

Carotidynia: A Pain Syndrome

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Carotidynia is a common neck pain syndrome first described by Temple Fay in 1927. The pain is typically dull, throbbing, continuous, and localized over the carotid bifurcation, but may radiate to the ipsilateral mandible, cheek, eye, or ear. Symptoms are frequently aggravated by swallowing, chewing, and contralateral head movements. The cardinal physical finding is tenderness

on palpation of the carotid bulb, sometimes accompanied by prominence or throbbing of the carotid pulse. Although several serious conditions should be excluded, most cases follow a benign course.

Key words. Carotidynia; migraine; carotid arteries; arteritis; temporal arteritis. (*J Fam Pract* 1994; 39:71-75)

Carotidynia is a symptom of unilateral vascular neck pain which was first described by Temple Fay in 1927.¹ The cardinal physical finding is tenderness on palpation of the carotid artery. The incidence and prevalence of carotidynia are unknown. Most authors agree that it is common but frequently unrecognized. It may be two or three times as common as cluster headaches.²

Lovshin³ reported a series of 100 cases in patients who ranged in age from 10 to 78 years with a median age of 55 years. Female patients were affected four times as frequently as male patients. In Lovshin's series, as in many others, no serious organic conditions were found among patients presenting with carotidynia, and all were treated symptomatically.

We present three case reports, followed by the diagnosis, possible causes, and various treatment modalities for this syndrome.

Case Reports

Case 1. A 42-year-old man presented with a history of pain in the left side of his neck for the previous 2 weeks. He was in normal health except for a history of migraine headaches, which had occurred once or twice a year since he

had been 18 years old. The patient indicated that his pain was moderate in intensity, transmitted slightly to the left ear, and was exacerbated by swallowing. His most recent headache had occurred approximately 1 month before the onset of the neck pain. No abnormalities were found on examination of the ears, nose, mouth, and throat. Indirect laryngoscopy was normal. Although palpation of the neck revealed no abnormal lymph nodes, organ enlargement, masses, or deformities, the left carotid artery was moderately tender just below the bifurcation. The patient was treated for 2 weeks with a nonsteroidal anti-inflammatory agent. The pain gradually diminished and resolved, although he had mild recurrences for several months.

Case 2. A 30-year-old female patient presented to the clinic with a 2-week history of persistent pain and fullness in her face. She had a history of mild episodes of sinusitis and allergic rhinitis. Her vital signs were: pulse 64 beats per minute, blood pressure 130/90 mm Hg, temperature 97.4°F, and weight 121 lb. Physical examination showed normal tympanic membranes. Her nasal mucosa was edematous and red, and the area over both maxillary sinuses was tender to percussion. Neck, chest, cardiovascular, and abdominal examinations were all normal. She was treated with amoxicillin 500 mg three times a day for 14 days and a combination antihistamine and decongestant.

She returned to the clinic 1 month later complaining of persistent pain in the left side of her neck and in her left ear. The sinusitis had subjectively resolved by this time. Vital signs were: pulse 88 beats per minute, blood pressure 120/70 mm Hg, and temperature 96.0°F. On ex-

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amination, the tympanic membranes were found to be clear. There was no sinus tenderness to percussion. Nose, throat, and oral examinations were normal. Her neck was supple with no adenopathy, organomegaly, or masses. A tender, throbbing carotid artery on the left side was noted. The remainder of the examination was normal. She was treated with a nonsteroidal anti-inflammatory agent, and the pain resolved during the next 1 to 2 weeks.

Case 3. A 36-year-old woman presented to the emergency department with a 3-day history of sore throat, fever, stiff neck, and right-sided headache extending from behind the angle of the jaw to the entire right hemicrania.

Since the age of 18 years, the patient had experienced episodes of severe, usually unilateral pounding headaches, which were accompanied by vomiting and sometimes preceded by scintillating contralateral visual scotomata. The headaches frequently resolved with sleep.

The patient's temperature was 104°F. She appeared to be in pain and was photophobic. The throat was deeply hyperemic and the tongue thickly coated. The right carotid bulb pulsed prominently and was exquisitely tender to touch. The sternomastoids and cervical trapezius were moderately tense and tender, and flexion of the neck was limited to 45°. There was no adenopathy or thyromegaly. The neurological examination was unremarkable.

The white blood count was 8200/ μ L, with 30 segmented neutrophils, 47 polymorphous nuclear leukocytes, 19 lymphocytes, and 4 monocytes. Spinal fluid protein was 31 mg/dL, glucose 77 mg/dL, and cell count 7 mononuclears/ μ L. The sedimentation rate was 63. A repeat sedimentation rate at 2 weeks follow-up was 13. All cultures and other laboratory tests were negative.

The patient was treated with intravenous antibiotics and narcotic analgesics. The sore throat and fever gradually resolved during the ensuing 5 days. The headache and neck pain resolved with analgesia within the first 48 hours, but an exacerbation occurred on the day before discharge, which again resolved with symptomatic treatment. She continues to suffer from frequent episodes of pain that involve either or both sides of the head or neck and often require narcotic analgesics.

Discussion

Clinical Characteristics

The pain of carotidynia is most often unilateral and localized to the neck, although radiation to the face, ear, or malar region is not uncommon. It is frequently described as dull and throbbing in character and continuously present, although day-to-day or hour-to-hour exacerbations and remissions are common. Severity varies from

Table. Differential Diagnosis of Unilateral Neck Pain

Carotidynia
Acute pharyngitis
Peritonsillar abscess
Dental disease
Temporomandibular joint syndrome
Lymphadenitis
Submandibular gland disease
Myositis/myalgia
Histamine cephalgia
Sinusitis
Thyroiditis
Tumor of tongue, salivary gland, or larynx
Neuralgia

mild to agonizing, and the pain is frequently aggravated by swallowing, yawning, coughing, sneezing, or elevating the head while moving it toward the contralateral side. A history of migraine may be present (eg, the first and third cases). In other cases, a history of previous pharyngitis, tonsillitis, upper respiratory tract infection, oral conditions, or recent dental procedures is reported (eg, the second and third cases). Anxiety or fear of cancer is often present.^{3,4}

Physical examination is usually normal except for mild to severe tenderness and sometimes prominent pulsing at the carotid artery bifurcation. Tenderness has also been reported over the proximal 6 cm of the internal carotid or the facial artery. Fay's sign was originally described as follows: "If the thumbs are placed on the common carotid artery just below the bifurcation, and the structures pressed back against the transverse cervical processes with a rolling movement, a severe reaction of pain is produced on the side of the atypical neuralgia. This response I have termed 'carotidynia.' . . ."⁵

Differential Diagnosis

Many other conditions, some potentially serious, can cause unilateral neck pain (Table). Most of these conditions may be excluded by a careful examination of the head and neck, supplemented when indicated by laboratory or radiology tests. Acute pharyngitis, peritonsillar abscess, acute sinusitis, dental abscess, temporomandibular joint syndrome, and cancer of the tongue, salivary glands, or larynx can be excluded by a combination of observation, palpation, and percussion of the structures of the mouth, face, and throat accompanied by direct or indirect laryngoscopy.

The pain in Eagle's syndrome⁶ (facial pain caused by an elongated styloid process) can be reproduced by finger pressure along the base of the tongue. The tenderness of thyroiditis and tracheitis are located more centrally and symmetrically in the neck. It is important to exclude an-

terior cervical adenopathy by palpation, as this is probably the most common incorrect diagnosis. Cervical muscle spasm can be excluded by palpation of the muscles involved. The distinctive patterns of radiation of cervical disc disease and degenerative cervical arthritis coupled with the absence of Fay's sign should direct attention to these conditions.

Pathogenesis and Origin

Carotidynia is a symptom based on a characteristic complaint accompanied by a unique physical finding. The International Headache Society classifications include "carotidynia (idiopathic)."⁷ However, in the literature, carotidynia is not described as a clinical entity in and of itself, but rather as a symptom of another process. We concur with the authors of *The Headaches*⁸ that "idiopathic carotidynia has not been described and that it is a syndrome of neck pain." Since it is not a disease, there are several possible causes.

A local process involving the carotid artery appears to cause local pain and tenderness as well as referred pain to the mandible, face, eyes, ear, and head. The pain is produced by stimulation of the nerve endings of the carotid plexus as described by Fay.^{1,5} He reproduced the pain's characteristic radiation patterns by electrical stimulation (Figure), and based on his observations of patients undergoing nerve resection for intractable pain, deduced that the vagus nerve and the cervicothoracic spinal nerve roots must be the neurological pathways to the brain.

Reported causes of carotidynia include migraine,⁹ viral or postinfection,¹⁰ giant cell (temporal) arteritis,¹¹ carotid artery dissection,¹² carotid artery aneurysm,¹³ and total carotid artery occlusion.¹⁴ The first two causes are the most common and the most benign. Treatment and prognosis depend on the cause. Physicians can distinguish benign causes from those that are serious by inquiring about neurological symptoms, performing a neurological examination, and considering the results of a sedimentation rate. Unusual or suspect presentations may require further laboratory or imaging studies.

MIGRAINE

The most commonly ascribed cause is migraine. Lovshin³ noted a high incidence of vascular headache in both family and personal histories before the onset of carotidynia. He also observed that many of his patients had a "migraine personality" (sensitive, conscientious, and compulsive). His patients tended to suffer most from carotidynia when fatigued, frustrated, or under stress, but also during periods of relaxation after stress. Raskin and Prusiner⁹ made the important observation that Fay's sign is positive in

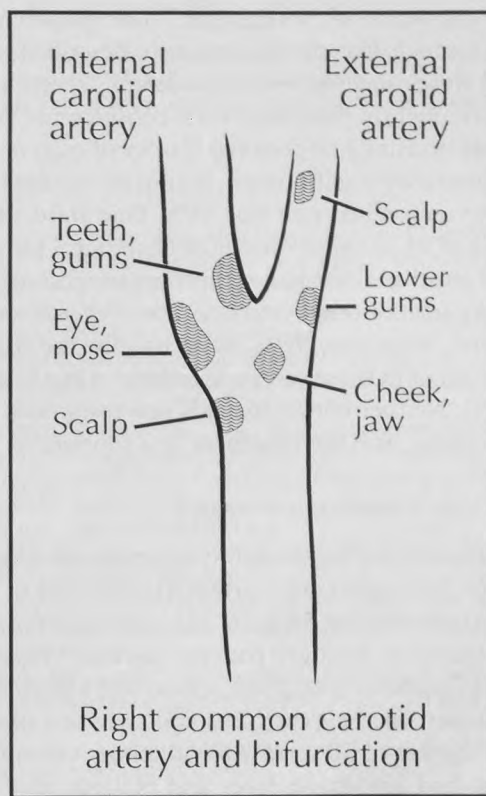


Figure. Right common carotid artery and bifurcation showing radiation of pain by electrical stimulation. Adapted, with permission, from Fay T. Atypical facial neuralgia, a syndrome of vascular pain. *Ann Otol Rhinol Laryngol* 1932; 41:1030-62.

more than 50% of patients experiencing at least one vascular headache per week. The association is so striking as to lead one author to advocate that "carotidynia is such a frequent accompaniment of migraine that part of the routine examination of these patients should include palpation of the neck."¹⁵

Carotidynia associated with migraine tends to recur over periods of months to years with exacerbations sometimes lasting up to several weeks. The pain in these cases is generally described as dull and deep-seated, aggravated by swallowing, stooping, or straining, and accompanied by a mild earache.^{2,9} The pain in this variant of carotidynia usually responds to drugs commonly prescribed for migraine, such as ergonovine, tricyclic antidepressants, beta blockers, calcium channel blockers, or methysergide.

INFECTION OR POSTINFECTION

Carotidynia is also known to follow cases of pharyngitis, infected aphthous ulcers, and viral upper respiratory tract infection. Roseman¹⁰ reported a series of 33 younger patients (mean age, 37 years) with first attacks of carotidynia. The patients were equally divided between male and female patients. These patients frequently presented

with the complaint of "sore throat." The symptoms were otherwise much like those previously described, except that they were usually severe or moderately severe and the patients frequently presented with considerable anxiety. There was no family or personal history of migraine, and the symptoms were self-limited, lasting an average of 11.6 days. The recurrence rate was 10%. One third of these patients had objective evidence of pharyngeal inflammation and another third had symptoms suggesting upper respiratory tract or other viral infection. Fever was usually absent and laboratory tests were usually normal. The causative agent in these cases was unknown but suspected to be viral. Neither corticosteroids nor antibiotics modified the course, and the treatment was supportive.

GIANT CELL (TEMPORAL) ARTERITIS

Carotid arteritis is a less likely but more serious diagnostic possibility. Although cranial arteritis occurs almost exclusively in patients over 55 years old, carotid arteritis frequently occurs in younger patients. Swann¹⁶ reported a series of 17 patients with giant cell carotid arteritis, three of whom were in their early 20s and only one of whom was over the age of 50 years. All had tender carotid arteries, many had low-grade fever and malaise, and all responded promptly to corticosteroids. Two other case reports described patients with this variant of giant cell arteritis who were younger than 50 years of age.^{15,17}

Most cases of giant cell (temporal) arteritis resolve spontaneously within 2 to 30 months. Neurological damage, including visual loss, is usually permanent, and if untreated, has an overall mortality rate of 12%. Therefore, an erythrocyte sedimentation rate is mandatory for all patients presenting with carotidynia regardless of age. Corticosteroid therapy is the treatment of choice. The diagnosis can sometimes be confirmed by biopsy, but temporal artery biopsy is usually negative. Paulley and Hughes¹⁸ favor facial artery biopsy if needed to confirm the diagnosis.

CAROTID ARTERY ANEURYSM

Chambers and colleagues¹⁵ reported two patients with carotid aneurysms who presented with carotidynia. Their review of the literature up to that time (1981) noted that in 39 of 113 previously reported cases of carotid aneurysm, the patients had suffered from head and neck pain. They cited Countee's observation that carotidynia was "the most common presentation" of carotid aneurysm.¹³

A carotid artery aneurysm can be located anywhere along the common or internal carotid arteries and may manifest solely as a tender carotid artery. A mass may or may not be palpable, and even if present, it may not be pulsatile. Arteriography is the most reliable means of

reaching a definitive diagnosis. Although arteriosclerosis and trauma are the most common causes, Marfan's syndrome, cystic medial necrosis, ionizing radiation, pyogenic infections, previous carotid artery surgery, congenital defects, and syphilis are also associated. Although rupture of the artery is unusual, neurological complications such as stroke, amaurosis fugax, transient cerebral ischemia, syncope, and coma are possible. Neurological signs and symptoms indicate the need for further diagnostic testing. Surgical resection is the treatment of choice.

CAROTID ARTERY DISSECTION

Although rare, carotid dissection is a leading cause of stroke in children and young adults. It typically presents with the acute onset of cervical, facial, or head pain, followed within hours or days by ischemic symptoms. Biousse and colleagues¹² reported a posttraumatic carotid dissection presenting with carotidynia as its sole manifestation. Symptoms also may include Horner's syndrome, pulsatile tinnitus, or amaurosis fugax. The artery may recanalize with time. Carotid duplex ultrasonography, magnetic resonance imaging (MRI), and carotid arteriography are the diagnostic tests of choice. The usual treatment is intravenous heparin followed by 6 months of warfarin therapy. Surgical intervention may be required in cases involving increasingly frequent or severe ischemic symptoms or other signs of neurological deterioration.

CAROTID ARTERY OCCLUSION

Because of the efficacy of the collaterals of Willis, total occlusion of one carotid artery may be neurologically silent. Donnan and Bladin¹⁴ described two cases of carotidynia caused by incomplete obstruction of the internal carotid artery by long intraluminal clots originating from subintimal hemorrhages beneath atheromatous plaques. The symptom of carotidynia was attributed to local vascular trauma. Both patients presented with neurological symptoms, as in all of the reported cases of carotid aneurysm with or without dissection. Clearly, the presence of ischemic or neurological signs or symptoms indicates the need for color duplex Doppler, MRI, or arteriogram.

Conclusions

Carotidynia is a painful symptom rather than a disease. Proper management of this condition depends on the underlying cause. The most important step to take when a patient presents with unilateral neck pain is to make a positive diagnosis. In the case of carotidynia, this involves eliciting a history of medical conditions, such as migraine or upper respiratory tract infection, and other symptoms

(especially neurological or ischemic). A careful examination including oral and dental examination, neck palpation, and possibly indirect or fiberoptic laryngoscopy, is mandatory.

If Fay's sign is present, the initial laboratory evaluation should include a sedimentation rate. If neurological signs or symptoms are present, a duplex color Doppler, and depending on the result as well as the presentation of the patient, either MRI or arteriography should be performed. In most cases, the sedimentation rate and neurological history and examination will be normal. In these cases, a diagnosis of benign carotidynia can be made.

Patients with a history of migraine and a prolonged but mild course of carotidynia may be treated with appropriate migraine therapy. Younger patients with a more acute course, no history of migraine, and symptoms suggesting viral infection may be treated symptomatically without further evaluation other than a sedimentation rate. Most of these cases, especially those with an underlying viral etiology, will respond to heat, reassurance, anti-inflammatory agents, and time. Reassurance is important because unilateral neck pain of some duration is often sufficient to cause a great deal of anxiety in some patients and their physicians because of its unknown nature and the fear of cancer.

The ultimate treatment in severe and recalcitrant cases may be denervation of the carotid bulb. In a case reported by de Vries and colleagues,¹⁹ the pain was so severe that the patient had been unable to eat, and medical management had failed. Surgical denervation resulted in immediate and lasting relief. Fay¹ also recommended stripping of the common carotid artery and bulb in refractory cases. This obviously represents a radical approach that should be undertaken only in rare cases with no other option.

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