PHOTO ESSAY

Cerebral Venous Thrombosis with Papilloedema Secondary to Skull Base Plasmacytoma

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ABSTRACT

A 60-year-old woman with history of multiple myeloma was in remission after stem cell transplant 6 years prior. She was undergoing work-up for headaches that were thought to be secondary to a right mastoiditis seen on magnetic resonance imaging (MRI). On routine eye exam, papilloedema was noted. A lumbar puncture was performed, with elevated opening pressure with normal constituents. She was an atypical age for idiopathic intracranial hypertension, and her mastoiditis raised concern for secondary cerebral venous sinus thrombosis. Magnetic resonance venography (MRV) was performed showing poor flow in the right sigmoid sinus, and computed tomography venography (CTV) showed lack of contrast enhancement distal to the right sigmoid sinus, consistent with occlusion. There was also an enhancing mass inferior to the right occipital bone. Biopsy confirmed recurrent plasma cell myeloma. She was treated with chemotherapy, radiation, and warfarin for presumed cerebral venous sinus thrombosis.

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Cerebral venous sinus thrombosis; papilloedema; plasmacytoma; skull base

Case presentation

A 60-year-old woman with history of multiple myeloma was in remission after stem cell transplant 6 years prior. She was undergoing work-up for headaches and had magnetic resonance imaging (MRI) of the brain that showed right mastoiditis. At that time, she was already under the care of an otolaryngologist for symptoms of “stuffy ear”. Laboratory tests, including complete blood count, angiotensin-converting enzyme (ACE), and Lyme titres, were normal.

Two months later, she saw an ophthalmologist for routine eye examination, who observed papilloedema. Brain MRI was repeated showing worsening mastoiditis. She was sent for lumbar puncture, which returned elevated cerebrospinal fluid opening pressure of 47 cm H₂O but no laboratory abnormalities, including white blood cell count, fungal cultures, ACE, acid-fast bacteria, oligoclonal bands, and Gram stain. She was started on acetazolamide. MRI was repeated 2 months later, with stable findings.

She was referred for neuro-ophthalmic evaluation due to persistent papilloedema and headaches. Frisen grade 2 papilloedema without haemorrhages was confirmed, and her Humphrey visual field showed inferior constriction in the right eye but diffuse constriction in the left. There was concern for possible secondary venous sinus thrombosis related to chronic mastoiditis. Magnetic resonance venography (MRV) showed poor flow in the right sigmoid sinus (Figure 1). Computed tomography venography (CTV) showed lack of contrast enhancement distal to the right sigmoid sinus consistent with occlusion as well as an enhancing mass inferior to the right occipital bone and lateral to the occipital condyle compressing the right sigmoid sinus (Figures 2). She was referred to otolaryngology and stroke neurology for evaluation and management.

Two otolaryngologists felt the mastoiditis to be mild and insufficient to account for cerebral venous thrombosis (CVT). An ear tube was placed for presumed infectious mastoiditis but did not reveal any fluid.

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Figure 1. MRV showing thrombosis of the distal right sigmoid sinus and right jugular bulb (white arrow).

Right posterior neck lymph node and right skull base mass biopsies revealed increased population of plasma cells, diagnostic of recurrent plasma cell myeloma (Figures 3). She underwent chemotherapy and radiation for her tumour and started warfarin for presumed CVT.

Discussion

Plasmacytomas are malignant bony tumours and represent 1% of all head and neck tumours.^{1,2} Eighty percent affect the upper airways and paranasal sinuses, with rare involvement of the skull base. In the skull base, they most often affect the clivus or

Figure 3. High-power view of lymph node biopsy showing sheets of plasma cells. Plasma cells were kappa light chain restricted by in situ hybridisation studies (not shown).

petrous apex where they can cause compression of cranial nerves.³⁻⁵ The most common presenting symptoms are headache, visual deficits due to cranial nerve palsies, and endocrinologic abnormalities. Although there are case reports of falcotentorial plasmacytomas causing elevated intracranial pressure and papilloedema,⁶⁻⁸ no reports have been made in the literature of secondary venous sinus thrombosis from a skull base plasmacytoma.^{1,2}

CVT is a rare type of stroke, most common in women under the age of 50.⁴ The most common causes of CVT are prothrombotic conditions, and the most commonly involved sinus is the superior sagittal sinus, whereas transverse and sigmoid sinus thromboses are commonly caused by parameningeal infections, including middle ear infection and mastoiditis.⁴ In the International Study on Cerebral Venous and Dural Sinus Thrombosis, researchers

Figure 2. MRI, May 2016. (A) Axial T1 pre-contrast, (B) axial T1 post-contrast, and (C) coronal T1 post-contrast showing right occipital condyle enhancing lesion compressing the right sigmoid sinus (white arrows).

found that 7.4% of cases of CVT were associated with cancer, particularly haematologic malignancies. Potential mechanisms include direct tumour compression, tumour invasion of cerebral sinuses, and increased risk of thrombosis,⁴ all three of which may have played a role in our patient.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

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