A Case of Prethoracic Pain Radiating Upward and Initiating Nervus Intermedius Neuralgia and Migraine Headache: Could Epicrania Fugax Pain Start in the Upper Body?

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Epicrania fugax (EF) was recently classified as a primary headache in the Appendix of the International Classification of Headache Disorders, third edition (ICHD-III). It is characterized by a paroxysmal pain rapidly radiating forward or backward along a linear or zigzag trajectory on the surface of the head. This article reports a 76-year-old woman who newly developed a paroxysmal EF-type pain distributed not only in the territories of the trigeminal and occipital nerves, but also in the territories of the cervical and thoracic nerves. This EF-type pain started in a point on the prethoracic area, radiated along the ipsilateral neck, face, auditory canal, and head surface in a linear trajectory, and finally initiated attacks of nervus intermedius neuralgia (NIN) and migraine without aura (MWA). Treatment with a low dose of carbamazepine was associated with decreased intensity of EF-type pain and fewer NIN and MWA attacks, while a higher dose of carbamazepine was associated with complete termination of EF-type pain and NIN and MWA attacks. This case report expands the clinical spectrum of EF and may also be helpful in understanding its pathophysiology. *J Oral Facial Pain Headache 2017;31:398–401. doi: 10.11607/ofph.1772*

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Epicrania fugax (EF) has recently been classified as a primary headache in the Appendix of the International Classification of Headache Disorders, third edition (ICHD-III).[1] EF is characterized by a paroxysmal pain starting in a particular area of the head and rapidly radiating forward or backward along a linear or zigzag trajectory on the surface of the head innervated by different nerves.[2,3] Recently, Cuadrado et al reported a very probable EF variant in which the paroxysmal EF-type pain started in the lower face, in the territories of the second or third branch of the trigeminal nerves, and radiated upward.[4] Up to the present, all reported EF pains were in the territories of the trigeminal and occipital nerves, and it had never been reported that spreading of EF pain could initiate neuralgia involving other peripheral nerves. This article reports a case of a paroxysmal EF-type pain that was distributed not only in the territories of the trigeminal and occipital nerves, but also in the territories of the cervical and thoracic nerves. This EF-type pain could also often initiate attacks of nervus intermedius neuralgia (NIN) and migraine without aura (MWA) while it was radiating upward along the ipsilateral neck, face, auditory canal, and head surface.

Case Report

A 76-year-old woman had newly developed episodic headache attacks for 3 months. Each attack would begin with a brief moderate-to-severe prethoracic pain (rated as 5 to 8 in severity on a 0 to 10 visual analog scale [VAS]) that started at a point above the left nipple and then moved upward following a linear trajectory along the ipsilateral neck, face, auditory canal, and temporal region, reaching the parietal region in 2 to 3 seconds. Five times within 3 months, this brief upward-radiating linear pain occurred independently and did not evolve into a different type of

pain. However, apart from these five times, once this radiating linear pain reached the left auditory canal, the patient would suffer an abrupt onset of severe stabbing pain (8 to 9 on the VAS) localized in the depth of the auditory canal and thereafter a pulsating headache (8 to 9 on the VAS) of the ipsilateral hemisphere accompanied with dizziness, nausea, and phonophobia, but no vomiting. One episode of auditory canal pain would last for 5 to 10 seconds and be relieved spontaneously, one episode of the headache attack would last for 4 to 5 hours and be relieved spontaneously, and clustered episodes continued for 4 to 5 hours and were relieved spontaneously. The pain, including the linear pain, auditory canal pain, and headache, would occur at a frequency of 2 to 3 times a day, with remission lasting for 1 to 2 days once every 15 to 20 days. The patient had tried analgesics consisting of aminopyrine, phenacetin, caffeine, phenobarbital, and indomethacin, but none of these relieved the pain in frequency or severity. The patient had no personal or familial history of similar head pain or migraine headache. No abnormality was revealed in neurologic examination, electroencephalogram (EEG), or blood tests, including erythrocyte sedimentation rate (ESR) and myocardial enzyme spectrum. An electrocardiogram (ECG) showed changes in ST segment and adjacent T wave (ST-T). Cardiac ultrasonography was normal. Brain magnetic resonance imaging (MRI) revealed lacunar infarction in the white matter around the bilateral ventricles.

Carbamazepine was administered at a dose of 0.1 g twice a day, and the linear pain severity was reduced from moderate or severe pain (5 to 8 on the VAS) to mild pain (2 to 3 on the VAS), but its frequency was unchanged. For the auditory canal pain and the hemispheric headache, the frequency was reduced from 2 to 3 times a day to 1 to 2 times a day, but the pain severity was unchanged. This indicated that the linear pain might develop independently without evolving into auditory canal pain or hemispheric headache after a low dose of carbamazepine. Then, 3 days later, the carbamazepine dose was increased to 0.2 g twice a day, and the linear pain, auditory canal pain, and headache were all relieved completely. During the 1-month follow-up, no pain recurred while the patient was continuing on carbamazepine, except for three instances of pain attacks within 2 days when carbamazepine was discontinued.

Discussion

In this patient, the linear pain features—including paroxysms, short duration (2 to 3 seconds), linear trajectory, radiation, and commencement and termination in territories of different nerves—were identical to that of EF and fulfilled most of the ICHD-III criteria for EF, except for the much longer pain trajectory (Table 1). The upward radiation from an area outside the head and the transverse radiation of this linear pain may be consistent with former descriptions of EF variants stemming from the lower face or from multiple directions, including transversely.[4–6] The pain pattern of a long-distance, line-shaped pain area including the head surface, the short duration (2 to 3 seconds), and the response to carbamazepine make this linear pain obviously different from radiating pain due to ischemic heart disease, although the pain did stem from an area corresponding to the heart, and there were ECG (ST-T) changes possibly indicating myocardial ischemia. However, it is especially difficult to explain how such a transient (2 to 3 seconds) radiating pain of ischemic heart disease could trigger a long-lasting auditory canal pain and hemispheric headache, and the normal myocardial enzyme spectrum might further support the view that the prethoracic pain and its radiation were not due to ischemic heart disease. The linear pain was mainly defined by topographic criteria, as are EF, nummular headache, and linear headache.[7–10] Nummular headache is a coin-shaped head pain and thus obviously different from the current linear pain. Linear headache pain is also distributed in a line-shaped area, but its long duration (hours to days) and nonradiation features make it significantly different from the linear pain experienced by the present patient.[7–10] Thus, the paroxysmal linear pain did not fit any other pain diagnosis and would most probably correspond to an atypical variant of EF. The recurrent paroxysms of severe and short-duration stabbing pain in the depth of the auditory canal following the EF-type pain fulfilled the ICHD-III criteria for NIN (Table 1). The hemispheric headache following the EF-type pain and NIN pain might be considered a radiating pain of NIN, since NIN pain sometimes radiates to the parieto-occipital region; however, the accompanied symptoms of dizziness, nausea, and phonophobia obviously made the headache different and in fact fulfilled the ICHD-III criteria for MWA (Table 1). The occasional independent EF-type pain attack that did not evolve into NIN or MWA before carbamazepine administration and the increased independence of EF-type pain after carbamazepine administration indicated that three pain entities existed in this patient: the EF-type pain, NIN, and MWA.

Many EF patients also have migraine or another type of headache, but there is no temporal relationship between EF and other headache disorders.[2,3] Recently, there have been reports of migraine attacks that progressed from typical EF.[11,12] In the present case, the temporal relationship between EF-type pain attacks and the onset of NIN and MWA, as well

Table 1 International Classification of Headache Disorders Third Edition (ICHD-3) criteria for Epicrania Fugax, Nervus Intermedius Neuralgia, and Migraine Without Aura

Epicrania Fugax	Nervus Intermedius Neuralgia	Migraine Without Aura
A Recurrent stabbing head pain attacks lasting 1–10 seconds, fulfilling criterion B.	A At least three attacks of unilateral pain fulfilling criteria B and C.	A At least five attacks fulfilling criteria B–D.
B The pain is felt to move across the surface of one hemicranium in a linear or zig-zag trajectory, commencing and terminating in the territories of different nerves.	B Pain is located in the auditory canal, sometimes radiating to the parieto-occipital region.	B Headache attacks lasting 4–72 hours (untreated or unsuccessfully treated).
C Not better accounted for by another ICHD-3 diagnosis.	C Pain has at least three of the following four characteristics:	C Headache has at least two of the following four characteristics:
	1 Recurring in paroxysmal attacks lasting from a few seconds to minutes	1 Unilateral location
	2 Severe intensity	2 Pulsating quality
	3 Shooting, stabbing, or sharp in quality	3 Moderate or severe pain intensity
	4 Precipitated by stimulation of a trigger area in the posterior wall of the auditory canal and/or periauricular region	4 Aggravation by or causing avoidance of routine physical activity (eg, walking or climbing stairs)
	D No clinically evident neurologic deficit.	D During headache, at least one of the following:
		1 Nausea and/or vomiting
		2 Photophobia and phonophobia
	E Not better accounted for by another ICHD-3 diagnosis.	E Not better accounted for by another ICHD-3 diagnosis.

as the nonrecurrence of NIN and MWA attacks due to the termination of the EF-type pain following the administration of carbamazepine, indicated that the NIN and MWA attacks were triggered by the EF-type pain. The initiation of EF has been postulated to involve the trigeminal and occipital nerves.[2] The present case was unique in that the EF-type pain was distributed not only in the territories of trigeminal and occipital nerves, but also in the territories of cervical and thoracic nerves; ie, this pain was initiated within the territories of thoracic nerves and terminated within occipital nerves. Another unique feature of this case was that the EF-type pain triggered NIN while it was spreading over the nervus intermedius.

The pathophysiology of EF-type pain is unknown, since the underlying mechanism of typical EF pain is unclear. The radiation characteristics, the triggering of NIN while the pain was spreading over the nervus intermedius, and the response to carbamazepine indicated a peripheral origin of this EF-type pain in the present case. This is supported by a recent case report of symptomatic EF caused by a meningioma compressing the trigeminal nerve.[13] But the EF-type pain trajectory in the present case was not topographically in line with the anatomical features of thoracic, cervical, trigeminal, and occipital nerves; thus, pain-processing regions in the brainstem and spinal cord might be involved in the formation of the long-distance trajectory of EF-type pain, as proposed for typical EF.[14,15] Previous studies have demonstrated the convergence of orofacial and cervical afferents onto nociceptive neurons in the trigeminal brainstem subnucleus caudalis and upper cervical spinal dorsal horn (the trigeminocervical complex [TCC]).[16,17] This convergence of nociceptive afferents from trigeminal (orofacial) and spinal nerves and further nociceptive processing in the TCC may be an integral underlying mechanism of the line-shaped trajectory of EF-type pain. The succeeding migraine headache attacks further support the involvement of the TCC, since neurons in the TCC are also the major relay neurons for nociceptive afferents from the meninges (trigeminal) and cervical structures (spinal), and the convergence of meningeal and cervical afferents and the sensitization of TCC are considered to contribute to the neural substrates of migraine headache.[18] Thus, activation of TCC neurons through trigeminal and cervical nerve pathways in EF-type pain may or may not lead to migraine headache attacks, probably depending on the intensity of the EF-type pain. This view seems to be supported by the decreased intensity of EF-type pain after treatment with a low dose of carbamazepine in

the present case initiated fewer attacks of NIN and MWA, while an increased dose of carbamazepine terminated EF-type pain and, as a consequence, the attacks of NIN and MWA were relieved completely. Finally, this view is also consistent with the evidence that peripheral afferents can be involved in the generation of migraine headache.[19]

Conclusions

EF may have a variant whose pain starts from the territories of thoracic nerves and that might be a trigger for neuralgia and migraine attacks. This case description expands the clinical spectrum of EF and may also be helpful in understanding its pathophysiology.

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