CHAPTER 42

THE PATIENT WITH NECK PAIN AND HEADACHE

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Case History

This patient was a 21-year-old nurse when her headaches started. She reported that the headaches were precipitated by a period of work during which she assisted in a radiology department and had to wear lead aprons for some 4 to 5 hours per day, 4 days a week for about 1 month. She developed severe neck pain and headache, associated with a decreased range of movement of her neck. She was relieved of work for 2 weeks, but upon resuming work she had to stop after 10 days because the pain had become worse. She presented some 2 years after the onset of her pain.

On presentation, her pain was felt high in the right posterior cervical region and radiated into the right occipital region and orbit, and sometimes to the right shoulder, right arm, and forearm. The symptoms were mainly on the right but sometimes passed to the left. The quality of the pain was dull and aching. It was present daily and aggravated by lifting, turning her head, sudden movements of the head, getting into awkward positions, and by looking down. The pain was relieved somewhat by application of heat and by lying down. Irregularly, the pain was associated with tingling over the right periorbital region and cheek.

She reported an episode some 3 months prior to presenting in which she was craning forward and felt a "clunk" in the back of her neck, which was followed by spasm of her neck muscles and very severe pain which obliged her to take to her bed. The next morning she felt that her neck was still "out of place" and was very painful. This persisted for some days until, when she was picking up something from her lap, she lifted her head backward and again felt a "clunk," after which there was a dramatic relief of her pain.

She had no family history of illness. Her own medical history included appendicitis and conjunctivitis, both resolved. A systems review revealed no symptoms of concurrent illness. Neurologic examination revealed no sensory or motor deficits in the distribution of the cranial or spinal nerves. Specifically, there was no numbness or hyperesthesia in the distribution of the trigeminal nerve or C2 spinal nerve.

Questions about This Case

- What are your diagnostic considerations in this case?
- Why have her headaches continued?
- Do you wish to know more about this particular case; if so, what pertinent information would you like to know?
- How would you have investigated this patient's headaches, and what specific therapies would you suggest?
- What long-term management strategies would you suggest for her case?

Case Discussion

Several features in this case implicate the neck as the source of pain. Headache is the predominant complaint, but the patient also complains of neck pain. The orbital region is involved but the pain spreads from the posterior upper neck to the orbit, rather than arising in the orbit. Neck movements aggravate the pain, as do activities of daily living that involve moving or loading the neck, such as lifting.

The headache suffered by this patient lacks the periodicity and associated features of migraine. It lacks any vascular features. The relationship to neck movements and activities denies a diagnosis of chronic tension-type headache. The unilateral distribution of the headache and the lack of systemic illness or neurologic signs, and the lack of progression exclude intracranial diseases.

Prior to presenting, she had seen a variety of health professionals, each of whom inferred that the neck was involved, although none offered a specific diagnosis. Nevertheless, therapy was applied empirically to the neck. Treatment by a chiropractor did not help; manipulative therapy by three different physiotherapists gave the patient some intermittent relief, as did acupuncture; but nonsteroidal anti-inflammatory drugs afforded no particular benefit. The challenges in a case like this are whether the headache stems from the neck, and if so, from where exactly?

One of the manipulative therapists who saw the patient reported signs of abnormal joints at C2-3 and C3-4, consisting of abnormal quality of movement and abnormal end-feel, with reproduction of the patient's pain upon passive intersegmental motion. An orthopedic surgeon requested plain radiographs of her neck. These showed slight (grade 0.5) spondylolisthesis at C2-3 and C3-4, but this grade of translation is within the normal limits of motion at these levels.

Medical imaging is notoriously useless in determining a diagnosis of cervical headache. Tumors, infections, and inflammatory diseases of the cervical spine are rare causes of headache. In the vast majority of patients there is nothing that might possibly be evident on radiographs of the neck that either implicates or refutes a diagnosis of cervical headache. Spondylosis is not a cause of headache, because the same spondylotic changes are evident in the same proportion of patients as in asymptomatic individuals of the same age.

Manipulative therapists believe that they can pinpoint abnormal and symptomatic joints in the neck by careful manual examination. This has been validated in the case of one manipulative therapist but the results of that study cannot be generalized. It has not been shown that manipulative therapists at large are either reliable or valid in their claims of diagnostic acumen. Serendipitously, the manipulative therapist in the present case proved to be correct.

Diagnostic blocks are the only available, valid means of pinpointing a source of neck pain or cervical headache. Various joints and ligaments in the neck, that might be the source of the pain, are simply not palpable, but they are accessible to needles. Under radiographic control, needles can be used to anesthetize selected joints, ligaments, or nerves, in order to test if that structure is the source of the patient's pain, or in the case of nerves, if that nerve mediates the patient's pain.

At the request of her orthopedic surgeon, she underwent a diagnostic block of her right C2-3 zygapophysial joint. Upon injection of contrast medium into that joint her pain was aggravated, but upon injection of local anesthetic and corticosteroid, her pain was promptly relieved. This provided prima facie evidence that the C2-3 zygapophysial joint was the source of her headache, and this preliminary diagnosis was subsequently explored.

The C2-3 zygapophysial joint is innervated by the third occipital nerve. A right third-occipital-nerve block, using 0.5% bupivacaine, promptly and completely relieved her of her headache for several hours. Subsequently an intra-articular injection of 0.8 mL of a mixture of 1 mL 0.5% bupivacaine and 1 mL (5.7 mg) of betamethasone provided complete relief of her headache for some 12 weeks. A repeat injection of 0.5 mL of betamethasone alone resulted in a further period of 12 weeks of relief, on each of two subsequent occasions.

Although grateful for the quality of relief that these injections afforded, the patient wanted to obtain longer-lasting relief. She had been informed of the prospect of percutaneous radiofrequency neurotomy, and she wanted to pursue this.

A prognostic block of her right third occipital nerve was performed and, again, completely relieved her headache. On the basis of this response and her consistent response to third-occipital-nerve blocks and to intra-articular steroids, she was scheduled to undergo percutaneous radiofrequency neurotomy of her right third occipital nerve.

This was performed and she obtained complete relief of her headache. The relief lasted for some 9 months, but she suffered two side effects, each predicted from the prognostic blocks. She had numbness in the cutaneous territory of the third occipital nerve, which was not a problem, but she also had ataxia. This gave her trouble walking down stairs, but she could cope if she held the banister and looked straight ahead instead of at her feet. The ataxia and the numbness, however, resolved as her headache returned.

As the relief of her headache waned, she asked if she could have the neurotomy repeated. Told that if she had a repeat operation the ataxia would recur, and asked if she wanted to suffer that side effect again, she responded, "Are you kidding? I would swap these headaches for unsteadiness any day."

Ataxia is a regular side effect of third occipital neurotomy and occurs temporarily when that nerve is blocked with local anesthetics. It arises because of interference with tonic neck reflexes, ostensibly because of block of conduction in proprioceptive afferents from muscles innervated by the third occipital nerve.

Ten months after the initial neurotomy she underwent a repeat neurotomy, and regained complete pain relief. Eleven months later the pain recurred, and she underwent another repeat neurotomy. This afforded her complete relief again, this time lasting some 12 months.

Almost as regularly as clockwork she has enjoyed complete relief every year since, with pain recurring at about

12 months, only to be relieved by a repeat neurotomy. On February 22, 1997, she celebrated the seventh year of relief from her headaches.

During this period her life was rehabilitated. After having been unemployed and disabled for some 3 years, the pain relief allowed her first to complete a university degree, and maintain part-time employment. Upon graduating she was able to assume full-time employment, which she has maintained since. The only interruption has been the 1 day a year that she requires to have a repeat neurotomy, and 5 days' leave for recuperation. The one side effect that she has developed is addiction to the pain relief she has obtained. She cannot countenance going back to the way she was, if radiofrequency neurotomy was no longer to be available.

Management Strategies

- Patients with suspected cervical headache require a precision diagnosis.
- A diagnosis might be established using the skills of a well-trained manual therapist, but the validity and reliability of such skills in the community at large has not been established.
- Manual therapy might be tried, but there is little guarantee that it will work at all, or if it does seem to work, that it will have any profound or lasting effect.
- For many sources of cervical headache, diagnostic blocks are available for precision diagnosis. The putatively painful joint or its nerve supply can be selectively anesthetized under radiographic control. Complete relief of pain pinpoints the offending structure.
- Oral drug therapy is of no proven benefit for cervical headache, and has not even attracted anecdotal support.
- Intra-articular steroids are potentially of benefit for some patients, but most patients do not obtain lasting relief. Pain recurs within 1 to 2 weeks in more than 80% of patients.
- The only treatment known to have any lasting effect on cervical headache is radiofrequency neurotomy, but this procedure has not been proved in a controlled trial.

Case Summary

- This patient suffered cervical headache which was pain referred from her right C2-3 zygapophysial joint to her occiput and orbit.
- The pain was repeatedly and consistently relieved by local anesthetic blocks of her third occipital nerve or by intra-articular injections of a corticosteroid.
- This pain relief, however, was only temporary, lasting hours with local anesthetic and only weeks with steroids.
- Percutaneous radiofrequency third-occipital neurotomy abolished her headache for periods of up to 1 year and beyond.
- Despite side effects of numbness and ataxia, she craved and insisted on relief from her pain.
- As the effects of the operation wore off, so did her side effects.
- Repeat neurotomy has reinstated her relief regularly for 7 years.
- Relief of her pain has restored her life.

Overview of Cervical Headache

Diseases or injuries of the upper cervical spine can cause local pain and pain referred into the head. The referred pain is perceived in the occiput and may extend through the parietal region and into the frontal and orbital regions.

The anatomic basis of cervical headache appears to be convergence between cervical and trigeminal afferents in the trigeminocervical nucleus, which consists of the pars caudalis of the spinal nucleus of the trigeminal nerve and the dorsal horns of the first three cervical spinal cord segments. Exactly where the referred pain is felt depends on which nerves converge in this nucleus. Convergence between cervical joint afferents and the first division of the trigeminal nerve will result in referral to the frontal region and orbit. Convergence between joint afferents and fibers of the C2 spinal nerve will result in referral to the occiput.

Although the specific neuroanatomy has not been demonstrated, several clinical experiments in normal volunteers have demonstrated the capacity of the neck to refer pain into the head.

Campbell and Parsons and Feinstein et al. showed that stimulating the upper posterior neck muscles could produce headache in normal volunteers. Dwyer et al. show the same for the C2-3 zygapophysial joint, and Dreyfuss et al. for the atlanto-occipital and lateral atlantoaxial joints.

In patients with headache, Ehni and Benner reported temporary relief of pain following periarticular blocks of the lateral atlantoaxial joints, and McCormick reported relief following intra-articular blocks of these joints. Bogduk and Marsland reported relief following blocks of the C2-3 zygapophysial joints, later confirmed by Lord et al. using double-blind, controlled blocks.

Lord et al. established that in patients with chronic headache following whiplash, the headache could be traced to the C2-3 joint in some 50% of cases. Joints below C2-3 are uncommonly the source of headache, if at all.

Jull et al. reported that an expert manual therapist was capable of accurately diagnosing cervical zygapophysial joint pain. The diagnostic features of a symptomatic joint

were abnormal quality of movement, abnormal end-feel and reproduction of pain upon passive movement of the joint. These findings, however, have not been followed by subsequent studies to show that any other manual therapist is equally accurate.

Nevertheless, using Jull as the diagnostic instrument, Treleavan et al. found that many cases of so-called post-concussional headache could be traced to the C1-2 or C2-3 joints.

Ironically, cervical headache is one of the best defined types of headaches, yet the least respected. The definition of cervical headache and its diagnostic criteria are far more explicit and objective than those for tension-type headache, yet the latter entity is honoured far more than cervical headache.

The definition of cervicogenic headache proposed by the North American Cervicogenic Headache Society is that the headache is referred pain that stems from a structure innervated by one or other of the cervical spinal nerves. This definition expressly differentiates cervical headache from another type of headache, some of whose clinical features might be shared. But the diagnosis of cervical headache rests not on recognizing clinical features; it rests solely on demonstrating a cervical source of pain, by whatever means, provided that those means are valid. At present, controlled diagnostic blocks are the only valid means.

Opposition to the concept of cervical headache seems to stem largely from lack of familiarity with the concept, or lack of ability or reluctance to apply or obtain the necessary diagnostic procedures. Because the diagnosis cannot be established by conventional clinical examination, neurologists and others are not equipped or trained to make the diagnosis. For this reason, the entity has been embraced far more strongly by anesthesiologists in pain medicine, and by radiologists who have at their disposal the needle skills and imaging facilities required to pursue the diagnosis.

Although the earlier literature relied on uncontrolled diagnostic blocks of cervical joints or cervical nerves in the pursuit of cervical headache, controlled blocks are now mandatory. Controlled blocks guard against false-positive responses due to placebo effects. Controls are required in each and every case.

Placebo controls pose ethical and logistic problems. In most countries, impromptu, single-blind, placebo injections are unethical without informed consent. Therefore, if placebos are to be used, a series of three diagnostic blocks is required to make a diagnosis of cervical headache. The first block must be with an anesthetic agent in order first to establish that the target joint is, indeed, painful. Unless that is shown, it would be a waste of time and resources to complete the series on a joint that is not symptomatic. The second block cannot routinely be the placebo, for a mischievous patient would know that the second block was always the dummy. In order to maintain blinding and the element of chance, the second block must be either a placebo or an active agent. To complete the series the agent not used on the second occasion is administered as the third block. A positive response would be one in which the patient obtains complete relief of their headache whenever a local anesthetic was used, but no relief when normal saline was used.

A more practical form of control is a comparative local anesthetic block. This circumvents the ethical and logistic problems of placebo-controlled blocks. On separate occasions the diagnostic block is performed using different local-anesthetic agents. A positive response is deemed to have occurred if on each occasion the patient obtains complete relief of their pain but short-lasting relief when a short-acting agent is used, and long-lasting relief when a long-acting agent is used.

Lord et al. have shown that these criteria are robust against placebo-challenge; they are highly specific, but lack optimal sensitivity. Some patients who obtain complete relief but inordinately prolonged relief with lignocaine defy the diagnostic criteria of comparative blocks, but nonetheless do not respond to placebo. Such patients may fail to be included as positive if the diagnostic criteria for comparative blocks are rigidly applied.

Diagnostic blocks of many upper cervical structure are not difficult to perform. Therefore, in principle few possible sources of cervical headache can escape pursuit. What is required is the willingness to entertain these sources, and the skills and technical resources to pursue them. Blocks can be performed of the atlanto-occipital, lateral atlantoaxial, and zygapophysial joints, as well as the C2 and C3 spinal nerves. Only the median atlantoaxial joint is inaccessible for blocks.

What remains unknown is the extent epidemiologically to which cervical headaches stem from the atlanto-occipital, atlantoaxial, C2-3 zygapophysial joints and other cervical structures that are responsible for headaches. To date, the only index is that the C2-3 joint is a very common source of headache after whiplash.

The pathology of cervical headache is unknown. Presumably it arises as a result of some form of injury to one or other cervical structure. Frustratingly, contemporary medical imaging cannot resolve such injuries even though they may be present. Postmortem studies have shown that injuries to the atlantoaxial, atlanto-occipital, and cervical zygapophysial joints, evident on dissection, are invisible on radiographs of the specimen, even on retrospective review.

In the case described in this chapter, 1-mm CT scans were taken perpendicular to the surface of the C2-3 and

other zygapophysial joints. The joints at C2-3 were extremely narrowed, to less than 1 mm, implying virtually no articular cartilage. This feature was not evident at lower joints and intuitively must be abnormal for a 21-year-old woman. However, there are no data against which this intuition might be tested. Not only do we not know what the CT appearance is of painful zygapophysial joints, we do not even know what their normal appearance is at different ages. Until that normative data is to hand, CT scanning cannot be used to investigate the cause of zygapophysial joint pain.

Treatment of cervical headache remains at the frontier of medical research. No conventional therapy is known to work. There are no data on the utility of arthritis drugs, and no compelling data on physical therapy or manipulative therapy. No one has yet conducted a trial of any therapy for patients with proven cervical headache.

Intra-articular steroids might be countenanced as an upmarket medical intervention, but the study carried out by Barnsley et al. showed that few patients respond, and that steroids confer no benefit over local anesthetic alone.

Radiofrequency neurotomy remains an unproven treatment, and applicable only for cervical headache stemming from the C2-3 zygapophysial joint. Lord et al. found that radiofrequency neurotomy works for zygapophysial joint pain at levels below C2-3 for the treatment of neck pain, but they encountered technical difficulties when treating pain from C2-3 which limited application of the procedure. Whereas some patients responded outstandingly, as in the present case, others responded dismally. The flaw did not lie in the diagnosis; it lay in the operative technique. If the third occipital nerve is effectively coagulated, and remains coagulated, the patient reliably obtains relief. In essence, if they go numb, they lose their pain; but if the nerve is missed, and they do not go numb, their pain remains.

Current research is focused on perfecting the technique for third occipital neurotomy. For joints other than C2-3, some form of definitive therapy still needs to be conceived, let alone evaluated.

Selected Readings

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Editorial Comments

The relationship of headache to neck disorders and neck pain has long been a source of debate, with wide and varied views put forth by many individuals. Dr. Bogduk presents an important case, with a diagnosis and management strategy supported by his considerable knowledge and experience with similar cases. The concept of convergence of cervical and trigeminal pathways makes intuitive sense, both anatomically and clinically. Diagnostic and therapeutic blocks can be harder to accept, except in Dr. Bogduk's hands. There is much for the reader to learn from this case, whether you agree with the diagnosis and treatment or not. Even if one does not accept the entity of cervicogenic headache, that diagnostic possibility should be considered and investigated in cases such as these.