CHAPTER 22

THE PATIENT WITH "ICE-PICK" PAINS

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Case History

This patient is a 46-year-old woman who began experiencing recurrent headaches at the age of 13. They were infrequent during her teenage years. There were no premonitory symptoms. The pain was described as a unilateral or bilateral aching pain which would gradually build up in intensity. Eventually this developed into a severe throbbing pain over the entire head. This was associated with nausea, photophobia, sonophobia, and vomiting if the pain was very severe. Sometimes the headaches occurred around her menstrual periods but not consistently. Hunger and decreased amounts of sleep often triggered them.

The frequency and severity of the patient's headaches increased during her twenties. However, during her pregnancy the headache almost completely subsided only to return after delivery.

The headache frequency decreased during her thirties. She then noticed the development of a sharp head pain usually over the frontal regions or in the eye prior to the onset of her regular headache. Occasionally she would experience several of these sharp pains during the entire day and would eventually develop one of her usual headaches. This sharp jabbing pain lasted only for 15 to 20 seconds but was very severe. It virtually jolted her each time it occurred. These painful episodes did not have any specific pattern or cyclical features. There was no conjunctival injection, tearing, or nasal congestion.

In her forties, the lifelong severe headache almost completely subsided, however, she noticed several of the sharp jabbing pains on an almost daily basis. The frequency fluctuated but she generally had 20 to 30 of these on a single day. The pain still lasted only for 15 to 20 seconds. The location of the pain was either in the eyes or in the temporal regions, however, it did occur in other parts of the head. The pain did not awaken her from sleep. She did not observe any specific triggering factors.

She had no other medical problems. There was a history of recurrent headache in her mother, sister and daughter. None of them experienced the sharp jabbing pains. A magnetic resonance imaging (MRI) scan of the brain was normal.

Ergotamine tartrate had been used effectively in the past to relieve her regular headache. She was treated prophylactically in her twenties with beta-blockers which reduced the frequency of the headaches. They were reintroduced to control the sharp pains but with no benefit. She tried taking pain medications round the clock without benefit. She was subsequently treated with indomethacin, 25 mg three times a day, resulting in a 50% reduction of the sharp pains. When the dosage was increased to 50 mg, three times a day, the headache became very infrequent. Attempts to lower the dose subsequently lead to increased frequency. She has remained on this dose with intermittent unsuccessful attempts at withdrawal.

Questions about This Case

- What is your diagnosis?
- What are the differential diagnostic considerations?
- Are there any other investigations you want to undertake in this patient?
- How would you manage this patient on a long-term basis?

Case Discussion

This patient developed intermittent headache at the age of 13 that fluctuated in intensity and frequency over the years. Overall, as she grew older there was a gradual reduction in the frequency and severity and in her forties the headaches almost completely subsided. They were typical of migraine without aura as defined by the International Headache Society (IHS). The major features included a family history of headache in her mother, sister, and daughter, intermittent unilateral headache which eventually became bilateral with a pulsating quality, and associated nausea, vomiting, photophobia, and phonophobia. The history of headache triggered by hunger, change in sleep pattern, relationship to menstrual periods, and subsidence during pregnancy are all typical of migraine headache. Generally speaking the frequency and the severity of the headache decrease as patients grow older. In this particular patient these headaches subsided substantially by the time she was 40.

In addition to the migraine, she also noticed the sharp jabbing pains in her thirties. Initially these were associated with the migraine headache. They were either isolated or would occur as multiple episodes prior to experiencing a typical migraine. This is a very common phenomenon and often is overshadowed by the migraine itself because these episodes are very brief and infrequent. However once the migraine headache subsided she began having 20 to 30 of these in a single day. The pain lasted only for 10 to 20 seconds. It was located most frequently in either eye or the temporal regions even though it could be located in other parts of the head. There were no symptoms of conjunctival injection, lacrimation, ptosis, or rhinorrhea associated with these head pains. There was an excellent therapeutic response to indomethacin. This type of head pain is typical of "idiopathic stabbing headache," formerly known as ice-pick pains. The headache is described in the IHS headache classification (4.1) as transient stabs of pain in the head that occur spontaneously in the absence of organic disease of underlying structures or of the cranial nerves. The IHS diagnositc criteria for this type of headache are:

1. The pain is confined to the head exclusively or is predominantly felt in the distribution of the first division of the trigeminal nerve (orbit, temple, and parietal area).
2. The pain is stabbing in nature and lasts for a fraction of a second. It occurs as single stabs or a series of stabs.
3. It recurs at irregular intervals (hours to days).
4. The diagnosis depends on the exclusion of structural changes at the site of the pain and in the distribution of the affected cranial nerve.

Comments: Stabbing pains are more commonly experienced by people subject to migraine headache, in which case they are felt in the site habitually affected by headache in about 40% of patients and tend to be more frequent at the time of headache. They commonly subside with the administration of indomethacin, 25 mg orally three times daily.

This patient's head pain fits all the diagnostic criteria outlined above including the response to indomethacin.

There is no other condition which causes such sharp pains of short duration without any other neurologic signs and symptoms. However, it is appropriate to discuss some of the other clinical entities that may resemble this particular syndrome especially if the history of headache available is not adequate and the reader is not familiar with these other disorders.

Chronic and episodic paroxysmal hemicrania (CPH, EPH) is a rare disorder resembling cluster headache but predominantly occurring in females as opposed to the male predominance of cluster headache. Each headache episode lasts for a very short time, generally 2 to 5 minutes but according to IHS classification could be as long as 45 minutes. The frequency of the attacks is more than 5 per day but typically up to 30 attacks per day is more the norm. The headache has the same periorbital or retro-orbital distribution as cluster headache. The pain is associated with all the typical autonomic accompaniments including conjunctival injection, lacrimation, nasal congestion, rhinorrhea, and rarely ptosis, and eyelid edema. The headache is always unilateral and responds to indomethacin. Some patients will have cycles of these headaches which are then classified as EPH as opposed to CPH which is the most common type of presentation.

Short-lasting, unilateral, neuralgiform headache with conjunctival injection and tearing (SUNCT) is a rare syndrome. It occurs more commonly in men. The duration of the pain is from 30 to 120 seconds and is associated with conjunctival injection and tearing just like in cluster headache and CPH. Pain can often be precipitated by turning the head from side to side. The frequency of the headache varies. No specific drug appears to control it consistently.

In trigeminal neuralgia the pain is confined to one of the divisions of the trigeminal nerve, most commonly the mandibular division. The pain is extremely brief or momentary and has a sharp shooting quality. There are often paroxysms of pain, one after the other like a "machine gun peppering." There are no autonomic accompaniments. Trigger points are often present. Touching, cold air blasts, talking, and eating are the most

common triggering factors. The examination is normal unless the neuralgia is one of the rare symptomatic ones. Medical treatment using drugs like carbamazepine is usually very helpful in the majority of patients.

All patients with this type of headache syndrome have a normal neurologic and physical examination. There is no reason to investigate these patients beyond the history and examination unless there are atypical features which raise doubts in the examiner's mind regarding the diagnosis.

Management Strategies

Patients who have infrequent jabbing headache in association with migraine or other benign headaches do not require any specific therapy other than treating the accompanying major headache problem. Generally, if one is able to control the migraine headache, the jabbing pains also come under control. However, if the frequency increases and they seem to occur independent of any other headache syndrome, specific therapy may be required.

Indomethacin appears to be the first choice in treating this syndrome. The majority of patients show significant improvement and some patients attain complete resolution. However, the response is not "absolute" as described in other headache syndromes like CPH. Generally a dosage of 25 mg, taken three times daily, is effective. The dosage could be doubled if there is no response. Gastric problems are the most common side effects and these should be watched for and treated if necessary. It is possible to evaluate the effectiveness within 2 to 4 weeks. The drug should be discontinued if there is no response within this period of time because of the potential for gastrointestinal side effects in a large proportion of patients. Other nonsteroidal anti-inflammatory agents and antimigraine agents like calcium channel blockers have also been found to be effective. Tricyclic antidepressants could also be tried. There have not been any controlled trials of any of these drugs so far.

Case Summary

- This patient has migraine without aura.
- She experienced infrequent "jabs and jolts" either preceding or during a migraine headache.
- The "jabs and jolts syndrome" became the most prominent symptom after resolution of her migraine headache.
- There was significant response to treatment with indomethacin as seen in the majority of these patients.

Overview of Benign Idiopathic Stabbing Headache

This syndrome has been described under various terms including "jabs and jolts syndrome," "ice-pick like pain", "sharp stabbing head pain," and "needle-in-the-eye syndrome." The name suggested by the IHS is "idiopathic stabbing headache."

The exact incidence of this type of headache is unknown. The coexistence of isolated episodes of stabbing pain in patients with other types of headache is fairly common but patients rarely ever discuss or mention this unless they become frequent. This reportedly occurs in 40% of patients with migraine and often is located in the same territory as the patient's migraine. The frequency increases just prior to or during a migraine headache. It has also been reported with cluster, tension, and posttraumatic headaches. In some patients, however, this type of headache occurs as an independent entity with the frequency varying considerably. A patient may experience one or two episodes a day or there could be as many as 50 a day. Pain may sometimes occur in a cyclical fashion and rarely is there any fixed pattern as seen in cluster headache. There is no sex predilection as seen in cluster headache or CPH

The duration of the pain is often a few seconds and almost always for less than one minute. However, many patients will describe a very low-grade pain and occasionally tenderness in the same location for minutes to hours after an intense episode of pain. The location of the pain tends to change even though several episodes may occur at the same site. More often pain occurs at random locations. Temporal, orbital, and supraorbital locations appear to be most common. The intensity is moderate to severe and often jolts the patient especially because of the unexpected occurrence and partly due to the intensity. Each individual episode may not be disabling, but when multiple episodes occur on a daily basis the problem becomes intolerable and patients seek help.

Most of the painful episodes occur without any provocation. Some of Raskin's patients reported that a sudden change in posture, physical exertion, dark-light transition, and head motion as probable triggers.

In the vast majority of patients no specific treatment is required. Management of the associated headache syndromes often relieves the symptoms. Indomethacin appears to be effective in the majority of patients who suffer from the independently occurring episodes. The response is not thought to be absolute as seen in CPH patients. Other nonsteroidals or vasoactive agents similar to those used in the prophylaxis of migraine and cluster headache may also be beneficial in cases where indomethacin fails.

Selected Readings

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Editorial Comments

The spectrum of symptoms in patients with migraine is remarkable despite relatively precise IHS criteria. Migraine can express itself in a myriad of simple and complex symptomatologies, and this case reported by Dr. Vijayan is no exception. The differential diagnosis of such brief, frequent, paroxysmal, painful events is likewise fascinating. Pathophysiologically, one wonders what neuronal or vascular perturbations occur to result in "jabs and jolts," and presumably they are part of the "nerve storms" originally described by Livieng in his book On Megrim. *Much is to be learned form this case, in large part due to the author's clear and concise narrative.*