Occipital Neuralgias: Clinical Recognition of a Complicated Headache. A Case Series and Literature Review

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The objective of this study was to assist clinicians in the diagnosis of the occipital neuralgia syndrome by describing its clinical characteristics. Bibliographies and clinical descriptions of occipital neuralgia syndrome were identified through a review of literature published between 1966 and 1993. A prospective case series was performed by the authors in a university emergency department during a 1-year period. Patients with unilateral aching pain of the head, coupled with pain in the distribution of the occipital nerve, Tinel's sign, and relief of pain after local anesthetic injection, were included. Patients rated pain relief on a 10-point scale. Twelve patients met the criteria for occipital neuralgia and were included in the study. All patients reported at least 80% decrease of pain after injection, and 42% had complete relief. Clinical features, other than headache, that were common in patients included tinnitus in 33%; scalp paresthesia, 33%; nausea, 42%; dizziness, 50%; and visual disturbances, 67%. Occipital neuralgia is a benign extracranial cause of headache, and it may be confused with other more serious headache syndromes. Recognition depends on an understanding of the symptoms along with a careful history and physical examination. Local anesthetic injections produce significant relief of the headaches and can aid in the diagnosis of the syndrome.

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"Neuralgia" pain is generally described as a brief, sharp, lancinating pain in the distribution of the relevant nerve, and it usually lasts several seconds. There may also be a continuous aching component that may be present for days or weeks in the area of the affected nerve. The Headache Classification Committee of the

International Headache Society described occipital neuralgia as "a paroxysmal jabbing pain in the distribution of the greater or lesser occipital nerves, accompanied by diminished sensation or dysaesthesia in the affected area. It is commonly associated with tenderness over the nerve concerned."4 The International Headache Society's diagnostic criteria include "pain in the distribution of the greater or lesser occipital nerve; pain that is stabbing in quality, although aching may persist between paroxysms; affected nerve is tender to palpation: and condition is eased temporarily by local anesthetic block of the appropriate nerve."4 Hypesthesia or dysesthesia during or after the clinical episode, coupled with the presence of reproducible extracranial tenderness or Tinel's sign over the nerve, are hallmark features of the occipital neuralgias.5 Resolution of pain through anesthetic block of the greater occipital nerve can be used to confirm the diagnosis of occipital neuralgia.6 The aim of the present study was to report a literature review of the occipital neuralgias along with a case series of 12 patients to aid the emergency physician in the recognition of this clinical syndrome.

Literature Review

A MEDLINE bibliographic database search of English language literature was undertaken for all publications related to the topic and published during the period 1966 through June 1993. Additional articles were identified from the bibliographies of articles retrieved and from the bibliographies of chapters of textbooks on neurology and headache. Articles and letters (1949 through 1993) dealing with occipital neuralgia, auricular neuralgia, craniofacial pain, or headache produced by lesions in the upper cervical roots were obtained. Articles were critically assessed independently by the authors. No controlled studies dealing with the treatment of occipital neuralgia were found. Most of the published articles on occipital neuralgia are case reports or case series studies.

Materials and Methods

A 10-year retrospective chart review of outpatients and inpatients seen at the Medical College of Georgia (MCG), Augusta, GA, a large tertiary referral center and teaching hospital, yielded only three patients with occipital neuralgia. Because of this small number, the retrospective approach was abandoned. Instead, a prospective identification and case series of patients who presented with headache to the MCG emergency department from November 1992 through October 1993 (12 months) was undertaken. Permission was granted for the study by the Human Assurance Committee. The emergency department at MCG examines approximately 40,000 patients per year and is a referral resource for most of the state of Georgia. All patients presenting to the emergency department with the complaint of headache had a general review of systems and a general physical examination with special focus on neurologic symptoms and the neurologic examination. A headache history was also obtained. Patients presenting with a neuralgia-type headache were examined for tenderness over the occipital nerve. Patients meeting criteria for occipital neuralgia were examined by one of the authors. The authors interviewed each patient and recorded a general headache history. Special attention was paid to a history of any minor or major head or neck trauma. "Occipital neuralgia" was diagnosed by the authors if the patients had a history consistent with neuralgia-type pain, along with tenderness over the related nerves; Tinel's sign, causing worsening of the headache; and prompt relief of the headache with local anesthetic block of the greater or lesser occipital nerves using 1 to 3 mL of 0.5% bupivacaine hydrochloride with epinephrine over the area where the nerve travels over the occipital protuberance. No attempt was made to blind or control these injections. All patients were referred to the MCG neurology clinic for follow-up care. In December 1993, all patients were telephoned to discover if they had sought follow-up care, had persistent headaches, or had been involved in additional diagnostic studies. The charts on patients who attended follow-up visits in the neurology clinic were retrospectively pulled; the original diagnoses were compared with the final diagnoses from the neurology clinic. No attempt was made to reexamine the patients whose symptoms had resolved.

Results

The authors prospectively identified 12 patients who met the aforementioned criteria for the diagnosis of occipital neuralgia and who presented to the emergency department during a 1-year period. Information on history and demographics was gathered at the time of presentation. Follow-up telephone calls were made to all patients; it was possible for 10 of the 12 to be contacted to check Kuhn et al

Patient	Age (y)	Sex	Duration	Trauma history	Pares- thesia	Tinnitus	Nasal symptoms	Visual symptoms	Dizziness/ vertigo	Nausea	Tinel's sign
1	23	F	3.5 h	No	No	No	No	Yes	No	No	Yes
2	27	F	5 d	No	No	No	No	Yes	No	No	Yes
3	25	F	3 d	No	No	No	No	Yes	No	No	Yes
4	26	F	2 h	Yes	No	No	No	No	Yes	Yes	Yes
5	49	F	1 wk	Yes	No	No	No	No	No	No	Yes
6	48	M	3 m	No	No	No	No	Yes	Yes	Yes	Yes
7	49	E	3 d	No	No	Yes	No	Yes	Yes	Yes	Yes
8	51	M	4 d	No	Yes	Yes	No	No	No	No	Yes
9	44	M	3 mo	No	Yes	Yes	Yes	Yes	Yes	No	Yes
10	21	M	2 wk	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes
11	58	M	2 wk	No	Yes	No	Yes	Yes	No	No	Yes
12	30	F	4.5 d	No	No	No	No	No	Yes	Yes	Yes

Table 1 Initial Symptoms in Patients With Occipital Neuralgia

the validity of the initial information and to discover whether the headache had persisted or returned and whether they had sought follow-up care. Several patients returned to the emergency department with persistent headaches and were reexamined by the authors. The information that was gathered is presented in Table 1.

Of the 12 patients identified, 50% were white, 50% were black, and 60% were female. The duration of the headache ranged from several hours to 3 months. One third of the patients reported scalp paresthesia, and one third had tinnitus (Table 2). Seventeen percent (2/12) had nasal symptoms, usually congestion or "stuffiness" with the headache. Two thirds (67%) of the patients reported ocular symptoms: either blurring of vision or ocular pain. Half of the patients (6/12) reported symptoms of dizziness or vertigo. Twenty-five percent were found to have a history of trauma to the head or neck that preceded the onset of the headache by days or months. None of these were considered serious injuries; most related a minor head injury or "cervical strain" from a fall or motor vehicle accident in the distant past. Slightly less than half (42%) of the patients reported nausea with their headaches. All had Tinel's sign over the occipital nerve, which was a criterion for the diagnosis of occipital neuralgia.

Nearly one third of the patients had seen other physicians before presenting to the emergency department. Prior to presentation to the authors of the present study, one patient had made five visits and another had made four visits to multiple physicians without recognition or diagnosis of occipital neuralgia.

Although the intention of the study was not to report on the efficacy of local anesthetic injection over the occipital nerve, all of the patients had sig-

Table 2 Frequency of Symptoms Identified

	Patients	Patients (n = 12)		
	Number	Percent		
Scalp paresthesia	4	33		
Tinnitus	4	33		
Nasal congestion	2	17		
Visual blurring, occular pain	8	67		
Vertigo/dizziness	6	50		
Nausea	5	42		
Tinel's sign over occipital nerve	12	100		

nificant, immediate, and sometimes dramatic relief (within 20 minutes) of their headache following injection of 0.5% bupivacaine hydrochloride and epinephrine over the nerve. Patients who obtained relief from the local anesthetic, thus confirming the diagnosis of occipital neuralgia, were given a corticosteroid injection at the site. Patients rated this relief on a 10-point numerical scale, with "10" being the worst headache of their lives and "0" representing absence of any headache or head pain. All patients reported a minimum of 80% relief of acute pain after injection, and several had 100% relief. No patients required the acute administration of narcotics for their headaches after the local injection.

From the 10 of 12 successful follow-up phone calls it was found that two patients had no return of their headaches after initial presentation (Table 3). The other eight patients reported return of the headache 3 to 28 days after their visit: all reported that the recurrence of the headache (rated on a 10point numerical scale) was milder than the original headache for which they had sought care in the emergency department. None of the patients con-

Patient	Degree (%) of headache relief 20 minutes after injection (based on numerical score)	Time after injection until headache returned	Does patient have chronic, persistent headaches at follow-up?		
1 100		Not available	NA		
2	100 (7 to 0)	No return at 2.5 months	No		
3	100 (10 to 0)	2 weeks	No		
4	80 (10 to 2)	1 week	No		
5	90	1 week	No		
6	100	1 week	Yes		
7	80	1 month	Yes		
8	80	2 weeks	Yes		
9	100	2 weeks	Yes		
10	100	3 days	No		
11	100	No return at 4 months	No		
12	80 (10 to 2)	Not available	NA		

Table 3	Results After	Injection	With Lo	ocal Anesthetic,	Steroid	Combination	
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tacted reported any significant relief of their headaches with medication, including ibuprofen, which had been prescribed by the authors, or narcotic analgesics, which had been prescribed by other physicians.

Discussion

Several authors have described the clinical syndrome of occipital neuralgia. Occipital neuralgia is not an uncommon cause of protracted or severe intractable headache. It occurs more frequently in women,⁷ and unlike many other types of headache, it is not related to menstruation, emotional factors, physical fatigue, change in weather, alcohol, tobacco, or food allergies8; it is often present on awakening in the morning with no prodrome. The frequency of an attack can vary from weeks to months, and the duration of a headaches can vary from days to weeks.8 The pain is true neuralgia with intermittent, brief episodes of excruciating, sharp, lancinating pain, which is sometimes accompanied by a dull, persistent ache in the dermatomal area of the nerve. Scalp and neck tenderness or paresthesia in the distribution of the second cervical dermatome are usually present during painful episodes.9 Often the pain is exacerbated by flexion of the neck.6 However, in occipital neuralgia, there is no limitation of or resistance to passive neck movements, no changes in neck muscle contour, and no abnormal tenderness of neck musculature as in cervicogenic headaches.4 The attack is not terminated by sleep, ergot compounds, or sedatives. Blurred vision, retro-orbital pain, lacrimation, and facial flushing is a frequent complaint and may lead the patient to consult an ophthalmologist.^{8,10} Dizziness and occasional tinnitus are also described as well as nasal congestion not associated with nasal or sinus pathology.

Pascual-Leone and Pascual-Leone Pascua² described 12 patients with occipital neuralgia. All of the patients had nausea, vomiting, and photophobia, mimicking migraine headache. Eleven had blurry vision, seven had vertigo or dizziness, and five had nasal congestion with their headaches. Seven patients had complete relief of their symptoms 30 minutes after nerve block with 1% lidocaine, and the others had significant but incomplete relief.² Auricular neuralgia shares many of the same characteristics with occipital neuralgia but also involves pain referral to the ear, cheek, side of face, and, in some instances, within the ear canal itself and area of the temporomandibular joint, mimicking pathology of the ear or temporomandibular disorders.¹⁰

The neuroanatomy of the occipital nerves help define the clinical syndrome associated with these neuralgias. The greater and lesser occipital nerves are a continuation of the second and/or third cervical roots. The roots receive branches from the spinal accessory nerve and the superior sympathetic ganglion. There is also communication between the roots and the trigeminal ganglion and probably the acoustic and vestibular nerves, which may explain some of the confusing symptoms seen with occipital neuralgia (blurred vision, nasal stuffiness, tinnitus, and dizziness).¹¹ The greater occipital nerve passes backward between the atlas and the axis in the space between the inferior oblique capitis and the semispinalis muscle (Fig 1). The nerve pierces the semispinalis muscle and passes through a small aperture in the aponeurosis of the trapezius muscle to emerge over the occiput as the greater occipital nerve.7

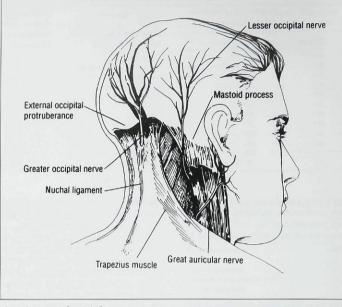


Fig 1 Anatomy of occipital nerves.

The occipital nerve supplies sensory distribution to the posterior aspect of the scalp, the side of the neck, the shoulders, and portions of the face.8 The great auricular nerve produces sensory distribution of the temporomandibular joint; external meatus; tympanic membrane; superior half of the pinna; and the skin and fascia of the cheek, temple, and scalp.12 Because these nerves are quite superficial as they exit from the posterior cervical musculature to enter the scalp tissues, they are readily stimulated by palpation.13 Tenderness of the lesser occipital nerve may be elicited by manual pressure in the depression that lies about 3 cm superomedially to the tip of the mastoid process. Tenderness of the greater occipital nerve may be elicited over the occipital protuberance or by digital pressure in the depression just lateral to the insertion of the trapezius muscle into the occipital bone.14 Cox and Cocks7 reported the resection of the occipital nerve in 95 patients seen for chronic pain. Many of these nerves showed pathologic changes (79%), including "neuromata, scarring around the nerve, neural fibrosis and densely adherent occipital lymph nodes." They also noted that "the aperture through which the nerve passes at the tendinous upper end of the trapezius muscle was frequently found to be compressing the nerve at that point."⁷

It is important to consider other causes of occipital pain when diagnosing occipital neuralgia.14 Migraine and cluster headaches as well as disease in the cervical spine, especially diseases that involve the C-1, C-2, and C-3 nerve roots, may easily be confused with occipital neuralgia. Other sources of pain in and around the vertebral column proper may include the apophyseal joints and the synovial joints of the occipitoatlanto and atlantoaxial junctions; the annulus fibrosus of the intervertebral discs: the ligaments of the spinal column and the periosteum of the vertebral bodies; the cervical muscles (including myofascial trigger points¹⁵) and their attachments to bone; the cervical nerve roots and nerves; and the vertebral arteries.16 The pain-sensitive structures may be stimulated by a variety of pathologic processes, including osteoarthritis, rheumatoid arthritis, temporal arteritis,17 trauma, abnormal posture,9 infection, tumor and Paget's disease. Causes of occipital pain that may mimic occipital neuralgia include C1-2 arthrosis^{18,19}; direct trauma to the occiput; inflammations, such as herpes zoster or postzoster neuralgia; compression between the atlas and the axis, as in traumatic hyperextension of the neck or atlantoaxial dislocation; entrapment between traumatized muscles; compression from caudally displaced tonsils of the cerebellum, as in the Arnold-Chiari malformations²⁰; primary and metastatic tumors, especially of the posterior fossa; mastoid and intraspinal infections; and metabolic disorders, including diabetes and gout.

Edmeads¹⁶ listed seven features of headache that suggest a cervical origin and necessitate a thorough radiologic examination of the cervical spine and craniovertebral junction. Radiologic evaluation should be considered for patients without a typical history of neuralgia-type pain, tenderness over the nerve, and relief of headache with local anesthetic injection or with persistent or recurrent pain after local anesthetic injection. Edmeads' criteria¹⁶ for radiologic evaluation include (1) headaches with a persistent occipital or suboccipital component, especially unilateral, (2) reproduction or alteration of the headache with movement of the neck, (3) abnormal posture of the head and neck, (4) suboccipital or nuchal tenderness, (5) abnormal motility at the craniocervical junction, (6) significant and painful limitation of neck movements, and (7) signs and symptoms referable to the lower medulla, upper cervical cord, or upper cervical nerve roots. For interpretation of the radiographs, it should be remembered that certain findings, including loss of lordosis, osteophytes, narrowed disc space, foraminal narrowing, and even disc protrusions shown on myelograms or magnetic resonance imaging scans are common in people older than age 4016 and may not indicate the cause of the symptomatology. In 1947, Perelson¹⁴ described palpable tenderness of the occipital nerves with multiple organic headache etiologies. The presence of any associated focal neurologic finding other than hypesthesia over the occipital nerve requires a more extensive evaluation, including computerized tomographic scanning or magnetic resonance imaging. Occlusion of the vertebral arterial system in the neck has been suspected to be a source of occipital headache in patients with vertebrobasilar insufficiency. The vertebral arteries are closely associated with the first six cervical vertebrae, rendering them susceptible to compression from trauma, including direct blows, attempted strangulation, or aggressive cervical manipulation.¹⁰ If a bruit is present, consideration should be given to vertebral angiography.21

Because there are no specific laboratory or radiologic findings to confirm the diagnosis of occipital neuralgia, the diagnosis and treatment rests on a careful and accurate history and physical examination. The best method of confirmation remains tenderness of the associated nerves. along with the characteristic pain syndrome and relief of the symptoms with local anesthetic blockade of the associated nerve. Blockade of the greater or lesser occipital nerve with local anesthetic should provide prompt and sometimes sustained relief from the headache. Because these nerves do not supply intracranial structures, headaches that are relieved following nerve block are considered to be caused by extracranial factors,7 with the possible exceptions of pain relief on the basis of either a placebo effect or a temporary redistribution of the anesthetic to the central nervous system.²²

Once the diagnosis of occipital neuralgia is confirmed, several modes of therapy may prove useful. Initial therapy should consist of reassurance and symptomatic treatment with deep heat, physical therapy, restriction of activities, and a mild analgesic.²¹ The nonsteroidal anti-inflammatory medicines, such as indomethacin, may be useful in some patients. Baclofen has also been advocated by some authors. It has been additionally noted that some patients may respond to transcutaneous electrical nerve stimulation (TENS).^{10,23,24} Cervical traction is not helpful and is not indicated. The application of cervical collars is controversial. Some authors advocate collars for pain relief. Others state that they should be avoided because they may exacerbate the pain by compression of the occipital nerve as it travels over the superior nuchal line.21 When pain is refractory to the above measures, a local block of the occipital nerve with a long-acting local anesthetic, such as bupivacaine hydrochloride with epinephrine, in combination with a corticosteroid (methylprednisolone acetate) often results in significant temporary or even protracted relief of the pain.24 Some authors recommend that this be done repeatedly.23 The treatment of recalcitrant pain is controversial. Several authors^{21,25} have advocated alcohol block of the nerve, nerve section, avulsion, or decompression of the nerve. Other authors6,12,23,26,27 have suggested that these procedures do not work in a large majority of instances, and they may lead to permanent pain and significant disability from denervation dysesthesia. Carbamazepine is the drug of choice for other forms of neuralgia and may be tried in occipital neuralgia.¹⁰ The initial

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dose of 100 mg per day can be given, and this is gradually increased to 800 mg per day in divided doses. Some patients may respond to lower doses of carbamazepine, and others may require more. However, most of the literature is anecdotal and none of the above drugs have been rigorously studied in patients with occipital neuralgia.

Conclusion

Occipital neuralgia is a clinical entity that may be encountered in patients presenting to an emergency department. The diagnosis is often straightforward from the history and clinical examination if it is considered by the examining physician. The headache frequently has two components, a recurrent, brief, lancinating component and a chronic, dull ache over the area of the nerve distribution. Only patients with typical neuralgia pain, tenderness over the nerve, Tinel's sign, and relief of pain with injection of local anesthetic were selected for the present study. The 12 patients were similar to those previously described in the literature in relation to type of headache and associated symptoms. Associated symptoms, beside headache, that were present in several of the patients included paresthesia in the distribution of the nerve, tinnitus, nasal congestion, visual symptoms, dizziness/vertigo, and nausea. There are no ancillary tests that can be used to aid in the diagnosis, except symptom relief from local anesthetic injection over the occipital nerve. All of the patients reported significant temporary relief of their symptoms within minutes of a local anesthetic injection, and several had longlasting relief. Although other more serious diseases may mimic the occipital neuralgias, a careful history evaluation and physical examination can usually exclude these if strict diagnostic criteria are followed. If any of the components of occipital neuralgia is not present, radiologic evaluation and referral is indicated. Because these neuralgias are often chronic, follow-up should be assured by someone knowledgeable in the care of these disorders. There have been no controlled studies dealing with the various options for pain control in patients with occipital neuralgia. Wellcontrolled studies are needed to evaluate the efficacy of various treatment options for patients with this disorder because the pain can be chronic and disabling. It is hoped that with increased awareness of the syndrome by clinicians, more patients with this problem can be recognized and treatment modalities can be evaluated.

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Resumen

Neuralgias occipitales: El reconocimiento clínico de una cefalea complicada. Series de casos y revisión de literatura

El propósito de este estudio fue el de asistir a los clínicos en el diagnóstico del síndrome de la neuralgia occipital, por medio de la descripción de sus características clínicas. Se identificó la bibliografía y las descripciones clínicas del síndrome de la neuralgia occipital a través de una revisión de la literatura publicada entre 1966 y 1993. Los autores revisaron series prospectivas de casos en un departamento de urgencias universitario, durante 1 año. Se incluyeron pacientes que presentaban dolor de cabeza unilateral, junto con dolor en la distribución del nervio occipital, signo de Tinel, y alivio del dolor después de la inyección de un anestésico local. Los pacientes evaluaron el alivio del dolor en una escala de 10 puntos. Diez pacientes caveron en la clasificación de la neuralgia occipital y fueron incluidos en el estudio. Todos los pacientes reportaron que el dolor había disminuido por la menos en un 80% después de la inyección, y el alivio total en un 42% de los pacientes. Los rasgos clínicos, fuera de la cefalea, que experimentaron comúnmente los pacientes incluyeron el tinnitus en un 33%; parestesia del cuero cabelludo en un 33%; naúsea en un 42%; vértigo, en un 50%; y disturbios visuales en un 75%. La neuralgia occipital es una causa extracraneal benigna de la cefalea, y puede ser confundida con otros síndromes de cefaleas mas serios. El reconocimiento depende del entendimiento de los síntomas junto con la ejecución de una historia y un examen físico cuidadosos. Las inyecciones con anestesia local producen un alivio significativo de las cefaleas y pueden ayudar en el diagnóstico del síndrome.

Zusammenfassung

Okzipitalneuralgien: klinische Erkennung eines komplizierten Kopfschmerzes. Ein Fallbericht und Literaturrückblick

Das Ziel dieser Studie war es, dem Kliniker in der Diagnose des okzipitalen Neuralgiesyndroms zu helfen, indem ihre klinischen Merkmale beschrieben werden. Bibliographien und klinische Beschreibungen des okzipitalen Neuralgiesyndroms wurden durch einen Rückblick der Literatur erhalten, weche zwischen 1966 und 1993 herausgegeben wurde. Ein prospektiver Fallbericht wurde von den Autoren in einer Universitätsnotfallabteilung während einer 1-Jahresperiode durchgeführt. Eingeschlossen wurden Patienten mit einseitigem Kopfschmerz, gepaart mit Schmerzen in der Verbreitung des Nervus occipitalis, Tinel's Zeichen und Abnahme der Schmerzen nach Injektion eines Lokalanästhetikas. Die Patienten schätzten die Schmerzlinderung auf einer 10 Punkte-Skala ein. Zwölf Patienten wiesen die Kriterien für eine Okzipitalneuralgie auf und wurden in die Studie aufgenommen. Alle Patienten berichteten über zumindest 80% Schmerzabnahme nach Injektion, und 42% hatten eine vollständige Abnahme. Klinische Merkmale ausser Kopfschmerzen, welche die Patienten gemein hatten, waren Tinnitus in 33%; Parästhesien der Kopfhaut, 33%; Nausea, 42%; Schwindel, 50%; sowie Sehstörungen, 75%. Die Okzipitalneuralgie ist eine gutartige extrakraniale Ursache für Kopfschmerzen, und sie kann vermischt sein mit anderen ernsthafteren Kopfschmerzsyndromen. Die Erkennung hängt vom Verständnis der Symptome zusammen mit einer sorgfältigen Anamnese und Untersuchung ab. Injektionen eines Lokalanästhetikas bewirken eine signifikante Abnahme der Kopfschmerzen und können bei der Diagnose des Syndroms helfen.

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