PHOTOESSAY

Atypical Chronic Headache and Recurrent Facial Ecchymosis: A Case Report

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ABSTRACT

We present the case of a 57-year-old woman with chronic paroxysmal headache and recurrent facial ecchymosis. The headaches are chronic, unilateral, sharp, electric shock-like, of short duration, without nausea nor vomiting, ptosis, miosis, conjunctival injection nor tearing. The facial ecchymoses have been mainly located in the periorbital and epicanthal areas. General, neurological and ophthalmological examinations were unremarkable. Work-up was unremarkable, including negative skin biopsy. The headache was difficult to include in any particular category given the atypical features, but the characteristics were suggestive of a variant of trigeminal autonomic cephalgia with lack of responsiveness to indomethacin. We discuss the possible pathogenetic mechanism of the occurrence of facial ecchymosis in primary headache disorders.

KEYWORDS facial ecchymosis, migraine, trigeminal autonomic cephalgia

CASE REPORT

A 57-year-old woman presented with severe headaches associated with recurrent episodes of periorbital and facial discoloration ipsilateral to the headaches. Symptoms started 9 years prior to presentation, following an acute transient episode of left leg tingling while walking, which progressed to involve the left arm and face. She was hospitalized for possible stroke. Work-up was non-contributory. Although the numbness resolved, she stated that she continued to drag her leg while walking for few months. During her hospital stay, and following the resolution of the hemibody numbness, she developed a severe left sided headache. The pain started over the occipito-nuchal area and gradually radiated to the left temple. The headache was described as severe excruciating stabbing, jabbing, electric shock-like. The pain lasted seconds and occurred up to 10 times a day, and was not associated with ptosis, miosis, eye tearing or redness, nor nasal congestion. There was no associated photophobia, phonophobia, nausea nor vomiting. The pain did not seem to vary during the day or with activity, sneezing, or coughing, and had no temporal or nocturnal pattern. No particular triggers were identified. Around 4 years later, she started developing intermittent, non-tender facial discoloration, in conjunction with and

ipsilateral to the headache. The discoloration was mainly periorbital, starting as a small bruise under the eye, and extending to the epicanthal area and the upper cheek, with occasional peeling of the skin at the site of the discoloration (Figure 1). Although the headaches were chronic, the facial ecchymosis occurred initially weekly, and the gradually decreased in frequency to monthly or bimonthly.

Her past medical history is notable for hypothyroidism. She smoked 1 1/2 packs of cigarettes a day for 30 years, and did not drink alcoholic beverages. There was no family history of migraines or other headache disorders. General, neurological, and ophthalmological examinations were unremarkable. Laboratory evaluation including routine blood count, basic metabolic panel, and erythrocyte sedimentation rate (ESR) was normal. Magnetic resonance (MR) imaging of the brain, MR angiography of the head and neck, and MR venography were unremarkable. Skin biopsy of the ecchymotic lesion showed no underlying vasculitis or amyloid changes.

She received indomethacin at doses up to 150 mg daily for 4 weeks, without clinical improvement. She did not respond to triptans, ergot products, lidocaine intranasal nor intravenous. She tried numerous preventive therapies, including valproic acid, gabapentin, lamotrigine, tricyclic antidepressants,

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FIGURE 1 Facial ecchymosis associated with atypical unilateral headache.

beta blockers, and calcium channel blockers without any relief. She is currently on topiramate and hydrocodone chronically, and reports decrease in intensity of the headache, and frequency of facial ecchymosis to bimonthly.

DISCUSSION

The headache characteristics, including chronicity, duration, and frequency are suggestive of a variant of trigeminal autonomic cephalalgias (TACs) associated with periorbital and facial ecchymosis in the absence of autonomic features, and unresponsiveness to indomethacin, with superimposed medication overuse headache.¹ TACs include cluster headaches; paroxysmal hemicrania; short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT); and short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA). TACs are primary headache disorders characterized by unilateral headache and ipsilateral cranial autonomic features, including ptosis, miosis, eye tearing or redness, or nasal congestion. Skin discolouration with eyelid oedema and periorbital ecchymosis has been described in primary headache disorders

including cluster headache and migraine.²⁻⁵ Recurrent leg and buttock ecchymosis have also been reported in a child with abdominal migraine.⁶ The pathogenesis of this phenomenon of facial discoloration is not understood; the ecchymosis may be accounted for by trigeminovascular activation and autonomic vascular dysfunction, and distension of the cutaneous vessels that occurs during the severe headache. Another factor may be superimposed coagulation changes previously documented during migraine attacks, with heparin release from mast cells and basophilic leukocytes, and extravasation of red blood cells during vessel dilatation. A skin biopsy during an attack may be helpful to further understand the mechanism of facial ecchymosis.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figure 1 of this article is available in colour online at www.informahealthcare.com/oph.

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