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Case Report

Acute Cerebral Venous Thrombosis with Papilledema and Complete Vision Loss

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Abstract

Introduction: The evaluation of optic disc edema can pose a diagnostic challenge. It is important to perform thorough clinical investigation to exclude vascular, infectious, inflammatory, toxic, metabolic, and compressive causes.

Case Report: Clinical records of one patient from University of Pittsburgh Medical Center were retrospectively reviewed and discussed in this case report. A 33-year-old male with history of stage IA germ cell testicular cancer presented to the emergency department with a three-day history of progressive, painless vision loss to No Light Perception (NLP) bilaterally. He was diagnosed with cerebral venous sinus thrombosis and promptly treated with anticoagulation, acetazolamide, high dose corticosteroids, venous thrombectomy and ventriculoperitoneal shunt. He achieved significant improvement of vision after treatment.

Discussion: The broad range of clinical characteristics can make the diagnosis of CVT challenging. Symptoms include headache, focal neurologic deficits, seizures, paresis, impaired consciousness, or visual disturbances. Increased intracranial pressure (ICP) and resulting papilledema is present in over 80% of patients with CVT [1]. Papilledema commonly causes transient visual impairment, but if left untreated, can lead to permanent vision loss due to compressive optic atrophy.

Conclusion: The etiology of cerebral venous thrombosis can be variable and multifactorial. Appropriate clinical suspicion for CVT assures early diagnosis and may prevent severe morbidity and mortality. In the setting of vision loss secondary to increased Intracranial Pressure (ICP), prompt treatment may help prevent irreversible optic neuropathy and irreparable vision loss. High dose corticosteroids may be considered as an adjuvant therapy in the setting of acute optic nerve damage.

Keywords: Cerebral Venous Thrombosis; Optic Disc Edema; Papilledema; Venous Sinus Thrombosis

Case Report

Introduction

The evaluation of optic disc edema can pose a diagnostic challenge to the clinician. In the setting of known primary neoplasm, the differential diagnosis for optic neuropathy should also include carcinomatous meningitis, orbital or intracranial metastasis, direct infiltration of optic nerves, paraneoplastic syndrome, and increased ICP.

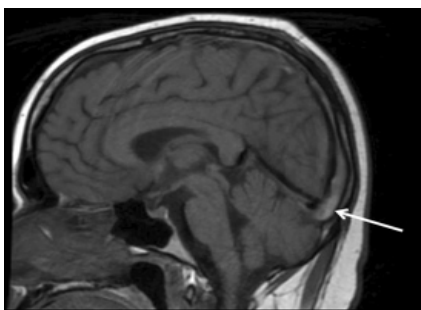
Clinical records of one patient from University of Pittsburgh Medical Center were retrospectively reviewed and discussed in this case report. The patient is a 33-year-old male with history of stage IA germ cell testicular cancer status post orchiectomy who presented to the emergency department three weeks after his first BEP (bleomycin, etoposide, cisplatin) chemotherapy cycle with reports of complete vision loss. He reports a progressive and painless vision loss in both eyes over three days. He denies prior ocular history. Review of symptoms was otherwise negative at

time of presentation. He denied headache or other focal neurologic deficits. At his initial presentation, ophthalmologic exam revealed no light perception vision in both eyes. Pupils were dilated and non-reactive to light nor to mydriatic drops. Dilated ophthalmoscopy revealed Frisén Grade III disc edema with obscuration of vessels, hemorrhage, and vascular tortuosity in both eyes. There was nasal edema with scattered pre-retinal hemorrhages in the macula in both eyes. Baseline testing was performed prior to discharge. Optical Coherence Tomography (OCT) showed supranormal retinal nerve fiber layer thickness. Manual Goldmann visual fields exhibited small paracentral islands in both eyes.

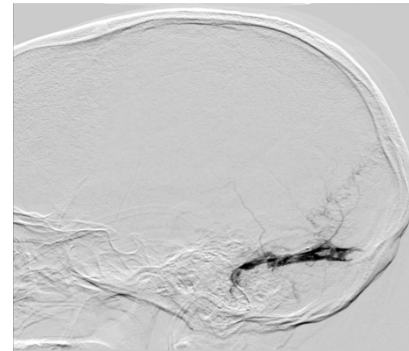
Magnetic Resonance Imaging (MRI) brain with and without contrast revealed cortical venous thrombosis associated with a small subarachnoid hemorrhage. There was evidence of superior sagittal sinus, right transverse sinus, and right sigmoid sinus thrombosis extending into the right jugular bulb and cervical internal jugular vein.

There was also prominent CSF within the optic nerve sheath complexes suggestive of elevated intracranial pressure. Extensive infectious, inflammatory, ischemic, toxic, and metabolic testing excluded alternate causes of severe optic neuropathy and disc edema and was unrevealing. Hypercoagulable work-up was performed and revealed elevated Factor VIII: C (1.78). Large volume lumbar puncture was performed and revealed a significantly elevated CSF opening pressure of 58 with bland fluid.

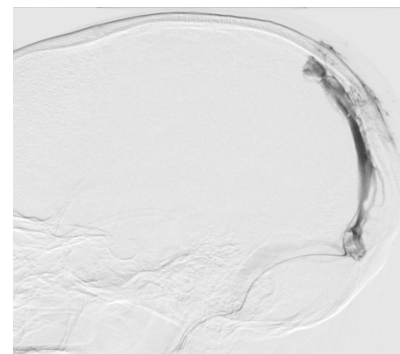
He was started on heparin with bridge to warfarin. He was also started on oral acetazolamide 500 mg three times per day. After initial infectious work-up was performed and negative, the patient was treated with high dose steroids: Intravenous (IV) methylprednisolone 1gram/day for 4 days. Digital Subtraction Angiography (DSA) and formal angiogram was performed and confirmed elevated intracranial pressure with thrombus. Urgent Venous thrombectomy was performed with significantly improved venous outflow. MRI and angiography imaging are shown in (Figure 1).



(a)



(b)



(c)

Figure 1: (a) Sagittal MRI showing thrombosis of the proximal superior sagittal sinus (arrow). Angiography showing venous flow through the superior sagittal sinus thrombosis before (b) and after (c) thrombectomy.

Given severity of vision loss due to ICP elevation, a ventricular peritoneal shunt was placed. Optic Nerve Sheath Fenestration (ONSF) was deferred given the initial suspicion of chemotherapy related toxic optic neuropathy as well as added risk of retrobulbar hemorrhage in the setting of anticoagulation. The patient's visual acuity improved to hand motion in each eye by discharge five days after admission.

He was closely followed by the neuro-ophthalmology service. Three months after initial emergent evaluation, his vision and disc edema significantly improved. Best corrected visual acuity was 20/200 in his right eye and 20/60+1 in his left eye. His pupils remained dilated and minimally reactive without APD. Dilated ophthalmoscopic exam revealed pale, gliotic optic nerves with blurred nasal margins. Optical Coherence Tomography (OCT) showed retinal nerve fiber layer atrophy. Manual Goldmann visual field testing showed significant improvement of central visual fields in each eye, with persistent, concentric, peripheral constriction in both eyes. Fundus photography at presentation and three-month follow-up is shown in (Figure 2).

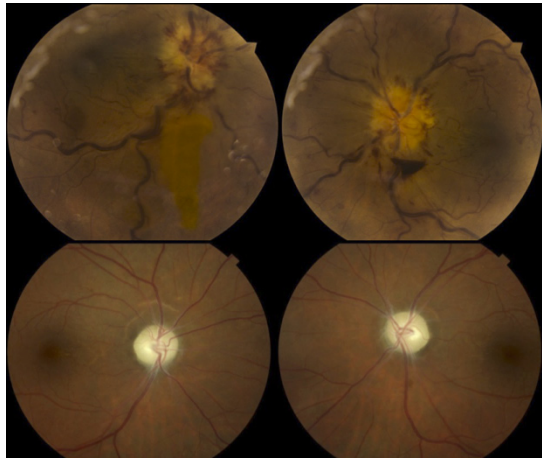


Figure 2: (a) Color fundus photograph on presentation revealed disc edema with obscuration of vessels, hemorrhage, and vascular tortuosity in both eyes. (b) Color fundus photograph three months after initial presentation revealing pale and gliotic optic nerves.

Discussion

We present a patient with initial presentation of bilateral no light perception (NLP) vision in the setting of germ cell testicular cancer and recent chemotherapy cycle. On exam he was found to have severe disc edema in both eyes. Imaging and lumbar puncture confirmed the diagnosis of severe papilledema secondary to cerebral venous thrombosis. An acute increase in intracranial pressure Cerebral Venous Thrombosis (CVT) accounts for approximately 0.5% to 1% of all strokes [1]. The estimated annual incidence of CVT is 2 cases per 100,000 people worldwide [2]. The etiology of cerebral venous thrombosis can be variable and multifactorial. The pathophysiology relates to stasis of blood, endothelial injury, and/or hypercoagulability. Risk factors may be acquired (surgery, trauma, exogenous hormones) or congenital (inherited thrombophilia). The most common location of thrombosis is the Superior Sagittal Sinus (SSS) which represents up to 60% of cases of CVT [1]. The broad range of clinical characteristics can make achieving the diagnosis of CVT challenging. Symptoms include headache, focal neurologic deficits, seizures, paresis, impaired consciousness, or visual disturbances. Headache is the most common presenting symptom with CVT. Although headache is present in up to 90% of cases [2], our patient denied headaches. Historically, the diagnosis of CVT had been made on autopsy with estimated mortality rates approaching 8% [1]. New imaging modalities may achieve an earlier diagnosis and improve the overall prognosis. Standard CT or MRI may be used to rule out acute stroke or mass lesions, but negative imaging does not rule out CVT. A venographic study (either CTV, MRV or dedicated venogram) is the diagnostic modality of choice [3].

The International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT) reports that 7.4% of cases of CVT are related to cancer [4]. The mechanism of CVT related to cancer can be variable and include direct tumor compression or invasion, hypercoagulable state associated with cancer, or attributed to chemotherapeutic and/or hormonal treatment [3]. Chemotherapy induced thromboembolic events are rare, but have been specifically described with bleomycin and cisplatin treatment for germ cell cancers [5]. These chemotherapeutic agents have been associated with acute and long-term vascular toxicity and, in very rare cases, cerebral venous thrombosis [5]. Our patient's hypercoagulability was multifactorial, secondary to germinal cell carcinoma, cisplatin/bleomycin treatment, and elevated factor VIII levels. Current guidelines recommend anticoagulation treatment with warfarin for 3-6 months with known cause of thrombophilia (or 6-12 months with unknown cause) [3].

Increased intracranial pressure and resulting papilledema is present in over 80% of patients with CVT [4]. Papilledema commonly causes transient visual impairment in the acute stage, but if left untreated, can lead to permanent loss due to optic atrophy. Vision loss initially presents as enlarged blind spot or nasal visual field loss. Central visual acuity may be preserved in indolent cases, without severe, rapid, or long standing intracranial pressure elevation. Management is dependent on the severity of the clinical sequela. Initial treatment consists of anticoagulation and acetazolamide. If there is severe or progressive vision loss, other therapies including serial lumbar punctures, shunts, endovascular intervention, and optic nerve sheath fenestration should be considered [3,4]. Our patient was treated with urgent venous thrombectomy, ventriculoperitoneal shunt, anticoagulation, Pulsed IV methylprednisolone IVMP, and oral acetazolamide.

The adjuvant treatment of acute optic neuropathy with corticosteroids continues to be debated amongst neuro-ophthalmologist. There are currently no large randomized controlled clinical trials in the literature, but some anecdotal evidence suggests improvement in visual function.

Conclusion

The etiology of cerebral venous thrombosis can be variable and multifactorial. Early clinical suspicion for CVT in the appropriate patient can help assure early diagnosis and prevent severe morbidity and mortality. In the setting of vision loss secondary to increased intracranial pressure, prompt treatment may help prevent irreversible optic neuropathy.

Declaration of Interest

None.

References

1. Bousser MG, Chiras J, Bories J, Castaigne P (1985) cerebral venous thrombosis: Review of 38 cases. *Stroke* 16:199-213.
2. Agrawal K, Burger K, Rothrock JF (2006) Cerebral Sinus Thrombosis. *Headache* 56: 1380-1389.
3. Saposnik G, Barinagarrementeria F, Brown RD Jr, Bushnell CD, Cucchiara B, et al. (2011) Diagnosis and management of cerebral venous thrombosis: A statement for healthcare professionals from the American Heart Association/American Stroke Association. *Stroke*: 42: 1158-1192.
4. Ferro JM, Canhao P, Stam J, Bousser MG, Barinagarrementeria F, et al. (2004) Prognosis of cerebral vein and dural sinus thrombosis: Results of the International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT). *Stroke*: 35: 664-670.
5. Weijl NI, Rutten MF, Zwinderman AH, Keizer HJ, Nooy MA, et al. (2000) Thromboembolic events during chemotherapy for germ cell cancer: a cohort study and review of the literature. *J Clin Oncol* 18: 2169-2178.