Hemicrania Continua and Hypoglossal Nerve Palsy: Signs of Intracranial Hypertension

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Abstract

A 50 year old male with obstructive sleep apnea developed de novo, daily, intense, continuous, left side pounding headache with superimposed stabbing pains, fluctuating in severity. Headache was associated with ptosis of the left eye, left hypoglossal palsy and eventual tongue hemiatrophy, in the absence of papilledema. His Brain and neck imaging procedures (MRI, MRA, MRV) and a left temporal artery biopsy were normal, while his cerebrospinal fluid (CSF) opening pressure was found elevated.

Introduction

Idiopathic intracranial hypertension (IIH) is a common syndrome characterized by chronic headache and in its classic form accompanied of bilateral papilledema [1]. Very rare examples of IIH with unilateral papilledema and pseudopapilledema, and of patients with headache, papilledema, and normal CSF pressure have been reported [2,3]. Conversely, IIH may occur without papilledema [4]. Visual symptoms in IIH can be striking and potentially of ominous significance, since untreated cases may progress into irreversible optic nerve damage and blindness. Patients with IIH may present with sixth nerve or seventh nerve palsy and exceptionally, with oculomotor palsy [1,5]. To my knowledge, IIH is not associated with hypoglossal palsy. Mateen, et al, described three patients with unilateral headache and idiopathic unilateral hypoglossal palsy in 2007 [6]. None of their patients were formally diagnosed with hemicrania continua (HC) or with new daily persistent headache (NDPH). In 2 of their 3 patients on whom lumbar puncture was done, elevation of the CSF pressure was not documented (i.e. “CSF studies were normal”).

Case Report

A 50 year old dentist reported the rapid development of a left side persistent and annoying daily headache with superimposed intense left frontal stabbing pain. He only had rare, mild, and very infrequent headaches in the past. He remembered the exact day he developed the headache in question. Within three days he had trouble speaking and his tongue deviated to the left when protruded. His left eyelid became droopy. He had no nausea or vomiting. He had no double vision or noticed any other visual disturbance. There were no apparent initial triggers including trauma, surgeries or major situational stress. He had no previous history of migraine and his family history was negative for neurological disorders including migraine. He had no history of drug dependence or of psychiatric illness. He complained of chronic back pain and of mild left eye photophobia. He had obstructive sleep apnea (OSA) appropriately treated with C-PAP, hypertension, irritable bowel syndrome, kidney stone, gastroesophageal reflux disease and lumbago. He was taking gabapentin 300 mg t.i.d., valsartan 40 mg q.d., hydrochlorothiazide 25 mg q.d., omeprazole 20 mg q.d. and oxycodone as needed for pain.

His general physical examination was unremarkable. His BMI was 34.2. He was afebrile and his blood pressure was normal on multiple occasions. His mental status examination was normal. He had mild left eye ptosis without pupillary changes or extraocular muscle weakness. He exhibited residual left tongue hemiatrophy with no weakness. Deep tendon reflexes were hypoactive. Imaging studies including brain and cervical MRI, MRA of the head and MRV were normal. MRA of the neck vessels showed no dissection or stenosis of the extracranial circulation. CSF studies including cultures for bacteria and fungi were all normal but his opening pressure was 26 cm of water on the lateral decubitus position with the patient completely relaxed. There were no oligoclonal bands in the CSF. A left temporal artery biopsy and blood sedimentation rate were normal. Lyme disease serology was negative. Epstein Barr virus (EBV) IgG antibody and nuclear antigen titer were positive indicative of a past infection. There was no serological evidence of recent infection with EBV. Serum testosterone levels were normal. Angiotensin converting enzyme (ACE) blood levels were mildly elevated but an axillary lymph node biopsy and a chest CT were negative for sarcoidosis. Bilateral blink reflex studies and nerve conduction velocities (NCV) showed no evidence of trigeminal or peripheral neuropathy. A trial of indomethacin, topiramate, and acetazolamide and a five day course of intravenous
methylprednisolone followed by a prednisone oral taper were ineffective. Gabapentin up to 1.800 mg a day ameliorated the intensity of the headache combined with rescue oxycodone. He has developed no new symptoms in over two years of regular follow up.

Discussion

Hemicrania continua (HC) is classified as a primary headache disorder, typically manifested as daily unilateral continuous headache with superimposed, normally severe, stabbing or throbbing pain, associated with cranial autonomic signs such as ipsilateral ptosis, increased lacrimation and rhinorrhea [7]. Along with chronic tension-type headache, chronic migraine and NDPH, HC is part of the diagnostic category of chronic daily headache (CDH) [7]. Secondary presentations of HC are infrequent. Isolated cases of HC secondary to lung malignant tumor, mesenchymal tumor of the sphenoid, left vertebral artery occlusion, lateral medullary stroke, cavernous internal carotid artery aneurysm, internal carotid artery dissection, prolactinoma and venous malformation of the masseter muscle have been reported in the literature [7]. It is of interest that this patient had features of NDPH except that his headache was obviously lateralized from the onset and associated to cranial nerve findings. The list of disorders causing secondary forms of NDPH is extensive and includes examples of cerebral venous thrombosis and of low or elevated CSF pressure (IH) [7]. On the other hand, cerebral venous thrombosis and IH are encountered in a significant number of patients with CDH [8]. Most patients with IH report headaches with features conforming with either tension-type headache or migraine, but not with the typical features of HC with cranial dysautonomia [1,7]. IH without papilledema may represent an under diagnosed factor in the progression or perpetuation of migraine and chronic tension-type headache [9]. This patient is exceptional, not only because his comorbid hypoglossal palsy, but because his HC may have been maintained, if not precipitated, by probably a chronically elevated CSF pressure.

IH often associates with obesity and pregnancy [1]. Association of IH and headache with obstructive sleep apnea was recently confirmed by Fraser, et al [10]. Although this patient had centripetal obesity, an isolated abnormal BMI is not responsible for CSF hypertension or for IH with or without papilledema, according to Bono, et al, comprehensive investigation [11]. Therefore, it is logical to assume that he had bona fide primary IH, albeit the pressure reading was in the low, but still abnormal range. His elevated CSF pressure was probably compounded by OSA. Why IH in the presence of a normal cerebral brain venography (MRV) is unclear.

The three patients reported by Mateen, et al, had hemicrania, although it is not clear from the report if their headaches were of abrupt onset and termination, neither their clinical history conformed the expected presentation of NDPH [6]. In my view, one of Mateen, et al, patient’s had symptoms compatible with episodic hemicrania, the second with chronic recurrent hemicrania and the third with HC. Other than hypoglossal paresis they had no autonomic cranial signs however. All the imaging procedures and tests conducted to discard autoimmune and rheumatic disorders were normal. Two had normal CSF studies but no mention is made in the report of their opening CSF pressure [6]. The differential diagnosis of unilateral hypoglossal paresis associated with headache is limited to cases of carotid dissection or carotid fibromuscular dysplasia with fusiform-like aneurysm formation [6]. Patients with Raeder syndrome exhibit facial pain and homolateral Horner syndrome and patients with “neck tongue syndrome” have unilateral neck and tongue pain but in neither of these two examples the patients have hypoglossal weakness or tongue hemiatrophy [12,13]. Of parallel relevance to the topic herein discussed, an exceptional patient with benign Raeder syndrome evolving into indomethacin-sensitive HC was reported by Koutsis, et al, while patients with OSA may experience an exacerbation of their apnea after the administration of indomethacin, even than the latter drug may substantially reduced intracranial hypertension [12,14,15]. Finally, although patients with Collet-Sicard (jugular foramen) syndrome may complain of unilateral head pain, they will exhibit multiple lower unilateral cranial nerve compromise on their neurological examination [16].

The pathogenesis of headache associated with idiopathic hypoglossal paresis is yet to be elucidated. It has been speculated that the cause of this unusual association occur on basis of a primary viral (herpetic, EBV?) infection, either of spontaneous origin or precipitated by trauma, as it is similarly theorized for cases of idiopathic Bell’s palsy. It is uncertain on this patient if he had subclinical acute mononucleosis at the offset of his headache, since he was seen by the author nine months after the abrupt initiation of symptoms. An alternative explanation advanced for these unusual cases is ischemia, developing in the vascular territory supplied by a small caliber branch of
the upper internal carotid artery that includes the hypoglossal nerve [6]. It is speculated that pain in these patients is mediated by posterior fossa meningeal sensory fibers traveling the twelve nerve making synapse in the dorsal horns of the upper cervical segments, and in the spinal descending nucleus of the trigeminal nerve. Low testosterone levels are commonly found in obese subjects with OSA and in some male (and female) patients with cluster headache [10,17]. The latter association is explained on basis of a presumptive disequilibrium between gonadotropin and corticotropin levels and the hypothalamic entrainment of melatonin release by the pineal gland. Because this patient had a normal testosterone blood level, an underlying hypothalamic dysfunction cannot be adduced as the cause for his persistent headache.

References

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