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Case Report

Tumor cerebri: Metastatic renal cell carcinoma with dural venous sinus compression leading to intracranial hypertension; a case report

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Abstract

Background: Pseudotumor cerebri (PTC), also known as idiopathic intracranial hypertension (IIH), is a condition associated with increased intracranial pressure (ICP) in the absence of radiographic findings such as mass lesions or cerebral edema.

Case Description: We describe a case of progressive headache and visual disturbances attributed to PTC that resulted from subacute superior sagittal sinus (SSS) stenosis by a metastatic tumor.

Conclusions: Venous outflow obstruction often presents with an acute symptomatology including infarcts, hemorrhages, and seizures, but only rarely does it cause the progressive development of raised ICP. The sinister presentation of our patient's pathology stemmed from local mass effect caused by a tumor that has hitherto not been reported to cause intracranial hypertension (IH) and was best elucidated using magnetic resonance venography (MRV).

Key Words: Idiopathic intracranial hypertension, papilledema, pseudotumor cerebri, renal cell carcinoma, venous sinus compression

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INTRODUCTION

Pseudotumor cerebri (PTC), also known as idiopathic intracranial hypertension (IIH), is a condition associated with increased intracranial pressure (ICP) in the absence of radiographic findings such as mass lesions or cerebral edema.[11] The concept, introduced by Nonne in 1904, attempted to explain patients whose presentations suggested an intracranial tumor, but clinical courses appeared to preclude this diagnosis.[16,22] IIH can lead to headaches, diplopia, papilledema, and progressive vision loss. It can affect patients of any age or size, but is predominantly found in obese women of childbearing age. The pathophysiology of IIH is not entirely understood, but it has been associated with venous sinus obstruction. In fact, some studies indicate that transverse sinus stenosis is seen in over 90% of patients with IIH.[9,31] Magnetic resonance venography (MRV) has been shown to be an appropriate screening tool in these patients with a sensitivity of 93–100%.[6,9] There are conditions that mimic IIH,[7,25,26,37] but the presence of non-acute, non-thrombotic superior sagittal sinus (SSS) compression leading to signs and symptoms resembling IIH is rare. We

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describe a case of metastatic renal cell carcinoma (RCC) with SSS compression presenting as PTC.

CASE DESCRIPTION

VH is a 50-year-old male who presented to an outside facility with complaints of diplopia and headache initially diagnosed as PTC. He was evaluated by ophthalmology and neurology before being transferred to us for further tertiary management of sagittal sinus thrombosis. The patient described mild to moderate headaches of 3 weeks duration with blurry vision and 1 week of double vision that seemed to be exaggerated with rightward gaze. The patient had no pertinent past medical history. His physical exam was remarkable only for bilateral papilledema and subtle right-sided abducens nerve palsy. He was otherwise alert and oriented, with full muscle strength and without myelopathy or other cranial nerve findings. Of note, he had a palpable, compressible soft tissue mass over the vertex of his skull.

Imaging demonstrated a midline parietal extradural mass with erosion through the skull and into the subgaleal soft tissues. Magnetic resonance imaging (MRI)/MRV demonstrated depression of the SSS with local stenosis in that region. Computed tomography (CT) of the chest, abdomen, and pelvis yielded a 4-cm solid mass on the upper pole of the right kidney. The patient was initially started on a heparin drip by the primary team which was discontinued after definitive imaging was obtained and prior to a diagnostic lumbar puncture (LP). After an opening pressure of 51 cm H_2O by manometer in the lateral recumbent position confirmed intracranial hypertension (IH), he was placed on steroids and acetazolamide prior to discussion and recommendation of surgical resection.

A biparietal craniectomy was performed with gross total resection of the mass as demonstrated in preoperative and postoperative MRI scans [Figure 1]. Recovery was uneventful with improvements in the patient's headache and gradual improvement in the patient's diplopia. A repeat LP performed 2 days after surgical resection showed an opening pressure of 4 cm H_2O and he was discharged on postoperative day number 3. Follow-up MRV demonstrated improved dural venous outflow [Figure 2]. Ophthalmology follow-up 2 weeks later documented improvement in his papilledema and a repeat LP at that time gave an opening pressure of 21 cm H_2O. He received a cytoreductive right laparoscopic radical nephrectomy 6 weeks after his craniectomy. Twelve weeks after his cranial surgery, he presented with right leg pain related to a metastatic lesion to his femur necessitating prophylactic intramedullary nailing (IMN). Seven days later, X-ray demonstrated a minimally displaced pathologic fracture within the lytic lesion. Unfortunately, prior to receiving adjuvant radiation and chemotherapy, he was found deceased at home 1 week later from an unknown cause, approximately 4 months after presentation.

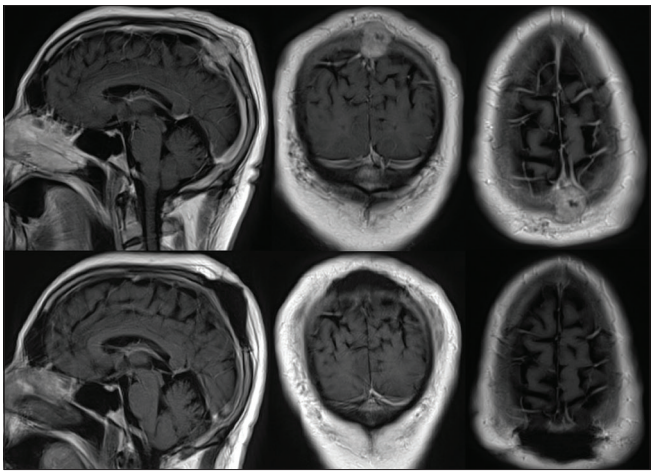

Figure 1: Preoperative (top) axial, coronal, and sagittal T1 magnetic resonance (MR) with contrast demonstrating an enhancing epidural mass extending through the calvarium and into the subgaleal space with compression of the superior sagittal sinus (SSS). Postoperative (bottom) axial, coronal, and sagittal T1 MR with contrast showing gross total resection of tumor with preservation of the SSS and resolution of sinus stenosis

DISCUSSION

The mechanism of increased ICP in IIH has not been fully elucidated, but the main concepts utilize the Starling resistor hypothesis and the Monro-Kellie doctrine. The latter doctrine explains that because the volume within the cranial compartment is fixed by the rigid confines of the skull, any increase in volume of one of the cranial constituents [brain matter, blood, and cerebrospinal fluid (CSF)] occurs at the expense of the others and the extent of which is at least in part determined by compensatory mechanisms in healthy individuals.[1,35] The former hypothesis elaborates that the collapsible terminal venous components reflect increased ICP to upstream dural sinuses and cerebral veins until they equal the ICP via a hydraulic mechanism.[5,33] In this case, it was assumed that impaired CSF reabsorption was secondary to the decreased CSF-venous pressure gradient across the arachnoid villi created by progressive stenosis of the sagittal sinus by the tumor.[32] Furthermore, the pressure response to increased CSF volume is not linear, but exponential.[5] Therefore, any increase in resistance to venous drainage will cause decreased upstream reabsorption of CSF leading to increased ICP, venous collapse, and exponential compounding until IH is developed.[33]

Papilledema and IH are well known phenomena that can occur with pathology of the dural venous sinuses.[9,17] In the setting of SSS involvement, symptoms of IH often

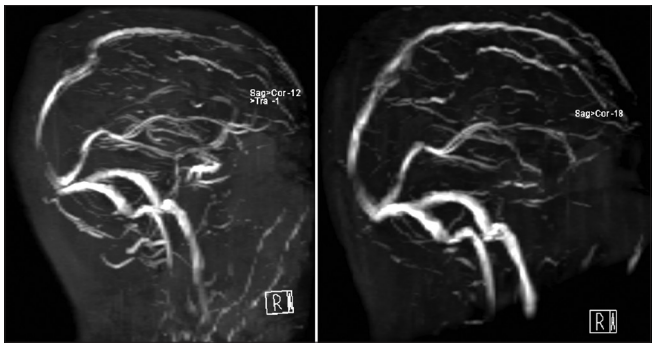

Figure 2: Preoperative (left) magnetic resonance venography (MRV) demonstrating displacement and attenuation of the superior sagittal sinus (SSS). Postoperative (right) MRV demonstrating near resolution of SSS compression

occur in the context of thrombosis[28] and trauma,[12,34] but seldom due to direct compression. However, the SSS is rarely involved in an acute/subacute setting in the absence of clinical features suggesting cortical vein involvement such as seizures, paresis, venous infarcts, etc.[27] Most cases of SSS occlusion associated with neoplasms are thought to be attributed to thrombotic complications secondary to hypercoagulability states.[13] It must also be noted that removal of the offending pathology, whether it be thrombus or mass effect, may not always result in permanent cure of the IH.[32]

IH has rarely been described in the context of tumors compressing a dural venous sinus. Case reports include Ewing's sarcoma, plasmacytoma, neuroblastoma, disseminated carcinoma of the breast, and prostate cancer.[19,21,27-29] Tumors involving the posterior aspect of the SSS and/or the torcular herophili, dominant transverse sinus, or jugular foramen have a tendency to cause more symptomatic venous compression.[2,4,19,36] Though slow growing tumors such as meningiomas have been associated with "pseudotumor-like" symptoms,[32] IH attributable to neoplasms are more commonly seen with rapidly growing masses. Slower growing tumors with sinus compression are thought to be better tolerated because of the development of collateral venous drainage.[14,18] To our knowledge; however, IH has not been described with metastatic RCC.

Patients with brain metastases from RCC have a poor prognosis. The average survival time is 3 months if left untreated and 2–9 months if treated with whole brain radiation therapy (WBRT).[10] Unfortunately, metastatic RCC responds poorly to both WBRT and stereotactic radiosurgery (SRS).[10] Longer survival may be predicted by the absence of IH, few metastatic foci and extracranial metastasis. Surgical resection does seem to improve patient survival,[3] and in patients with a single brain lesion, good performance status, and limited/controlled systemic disease, surgery has been shown to confer an even greater survival benefit.[15,24] However, it must be noted that local failure rates approach 60%.[15] Cytoreductive nephrectomy may improve survival in patients with RCC, though brain metastasis is a known independent predictor of worse overall survival[23] and its use has become controversial in this setting.[30] New immunotherapies are showing promise for extending survival time.[8]

CONCLUSION

In conclusion, when patients present with signs and symptoms resembling PTC, we must reiterate that intracranial mass lesions must first be ruled out as IIH is, by definition, a diagnosis of exclusion.[20] In addition, there is increasing evidence that non-traumatic, non-acute symptoms indicative of IIH such as papilledema, headaches, and visual disturbances may suggest sinus involvement and early vascular imaging should strongly be considered. Though symptomatic venous sinus compression from an intracranial neoplasm is a rare phenomenon, surgical resection can help alleviate symptoms attributable to elevated ICP.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Aaslid R, Lindegaard KF, Sorteberg W, Nornes H. Cerebral autoregulation dynamics in humans. Stroke 1989;20:45-52.
2. Angeli SI, Sato Y, Gantz BJ. Glomus jugulare tumors masquerading as benign intracranial hypertension. Arch Otolaryngol Head Neck Surg 1994;120:1277.
3. Bennani O, Derrey S, Langlois O, Castel H, Laquerriere A, Freger P, *et al*. Brain metastasis from renal cell carcinoma. Neurochirurgie 2014;60:12.
4. Brookes G, Graham M. Benign intracranial hypertension complicating glomus jugulare tumor surgery. Am J Otol 1984;5:350.
5. Chopp M, Portnoy H, Branch C. Hydraulic model of the cerebrovascular bed: An aid to understanding the volume-pressure test. Neurosurgery 1983;13:5-11.
6. Degnan A, Levy L. Pseudotumor cerebri: Brief review of clinical syndrome and imaging findings. AJNR Am J Neuroradiol 2011;32:1986.
7. Degnan A, Levy L. When Is 'Idiopathic Intracranial Hypertension' No Longer Idiopathic?. AJNR Am J Neuroradiol 2011;32:1986.
8. Derosa L, Albiges L, Massard C, Loriot Y, Fizazi K, Escudier B. Safety of available treatment options for renal cell carcinoma. Expert Opin Drug Saf 2016;15:1097-106.
9. Farb RI, Vanek I, Scott JN, Mikulis DJ, Willinsky RA, Tomlinson G, *et al*. Idiopathic intracranial hypertension: The prevalence and morphology of sinovenous stenosis. Neurology 2003;60:1418.
10. Ferrel E, Roehrig A, Kaya E, Carlson J, Ling B, Wagner A, *et al*. Retrospective study of metastatic melanoma and renal cell carcinoma to the brain with multivariate analysis of prognostic pre-treatment clinical factors. Int J Mol Sci 2016;17:400.
11. Friedman D, Liu G, Digre K. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology 2013;81:1159:65.
12. Fuentes S, Metellus P, Levrier O, Adetchessi T, Dufour H, Grisoli F. Depressed skull fracture overlying the superior sagittal sinus causing benign intracranial hypertension. Description of two cases and review of the literature. Br J Neurosurg 2005;19:438.

13. Gironell A, Martí-Fàbregas J, Bello J, Avila A. Non-hodgkin's lymphoma as a new cause of non-thrombotic superior sagittal sinus occlusion. J Neurol Neurosurg Psychiatry 1997;63:121.
14. Holgate R, Hsu C, Scott T. Occlusion of the transverse sinus by meningioma simulating pseudotumor cerebri. Neuro-Ophthalmology 1987;7:113-7.
15. Ippen FM, Mahadevan A, Wong ET, Uhlmann EJ, Sengupta S, Kasper EM. Stereotactic radiosurgery for renal cancer brain metastasis: Prognostic factors and the role of whole-brain radiation and surgical resection. J Oncol 2015;2015:636918.
16. Johnston I. The historical development of the pseudotumor concept. Neurosurg Focus 2001;11:E2.
17. Johnston I, Kollar C, Dunkley S, Assaad N, Parker G. Cranial venous outflow obstruction in the pseudotumour syndrome: Incidence, nature and relevance. J Clin Neurosci 2002;9:273.
18. Kashimura H, Arai H, Ogasawara K, Ogawa A. Persistent falcine sinus associated with obstruction of the superior sagittal sinus caused by meningioma--case report. Neurol Med Chir (Tokyo) 2007;47:83.
19. Kim A, Trobe J. Syndrome simulating pseudotumor cerebri caused by partial transverse venous sinus obstruction in metastatic prostate cancer. Am J Ophthalmol 2000;129:254.
20. Mathews M, Sergott R, Savino P. Pseudotumor cerebri. Curr Opin Ophthalmol 2003;14:364-70.
21. Mones, R. Increased intracranial pressure due to metastatic disease of venous sinuses. A report of six cases. Neurology 1995;15:1000.
22. Nonne M. Ueber Falle vom Symptomkomplex "Tumor Cerebri" mit Ausgang in Heilung (Pseudotumor Cerebri). Dtsch Z Nervenheil 1904;27:169-216.
23. Nunez Bragayrac L, Hoffmeyer J, Abbotoy D, Attwood K, Kauffman E, Spiess P, *et al*. Minimally invasive cytoreductive nephrectomy: A multi-institutional experience. World J Urol 2016;34:1651-6.
24. Patchell RA, Tibbs PA, Walsh JW, Dempsey RJ, Maruyama Y, Kryscio RJ, *et al*. A randomized trial of surgery in the treatment of single metastases to the brain. N Engl J Med 1990;322:494.
25. Pelton RW, Lee AG, Orengo-Nania SD, Patrinely JR. Bilateral optic disk edema caused by sarcoidosis mimicking pseudotumor cerebri. Am J Ophthalmol 1999;127:229.
26. Peng K, Fuh J, Wang S. High-pressure headaches: Idiopathic intracranial hypertension and its mimics. Nat Rev Neurol 2012;8:700.
27. Plant GT, Donald JJ, Jackowski A, Vinnicombe SJ, Kendall BE. Partial, non-thrombotic, superior sagittal sinus occlusion due to occipital skull tumours. J Neurol Neurosurg Psychiatry 1991;54:520.
28. Platas-Moreno I, Antón-Benito A, Pérez-Cid-Rebolleda MT, Rosado Sierra MB. Papilledema secondary to a superior sagittal sinus thrombosis. Mantle cell lymphoma paraneoplastic syndrome. Arch Soc Esp Oftalmol 2016;91:44-7.
29. Powers JM, Schnur JA, Baldree ME. Pseudotumor cerebri due to partial obstruction of the sigmoid sinus by a cholesteatoma. Arch Neurol 1986;43:519.
30. Rendon M. New surgical horizons: The role of cytoreductive nephrectomy for metastatic kidney cancer. Can Urol Assoc J 2007;1(2 suppl):S62.
31. Riggeal BD, Bruce BB, Saindane AM, Ridha MA, Kelly LP, Newman NJ, *et al*. Clinical course of idiopathic intracranial hypertension with transverse sinus stenosis. Neurology 2013;80:289.
32. Shah A, Ivan M, Komotar R. Pseudotumor-like syndrome and cerebrospinal fluid leak in meningiomas involving the posterior third of the superior sagittal sinus: Report of 4 cases. J Neurosurg 2016;125:62-6.
33. Ursino M, Lodi C. A simple mathematical model of the interaction between intracranial pressure and cerebral hemodynamics. J Appl Physiol 1997;82:1256-69.
34. van den Brink WA, Pieterman H, Avezaat CJ. Sagittal sinus occlusion, caused by an overlying depressed cranial fracture, presenting with late signs and symptoms of intracranial hypertension: Case report. Neurosurgery 1996;38:1044.
35. Wilson M. Monro-kellie 2.0: The dynamic vascular and venous pathophysiological components of intracranial pressure. J Cereb Blood Flow Metab 2016;36:1338:50.
36. Wilson M, Browne JD, Martin T, Geer C. Case report: Atypical presentation of jugular foramen mass. Am J Otolaryngol 2012;33:370.
37. Yri H, Wegener M, Jensen R. Syphilis mimicking idiopathic intracranial hypertension. BMJ Case Rep 2011;2011.