

Interesting Cases from The Headache Clinic Records



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Case No. 1

RS, F/28, presented with a history of recurrent headaches for the last three years, since her last delivery. She did not give a previous history of headaches and her pregnancy was uneventful. The head pain was mainly right hemicranial, throbbing in nature and was associated with vomiting. Interestingly, these attacks were clearly linked to her menstrual periods, and the periods were regular. The attacks would come on 3-4 days prior to her periods and inter menstrually, she was free of headaches. There was a positive family history of similar headaches in her mother also, who also had terrible headaches during her periods. Because these headaches were menstrually linked, she consulted her gynecologist who diagnosed it as pre-menstrual syndrome and prescribed painkillers and anxiolytics.

Questions

1. What is your diagnosis?
2. How would you treat this patient?

Discussion

Migraine affects women three times more than men and 60% of women often report an increased frequency of migraine in association with their menstrual period. This is referred to as menstrual migraine. Approximately 14% experience headaches exclusively with menstruation as in this patient when it is labeled Pure Menstrual Migraine.

It is well known that the female hormones estrogen and progesterone have a profound effect on the brain and there are changes in the levels of estrogen and progesterone in the blood during a menstrual cycle. During days 16-25, both levels are high and just before the next menstrual period, the levels of estrogen and progesterone dramatically fall. 'Pure' Menstrual migraine which occurs regularly at the start of the period or \pm 1-2 days prior and at no other time is triggered off by this rapid fall in level of estrogen. The fall of progesterone leads to the bleeding but does not contribute to the headache. Fall in estrogen influences chemicals in the brain such as serotonin, adrenaline, dopamine and endorphins. Changes in the activity of these brain chemicals likely trigger the development of migraine headache.

Because in this patient, the timing of menses is predictable, preventive treatments taken just before the periods are a viable alternative. There are usually started 1-2 days prior the expected date of the periods and continued for 5-10 days. NSAIDs, rofecoxib and magnesium supplementation have shown to be of benefit in women with menstrual migraine.

Hormone manipulation may be required for intractable, menstrual migraine prevention. Options have been studied and recommended. Adding estrogen in the form of a patch or a pill beginning 2-3 days before the menstrual period to smooth on the natural drop in estrogen can be beneficial.

Other therapies for prevention of menstrual migraine include diuretics, melatonin and feverfew. Preventive treatment of menstrual migraine is a challenging trial and error process, but persistence and good patient physician communication can yield worthwhile improvements in migraine control.

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Case No. 2

NR, M/39 came to our Headache Clinic with a history of recurrent headaches since 1992. At the onset, the pain was mainly localized to the right or left periorbital region, would come on 2-3 times/week and was associated with vomiting, and photophobia. The headaches worsened with exposure to the sun and when he missed his breakfast. There is a family history of similar headaches in his mother. He was diagnosed to be suffering from Migraine and was started on ergotamine tablets to control the acute attacks and was also given a course of Flunarizine 5 mg on a daily basis for 3 months. He stopped Flunarizine but his headaches continued. And so he started taking 1-2 tablets of Ergotamine whenever he had the headache. He got temporary relief but the frequency of his headaches gradually became worse.

The headaches later started coming on a daily basis and he had seen many neurologists who had tried different prophylactic drugs in increasing doses. But since he got maximum relief with Ergotamine tablet he would take these regularly. The headache that came on a daily basis did not however have all the features of migraine.

Questions

- a. Why did the headache worsen? And go on to becoming a daily headache inspite of treatment?
- b. What is the diagnosis?
- c. How should he be treated?

Discussion

This man started off with episodic attacks of Migraine without Aura. Because he was under the wrong impression that prophylaxis is a course for three months, he stopped the background medications and went on to gradually overdosing himself with ergotamine. His daily headaches with reduced migrainous features was due to Medication Overuse following on excessive consumption of Ergotamine. It is now well established that when there is overuse, routine prophylactics are ineffective and therefore just increasing the dosage of prophylactic drugs did not give him relief.

The right way to treat Medication Overuse Headache (MOH) is to

1. Withdraw the offending agent
2. Replace with an alternative drug to treat the acute attacks for e.g. Naproxen or one of the triptan drugs
3. Continue the prophylaxis. Topiramate has evidence as a good prophylactic choice in MOH
4. Amitriptyline is helpful whenever there is a daily headache
5. Once the offending drug was withdrawn, he showed improvement and the headache frequency and severity improved. He is now on minimum dose of prophylaxis.

Case No. 3

SD, F/37 was referred to our Headache Clinic for further evaluation of her intractable headache. She would mainly complain of a sudden, severe, bilateral, throbbing occipital headache lasting for just 10-15 secs. These attacks always came on when she laughed or stretched her neck and shoulders or



Fig. 1 : Sagittal T1 image of Chiari type I malformation showing descent of cerebellar tonsil below the level of the foramen magnum

coughed. They were sudden and so intense that she felt a sudden rush of blood to the head and felt she was heading for a stroke. This was followed for a few hours by a dull bearable head pain with nausea. Because of the onset with laughing, she had even stopped attending parties and social functions where she could be provoked into laughing. There were no visual complaints or neurological deficits. Clinical neurological and systemic examination were normal.

She also has a second type of mild bearable headache which she developed whenever she went in the sun, or had some physical or emotional stress or consumed red wine. There was a positive family history of migraine in her mother and grandmother. She came with a CT scan done earlier which was normal. She had received routine analgesics and was diagnosed to be suffering from Migraine but had never received optimal treatment. In view of the history we asked for an MR scan with request for special views.

Questions

1. Why did we ask for an MR scan, even though a CT Scan was normal? What was the specific abnormality we were looking out for on MR.?
2. What is the most important clue to the diagnosis of the cause

of her severe headache?

3. What are the main differences between the 2 different types of headache she has?
4. What are the treatment options available?

Discussion

This patient has 2 different types of head pain. In such situations when the patient has more than one type of headache, it is advisable to analyse the more severe type of headache first. In this patient, the sudden onset, severe unbearable short-lasting headaches are precipitated by laughing, stretching the neck or coughing. Whenever a maneuver aggravates or produces this type of headache, be it sneezing, weight-lifting, bending, straining at stool or stooping, it is defined as Cough Headache. This was originally described by Tinel in 1932 and later by Symonds in 1956; Rooke in 1968 proposed the broader term. "Benign exertional headache" for any headache that is precipitated by exertion, has an acute onset and is unassociated with structural central nervous system disease, thus combining cough and exertional headache.

This type of maneuver related headache may be benign or symptomatic. When it is benign the pain begins immediately or within seconds after coughing, sneezing or a Valsalva maneuver (lifting, straining at stool, blowing or crying or singing). The pain is severe, bursting, explosive or of splitting quality lasting for a few seconds or minutes. The headache is usually bilateral, maximal at the vertex or in the occipital region and is not generally associated with nausea or vomiting and the neurological examination is normal. The paroxysms may be followed by dull, aching pain that may persist for hours.

In contrast to this, symptomatic cough headache could be precipitated by laughing, crying, weight-lifting or head postural changes. Symptomatic cough headaches can be caused by hind-brain abnormalities, posterior fossa meningiomas and Arnold Chiari malformation. This patient's MRI was done with a specific request to rule out a hindbrain compressive problem. As seen in Fig. 1, she has an Arnold Chiari malformation type 1 with cerebellar tonsillar ectopia. Here the cerebellar tonsils extend below the foramen magnum and may cause a functional valvular obstruction at that level. They are therefore associated with or neck pain which is worsened by impulse related features like laughing, coughing or stooping. Many of these patients benefit from decompressive surgery once the problem is identified on MRI. With a positive family history suggestive of migraine, it is easy to diagnose the second type of trigger-linked bearable headaches that she gets. For these headaches, routine migraine prophylaxis will work.

In conclusion this patient has symptomatic cough headache with an associated Chiari malformation type 1 on a background history of migraine. She has been started an antimigraine prophylaxis and Indomethacin for the short-lasting headaches but we can expect good results in her case only if she agrees to decompressive surgery. It is worthwhile remembering here that other causes of short-lasting headaches should be ruled out before coming to this conclusion.

Case No. 4

AI, M/25 consulted us at the Headache Clinic with complaints of recurrent headaches for 10 years prior. He gave a history of moderate to severe right periorbital head pain, that was gripping in nature, lasting for up to one hour and coming on a daily basis for 2-3 months in a year. The pain would subside spontaneously and then he could be alright for 8-9 months of the

year. Alongwith the intense head pain he also had redness and watering from the right eye, intermittent right nasal stuffiness, photophobia, phonophobia, osmophobia and during the attack he felt nauseated, would vomit and only then did he feel better.

The pain would build up gradually and radiate hemicranially to the neck and he would feel totally drained out . He would get relief during the attack if he sat still in one position and did not bend or move. The attacks would come on more at night and he is woken up by the pain 2 hours after sleep. He definitely would get the attacks on the day he has a stomach upset or eats a heavy meal. There is a positive family history of headaches in his grandmother and aunt. Clinical neurological examination was normal.

Imaging studies done have been normal. He has seen chiropractors , has taken feverfew but nothing seems to help. Interestingly whenever he consumes alcohol during the two months of his 'migraine season' as he calls it, he develops a severe headache and he jokingly comments 'the only good thing that this headache has done to me is to make me give up smoking and drinking', otherwise nobody seems to understand why my head pains!''.

With this despondency, he visited us hoping for some relief of his head pain.

Questions

1. What is your diagnosis ?
2. How would you treat this patient ?

Discussion

This patient has features of both cluster headache and

migraine during the attack. The following features are suggestive of Cluster Headache : the periodicity with remissions, the predominant nocturnal frequency with the attacks having a sense of timing and the waking up with headache 2 hrs after he goes to sleep, the persistent right periorbital + hemicranial location, the lacrimation, the conjunctival injection, the nasal stuffiness , the duration of less than one hour and the worsening with alcohol if taken during the cluster period. The features of migraine in this patient are: the presence of a prodrome and postdrome with nausea and a tendency to vomit, the intense photophobia, phonophobia and osmophobia, the desire to sit still during the attack and the significant dyspeptic symptoms during the attack.

When a patient presents with chronic recurrent headache with combined features of 2 different primary headaches, it can be more difficult to treat the patient. After ruling out structural problems by an MRI scan, you need to identify features of the headache that are more dominant and treat with prophylactics according to the dominance. What this patient has is a rare combination which would fit the label of 'Cluster Migraine Syndrome'. We started him on prophylaxis with verapamil, lithium, a small dose of steroids for cluster headache and flunarizine for his migraine. His attacks subsided very soon and he went into remission. He was advised Sumatriptan for his acute attacks but he did not need it. It is now one month that he has had no headaches.

Although a lot of research is ongoing in this area and PET and functional imaging and neuropeptide estimations are throwing up new data we still do not have the final answer. We need a revision of terminologies that will convey the extent of overlap.