CASE REPORT

Two atypical cases of isolated migrainous paraesthesia

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SUMMARY

A large number of migraineurs have various transient neurological symptoms in the absence of headache at one time or another. Such transient neurological symptoms largely represent the typical migraine auras without headaches. Isolated visual aura is the most common aura in this category. Sensory aura is the second most common aura. However, the literature is sparse regarding isolated sensory aura without headache. Herein, we are reporting two cases of recurrent paraesthesia that fulfilled the International Classification of Headache Disorders (ICHD-3β) criteria of typical migrainous sensory aura without headache. Review of the literature suggests that upper limb involvement is almost universal (>90%) in the typical sensory aura. However, our both cases were unusual in this aspect. Case 1 had recurrent spontaneous paraesthesia predominantly on the left side of the face. Case 2 had recurrent spontaneous paraesthesia in either the left or right lower limb. Both cases responded to antimigraine drugs.

BACKGROUND

About 20% of migraineurs have various transient neurological symptoms in the absence of any headache at one time or another.[1] Many of them represent typical auras of migraine without headaches. About 42% patients with migraine with typical auras may have an isolated aura in the absence of headache.[2] Visual auras are the most common migraine auras, and it has been well reported even without headaches. Sensory auras are considered as the second most common auras.[2] However, the literature is silent regarding isolated migrainous sensory auras. Earlier, we have reported 14 cases of typical sensory auras in the absence of headaches.[3] Herein, we are reporting two additional cases where patients had atypical location of sensory auras without headaches.

CASE PRESENTATION

Case 1

A 32-year-old man had recurrent episodic paraesthesia over the left side of the face and head since 3–4 years. The paraesthesia was felt as unpleasant prickling (tingling) and burning sensation. The frequency was one episode at 2–3 months interval until 5–6 months back. After that, the frequency gradually increased to more than 10 attacks in a month. The attack was always on the left side. Most attacks had marching (spreading) of the symptoms. In a typical attack, the paraesthesia starts around the corner of the mouth and spreads upward to involve the upper two-thirds of the face (including frontal and temporal areas). However, it never involved the orbit. The marching of the sensory symptoms was slow and used to take 5–10 min to reach the upper part of the face and head. Most attacks were also accompanied by paraesthesia of the left side of the tongue. In about one-fourth of the attacks, he had similar paraesthesia in the left feet. Very rarely (in about 10% attacks), he felt symptoms in the left fingers and hand. The paraesthesia of the left foot and left hand was the extension of the marching phenomenon (ie, used to develop after the symptoms of the face and tongue). Overall, the paraesthesia used to persist for about 60 min. The paraesthesia of the left feet or left hand was never present in isolation (ie, It was always associated with paraesthesia of the face and tongue). The patient denied the paraesthesia on the right side of the face, right upper limb and right lower limb. Between the attacks, the patient did not feel any symptom in the face.

The patient had a history of migraine since adolescents, occurring at a frequency of 1–2 attacks per month. On asking, he admitted having occasional visual disturbances (seeing flashes of light) for a few minutes. However, he did not find any interrelation of paraesthesia to headaches and visual symptoms. He noted stress, sun exposure and change in the weather as precipitating factors for his paraesthesia. Jaw movements (eating, chewing and talking) had no relation in the initiation or aggravation of the symptoms. There was no nocturnal attack.

The patient visited a number of doctors, including dentists, otolaryngologists and neurophysicians. No physical abnormality was ever detected by any doctor. The patient was extensively investigated in the past and it included various biochemical parameters, MRI brain, MR angiography (MRA), MRI cervical spine, nerve conduction study (NCS) and EEG. No abnormality was noted in any investigations. Serial EEG was performed to find out any abnormality or temporal relation with paraesthesia. However, no abnormality was noted even during an episode of paraesthesia. A number of differential diagnoses were considered (trigeminal neuralgia, atypical facial pain, related to some intraoral pathology, psychiatry disorders etc).The patient received a number of drugs for his paraesthesia and it included: carbamazepine, oxcarbamazepine, lamotrigine, gabapentin, pregabalin, fluoxetine, clonazepam, tramadol and various multivitamins. However, he did not get much benefit with any of these treatments.

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We considered a possibility of migrainous sensory aura without headache. The patient fulfilled the International Classification of Headache Disorders (ICHD-3β) criteria of migrainous sensory aura without headache. (ICHD-3β code: 1.2.1.2) The patient was prescribed flunarizine at a dose of 10 mg. Three weeks later, he reported a marked reduction in the frequency of the attacks (just one attack in 3 weeks). Flunarizine was continued at the same dose. He was asked to maintain a headache diary to find out the auras and an interrelation between headaches and paraesthesia. He had just three attacks of paraesthesia during the next 4 months. The drug was withdrawn after that. However, he was asked to maintain the headache diary. In the next 12 months of follow-up, he had one attack at 2–3 months interval. In, at least two attacks of paraesthesia, he noted migrainous headaches (although relatively milder than his usual headache attacks.). He also noted visual auras (seeing flashes of light and blurring of vision) in some of his usual migraine headache attacks. However, he did not find visual symptoms with paraesthesia even in the follow-up.

Case 2

A 29-year-old man was referred to the neurology outpatient department for the evaluation of episodic paraesthesia (tingling sensation) in the lower limbs of 5–6 years duration. It was too infrequent in the earlier years (2–3 attacks in a year). However, the frequency gradually increased and it was 1–2 attacks per week for the past 9–10 months. Most attacks were limited to the left lower limb. Right lower limb was involved in about one-fourth of the attacks. However, he never felt paraesthesia in both lower limbs simultaneously. In a typical attack, the paraesthesia starts in the toes of one of the lower limbs, and thereafter it spreads upward to involve the whole lower limb on the same side within 5–10 min. On asking, the patient admitted having marching of this symptom on the same side of the upper limb (mainly fingers and the hand) in about 10% of the attacks. However, the paraesthesia of the upper limb was not as marked as of the lower limbs. The average duration of symptoms was about 30–45 min. However, a few attacks persisted for whole days. Between the attacks, the patient did not feel any symptom in the lower limbs.

Most attacks started spontaneously. The patient did not find any relation of paraesthesia with any posture or activity of the lower limbs. Walking has no effect on paraesthesia. There were no nocturnal attacks.

The patient had a history of migraine headache since the age of 15 years. He noted an increase in the frequency of migraine headaches in the recent past (3–4 attacks in a month for the past 1 year). On asking, the patient admitted having visual auras (seeing flashes of light) and vertiginous sensation with migraine headaches in about one-fourth of the attacks. He did not find any interrelation between headaches and paraesthesia. However, he noted that precipitating factors for both migraine headaches and paraesthesia were almost similar (skipping a meal, lack of sleep, weather changes, sun exposure, etc). He used to take paracetamol as abortive therapy. The patient did not receive any prophylactic medication for his migraine.

He visited a number of physicians, neurophysicians, neurosurgeons and orthopaedicians. However, no physical abnormalities were ever noted. He was extensively investigated over the years. Routine haematological and biochemical investigations were normal. MRI brain, MRA, MRI spine (performed twice), NCS (performed thrice) and EEG were normal. EEG did not reveal any abnormality even during an ongoing attack of paraesthesia.

He received numerous drugs (such as pregabalin, gabapentin, clonazepam, fluoxetine, multivitamins etc). However, he did not feel much improvement.

We suspected a possibility of migrainous sensory auras without headaches. The patient fulfilled the (ICHD-3β) criteria of migrainous sensory aura without headache. The patient was prescribed a combination of propranolol (40 mg daily) and flunarizine (10 mg).

The patient started showing improvement after 2 weeks. The frequency reduced to less than one attack per month. The same drugs were maintained for 8 months. After 8 months, both drugs were gradually tapered off. He was followed up for another 10 months. There was a slight increase in the frequency (1–2 attacks/month) after the withdrawal of the drugs. We did not start any drug and reassured the patient about the benign nature of the symptom.

DISCUSSION

The presence of paraesthesia in the face or in one limb highly suggests a possibility of secondary causes.[4] Both patients were extensively investigated over the years for paraesthesia. However, no abnormality was ever noted. Structural pathologies of the brain stem, spinal cord and radicles were ruled out by repeated neuroimaging in both cases. Multiple sclerosis may rarely have episodic paraesthesia in the extremities.[5] However, it was less likely as the symptoms were present for very long periods and there was long follow-up without developing any other symptoms, suggestive of multiple sclerosis. Moreover, MRI performed on a few occasions, did not reveal any abnormality. A possibility of peripheral nerve involvement (mononeuropathy or mononeuritis multiplex) was less likely as paraesthesia was not in the nerve distribution and repeated NCS testing did not reveal any abnormality.[4]

The symptoms were present for a very long periods in both cases (case 1: 3–4 years, case 2: 5–6 years) and there were many episodes in the past. The patients were absolutely normal between the attacks. It suggests some benign pathologies in both patients. Both patients had a history of migraine. It is suggested that migraine should be considered as an aetiology in any periodic or episodic neurological symptom.[1] Both patients fulfilled the ICHD-3β criteria for typical aura without headache (ICHD-3β code: 1.2.1.2).[6] However, a possibility of other episodic disorders should always be ruled out.[7] Epilepsy is the closest differential diagnosis in such circumstances.[7] However, a possibility of epilepsy was less likely because of the following reasons: marching and overall duration of an attack were too long, there were too many attacks in the past, EEGs were normal (even during the attacks of paraesthesia) and there were no responses of any antiepileptics used in the past in both patients.

A typical sensory aura is cheiro-oral or digitolingual in distribution and shows marching of the symptoms.[2] Classically, sensory aura starts in one of the hands and spreads up to involve the whole of the upper limb. In about two-third of cases, it jumps up to involve the ipsilateral face.[4 8] In case 1, the patient had paraesthesia localised mostly to the face and tongue. The marching was noted towards the upper part of the face. The involvement of the tongue is very specific of migraine. Migraine is considered as the most common cause of episodic tongue paraesthesia.[9] Occasionally with facial paraesthesia, he felt paraesthesia in the lower limb and upper limbs as well. This hemidistribution of paraesthesia was more reinforcing as a possibility of migrainous paraesthesia (rather than peripheral nerve

involvement). Case 2 showed side shifting paraesthesia in the lower limbs and on a few occasions, there was hemiparaesthesia. This was consistent with typical sensory aura. Moreover, in follow-up, when the patients were asked to maintain a headache diary, a few attacks of paraesthesia had a migraine-like headache. Precipitating factors for paraesthesia in both patients (skipping a meal, lack of sleep, weather changes, sun exposure, stress etc) were compatible with the usual precipitating factors of migraine. There was a temporal relation in the initiation of the drugs and reduction of the attacks of paraesthesia. With all these features we presume that paraesthesia in both patients was linked to migraine. We started flunarizine (both patients) and propranolol (one patient) for their episodic paraesthesia. We tried to avoid the drugs which were effective in both migraine and epilepsy (such as, antiepileptic drugs). It was done to minimise the diagnostic confusion between migraine and epilepsy in both patients. European Federation of Neurological Societies (EFNS) guidelines for migraine prevention considers both flunarizine and propranolol, as drugs of the first choice for migraine prevention (with level 'A' evidence).[10] A few studies have demonstrated that a combination therapy may be more beneficial than an individual drug.[11]

Both cases are unusual in two aspects: (1) location of paraesthesia and (2) paucity of migrainous headaches and other auras during the attacks of paraesthesia. Migrainous sensory auras may be noted in any part of the body. The hand (96%) or upper limb as a whole (90%) is the commonest site in a typical sensory aura.[2 8] All other sites are usually described in relation to hand or upper limb. In our both patients, there were a few instances where the patients felt pain in the upper limb, but largely it was absent and the patients were worried for paraesthesia at different sites. To the best of our literature search, sensory auras limited to the lower limb or localised mainly to the face have not been reported.

The ICHD-3β recognises typical aura without headache as a separate entity. However, review of the literature suggests that isolated migrainous aura (ie, without headaches) is almost limited for visual auras. Sensory aura is the second most common aura (30–54% of total aura).[2 8] Therefore, it has been hypothesised that even sensory aura without a headache may be found in some migraineurs. As the prevalence of visual auras is very common (80–99%), patients with sensory aura usually have a coexistent visual aura.[2 8 8] There are a few cases in the literature where patients had both visual and sensory auras together in the absence of headaches. In a series of 163 patients of migraine with aura, 5 patients had visual and sensory aura together in the absence of headache.[2]

However, a subset of patients may have only sensory aura (without any coexistent visual aura and headache). In our case series of 14 patients of sensory auras, only 36% had a history of visual auras.[3] Miller Fisher in his two seminal papers on late-life migraine accompaniments noted that 28% patients did not have any visual symptoms.[9 12] Several of these patients had episodic paraesthesia in combination with other neurological symptoms. Therefore, some auras may be even without headaches and without visual symptoms. Both our patients had a history of visual auras. However, it was not frequent and one patient noted it prospectively in the follow-up. In addition, there was no interrelation between sensory symptoms to visual auras.

The pathophysiology of auras is explained by the mechanisms of cortical spreading depression (CSD). CSD classically starts in the occipital cortex and spreads to the other lobes.[13] However, it is presumed that the CSD could start in any part of the brain. Depending on the involvement of the various part of the brain by CSD, the patients will have various symptoms (in isolation or in different combinations). We hypothesised that in both patients CSD was predominately involved in the parietal lobe.

CONCLUSION

Our case reports and review of the literature suggests that migrainous sensory auras may be without a headache. Moreover, paraesthesia may be predominantly in any part of the body. The marching of symptoms, an episodic symptom of 5–60 min and history of migraine headaches are clinical clues to detect it.

LIMITATION

Although all the possible causes described in the literature for episodic paraesthesia had been ruled out before giving a trial of antimigraine drugs, we cannot rule a possibility of other hidden secondary causes. In addition, with just case reports, we cannot conclude anything with confidence. Prospectively observations are warranted to confirm it.

Patient's perspective

Patient 2: surprised to know that it was because of migraine. Thank you doctor.

Learning points

- Just like the isolated visual aura, other migrainous auras may also present without headaches.
- Sensory aura typically starts in upper limb and spreads out to involve the face and other limbs. However, it may start in any part of the body.
- All migraineurs who have unexplained periodic sensory symptoms should receive a trial of antimigraine drugs.

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