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Neuro-ophthalmic presentations of clival plasmacytoma

Neurologically isolated cranial mononeuropathy (e.g., sixth nerve palsy) leading to diplopia is a common presentation to ophthalmologists. Although rare, clival lesions can cause isolated unilateral or bilateral abducens palsies with or without papilledema. Intracranial plasmacytomas (ICPC) are rare tumours that constitute less than 1% of intracranial neoplasms. They may present as a solitary plasmacytoma or may be part of a systemic malignant plasmacytosis, as in multiple myeloma (MM).

We describe 2 novel cases of ICPC presenting with abducens nerve palsies. The first patient had previously treated MM, but developed new binocular diplopia with fluid and decreased hearing in the right ear. The second patient presented with bilateral abducens palsies and associated right-sided V2 hypothesia and subsequently was diagnosed with both clival plasmacytoma and MM.

A 59-year-old African-American female presented with a 3-week history of horizontal binocular diplopia. She also had a headache and "fluid sensation" in her right ear causing mild hearing loss. Her medical history included MM successfully treated with chemotherapy 3 years ago, well-controlled hypertension, and hypothyroidism. Surgical history included spinal surgery and knee replacement for degenerative joint disease 8 years previously. She denied any drug allergies and was taking levothyroxine, amlodipine, amoxicillin, and low-dose prednisone. She denied alcohol, tobacco, or drug use. Her family history was significant for colon cancer in her sister, and diabetes, hypertension, and stroke in her father. Review of systems was otherwise negative. On neuro-ophthalmic examination, best corrected visual acuity (BCVA) was 20/25 OD and 20/20-2 OS. Ishihara colour plates were 14/14 OU. Humphrey automated visual fields were within normal limits OU. Pupils measured 4 mm in dark and 2 mm in light, and no relative afferent pupillary defect (RAPD) was noted. Intraocular pressure measured 19 mm Hg OU. There was no disk edema or pallor. Her ocular motility showed a −4 deficit of abduction OD. She had an incomitant 30 prism diopter (PD) esotropia (ET) in primary gaze, which increased in right gaze and decreased in left gaze. The remainder of her ophthalmologic examination was unremarkable.

Magnetic resonance imaging (MRI) of the brain with and without contrast showed a large enhancing lesion centred in the clivus measuring 3.2 × 1.9 × 3.1 cm (Fig. 1). The mass invaded the right cavernous sinus and extended through the posterior table of the sphenoid sinus on the right with invasion of the sellar floor and superior displacement of the pituitary gland. Heterogeneous enhancement throughout the calvarium was suspicious for infiltration, and a biopsy showed ICPC. Immunohistochemistry stained positive for CD38, CD138, CD56, and antilambda. Bone marrow biopsy showed 49% lambda light chain–restricted plasma cells consistent with plasma cell myeloma. The eye was patched for symptomatic relief, and she was treated with radiation therapy and bortezomib.

An 84-year-old female presented with headache, blurry vision, and progressive binocular horizontal diplopia over 2 weeks. Her medical history was significant for stroke, hypertension, coronary artery disease, congestive heart failure, aortic aneurysm, thyroidectomy, recent acute kidney injury, renal cysts, angiomyolipoma, hiatal hernia, gastrointestinal bleeding, multiple gastric polyps, and diverticulosis. Surgical history included coronary stenting and cholecystectomy. She denied any drug allergies and was on amlodipine, atorvastatin, carvedilol, dexlansoprazole, furosemide, meloxicam, potassium, and ranolazine. She denied tobacco, alcohol, and illicit drug use. Her family history was significant for fatal abdominal cancer in her mother and fatal cerebral aneurysm in her father. On neuro-ophthalmic examination, the visual acuity was 20/60 OD and 20/25 OS. Pupils were symmetric in light and dark without anisocoria or RAPD. Intraocular pressure measurements were 13 mm Hg OU. There was no disk edema or pallor. There was a 50 PD ET in primary position, which increased in both right and left gazes.

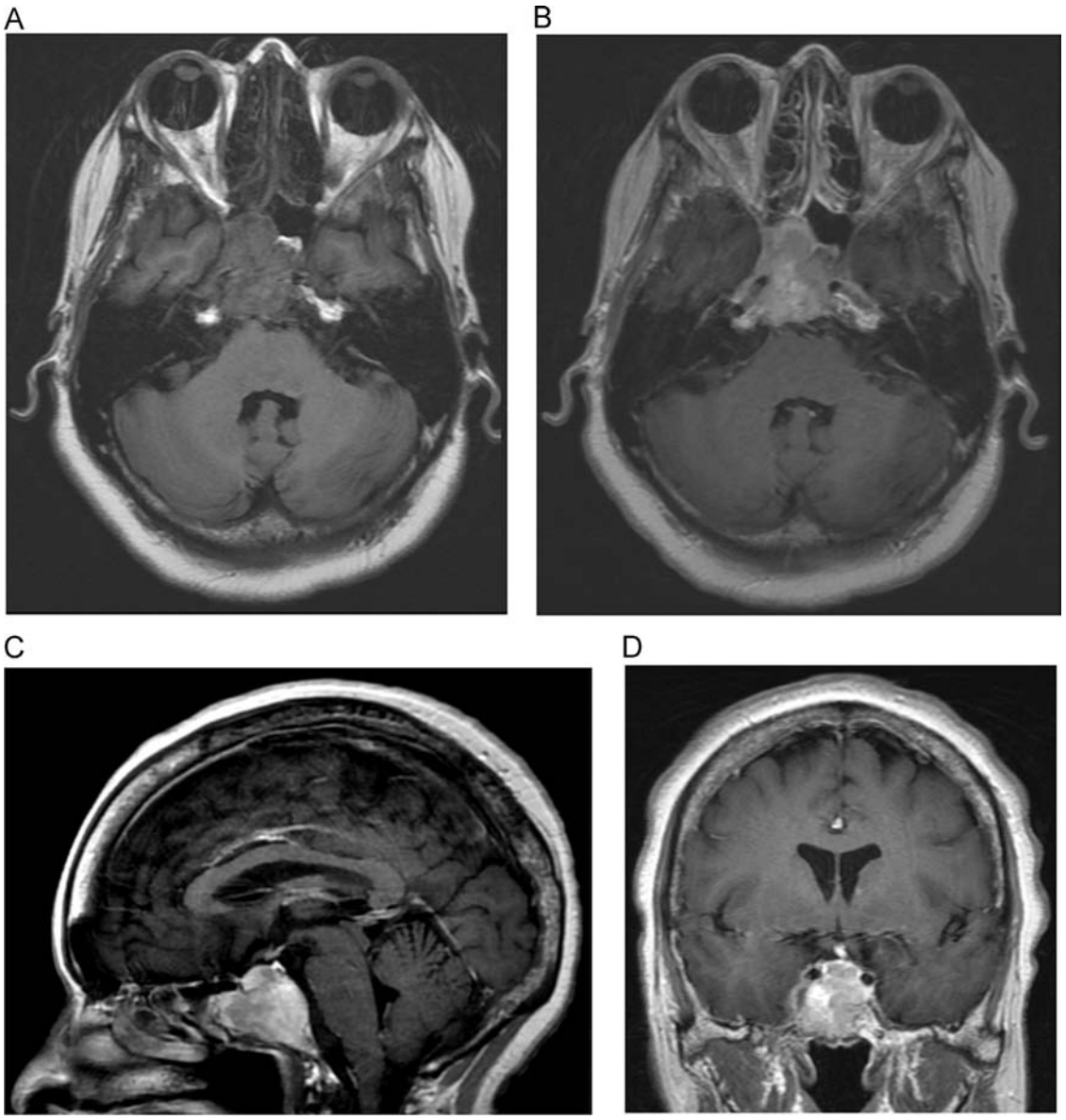

Fig. 1—Magnetic resonance imaging of clival plasmacytoma, case 1. Images demonstrate the heterogeneously enhancing clival-based mass with extension into the right cavernous sinus. T1 axial precontrast (A) and postcontrast (B), sagital postcontrast (C), and coronal postcontrast (D).

Motility showed −4 abduction deficits bilaterally. The remainder of her ophthalmologic examination was unremarkable.

Sensation to pinprick was decreased in the right V2 distribution. Findings were consistent with a bilateral abducens palsy and right V2 anaesthesia. MRI revealed a 3 × 4 × 6-cm clival mass extending into the sphenoid and posterior ethmoid sinuses (Fig. 2). Trans-sphenoidal biopsy showed abundant pleomorphic plasma cells in a hemorrhagic background with occasional Dutcher and Russell bodies, and immunohistochemical staining for CD138 and lambda light chain were diffusely and strongly positive, consistent with plasmacytoma. Bone marrow biopsy showed lambda-restricted, immunophenotypic aberrant monotypic plasma cells comprising 19% of the total cellularity. Findings suggested hyperdiploid MM. The patient was started on radiation therapy and bortezomib. She suffered from a fatal cardiorespiratory arrest during her hospitalization.

Clival masses may often be differentiated clinically and radiographically by location. Lesions may arise from the skull base itself (e.g., plasmacytoma, chordoma, chondrosarcoma, neuroenteric cysts, intraosseous lymphoma, and metastasis); the intracranial compartment with secondary contiguous involvement of the clivus (e.g., meningioma, craniopharyngioma, and pituitary macroademoma); or from the adjacent sinuses (e.g., nasopharyngeal carcinomas, mucoceles, or squamous cell carcinomas). Cranial nerve palsies have been identified in a subset of these skull base pathologies. Chordoma, chondroma, and

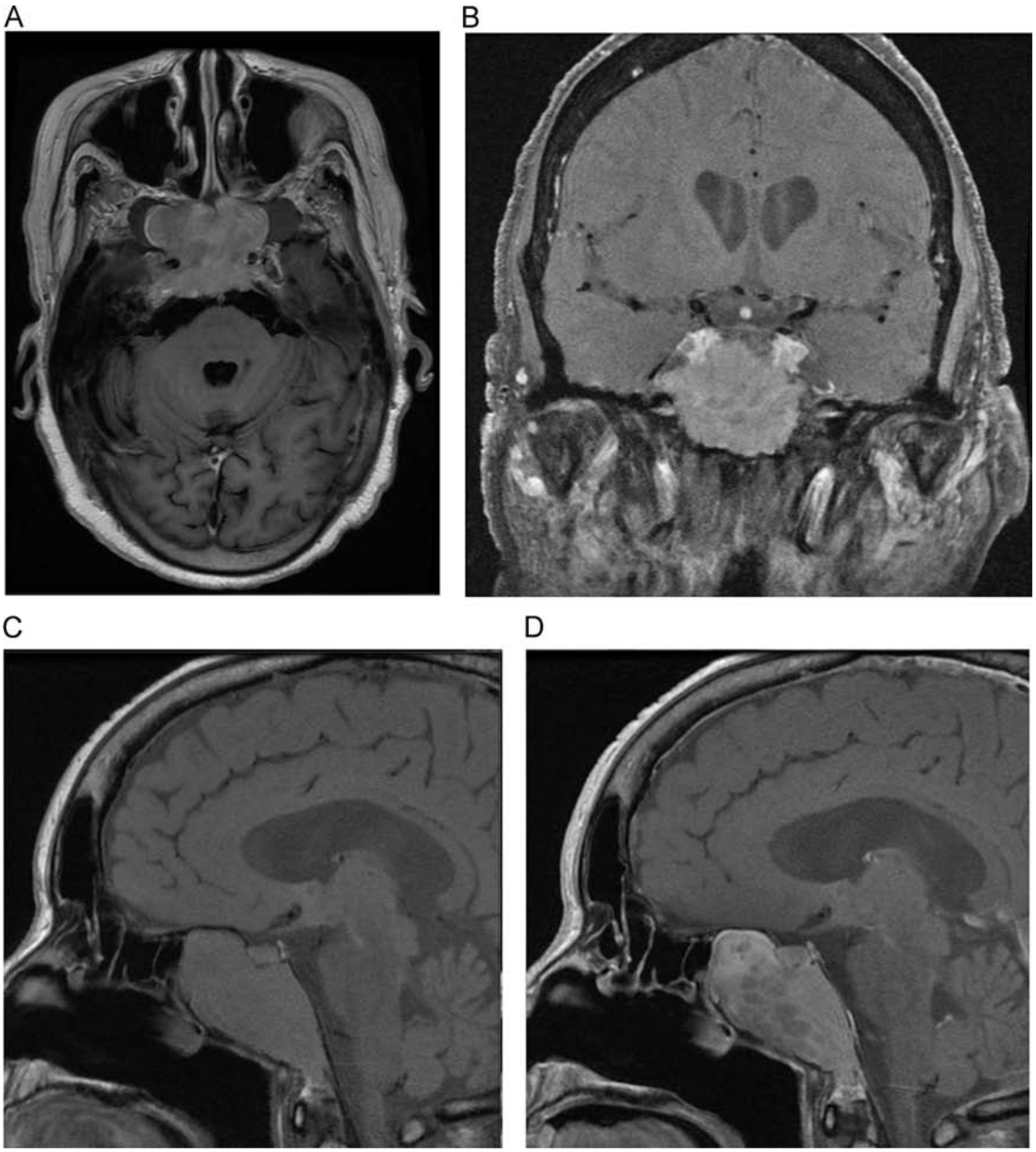

Fig. 2—Magnetic resonance imaging of clival plasmacytoma, case 2. Images show the large heterogeneously enhancing clival mass with greater involvement of the left than right cavernous sinuses. T1 axial postcontrast (A), coronal postcontrast (B), sagital precontrast (C), and sagital postcontrast (D).

chondrosarcoma have all been reported to produce sixth nerve palsy, and in some of these cases the diplopia spontaneously resolved.[1–8] The clinical presentations for these lesions varied from isolated sixth nerve palsy to sixth nerve palsy in association with headache, diplopia, facial numbness, and papilledema.[3–8]

It is often difficult to distinguish primary clival lesions (e.g., chordoma) from plasmacytoma, but chordomas typically occur in younger patients, whereas plasmacytomas are more prevalent in the elderly population. The presence of MM can also suggest the diagnosis of plasmacytoma. Although both plasmacytoma and chordomas extend from the clivus, the tumors may often be distinguished from imaging. A single trabeculated expansile lytic lesion accompanied by cortical thinning and destruction in the absence of sclerotic reactions is the hallmark of clival plasmacytoma. On computed tomography scan, an expansile lytic lesion with a thin cortex is similarly observable.[9] On MRI, a curvilinear low-intensity signal resembling sulci may be noted within the lesion. This finding has high specificity for plasmacytoma and may obviate the need for biopsy especially in patients with known systemic disease.[10] On MRI T2-weighted images, the plasmacytoma lesions are isointense to hyperintense, whereas chordomas demonstrate a high T2 signal.[11] Nevertheless, some patients still require tissue biopsy and

histologic and immunohistochemical confirmation of the diagnosis.

To our knowledge, these cases represent distinct presentations of ICPC. Our first patient presented with fluid in the ear, decreased hearing, and a new sixth nerve palsy after successful treatment for MM. Our second patient presented with V2 hypothesia and new onset bilateral sixth nerve palsy without a prior MM diagnosis. Two prior cases of plasmacytoma involving trigeminal parasthesias have been reported[12,13]; however, neither of these cases reported concurrence with bilateral sixth nerve palsy. ICPC as the underlying cause of sixth nerve palsy is uncommon, but at least 26 prior clival plasmacytoma cases presented with diplopia.[14–25] Of these 26 cases, 2 reported bilateral abducens nerve palsies.[22,23]

ICPC presentation may vary depending on the involved site. Raised intracranial pressure leading to papilledema, confusion, cranial nerve palsies, and seizures is sometimes observed in skull base lesions. These ICPC lesions have a high rate of progression to MM and a significantly worse prognosis than extramedullary plasmacytomas.[18,26] The rate of progression in these lesions may be as high as 50% with a variable 10-year survival rate ranging from 16% to 100%.[15,17] Systemic surveillance is generally recommended for at least 1 year after discovery of ICPC for the possible development of MM.[15] Fifteen (62.5%) of the 26 clival plasmacytoma cases harboured underlying MM,[14–23] as was also described in our second case. Although our second patient had multiple vasculopathic risk factors and no history of neoplasm, the atypical findings prompted neuroimaging. Clinicians should be aware that, although a sixth nerve palsy may be ischemic in a vasculopathic patient, neuroimaging may be warranted in patients with lack of improvement or progression over time, other neurologic signs/symptoms (nonisolated), bilateral involvement, or lack of vasculopathic risk factors. ICPC should especially be considered in patients with a prior diagnosis of MM.

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Horner syndrome in fibromuscular dysplasia without carotid dissection

A 62-year-old female was referred for right-sided lid ptosis and periorbital pain of a 1-month duration. She denied diplopia or any significant fluctuations in her ptosis, which had begun acutely 1 month before. She denied any history of trauma and had not noticed anisocoria previously. The patient had a complex history of chronic rhinosinusitis requiring endoscopic debridement with multiple revisions, the most recent of which was 6 weeks before her presentation. An episode of preseptal cellulitis heralded that operation and further complicated the etiology of her periorbital pain.

Upon ophthalmologic examination, her visual acuity was 20/20-1 OD and 20/30 OS. Unilateral right-sided ptosis was evident with an unequal upper marginal reflex distance (right 1 mm, left 5 mm) and asymmetric palpebral fissures (right 6 mm, left 10 mm). Levator function was symmetric (14 mm OU), and there was no fatigable component to the ptosis. Pupils were anisocoric (right 4.5 mm, left 6 mm) with a brisk response to light (right 2.5 mm, left 4 mm) although, interestingly, the anisocoria was not more prominent in the dark. There was no relative afferent defect. No periorbital swelling, facial rashes, or other skin findings were noted, and the remainder of the ocular examination was unremarkable.

To further evaluate for Horner syndrome (HS), 2 drops of 0.5% apraclonidine were instilled in each eye. This resulted in reversal of the anisocoria and improvement in ptosis, confirming a diagnosis of HS. Subsequent magnetic resonance imaging (MRI) of the brain showed only chronic small vessel disease and sinus pathology (mucosal thickening in the bilateral frontal, maxillary, and sphenoid sinuses and ethmoid air cells and small fluid levels in the maxillary sinuses) consistent with prior disease but no acute parenchymal infarct, hemorrhage, or mass. Brain and neck magnetic resonance angiography (MRA) showed no flow-limiting stenosis, aneurysm, or dissection. Neck MRA, however, showed ectatic distal internal carotid arteries (ICA), the right greater than the left, and multiple areas of narrowing and dilatation of the ICA and vertebral arteries bilaterally (Figs. 1 and 2). These findings are consistent with multifocal fibromuscular dysplasia (FMD).[1] Given the patient's smoking history, a lower neck mass or apical lung tumour affecting the right sympathetic chain was also considered, but follow-up imaging to the level of T2 revealed no such lesion. The neck MRI also confirmed the absence of ICA dissection. Although HS from an unidentified cause, with only coincidental radiological findings, could not be excluded, the more severe right-sided disease was the most likely etiology of the unilateral HS.

FMD is a nonatherosclerotic, noninflammatory vascular disease that may result in arterial stenosis, occlusion, aneurysm, or dissection.[1] A histological classification system, based on the affected vascular layer, was published by Harrison and McCormack in 1971, but the American Heart Association's proposed system, based on angiographic patterns, is more appropriate in this case.[2] Intimal fibroplasia causing focal arterial constriction characterizes "focal FMD," whereas multifocal disease, exhibited in our patient by the multiple areas of narrowing and dilation, indicates medial fibroplasia.[1] Traditionally, FMD was seen as a disease of young, premenopausal women but, although 90% of FMD cases occur in females, our 62-year-old patient was a typical age for presentation based on the most recent literature. In fact, the average age on diagnosis is 51.9 (13.4) years, with a range from 5 to 86 years.[3]

Secondary hypertension caused by FMD-related renovascular disease is a well-documented entity that can result in end-organ damage from hypertensive emergency and/or chronic hypertension, but FMD is also an important cause of cerebral aneurysm and subarachnoid hemorrhage as well as mesenteric ischemia and infarct.[4,5] The renal arteries are most commonly affected with involvement in as many as 75% of FMD cases; in the largest series, however, 32% had vertebral or ICA involvement, as in this case.[6,7]

HS is an uncommon manifestation of cerebrovascular FMD; it was the presenting sign in only 4.7% of patients in the U.S. Registry for FMD. Although it is not uncommon to have a carotid dissection due to FMD or for a dissection to present with HS, our review of the literature found no cases of FMD presenting as HS without concurrent ICA dissection.[3,8] Positive apraclonidine tests have been reported in HS caused by lesions of all 3 segments (central, preganglionic, and postganglionic) of the sympathetic pathway.[9] As such, radiologic studies could not be targeted on a particular nerve segment and a comprehensive imaging workup was initiated.

Catheter-based angiography is the diagnostic gold standard for FMD. Although MRA has been well validated for cases of renal FMD with a sensitivity of 97% and specificity of 93%, no such studies have confirmed the utility of MRA in cerebrovascular FMD.[1,10] In this case, carotid artery dissection had to be considered given the ipsilateral pain in conjunction with HS, and MRI and MRA are adequate means of assessing carotid or vertebral arterial dissection.[11,12] Interestingly, the patient had been seen regularly over the prior 2 years by