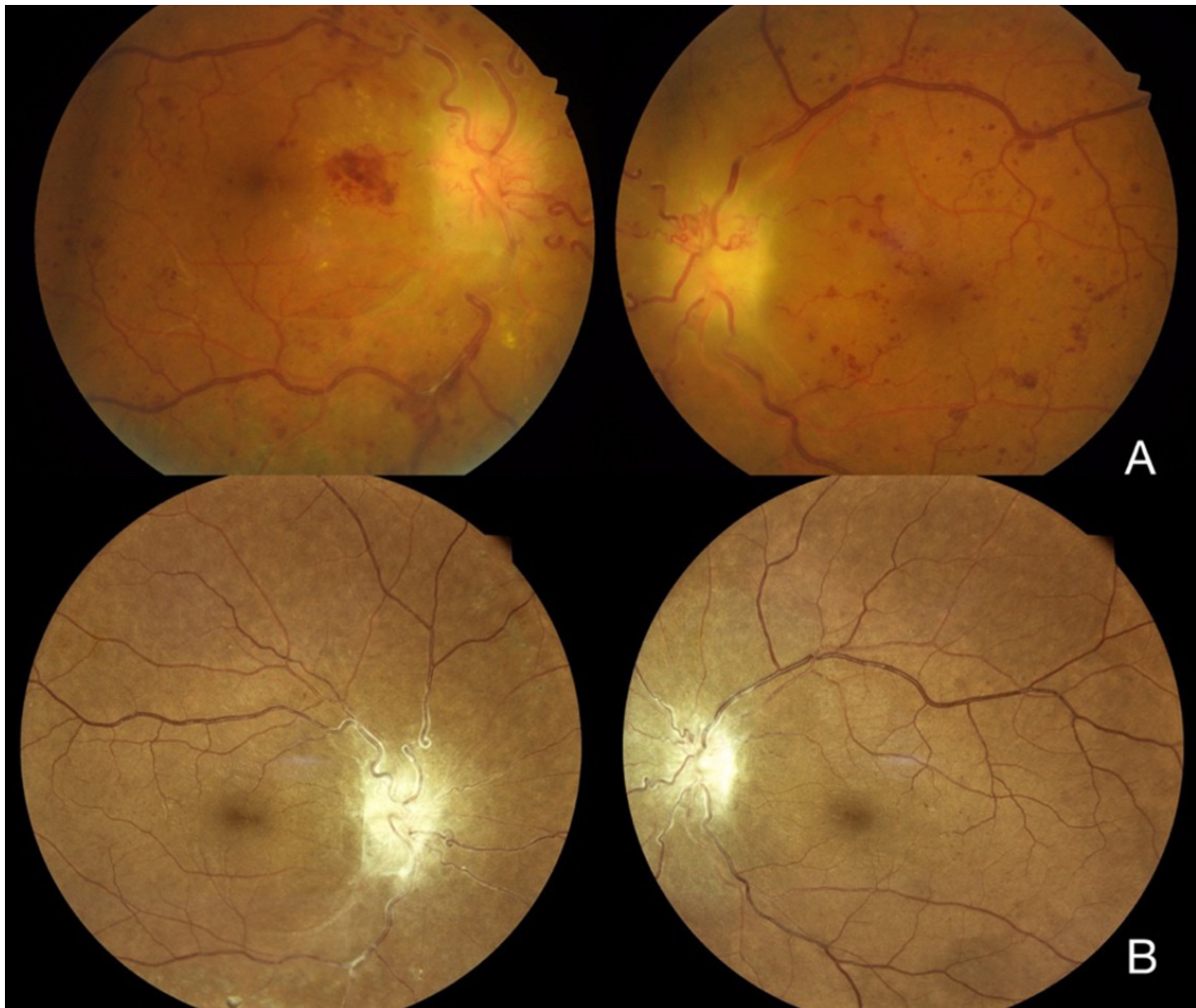
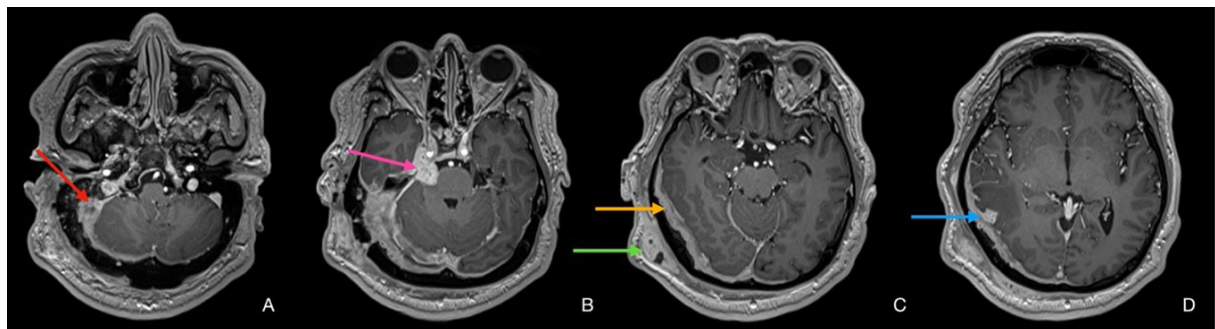


Figure 2: Axial T1-weighted gadolinium contrast-enhanced MRI sections. a) Section at ear canals showing enhancing inflammatory tissue extending from right periauricular soft tissue through abnormal modified right petrous temporal bone to posterior fossa (red arrow). b) Nodular deposit (pink arrow) extends into the right prepontine cistern, affecting right trigeminal nerve. c) Plaque-like enhancing dural thickening extending cranially to surround the right temporal lobe (orange arrow). Sub-galeal nodular enhancing soft tissue (green arrow) in the temporoparietal area. d) Temporal nodular dural focus with surrounding cerebral oedema and midline shift (blue arrow).



commonly causes infection in immunocompromised patients.⁴ *C. acnes* has also been reported to be the causative organism in a small number of SBO cases complicated by cranial neuropathy.⁵ The facial nerve is the most implicated cranial nerve in SBO due to its course through the stylomastoid foramen.⁶ Optic nerve involvement is much less common, and in this case the involvement was indirect through intracranial hypertension. The case highlights important aspects of SBO,

particularly 1) external ear swab results can be misleading, especially after antibiotic treatment;⁷ 2) a prolonged relapsing course from treatment failure is well recognised;⁸ thus 3) sterile-site microbiological samples should be prioritised in the workup of SBO, after 48 hours without antimicrobials; and finally 4) among the catastrophic complications of SBO, this is the first report of intractable blinding papilloedema despite optic nerve sheath fenestration.⁹

COMPETING INTERESTS

Nil.

AUTHOR INFORMATION

James Corbett: Ophthalmology Registrar, Wellington Regional Hospital.

Nigel Raymond: Infectious Disease Specialist, Wellington Regional Hospital.

Rebecca Garland: Otorhinolaryngologist, Wellington Regional Hospital.

Andrew Parker: Neurosurgeon, Wellington Regional Hospital.

Jesse Gale: Ophthalmologist, Wellington Regional Hospital.

CORRESPONDING AUTHOR

Jesse Gale: Ophthalmologist, Wellington Regional Hospital, Private Bag 7902, Wellington South 6242.

E: Jesse.gale@ccdhb.org.nz

URL

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