



NORD (National Organization for Rare Disorders)

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Arachnoiditis

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Synonyms of Arachnoiditis

- arachnitis
- spinal arachnoiditis

Subdivisions of Arachnoiditis

- adhesive arachnoiditis
- arachnoiditis ossificans
- cerebral arachnoiditis
- hereditary arachnoiditis
- neoplastic arachnoiditis
- optochiasmatic arachnoiditis
- postmyelographic arachnoiditis
- rhinosinusogenic cerebral arachnoiditis

General Discussion

Arachnoiditis is a disease characterized by an acute inflammatory stage that occurs in the dura (exterior) and the arachnoid (interior), two of the three membranes that cover and protect the brain, the spinal cord and the nerve roots. The arachnoid contains the cerebrospinal fluid which circulates from the brain to the sacral area, about every two hours; it filters any invasion and usually responds first by inflammation and follows with a chronic stage life-lasting phase characterized by scarring and fibrosis. As a result, abnormal adhesion of nerve roots to the dural sac or to each other (clumping) occurs in a variety of configurations that alter significantly the function of the roots and the spinal cord. This causes a variety of neurological deficits and severe chronic neuropathic pain usually located in the area affected. In the pre-antibiotic era,

severe cases of tuberculosis or syphilis invaded the spine causing arachnoiditis; currently these infections are rare, but it is important to mention that arachnoiditis will result in most patients affected by fungal meningitis from attempted epidural injections of tainted steroids.

Signs & Symptoms

The severity of the symptoms usually depends on the extent and location of the injurious event, direct injury to the spinal cord or to the nerve roots resulting in immediate, severe pain in the corresponding area of innervation (i.e. the area that is supplied by the affected nerves). Chronic severe pain is mostly localized in the lower back, perineum, legs and feet; it may appear weeks after a spinal surgical intervention or an injection into the neuroaxis that went astray. In most cases the pain is intense, accompanied by tingling or burning on the legs and feet; skin sensations like “bugs crawling” or “water dripping”.

Frequently patients complain of severe pain radiating to the lower extremities, muscle cramps, gait abnormalities and alterations of proprioception. Moreover, patients suffer from severe headaches, vision disturbances, hearing problems, dizziness and nausea. Bowel, bladder and sexual dysfunction as well as “electric shocks” type of pain are common in patients with severe arachnoiditis. All the symptoms are caused by the alteration and impediment of the CSF circulation resulting from clumped nerve roots, scar tissue and fibrosis.

Severe complications from multiple operative procedures in the spine may ensue in spinal cord damage, like “softening” (myelomalacia) or elongation of the nerve roots as in cases when the nerve roots and/or the spinal cord adhere to the dural sac wall. Some patients may develop arachnoid cysts and or syringomyelia. (For more information on these conditions, choose “arachnoid cysts” or “syringomyelia” as your search term in the Rare Disease Database.)

Most symptoms are initially due to progressive inflammation around where damage to the arachnoid membrane was produced. Because in the early stages, arachnoiditis is usually a progressive disorder, first it is characterized by inflammation of the arachnoid membrane and invasion of the subarachnoid space. Eventually, the nerve roots undergo an inflammatory stage in the affected areas and after three months, they begin to adhere to each other and to the inner layer of the arachnoid initiating the adhesive phase of arachnoiditis. Subsequently, these adhesions can exert undue pressure upon the nerve roots or the dural sac wall and eventually result in scarring and fibrosis which in turn may restrict blood flow to the affected area and eventually also impede the free flow of the CSF. In this manner of distribution, the rotation of the CSF can spread infections, disseminate malignant tumors, and disperse fungi.

Causes

There are numerous causes of arachnoiditis:

- a) Direct mechanical injuries (dural cut/tear) caused by surgical interventions, especially repeat spinal operations, after fusions or minimally invasive procedures requiring corrective surgeries.
- b) Trauma to the spinal cord.
- c) Epidural disc prolapse.

- d) Repeat manipulation during catheter or leads insertion.
- e) One or multiple spinal taps.
- f) Several steroid epidural injections.
- g) Injections of epidural steroids gone intrathecal.
- h) Difficult epidural blood patches (injection >20ml).
- i) Injection of myelogram dye into the spinal cord.
- j) Infections that may cause meningitis (viral, fungal or bacterial).

Affected Populations

Arachnoiditis is a rare disorder affecting more females than males, probably because two thirds of the pregnant women in the USA, Latin America and most other countries receive spinal or epidural anesthesia for the delivery. (It is estimated that about 4% of them develop arachnoiditis due to complications of anesthesia).

Individuals who have had spinal surgery, intrathecal injection of toxic fluids (i.e. hyaluronidase, blood, dyes, steroids and local anesthetics with preservatives) into the spinal dural sac, or have had injuries to the spine or head, may be at greater risk to develop this disorder. However, the precise prevalence and incidence of arachnoiditis is unknown. According to one estimate, approximately 11,000 new cases occur each year in the USA; however the cause varies, ranging from back surgery, pain relief procedures and diagnostic interventions mostly performed in North and South America, Europe and Asia, with an undetermined number in Africa. Obviously, the greater number of surgical and anesthetic spinal interventions has increased the number of cases considerably. For different reasons, some cases of arachnoiditis may go misdiagnosed or undiagnosed, making it difficult to determine its real frequency in the general population. Diagnosis of arachnoiditis by MRI with intravenous contrast is preferred. The intrathecal injection of some dyes is potentially hazardous and should be avoided.

Related Disorders

There are numerous conditions characterized by signs and symptoms that are similar to those found in arachnoiditis, however, only a few will be listed. They include the failed back surgery syndrome, multiple sclerosis, fibromyalgia, reflex sympathetic dystrophy, chronic pain syndrome, cauda equina syndrome, syringomyelia, and some spinal cord tumors. In many cases, certain conditions can arise as complications of arachnoiditis, further rendering the diagnosis, more complex. Occasionally, the symptoms manifested by some individuals with arachnoiditis may be dismissed as psychosomatic and these patients may be branded as "pain is in your head" cases.

Diagnosis

A diagnosis of arachnoiditis is based upon the antecedents of having had an invasive procedure or serious illness within the spine and a complete clinical exam, a detailed patient history, identification of key

symptoms and a variety of specialized tests of which radiological confirmation is essential. Since most cases occur in the lumbosacral spine, an MRI with contrast is the optimal examination to verify the extent of the illness. A complete history and physical may define the extent and severity of the illness, including the injury caused by prior trauma, surgery and other interventions into the spine. The symptoms of arachnoiditis resemble those associated with other spinal conditions including back pain, muscle spasms, referred pain, headache, dizziness and alterations of balance. When the intensity of the pain increases post surgery or certain procedures such as manipulations or punctures of the spine are followed by intense headache, back pain and or by visual disturbances, proper radiological studies are indicated, nerve conduction studies are non-contributory.

Clinical Testing and Work-Up

Of preference, a magnetic resonance imaging (MRI) of the lumbosacral spine with contrast is ideal, more precise and less injurious. The MRI uses a magnetic field and radio waves to produce cross-sectional images of particular organs and bodily tissues. To obtain the best results is imperative that an intravenous dye is administered only a few minutes before the test in order to be able to have the optimal images, especially within the dural sac, ideally obtaining axial and sagittal views of the lumbosacral region. However, the presence of certain non-titanium metal objects (screws, wires, etc.) makes the MRI contraindicated since it may heat certain metals. If an MRI is contraindicated, the diagnosis of arachnoiditis will have to be made using a computed tomography (CT) scan, which requires the administration of contrast media into the intrathecal compartment; however the images are not as informative as those obtained from an MRI. The intrathecal injection of a dye (myelogram) presents one more obstacle, as it may be hazardous, especially in cases of infections. An intrathecal injection refers to injecting a substance directly into the cerebrospinal fluid that surrounds the brain and spinal cord.

Physical examinations of patients with arachnoiditis can reveal changes in reflexes, sensory deficits, strength, and/or weakness. Furthermore it confirms the physical findings between the right and the left side when compared with radiological images. Of great use is the exploration of proprioception which confirms the alteration of balance.

Standard Therapies

Currently, there is no complete cure for arachnoiditis, but there are therapies which, if implemented, may lessen pain and suffering. Multimodal medication management with Schedule III drugs should be employed for chronic indications. Any Schedule II opiates are to be used with great caution as there is a risk of dependence and development of side effects of continuous tolerance will require progressively higher dosages.

Mild physical therapy is recommended for affected individuals to restore motion, preserve function and to help them remain active. It may include massage, mild exercise (without producing pain), hydrotherapy, and hot or cold compresses.

Psychotherapy may be helpful for most patients with arachnoiditis, as there is a strong element of depression. Patients need to be able to cope with their disease problems and the detrimental effects of continuous severe pain.

Surgery as a treatment of arachnoiditis is generally not recommended because of the possibility of scar

tissue and fibrosis formation.

For information about treatment options for arachnoiditis, please write to

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Investigational Therapies

Information on current clinical trials is posted on the Internet at www.clinicaltrials.gov. All studies receiving U.S. government funding, and some supported by private industry, are posted on this government web site.

Repeated therapeutic infusions provide patients considerable pain relief.

For information about clinical trials being conducted at the NIH Clinical Center in Bethesda, MD, contact the NIH Patient Recruitment Office:

Toll-free: (800) 411-1222

TTY: (866) 411-1010

Email: prpl@cc.nih.gov

For information about clinical trials sponsored by private sources, contact:

www.centerwatch.com

Supporting Organizations

- [American Chronic Pain Association](#)
 - P.O. Box 850
 - Rocklin, CA 95677 USA
 - Phone: (916) 632-0922
 - Toll-free: (800) 533-3231
 - Email: ACPA@theacpa.org
 - Website: <http://www.theacpa.org>
- [American Pain Society](#)
 - 8735 W. Higgins Road
 - Suite 300

- Chicago, IL 60631
- Phone: (847) 375-4715
- Email: info@americanpainsociety.org
- Website: <http://www.ampainsoc.org>
- [BackCare, the Charity for Healthier Backs](#)
 - 16 Elmtree Road
 - Teddington, TW11 8ST United Kingdom
 - Phone: (208) 977-5474
 - Toll-free: (845) 130-2704
 - Email: contact@backcare.org.uk
 - Website: <http://www.backcare.org.uk>
- [Genetic and Rare Diseases \(GARD\) Information Center](#)
 - PO Box 8126
 - Gaithersburg, MD 20898-8126
 - Phone: (301) 251-4925
 - Toll-free: (888) 205-2311
 - Website: <http://rarediseases.info.nih.gov/GARD/>
- [National Spinal Cord Injury Association](#)
 - 75-20 Astoria Blvd
 - Jackson Heights, NY 11370 USA
 - Phone: (718) 803-3782
 - Toll-free: (800) 962-9629
 - Email: info@spinalcord.org
 - Website: <http://www.spinalcord.org>
- [NIH/National Institute of Neurological Disorders and Stroke](#)
 - P.O. Box 5801
 - Bethesda, MD 20824
 - Phone: (301) 496-5751
 - Toll-free: (800) 352-9424
 - Website: <http://www.ninds.nih.gov/>

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