

VASCULAR NECK PAIN—A COMMON SYNDROME SELDOM RECOGNIZED

Analysis of 100 Consecutive Cases

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A YOUNG or middle-aged woman reports to her physician because of a sore throat, without fever or other constitutional manifestations, which may have been present for weeks or months. The patient believes that the glands in the neck are swollen. These "swollen glands" are said to act strangely: sometimes the swelling lasts only a few hours, sometimes it persists for weeks; it disappears mysteriously and recurs frequently. During the course of the physical examination the physician finds no abnormality, but when he re-examines the neck and follows the directions given by the patient, he finds a tender swelling that *could* be an inflamed lymph node.

The patient then is treated with a sulfonamide or with penicillin, and, when no relief ensues, a course of one of the broad-spectrum antibiotics is administered. This therapeutic program also is unsuccessful, and the harried physician begins to think of other possibilities. Since the patient has "swollen glands," feels weak, tired, and run-down, and antimicrobial therapy has not helped, a diagnosis of infectious mononucleosis may be considered. But, results of a heterophil antibody test are negative, and the diagnosis is changed to possible viral infection.

After several weeks or months of having diagnoses changed, the nervous patient can sense that her physician is uncertain, and she begins to worry about the looming possibility of cancer. She keeps poking in the region of the soreness, and the area becomes even more tender. In desperation, further investigations are carried out and nothing definitely abnormal is found. Teeth are removed or the tonsils are excised; and sometimes the pain disappears for a while, only to return and persist for days or weeks. Occasionally the thyroid gland is suspect, and a diagnosis of subacute thyroiditis is made; however, treatment with roentgen rays or steroids gives no relief.

This patient has vascular neck pain, a condition analogous to extracranial vascular headache. The syndrome of vascular neck pain is not due to structural change; it is benign and reversible; it is as real as migraine headache. There is no inflammation or adenopathy. What is so often described as a swollen gland is merely a tender, distended segment of the carotid artery. This distention is a manifestation of dysfunction of the autonomic nervous system, and is closely related to the painful vasodilating syndromes of the face and the head. Physicians are well aware of the nuchal radiation that occurs during an attack of migraine, when pain commonly radiates to the anterior portion of the neck, and tenderness develops in the carotid area. It is when the vascular neck pain occurs without headache that there is so much diagnostic difficulty.

Watson and Thorn¹ in 1952 listed 43 causes of neck pain, and discussed the differential diagnosis in considerable detail. They recognized only organic conditions, many of them rare, and did not mention the extremely common functional neck pains of carotid or muscular origin. Fay² coined the term "carotidynia" in 1927, but he used it in describing the tenderness of the carotid artery in association with particular forms of atypical facial neuralgia. Hilger,³ in 1949, in discussing the general aspects of carotid pain, apparently first described the syndrome of vascular neck pain: "The patient usually complains of the recurrent and abrupt onset of unilateral sore throat with the appearance of a 'gland' in the neck. There is no evidence of inflammation. The bifurcation is palpable as a firm, extremely tender mass at the hyoid level. It pulsates—an important diagnostic point. Usually the tonsils have been removed in previous efforts to alleviate the recurrences of sore throat." Unfortunately, little attention has been paid to this succinct paragraph, and the syndrome of carotid neck pain remains unrecognized generally.

Wolff⁴ has stressed the interrelationship of the various vascular pains in the head, and considers the underlying mechanisms to be the same in each clinical pattern. Thus, a vasodilating hemicrania preceded by visual scotomata and associated with nausea and vomiting is called migraine. A vasodilating headache without all of the typical characteristics of migraine may be called atypical migraine or tension vascular headache. The vascular headache occurring with hypertension, but not directly related to it, is another variant. Histaminic cephalalgia, described by Horton, MacLean, and Craig⁵ in 1939, was thought to be a unique form of vascular headache related to hypersensitivity to histamine, or to an overproduction of histamine; they outlined a method of treatment based on desensitization to histamine. After extensive clinical trials, most observers have concluded that such desensitization is not a specific therapy for histaminic cephalalgia. This headache is now commonly regarded as a variant of migraine. For many years there has been confusion in the classification of atypical facial pain—and the syndromes of atypical facial neuralgia, sphenopalatine ganglion neuralgia, and vidian neuralgia were described. There is considerable evidence that all of these are vascular pains of the face, in which there is involvement of the facial branches of the external carotid artery, chiefly of the maxillary branch. When the distention and relaxation affect the carotid trunk alone, the syndrome of vascular neck pain results.

Clinical Characteristics

Vascular neck pain has been observed in children as young as 10 years and in adults as old as 75 years of age. It occurs more commonly in females than in males, the ratio being about 4 to 1. A carefully taken family history almost always indicates that one of the grandparents, parents, or siblings has suffered from one of the vascular syndromes. The patient's history is significant; many times migraine or other vascular headaches have occurred. Headache may have alternated with the bouts of neck pain, but usually the headaches become less frequent after the neck pain develops.

Usually the patient's temperament is similar to that commonly described for the person who suffers from migraine, namely, sensitive, conscientious, and perfectionistic. He is likely to be rigid, reserved, and strives to be well thought of. The neck pain may occur in a setting of fatigue, frustration, or tension; but also it may occur, as does migraine, with relaxation after a period of stress. A week-end neckache is just as diagnostic of vascular neck pain as a week-end headache is of vascular headache. The neckache is dull and feels deep-seated, and usually is not throbbing, but a throbbing component can be evoked by stooping, bending, or straining.

The discomfort is difficult to locate exactly, and though it often is described as being a sore throat, close questioning will elicit the fact that basically it is a sore neck. The thyroid gland is not tender. Usually the pain is centered about the carotid bulb and radiates upward along the course of the external carotid artery, behind the mandible, and up into the postauricular area. At times, the patient has a mild earache on the affected side. Most often the pain is unilateral, but it may move from one carotid area to the other and become bilateral. The pain in the region of the carotid artery is similar to that produced in cervical adenitis, so that confusion in diagnosis often occurs.

Usually the pain is not severe and it does not strike with the force noted in some types of vascular headache. There may be mild nausea but no vomiting. In general, the duration is longer than that of vascular headache; an almost continuous aching for from two to eight weeks is usual. The pain engenders a considerable emotional reaction. Since many of the patients with vascular neckache have a worrisome nature and are rather sensitive, they intuitively know when the physician is groping for a diagnosis and an effective method of treatment. Their intuitions give rise to more apprehension and worry; often cancerophobia develops.

Physical Findings

The findings on physical examination are normal except for the presence of a tender, throbbing carotid artery on the painful side. The tenderness is greatest at the carotid bifurcation or under the mandible. Of particular significance is the absence of injection of the pharyngeal mucous membrane; the tonsils are not inflamed; and there is no exudate. Adenitis is absent. The sternocleidomastoid muscle often is tender. This tenderness most likely is due to reflex muscle spasm. A tender carotid artery is easily demonstrated by pressing it posteriorly against the transverse processes of the cervical vertebrae and rolling the vessel under the fingers. All carotid arteries are somewhat tender, but these affected arteries are unduly so. The dilatation may be extreme, and at times may suggest a diagnosis of carotid aneurysm or carotid body tumor.

Often there are other evidences of dysfunction of the autonomic nervous system. Motion sickness and vertigo are more prevalent in persons afflicted with vascular pain than in the general population.

Analysis of a Series of 100 Cases

The Cleveland Clinic records of 100 consecutive patients with diagnoses of vascular neck pain (1954 through 1956) were analyzed. There were 82 females and 18 males; 62 patients had a history of previous vascular headache; 45 patients volunteered the information that the glands in the neck had been swollen. At the time of the examination, 73 of the 100 patients had tenderness of one or of both carotid arteries. The 27 who did not have demonstrable tenderness were not having vascular neck pain at the time of examination.

Age. The age range of the 100 patients was from 10 to 79 years; most of the patients were in the fifth decade of life. The age distribution of the 100 patients was: 10 to 19 years (1 patient), 20 to 29 years (17 patients), 30 to 39 years (21 patients), 40 to 49 years (31 patients), 50 to 59 years (19 patients), 60 to 69 years (9 patients), 70 to 79 years (2 patients).

Previous diagnoses. Although more than half of the patients had sought previous medical care, in no case had the correct diagnosis been made. Eleven patients had been told the neck pain was due to tonsillitis or infected throat. For six patients, the pain had been thought to result from carotid tumor or aneurysm, and three of those patients were referred for consideration of vascular surgery. Six patients were told they had thyroid disease, four of whom were referred directly to a thyroid surgeon. Infectious mononucleosis was the diagnosis for two patients. The others were given no definite diagnosis, or could not recall what they had been told.

Previous therapy. Courses of antibiotics had been given to 13 patients; 9 had dental extractions; 2 underwent subtotal thyroidectomy; 2 received a course of cortisone; and 1 patient had the jawbone scraped. Since the patients were still having neck pains, it can be assumed that treatment was not successful.

Treatment

Basically, the treatment for vascular neck pain is the same as the treatment for the other painful vasodilating syndromes of the head. Since the severity of the pain is less than in migraine or histaminic cephalalgia, and since the underlying neurosis (if any) is milder than that occurring in patients with vascular facial pain, the result of treatment of vascular neck pain is more satisfactory than that of migraine, histaminic cephalalgia, or vascular facial pain.

Because pain is a subjective phenomenon, and the occurrence of vascular pain is periodic and unpredictable, it is difficult to evaluate the result of a particular therapeutic agent in this group of conditions. The best therapists are enthusiasts; however, it is this same enthusiasm that when expressed in medical writing, often results in uncritical evaluation. The fact that more than 400 ways of "curing" vascular headache have been reported in the medical literature, indicates that no really successful therapy has been discovered. The study of any specific treatment undertaken without proper control, and evaluated without careful follow-up of at least two years' duration would only add to the confusion.

The following discussion of treatment used in the 100 cases of vascular neck pain offers no new therapeutic agent, but it summarizes a satisfactory program for handling the problem.

Psychologic treatment. Treatment begins with a painstakingly careful physical examination. Consultations must be obtained when necessary, but the diagnosis is made by utilizing positive evidence and not merely by the elimination of the possibility of organic disease. In a benign condition such as vascular neck pain, especially when the pain is not severe, reassurance is sometimes all that is needed. Many patients seek medical aid not because of the aching neck, but because of the fear engendered by the aching. Therefore the diagnosis must be made in a positive manner, and a thorough explanation of the mechanism of the pain must be offered to the patient. Often when the fear of serious disease is removed, the discomfort can be tolerated, although sometimes simple analgesics are needed for control of the pain. When the neck pain is but one manifestation of a severe underlying neurosis, intensive psychotherapy is required.

Drug therapy. In general, the medications used in the treatment of vascular neck pain are the same as those used for the control of vascular headache. These are administered to relieve pain, to prevent attacks, and they comprise four main groups: (1) ergotamine tartrate in its various forms; (2) analgesics; (3) sedatives; and (4) drugs that affect mood, such as the tranquilizers and amphetamines. The pharmacologic treatment is divided into two phases: relief of pain, and prevention of subsequent attacks.

To relieve pain, the simplest effective analgesic should be used. Sometimes aspirin is sufficient. Usually, mixtures of aspirin, phenacetin, and caffeine are efficacious. These mixtures with the addition of codeine give even greater analgesic effect. Darvon,* 32 mg., added to an aspirin-phenacetin-caffeine mixture works satisfactorily in many cases. Ergotamine tartrate is best administered orally in the form of Cafergot† or Cafergot P-B tablets. Since vascular neck pain does not occur in short, severe attacks, the Cafergot dosage is somewhat different from that used in the treatment of vascular headache. Four tablets per day may be given for several days, if necessary. It is best to use an analgesic tablet in conjunction with the ergotamine tartrate. Since there is little nausea and no vomiting associated with vascular neck pain, the parenteral use of ergotamine tartrate is unnecessary.

To prevent attacks, if the periods of neck pain have been frequent or if the aching has persisted for a long time, once the pain has been brought under control, a prophylactic program may be initiated. Often, 4 Bellergal‡ tablets per day, for from four to six weeks, will prevent recurrence. To simplify the administration of Bellergal, the long-acting tablets (Spacetabs) can be used: 1 tablet is taken on arising and 1 tablet again at bedtime. Four Benadryl§ capsules (each capsule 50

* Darvon (dextro propoxyphene hydrochloride), Eli Lilly and Company.

† Cafergot, Sandoz Pharmaceuticals.

‡ Bellergal, Sandoz Pharmaceuticals.

§ Benadryl hydrochloride (diphenhydramine hydrochloride), Parke, Davis & Company.

mg.) per day may be used for this purpose. Benadryl seems to be more beneficial than are the other antihistaminics, probably because of its sedative or tranquilizing action. In some instances, one of the tranquilizing agents may be administered for several weeks or months to prevent further attacks.

No surgical treatment. Surgical treatment is not recommended. Unnecessary elective procedures, such as thyroidectomy, tonsillectomy, and tooth extraction, should be avoided.

Physical therapy. Prolonged vascular neck pain produces reflex muscle tension that results in soreness of the sternocleidomastoid or posterior nuchal muscles. The application of heat and the use of massage may relieve the secondary muscular soreness in some cases.

Total treatment. A combination of the psychologic and the pharmacologic treatment is most commonly used. A detailed explanation of the origin of the pain and what is known about the mechanism of vascular pain is presented in a simplified form to the patient. The presence of a hypothetic chemical substance that is responsible for overdilatation and relaxation of the carotid artery is postulated, and the possible effects of nervous tension and emotion in the production of this hypothetic vasodilating substance is pointed out. The patient is at least reassured that the pain is not imaginary and that something is known of the cause. The patient nearly always fears the presence of cancer, and sufficient time must be taken to give strong reassurance and to explain why the condition could not be due to a malignant tumor.

A prophylactic medical program is outlined. This has many variations, but one of the most satisfactory combinations has been the administration of one Bellergal Spacetab each morning and at bedtime, along with three Compazine* (prochlorperazine) tablets (10 mg., each tablet) per day. For the pain itself, one of the simple analgesic mixtures is prescribed. If codeine is deemed necessary, the patient should be reassured that a few analgesic one-half-grain codeine tablets will not cause addiction. The patient is advised to take the medication for six weeks and then to return for examination if relief is not obtained. Morphine or meperidine hydrochloride never should be used to control vascular neck pain.

Results

After a follow-up period of at least one year, the results of therapy may be grouped in the following four categories relative to the original vascular neck pain: (1) the pain is much less severe or entirely absent; (2) the pain is about the same, but the patient no longer worries about it and is able to tolerate it; (3) the pain is unchanged; the patient complains bitterly; he either has not accepted or has not understood the explanation, and he has seen several other doctors in his search of another diagnosis; and (4) the neck pain has been supplanted by vascular headache. Fortunately, the results in most of the patients who have been seen at least one year after the initial examination, fall into categories 1 and 2.

*Compazine, Smith Kline & French Laboratories.

Report of Illustrative Cases

Case 1. A 41-year-old housewife was examined here because of sore throat. Ten years previously she was examined here and the diagnosis was chronic nervous exhaustion. The patient stated at the time of the second examination that all her life she had been having periodic sick headaches accompanied by some soreness in the neck. However, during the past year she had frequent bouts of sore throat and no headache, each episode lasting for a few days or a few weeks. The pain was located chiefly in the left side of the neck and was aggravated by bending, jarring, and nervous tension. At the time of the first attack of sore throat, she was treated by her family physician for a cold, and was given a course of penicillin. She was next told she had a goiter and was referred to a surgeon, who found no enlargement of the thyroid gland, and who diagnosed the condition as chronic tonsillitis. A consulting otolaryngologist disagreed with the diagnosis of tonsillitis and treated her for allergy.

General physical examination here revealed normal findings except for the presence of a tender carotid artery on the left side; the thyroid gland was not enlarged; the tonsils were small and not inflamed, and there was no evidence of organic disease of the ears, the nose, the throat, or the sinuses. Laboratory findings were normal. The diagnosis was: chronic nervous tension state and vascular neck pain.

Case 2. A 28-year-old man, a crime investigator for the Bureau of Internal Revenue, was examined here because of headache and swollen glands. An intelligent, conscientious, tense, high-strung person, he was working at a job he thoroughly disliked, since he was temperamentally unsuited for the task of investigating and prosecuting tax evaders. He had feelings of guilt about his work.

The history indicated that the patient had been in good health until three years previously, when he began to have swelling and soreness of the "submaxillary glands," usually of one gland at a time, but sometimes of both glands. The pain and soreness would extend into the jaws and the side of the face. Each time this difficulty persisted for about three months; there was one pain-free interval of 14 months. During those three years, the patient was examined by many otolaryngologists and other physicians, and various diagnoses were suggested. Although a diagnosis of anxiety neurosis had been made, no one could explain the mechanism of the pain in the neck and face associated with the "swollen glands." More recently a diagnosis of tic douloureux had been considered, and the patient was treated with injections of alcohol but no relief was obtained.

The attacks of headache lasted for from one day to one week. The onset of the headache had caused great apprehension, and it was obvious that the young man had read published details of subarachnoid hemorrhage due to ruptured cerebral aneurysm.

When examined here, in addition to having the neck and face pain of long standing, the patient also had headache in the right occipital, temporal, and frontal areas, associated with conjunctival injection of recent onset. The patient was disgruntled, apprehensive, and overconcerned. The right carotid artery was tender, dilated, and throbbing; other physical findings were normal, as were the results of all laboratory tests and the roentgenograms. A neurosurgeon and an otolaryngologist found no evidence of organic disease. The diagnosis was chronic anxiety tension state with vascular neck pain, and associated vascular head and facial pain of recent occurrence.

The patient's insight was poor and treatment was unsuccessful. One and one-half years after the examination, the patient stated that further medical reading had enabled

him to arrive at his own diagnosis of "temporal arteritis," and he was seeking a physician who was adept at treating this condition.

Case 3. A 40-year-old housewife was examined here because of aching in the throat which had persisted for one year. Ten years before the onset of pain, shortly after the birth of her only child, she had a nervous breakdown and was confined to a rest home for two months. After she was discharged she still was nervous and rundown. Her mother had been subject to sick headaches. The patient herself had periodic headaches for many years. The soreness in the neck was located in the carotid area, chiefly on the right side, the patient stating that the soreness felt as though the glands were swollen. The pain and tenderness appeared once or twice each week for about one day, accompanied by a feeling of tightness and choking in the throat. A diagnosis of thyroid dysfunction had been made and for three months each day she took orally three 50-mg. capsules of propylthiouracil. General physical findings were essentially normal; the thyroid gland was palpable, but neither enlarged nor tender, although there was a tender, throbbing carotid artery on the right side. An endocrinologist found no hyperthyroidism or subacute thyroiditis; an otolaryngologist ruled out the presence of organic disease of the ears, the nose, and the throat. A diagnosis of chronic anxiety tension state with vascular neck pain was made.

Comment

The syndrome of vascular neck pain is certainly not new, and, like headache, must be as old as civilization itself. Although it may have been recognized by a few physicians, little has been written about it, and the concept of "vascular headache in the neck" has not been widely disseminated. Although the underlying mechanisms of all the painful vasodilating syndromes of the head and neck probably are the same, the clinical patterns are so different that the division into various groups is logical and of practical value. However, despite the compartmentation of this group into migraine, tension vascular headache, histaminic cephalalgia, hypertensive migraine syndrome, vascular facial pain, and now vascular neck pain, some of the clinical variants are difficult to classify. Thus, whereas a headache with the typical features of migraine or of histaminic cephalalgia is easy to label, there are some head pains that have the characteristics both of migraine and of histaminic cephalalgia. The headache appearing in the presence of benign hypertension may be indistinguishable from migraine, it may have the pattern of an occipital throbbing headache occurring early in the morning, or it may take the form of vascular facial pain. This lack of a single specific form should not disconcert the clinician, since a mixed or changing pattern offers additional evidence of the basic interrelationship.

A carefully taken history often will indicate that the clinical pattern has changed during the years. When migraine does not entirely disappear in the older patient, it often continues in a form that is similar to tension vascular headache, or in the form of typical "hypertensive" headache, even though hypertension is not present. The carotid artery is almost always sore and tender during the course of a migraine attack, vascular facial pain, or tension vascular headache. It is only when the neck pain is unaccompanied by head or facial pain that the diagnosis is not suspected.

Summary

One of the most common causes of pain in the neck is related to overdistention, relaxation, and increased pulsation of the carotid artery. The syndrome of vascular neck pain is closely related to the various forms of extracranial vascular headache. Vascular neck pain has been referred to only occasionally, and apparently never has been described in detail in the medical literature. Most physicians have not made this diagnosis, although the condition is not rare.

Analysis of 100 consecutive cases of vascular neck pain has revealed that it is more common in women than in men in a ratio of about 4 to 1; it occurs at any age, but mostly in the fourth and fifth decades; and usually there is a history of previous vascular headache. The only abnormality revealed by physical examination is the presence of a tender, throbbing carotid artery. The condition is frequently misdiagnosed and therefore is not properly treated. The preferred treatment is similar to that for the other painful vasodilating conditions of the head.

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