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Migraine presenting as isolated facial pain: A prospective clinical analysis of 58 cases

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Abstract

Background: Sparse evidence has detailed the clinical phenotype of migraine presenting as isolated facial pain.

Objective and methods: This was a prospective audit, part of our multidisciplinary facial pain service evaluation, aiming to phenotype patients with migraine presenting as isolated facial pain who attended our service between 2013 and 2018.

Results: Fifty-eight patients were diagnosed with migraine with isolated facial pain (F = 46, 79.3%; mean age: 49.0 years, ± 9.85). Sixty-six percent of patients met the criteria for episodic migraine. The pain was strictly unilateral in 79% and located over the maxillary region in 85% of patients. Associated cranial autonomic signs/symptoms were reported by 45% of our cohort. A percentage of 77% of patients was triptan responders.

Conclusions: Migraine presenting as isolated facial pain is a rare but treatable condition with some distinct demographic and clinical characteristics. It is a diagnosis of exclusion that should be evaluated in specialised multidisciplinary facial pain clinics.

Keywords

Migraine, facial pain, orofacial migraine, atypical facial pain

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Introduction

Chronic orofacial pain encompasses numerous conditions including dental and non-dental causes, including primary headache disorders that can present with facial pain (1,2). Migraine with head pain episodes radiating to the facial territories is considered rare but it is sometimes reported (3–7). Migraine presenting with isolated facial pain (pain in V2 and or/V3) is considered extremely rare and its phenotype has not been described in full (8–10). In view of the uncommon pain location, a high proportion of these patients are misdiagnosed with dental or sinus-related conditions, frequently resulting in inappropriate surgical and medical treatments that themselves may complicate the presentation of the pain by changing its phenotype and further complicating diagnosis and timely management (11,12).

Here we analysed the demographic, clinical characteristics and response to treatments of patients who attended our facial pain clinic, diagnosed with migraine presenting as isolated facial pain.

Methods

This is a prospective clinical audit, part of a service evaluation, that took place at the King's Health Partner multidisciplinary facial pain service, which is a clinical and academic entity that includes the

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Orofacial Pain and Skull Base Neurosurgery services at King's College Hospital and the Headache Service at Guy's and St Thomas' Hospital. This service is dedicated to the diagnosis and treatment of complex facial pain patients. For this audit, data were collected from consecutive patients referred to our service between June 2013 to January 2018.

As per our service pathway, the initial clinical assessment of referred patients was performed by the orofacial pain clinicians. Odontogenic, inflammatory, and infective dental causes were excluded following clinical assessment and radiographic tests. When odontogenic causality was excluded, patients were assessed by the headache neurologist of our facial pain service (GL). Patients meeting the International Headache Society (IHS) criteria for migraine presenting with isolated V2 and/or V3 trigeminal intra- and/or extraoral facial pain were carefully phenotyped and included in the analysis. The diagnosis was reached after one or more face-to-face clinical assessments were conducted, and with the aid of headache diaries. Further investigations were carried out when appropriate, and the outcomes of certain medication trials were obtained. The definition of facial pain proposed by the International Classification of Headache Disorders 3rd edition (ICHD-3) was used (1). We also included patients with a previous personal history of episodic migraine with head pain, patients in whom the pain territory historically included V1, but which at the time of the assessment was exclusively present in V2 and/or V3, and patients with unilateral V2 and/or V3 migraine pain and other contralateral/bilateral tension-type headache provided that the patient was able to clearly discriminate between the two conditions. Conversely, patients with migraine pain centred over V2 and/or V3 and pain radiation to V1 distribution were excluded. Data were collated in an electronic password-protected spreadsheet.

Audit under current national guidelines does not require research ethics committee review (http://www.hra-decisiontools.org.uk/research/).

Results

Over the audit period, 3900 patients were assessed, of which 1248 (32%) were diagnosed with a primary headache disorder: A trigeminal autonomic cephalalgia (TAC) in 72 patients (5.8%), some of whom have been described elsewhere (13), and migraine in the remaining patients (n = 1176). Of these, 58 (4.9%) had migraine presenting as isolated facial pain and were included in the analysis.

Demographic characteristics

The patients were referred by dentists (n = 28, 48.3%), general medical practitioners (n = 18, 31.0%) and endodontists (n = 7, 12.1%). The remaining five patients were referred by secondary care specialists. Table 1 summarises the demographic and clinical characteristics of the facial pain in our patients.

Our patients were predominantly female (F = 46, 79.3%), of Caucasian ethnicity (n = 38, 65%). Apart from migraine, the medical history of our patients was remarkable for hypertension (n = 8), irritable bowel syndrome (n = 6), hypothyroidism (n = 4) lower spinal pain (n = 1) and endometriosis (n = 1). A previous personal headache history was reported by 37% of patients (n = 29). A percentage of 19% (n = 11) of

Table 1. Demographic and clinical characteristics of 58 migraine patients presenting with isolated facial pain.

	n (%)
Gender	
Male	12 (21%)
Female	46 (79%)
Caucasian ethnicity	38 (66%)
Mean (±SD) age in years (range)	49 (±9.9) (17–73 years)
Mean (±SD) duration of migraine in years (range)	4.9 (±12.2) (3 months – 30 years)
Headache onset	
Spontaneous	31 (53%)
Post-traumatic	27 (47%)
Headache course	
Episodic	20 (34%)
Chronic	38% (66%)
Laterality	
Unilateral	46 (79%)
Bilateral	12 (16%)
Pain distribution	
V2 only	49 (85%)
V3 only	3 (5%)
V2-V3	6 (10%)
Cranial autonomic features	Lacrimation: 12 (20%) Conjunctival injection: 8 (14%) Ptosis: 2 (3%) Miosis: 8 (14%) Rhinorrhoea/nasal blockage: 23 (40%) Facial redness: 2 (3%)
Triggers/worsening factors	Not apparent: 12 (20%) Stress: 16 (28%) Menses: 16 (28%) Alcohol: 3 (5%) Chewing: 11 (19%)

n: number; SD: standard deviation.

patients reported ongoing headaches fulfilling the IHS criteria for episodic tension-type headache (1); 31% of patients (n = 18) reported no personal previous or current history of headache. A percentage of 24% (n = 14) reported temporo-mandibular joint disorder (TMJD) as a comorbid pain condition to migraine.

Facial pain onset: The pain initiated spontaneously, without any apparent precipitant factors in 53% of patients (n = 31). In the remaining patients (47%, n = 27), the pain began in temporal connection to surgical interventions, namely dental extractions (n = 13), ENT procedures (n = 6), dental implants (n = 3), facial trauma, dental root canal treatment (n = 4) and whiplash injury (n = 1).

Episodic or chronic pattern: A percentage of 66% (n = 38) of patients met the IHS criteria for episodic orofacial migraine, reporting pain episodes lasting between 4 hours and 3 days, to a maximum of 4 days. The remaining patients (34%, n = 20) reported a daily facial pain with migrainous exacerbations, meeting the criteria for chronic orofacial migraine (2).

Laterality of pain and site(s): The pain was unilateral side-locked in 79% (n = 46) and bilateral in 16% of patients (n = 12). The majority of patients reported unilateral left-sided pain (67%, n = 32). Pain was localised mainly in V2 territory (84.5%, n = 49), followed by V2-V3 (10.3%, n = 6) and V3 (5.2%, n = 3). Four patients with pain in V2 reported temporal radiation.

Severity: In patients with non-constant pain, the reported severity ranged between 7–10 out of 10 on the verbal rating scale (VRS). In patients with constant pain, the background severity ranged between 3–5 out of 10, whereas the more severe exacerbations ranged between 5–10 out of 10.

Associated features: All but two patients reported at least one of the following: Nausea, vomiting, photo/phono- and/or osmophobia. Two patients experienced episodes of unilateral featureless throbbing facial pain without any associated symptoms. Thorough investigations ruled out dental, TMJD and other cranio-facial pathology. A meaningful response to triptans supported the migraine biology of these episodes. Cranial autonomic signs/symptoms were reported by 45% (n = 26) of our cohort. These ranged from just unilateral tearing (n = 8) to multiple autonomic signs/symptoms. No patients reported symptoms fulfilling the IHS criteria of aura.

Exacerbating and relieving factors: Pain was triggered by stress (n = 16) and alcohol (n = 3). Exacerbation of the pain during menses was reported by 16 of 46 female patients. Twelve patients could not identify a trigger. Pain was made worse by eating in 11 patients who also had comorbid TMJD.

Table 2 summarises treatments failed prior to referral to our service. Once the diagnosis of migraine with isolated facial pain was confirmed, triptans were tried in all episodic migraine patients (n = 30/38) apart from eight with a history of hypertension. Triptans were able to successfully and consistently abort the facial pain episodes in 77% of this group (n = 23/30). At the time of the assessment, 69% of patients were taking daily painkillers with poor benefit. Of these, 20 patients were having daily facial pain and 20 were having high-frequency episodic facial pain episodes. The discontinuation of daily painkiller intake led to an improvement of the facial pain symptoms in 40% (n = 16/40) of patients. All patients were offered preventive treatments with evidence of efficacy in migraine in view of the frequency of symptoms; however, only some decided to start them (Table 2). Indomethacin was used to exclude hemicrania continua in patients with unilateral side-locked facial pain with autonomic symptoms.

Table 2. Treatments tried before and after the diagnosis of migraine with isolated facial pain was confirmed in the 58 patients.

	n (%)
Patient-reported ineffective treatments prior to diagnosis of migraine being confirmed	Antibiotics: 55 (95%) Different pain-killers: 40 (69%) Endodontic treatments: 21 (36%) Multiple dental extractions: 17 (29%) Dental restorations: 3 (5%) Physiotherapy: 4 (7%) Functional endoscopic sinus surgery: 4 (7%) TMJ surgery: 1 (2%) SPG interventions: 1 (2%)
Patient-reported effective treatment once diagnosis of migraine was confirmed	Triptans: 30 (52%) Tricyclic antidepressants: 12 (21%) Propranolol: 4 (7%) Candesartan: 9 (16%) Topiramate: 4 (7%) Gabapentin: 3 (5%) Pregabalin: 1 (2%) Greater occipital nerve blocks: 4 (7%) Botulinum toxin type A: 4 (7%)

n: number; SPG: sphenopalatine ganglion; TMJ: temporo-mandibular joint.

Discussion

Migraine presenting as isolated facial pain is considered to be very rare, as shown in migraine population-based studies as well as in neurology clinical settings (4,7). The overlap of symptoms with dental and ENT pathologies may partly explain the high rate of misdiagnosis and the sparse attempts to describe the

clinical characteristics of migraine with this rare pain location (14). Furthermore, conflicting terminologies used to classify migraine with facial pain have certainly contributed to creating confusion among clinicians and scientists in systematically defining this group of patients. The recent effort of the International Classification of Orofacial Pain (ICOP) to standardise the clinical profile of orofacial pain conditions will hopefully create a common nosological language to be used in clinical practice and research settings (2). Our multidisciplinary facial pain team is a one-of-its-kind setup, including an orofacial pain specialist with a dental background, a headache neurologist, a neurosurgeon, and a pain specialist with an interest in facial pain. The level of clinical specialisation provided and the complexity of patients attending our service may explain why we have been able to assess and diagnose a large series of such patients. According to the ICOP, our patient group would be diagnosed as episodic or chronic orofacial migraine (2).

The demographic characteristics of our patients were similar to previous studies of facial pain in migraine (3). The late age of onset has been reported in other studies and simply interpreted as a normal delay due to the progressive change in pain location from the typical head/V1 territories to V2-V3 territories (3). However, it might also be possible that accumulation of minor but multiple V2-V3 nerve ending injuries due to dental and/or sinus conditions and/or surgical procedures may have led to the expression of the migraine phenotype in sensitised areas of the trigeminal system in subjects with a migraine biology (personal and/or family history of migraine).

The clinical phenotype of migraine patients with isolated facial pain that emerged from our analysis displayed some peculiarities that differ from migraine with head pain, namely the laterality of the pain and the proportion of patients with associated cranial autonomic features. The vast majority of our patients reported unilateral side-locked pain centred over the V2 trigeminal division, in particular the maxillary region, with infrequent radiation to the temple. Similar findings were observed in a series of 24 patients with migraine with isolated facial pain and subsequently in 76% of patients studied in a large population-based study (9,14), but not in a small series of seven patients of whom only two had pain limited to V2 (8); the latter contrasting finding may have been secondary to the small sample size. The high percentage of patients with V2 pain may also partly reflect a referral bias. Recurrent maxillary pain episodes are often misdiagnosed as "sinus pain" and referred to ENT specialists, who in turn refer them to facial pain clinics (15). Furthermore, when migraine affects the facial territories it seems to be more frequently associated with ipsilateral/bilateral cranial autonomic signs/symptoms compared to migraine with head pain, and hence is more often misdiagnosed as "sinus pain" (9). The presence of cranial autonomic signs/symptoms associated to the pain in our series (45%) was similar to that observed in a previous population-based study (48%) but also similar to studies conducted in non-neurological settings (3,4). It could be possible that parasympathetic overflow more often accompanies conditions characterised by strictly unilateral trigemino-vascular pain than those with bilateral pain distribution (16). This would explain why unilateral migraine and unilateral side-locked headache disorders like the TACs are more likely to be associated with ipsilateral cranial autonomic symptoms.

It has been shown that migraine with isolated facial pain largely responds to migraine-specific abortive treatments as well as to preventive treatments used in migraine prophylaxis (3,6,9,10). Our findings confirmed this trend. Apart from offering an effective abortive treatment option, the response to triptans could be helpful as a diagnostic aid in some cases with unclear presentations and, in cases where migraine with facial pain is comorbid with other facial pain conditions, complicate the diagnosis further.

This audit has limitations, including the less rigorous methodology typical of an audit, which includes real-world patients. However, the phenotyping and diagnostic processes were consistent and robust in each patient analysed and they were based on multiple clinical assessments over time, careful investigations, and use of drugs such as triptans and indomethacin as diagnostic trials to help clarify the diagnosis when required.

Conclusions

We refined the demographic and clinical phenotype of a very rare subtype of migraine in a large series of patients. The characteristics are largely similar to migraine with head pain apart from a high proportion of unilateral pain cases with associated cranial autonomic features. Migraine with isolated facial pain responds well to triptans and to the commonly used migraine preventive treatments. In view of its rarity, the diagnosis of orofacial migraine needs to be considered only when dental and sinus pathologies are excluded. The collaboration between neurologists and facial pain specialists is key to increasing awareness and education on this rare but treatable manifestation of an otherwise very frequent headache disorder.

Clinical implications

- Migraine with isolated facial pain is a rare but treatable form of orofacial pain, often misdiagnosed by dentists and ENT specialists.
- Migraine with isolated facial pain is often characterised by episodes of strictly unilateral pain centred over the V2 distribution associated with cranial autonomic features. The majority of cases present with an episodic pattern, but a chronic daily pattern can occur.
- Migraine with isolated facial pain responds to triptans and to migraine preventive treatments. Indomethacin tests are important to rule out indomethacin-sensitive headaches in unilateral cases with continuous pain and associated cranial autonomic features.
- The multidisciplinary approach is key for the diagnosis and treatment of this rare form of migraine and as an educational platform for physicians who treat patients with facial pain.

Contributors

All authors participated in the study design, implementation and/or conduct of the study. All authors contributed to the audit and approved the final manuscript.

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Declaration of conflicting interests

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