Paroxysmal hemicrania as the clinical presentation of giant cell arteritis

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Abstract

Head pain is the most common complaint in patients with giant cell arteritis but the headache has no distinct diagnostic features. There have been no published reports of giant cell arteritis presenting as a trigeminal autonomic cephalalgia. We describe a patient who developed a new onset headache in her fifties, which fit the diagnostic criteria for paroxysmal hemicrania and was completely responsive to corticosteroids. Removal of the steroid therapy brought a reemergence of her headaches. Giant cell arteritis should be considered in the evaluation of secondary causes of paroxysmal hemicrania; in addition giant cell arteritis needs to be ruled out in patients who are over the age of 50 years with a new onset trigeminal autonomic cephalalgia.

Introduction

Paroxysmal hemicrania (PH) is a rare primary headache disorder that typically begins in young adulthood with a mean age of 34 years and preferentially affects women.[1] It is one of the trigeminal autonomic cephalalgias (TACs) thus along with short lasting bouts of severe head pain, patients experience a multitude of cranial autonomic symptoms including lacrimation, conjunctival injection, nasal congestion/rhinorhea and the development of miosis and/or eyelid ptosis (Horner syndrome).[1] The other recognized TACs include cluster headache and SUNCT syndrome. The head pain of PH typically begins without warning, lasts 2 to 30 minutes in duration and can occur anywhere from 1 to 40 times per day. The pain of PH is severe in intensity, one sided and generally located in ocular, periorbital, temporal or upper facial regions.[1] Paroxysmal hemicrania is considered one of the indomethacin responsive headache disorders as the headaches are completed alleviated on indomethacin and return when indomethacin is tapered off and rarely if ever respond to any another other medication including other NSAIDs. Indomethacin responsiveness is actually required in the International Classification of Headache Disorders (second edition) to make the diagnosis of PH (Table 1).[1] Despite its rarity, a number of secondary causes of PH have been documented in the literature including intracranial tumors, infections, intracranial hypertension and aneurysms.[2] We now present the first ever case of PH as the clinical presentation of giant cell arteritis (GCA).

Case Report

A 56-year-old woman presented to the Emergency Department with a new type of headache beginning 6 weeks prior. She described a baseline every moment dull, persistent ache located to the left of vertex, and also reported daily short-lasting spikes of severe pain (10 out of 10 on VAS pain scale) occurring in the left temple and left parietal region which would occur intermittently throughout the day, lasting 15 min in duration. She would average between 5-10 attacks of pain exacerbation per day. These pain exacerbation periods were associated with cranial autonomic symptoms including left eyelid ptosis and left eye lacrimation. She also would experience migrainous associated symptoms including photophobia, blurred vision and nausea. In addition she also complained of short stabs of pain lasting 1-2 s in duration which would occur daily and also multiple times in a day and would mostly occur on the left side of her head but not in a specific location. She had a prior headache history for many years of intermittent more generalized headaches of mild intensity and without migrainous associated symptoms, which would alleviate within several hours of onset without medication. In addition to headache the patient on presentation felt overall very ill and lethargic. She did not however have jaw claudication by history.

On examination the patient had pain to palpation over the left greater occipital nerve, trochlear notch and supraorbital notch. She had a bounding left superficial temporal artery pulse but had an absent temporal artery pulse on the right side. She had a left supraclavicular and left carotid bruit on neurovascular examination. The remainder of her general and neurologic examination was non-focal. Laboratory testing revealed a normal sedimentation rate (14 mm/h: normal range: 0-15 mm/h) and low sensitivity C-reactive protein level (3 mg/L: normal range 0-5 mg/L). Brain magnetic resonance angiography (MRA) suggested diffuse intracranial vessel stenosis involving the basilar artery and bilateral intracranial carotid arteries thought more compatible with diffuse atherosclerosis but central nervous system vasculitis was in the differential. She had multiple stroke risk factors including hypertension, hyperlipidemia and she was a chronic smoker. Computed tomography angiography (CTA) was then completed to better define the arterial stenoses but this study did not denote any significant intracranial vessel abnormalities and there was no flow limiting arterial stenoses and nothing suggestive of vasculitis. However, CTA of the neck vessels demonstrated a very high-grade stenosis of the proximal left internal carotid artery, but no evidence of dissection.

The patient's headache symptomatology was consistent with a diagnosis of PH with associated idiopathic stabbing headaches, but because of her age and her general sense of feeling ill a secondary cause of PH was considered. Rheumatology evaluated the patient and felt GCA was high in the differential because headache was the main presenting symptom of her illness, in addition she complained of stabbing headaches which are part of the presentation of GCA and she had an absent temporal artery pulse on exam.[3] The decision was made to treat the headache with corticosteroids rather than indomethacin because of the possible morbidity that could result with holding corticosteroids in the face of GCA. On oral prednisone 40 mg per day the patient had a dramatic improvement in her pain becoming headache free within 24 h. Rheumatology diagnosed her with giant cell arteritis based on her robust response to steroids, however a left temporal artery biopsy was completed and this did not demonstrate arteritis. At the time of the biopsy she had been on prednisone for 3 days. The prednisone was tapered because of the negative biopsy results but her headaches immediately returned to a pre-steroid state, and then again alleviated once a higher dose of prednisone was achieved. Rheumatology felt her underlying condition

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was still GCA even with a negative biopsy. Over several months time, she intermittently tried to taper down her steroid dose but the headaches would immediately return. Upon increasing the steroid dose, her headaches and autonomic symptoms would again resolve. She has been on corticosteroids now for 8 months and remains pain free.

Discussion

PH is one of the indomethacin responsive TACs. As more cases of this unique primary headache are seen in the clinic more secondary mimics of the condition are discovered. This is important, as many of the secondary TACs clinically are indistinguishable from the primary forms and in many instances secondary indomethacin sensitive headaches respond in the same manner to therapy as the primary subtypes.[4] Any new secondary condition, which has not been noted previously for the TACs, should be documented in the literature because it broadens the diagnostic work-up for these headaches. In this specific case it is important to now note that GCA may present as a TAC specifically PH. GCA has no definitive presentation in regard to headache. Head pain is the most common complaint in GCA patients but the headache can occur anywhere on the head, not just the temples, be mild to severe in intensity and be dull to throbbing in quality; thus very amorphous in its presentation.[3] If there is associated jaw claudication and an elevated sedimentation rate or CRP level, then the diagnosis is highly suggestive of GCA; however, GCA cannot be completely ruled out even in the absence of these factors. Identification of GCA is critical because of the morbidity that can result if the disease goes untreated including vision loss and stroke. The diagnosis of GCA is confirmed by temporal artery biopsy; however this procedure can sometimes have high false negative rates because of the known skip lesions associated with this form of arteritis. There is now data to suggest that GCA may be present even with a negative biopsy.[5] In a large study from the United Kingdom specimen length of the biopsy sample was a crucial predictor of positive biopsy results. Specimen length of 0.7 cm or more had a significantly higher rate of positive results than smaller arterial samples. The subject of this case report had an approximately 2 cm arterial segment submitted to Pathology so specimen length was most likely not a factor in a possible false negative biopsy result. Another investigation using Bayesian methodology and data from studies which reported the results of bilateral temporal artery biopsies calculated that the sensitivity of a single temporal artery biopsy is 87.1%, thus there is a greater than 10% false negative result rate.[6] Finally, we are seeing a high rate of negative temporal artery biopsies at our institution in patients who clinically appear to have GCA. The departments of Rheumatology and Neurology are therefore questioning the biopsy technique being done by our vascular surgeons. Because not all patients have classic features of GCA (clinical and laboratory), it is important to be aware of the possibility of new clinical presentations. To date, there is no literature of GCA presenting as PH or any other TAC. Because PH is not common (occurring in 2 per 100,000 individuals) and typically occurs in younger adults (mean onset 34 years), an underlying secondary etiology should be considered especially in those who present with PH after the age 50 years.[7] We cannot state this patient absolutely had PH because the diagnosis depends on complete relief with indomethacin.[1] We can suggest however based on presentation this was a secondary form of a PH-like headache. We also cannot state for certain the case patient had GCA, but this was highly suggested based on her robust response to steroids and this was the diagnosis made by Rheumatology. Corticosteroids in single case reports have been shown to be somewhat effective in PH, but the dramatic response in the present case suggested steroid responsive GCA and thus steroid responsive GCA induced PH.[8] How GCA could present as a TAC can only be hypothesized. Positron emission tomography (PET) studies have demonstrated that PH is associated with significant activation of the contralateral posterior hypothalamus during attacks; thus the hypothalamus is a possible generator for this condition.[9] There is scant literature to support injury to the hypothalamus during the active phase of GCA, however this is felt to be a rare occurrence and thus may explain why we are reporting the first ever case of GCA presenting as PH and why it is not the typical headache presentation for GCA.[10] Of note the patient also had a very high-grade internal carotid artery stenosis on the same side as her headaches but this did not appear to play any role in headache pathogenesis, as after carotid endarterectomy with alleviation of the vessel stenosis there was no change in her headache pattern still requiring prednisone to remain pain free. Based on this case report GCA should be considered in the evaluation of secondary causes of paroxysmal hemicrania; in addition GCA needs to be ruled out in patients who are over the age of 50 years with a new onset TAC.

Table 1. Diagnostic criteria for paroxysmal hemicrania.

A.	At least 20 attacks fulfilling criteria B through D;
B.	Attacks of severe, unilateral, orbital, supraorbital, or temporal pain lasting 2 to 20 minutes;
C.	Headache is accompanied by at least one of the following:
	i) Ipsilateral conjunctival injection and/or lacrimation;
	ii) Ipsilateral nasal congestion and/or rhinorrhea;
	iii) Ipsilateral eyelid edema;
	iv) Ipsilateral forehead and facial sweating;
	v) Ipsilateral miosis and/or ptosis.
D.	Attacks have a frequency>5 per day or more than half of the time, although periods with lower frequency may occur;
E.	Attacks are prevented completely by therapeutic doses of indomethacin;
F.	Not attributed to another disorder.

References

1. Headache Classification Committee of The International Headache Society. The International Classification of Headache Disorders (second edition). Cephalalgia 2004;24 Suppl 1:1-160.
2. Favier I, van Vliet JA, Roon KI, et al. Trigeminal autonomic cephalgias due to structural lesions: a review of 31 cases. Arch Neurol 2007;64:25-31.
3. Rozen TD. Brief Sharp Stabs of Head Pain and Giant Cell Arteritis. Headache. 2010;50:1516-9.
4. Ertsey C, Jelencsik I. Cough headache associated with Chiari type-I malformation: responsiveness to indomethacin. Cephalalgia 2000;20:518-20.
5. Ypsilantis E, Courtney ED, Chopra N, et al. Importance of specimen length during temporal artery biopsy. Br J Surg 2011;98: 1556-60.
6. Niederkohr RD, Levin LA. A Bayesian analysis of the true sensitivity of a temporal artery biopsy. Invest Ophthalmol Vis Sci 2007;48:675-80.
7. Shapiro RE. The Man With the Red Eye. In: Purdy RA, Rapoport AM, Sheftell FD, Tepper SJ, eds. Advanced Therapy of Headache, 2nd ed. Hamilton: BC Decker Inc; 2005. pp. 311-315.
8. Boes CJ, Dodick DW. Refining the clinical spectrum of chronic paroxysmal hemicrania: a review of 74 patients. Headache 2002;42:699-708.
9. Matharu MS, Cohen AS, Frackowiak RS, Goadsby PJ. Posterior hypothalamic activation in paroxysmal hemicrania. Ann Neurol 2006;59:535-45.
10. Everett GD, Amanda JM, Woolf PD. Hypothalamic-hypopituitarism due to temporal arteritis. Arc Intern Med 1979;139: 474-5.