Adhesive Arachnoiditis: An Old Disease Re-Emerges

Authors: Martin J. Porcelli, DO, MHPE, PhD; Forest S. Tennant, DrPH, MPH, MD

Arachnoiditis Study and Education Project
Tennant Foundation
336½ So. Glendora Ave
West Covina, CA 91790

Dr. Forest S. Tennant

Dr. Martin J. Porcelli

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ABSTRACT

Lumbar-sacral adhesive arachnoiditis (AA) is a spinal canal inflammatory disease in which cauda equina nerve roots are adhered by adhesions to the arachnoid membrane of the spinal canal cover. It is a mass in which nerve roots that connect to the legs, bladder, intestines, and sex organs become impaired and dysfunctional. Severe intractable pain and multiple neurologic complications and premature death may result. In the last century AA essentially disappeared with the development of antibiotics and magnetic resonance imaging that eliminated the need for toxic myelogram dyes. Along with the increasing prevalence of back pain in this century, AA has re-emerged to the point that it is now regularly observed in clinical practice.

INTRODUCTION

Arachnoiditis (ARC) was defmed as inflammation of the arachnoid membrane in 1873.[1] The arachnoid is the middle layer of the meninges, the protective covering tissue that envelopes and encases the brain and spinal cord. The outer layer of the spinal canal covering is the dura and the inner most layer is the arachnoid membrane. Spinal fluid flows between the arachnoid membrane and pia mater,[2,3] In the 1800's ARC was often called a "Devil's Disease" because it was associated with severe pain, emaciation, suffering, and an early death. The usual causes were tuberculosis or syphilis.[4,5] There was no treatment. In 1855 Dr. Thomas Addison published his monograph on adrenal insufficiency and about one-third of his eleven cases had postmortem spinal cord pathology compatible with a diagnosis of ARC.[6]

In the early 1900's Horsley and Harvey described areas of adhesions that adhered or glued cauda equina nerve roots to the arachnoid membrane of the lumbar-sacral spinal canal cover.[7,8] Since this discovery the term adhesive arachnoiditis (AA) has been used to define the clinical condition whereby some cauda equina nerve roots are adhered by adhesions to the arachnoid membrane. Although inflammation of the arachnoid membrane can occur in the brain or upper levels of the spinal cord, lumbar-sacral AA is the condition now being observed with some regularity in clinical practice and is the subject of this report. Reported here are the results of our preliminary efforts to study and develop some diagnostic and clinical measures for lumbar-sacral AA.

THE RE-EMERGENCE

After treatment for tuberculosis and

CORRESPONDENCE:
Martin J. Porcelli, D.O., M.H.P.E., F.A.C.I.M., F.A.C.O.F.P., dist.
ppomando1@msn.com

syphilis were developed in the first half of the last century, AA essentially disappeared. [4,5] AA began to re-emerge, however, in the mid-1900's when some toxic dyes (e.g., Pantopaque™) were injected into the spinal canal to perform myelograms.[9,10] Unfortunately, a small percentage of people who received the dyes developed AA. Magnetic Resonance Imaging (MRI) was developed in the late 1980's, and AA again disappeared to a point where it was classified as a rare disease.[11] During this century AA has started to re-emerge. Although no widespread epidemiologic data is yet available, AA is now being observed in clinical practice.[12,13,14,15]

METHODS

Due to this paucity of epidemiologic and clinical information, an "Adhesive Arachnoiditis Study and Education Project'" has been established by the Tennant Foundation. The purpose of the project is to collect preliminairy demographic and clinical data to develop therapeutic measln·es and be a prelude to formal studies. To date, the main method used to collect data has been to collect data from cases documented by magnetic resonance imaging. An institutional review committee of the Foundation has reviewed and approved, and monthly reviews the project. Cases are voluntarily submitted to us be patients, families, and physicians. To obtain case review materials the project has published through AA social media groups that have developed in consonance with the re-emergence of AA. Cases have come to us for review from over 50 foreign countries. All cases have signed a release to use their information for educational purposes. There has been no financial charge or remuneration for persons with AA who have submitted their information for review and study. Table One shows basic information on eighty consecutive cases submitted to the project for review.

Clinical information and recommendations in this paper come primarily from a review of published literature.[10,12,16,17,18,19] Therapeutic suggestions are based on clinical observation and data collected in the clinical practice of one of us (Porcelli).

CAUSES OF AA

There are now several epidemiologic studies and reports showing that back pain has markedly increased in this century. The reason for this increase appears to be multifactorial and includes sedentary lifestyle, obesity, bucket seats, diet, and poor posture. The precise reasons for the re emergence of AA are somewhat unclear but the same factors that are causing the rise in back pain incidence may obviously be implicated. Some of the re-emergence of AA may be that cases are coming to attention because the new MRI techniques showing contrast and clarity between spinal fluid and solid tissues has made it possible to better identify the disease.[12] There are reports that herniated discs and spinal canal stenosis are precursor disorders of AA, and our observations which are described below support this notion. There appears to be some risk factors that predispose to AA and genetic connective tissue disease, autoimmune disorders, trauma, and anatomic spine defects such as scoliosis and spondylolisthesis. Medical procedures, especially epidural injections and surgery are highly associated with AA, but these procedures are done to treat the precursors and risk factors of AA. In summary, cases of AA seem to have multiple factors that precede its development in the century. Despite the fact that each case may have different causative factors, AA has clearly re-emerged as a clinical entity.

CLINICAL PROFILE

Table One shows the clinical profile of eighty consecutive cases of MRI-documented AA who were reviewed in our study project. Our information shows that the predominant group with AA is a middle-aged female who has an anatomic abnormality of the spine and who has undergone multiple epidural injections and spine surgery. Major symptoms have been constant pain relieved by standing and worsened lying flat or raising one's leg. Urination difficulties and blurred vision are common. An interesting symptom is that patients have sensations of insects crawling or water running down their legs. (Table 2)

COMPLICATIONS AND CONSEQUENCES

In the 1800's AA was often referred to as a "Devils Disease" because its complications may be profuond.[4,5] AA is an inflammatory adhesive mass inside the lumbar-sacral spine.[7,8] (Figure One) Its pain becomes extremely severe, debilitating, and requires the most potent pain relief measures. Nerve roots that become entrapped in the mass may connect to the bladder, intestine, stomach, and sex organs.[20] Some nerve root connections that go to the hips, legs, and feet are invariably trapped. Consequently, radiating pain that may be in waves causing lower extremity jerking, spasms, and burning sensations routinely occur. Weakness of legs requiring ambulation assistance with a cane, walker, or wheelchair are common in untreated patients. The intraspinal mass may impair spinal fluid flow and produce blurred vision, tinnitus,

migraine headaches, and possibly dementia if spinal fluid flow isn't normalized. Anorexia, malnutrition, and bed-bound state may occur. Premature spinal fluid leakage may occur because the inflammatory mass of AA may erode through the spinal canal covering, deaths likely occur due to sepsis and/or failure of the cardiovascular or pituitary-adrenal systems. Table Two summarizes complications and consequences that have been observed by us and reported by others.[9,10,12,15,16]

PHYSICAL EXAMINAITON

There is no physical sign that specifically identifies AA. A number of physical signs may, however, be present. (Table 2) There may be lower extremity weakness and a diminution of reflexes. Pain is usually elucidated on straight leg raising. Contractions and indentation of muscles and soft tissues is commonly seen on examination of the back. These anatomic distortions occur due to pain, splinting, and spinal fluid leakage into the soft tissues of the back which commonly occurs due to AA. The inflammatory, adhesive mass of AA erodes through the dura and allows tissue-toxic spinal fluid to enter the muscles and soft tissues between the spinal column and skin surface. (Figure 1)

LABORATORY TESTING

AA is an inflammatory disease.[1,8,20] It will often produce serologic evidence of excess inflammation.[20] White blood cell count, erythrocyte sedimentation rate, and C-reactive protein may be elevated. Cytokine panels are recommended to help confmn the diagnosis of AA, because a number of interleukins may elevate.[21,22]

STAGING AND CLASSIFICATION

AA is staged or classified as mild, moderate, severe, or catastrophic. Unfortunately, AA will usually progress tln·ough the stage as with any chronic, progressive disease. The mechanism of progression is a spread of inflammation inside the spinal canal with subsequent scarring and even calcification of nerve roots. In the late stages a person becomes seriously debilitated and immobile. Bowel and bladder become dysfunctional, and patients become immunocompromised and subject to serious systemic infections. (Table 3)

TREATMENT

There is neither a specific pharmacologic agent nor published guidelines for treatment of AA. Some of our treatment recommendations based solely on clinical observation and experience are given here. Treatment is directed toward chronic inflammation in the lower spinal canal. Only anti-inflammatory agents that cross the blood brain barrier, produce adequate spinal fluid concentrations, and bind to receptors on the cauda equina and arachnoid membrane are effective. Although there are no controlled studies, our experiences and review of cases has found these anti-inflammatory agents to have positive effects reported by patients: celebrex, meloxicam, indomethacin, and ketorolac. Our first choice for pain relief and suppression of inflammation is low dose naltrexone. Starting dosage is 1.0 mg given one or twice a day and then titrated upward overtime according to patient symptoms.[25] Intermittent, low dosage of methylprednisolone or dexamethasone are recommended. These two corticosteroids are known to have therapeutic effects in the central nervous system. Symptomatic pain relief may require both opioids and neuropathic agents. We advocate tissue regeneration strategies which may include polypeptides and hormones including dehydroepiandrosterone (DHEA) and nandrolone.[24] The latter carries a specific label for nutrition deficiency status, as AA qualifies. In addition to pharmacotherapy treatment, we emphasize flexibility and mobility of the lower extremities as AA often leads to paralysis of the legs and feet.

Osteopathic Manipulative Treatment (OMT) needs to be employed on this most difficult problem of AA. HLA is contraindicated for this disease due to spinal cord adhesions. Rather gentle OMT employing range of motion with stretching and motion to limits of pain production. Muscle Energy or resistive techniques need be employed in this disorder. Monthly visitations will usually suffice control of disease/pain.[26]

SUMMARY

AA is an old disease and we pray this paper has shed new light upon this disease. A specific medical cause of the intractable pain has been identified in the context of this paper.

Constant pain impacts physiological and/or mental function such as sleep, eating, hygiene, reading, concentration, and mobility, i.e., ADLs (activities of daily living).

Trials of standard medication and dosages with relevant medication, i.e., muscle relaxants, antiinflammatories, stimulants, anti-seizure medications, and low-dose opioids have been recommended for control of normalization of function. There is objective physical evidence of the causative disease or complications of the pain that can relate to blood

pressure changes, tachycardia, neurologic deficit, or anatomic structural abnormalities. There is an objective, diagnostic test result that documents these abnormalities of structural changes in pain such as the magnetic resonance imaging (MRI) abnormality, hormone deficiency, elevated autoimmune or inflammatory marker or abnormal electrophysiologic test.

Further research and studies are required for diagnosis and impactful treatment parameters.

AUTHORS DISCLOSURE

No relevant financial affiliations or conflicts of interest.

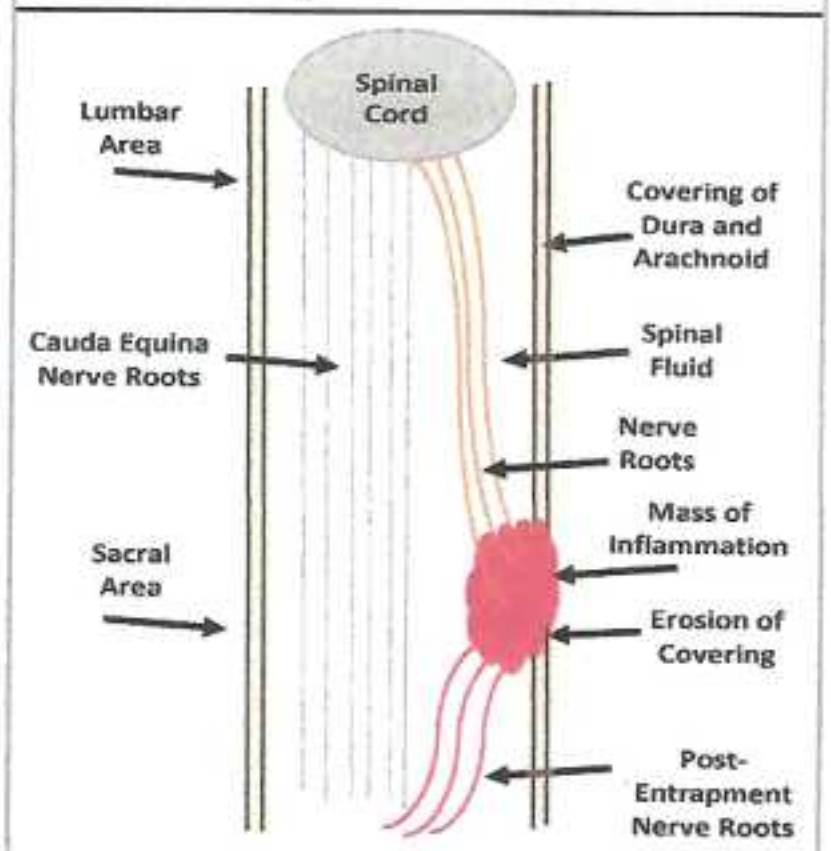
TABLE 1.
Clinical profile of 80 mri-documented cases of adhesive arachnoiditis

1. Females	65-81%
2. Males	15-19%
3. Age Range in Years	18 to 80
4. Mean Age ± S.D. in Years	48.9 ± 13.7
5. Number with a predisposing Spinal Condition*	61-76.3%
a. Herniated discs	44-55%
b. Spondylolisthesis	17-21.25%
c. Osteoporosis	6-7.5%
d. Spine arthritis	23-28.75%
e. Scoliosis	9-11.25%
f. Tarlov cysts	9-11.25%
6. Number with one or more Spine Surgeries	23-53.8%
7. Total number of Spine Surgeries in 43 Cases	91
8. Range of Surgeries in 43 Cases	1 to 8
9. Number who had two or more Spine Surgeries	22-27.5%
10. Number who had one or more Epidural Injections	69-86.3%
11. Total number of Epidural Injections in 69 Cases	236
12. Range of Epidural Injections in 69 Cases	1 to 20
13. Number reported over eight Epidural Injections	16-20%

TABLE 2.
Symptoms and Complications Reported by Over 55% of Cases

a. Pain relief on standing	70-87.5%
b. Standing too long Causes need to lie down	69-86.3%
c. Hurts to lie flat on back	67-83.8%
d. Pain always present	66-82.5%
e. Shooting pains, tremors, or jerking in the legs	64-78.8%
f. Burning pains/neuropathies in feet	63-78.8%
g. Cold hands or feet	58-72.5%
h. Crawling of insects on skin	58-72.5%
i. Water dripping/running down legs	53-66.3%
j. Difficulties starting urination/ defecation	51-63.8%
k. Leg raise hurts back	50-62.5%
l. Blurred vision	47-58.8%
m. Pain behind eyes (retrobulbar)	45-56.3%

FIGURE 1.
Diagram of entraped nerve roots in AA



The electricity normally conducted by the cauda equina nerve roots is trapped in the adhesive arachnoid mass and causes pain and inflammation.

TABLE 3.
Complications and consequences of adhesive arachnoiditis

Spinal Fluid Flow Obstruction	➢ Headache ➢ Blurred vision ➢ Tinnitus (Ringing in ears) ➢ Mental impairments (memory, attention, reading, and mathematics deficit)
Spinal Fluid Flow Leakage	➢ Contraction of paraspinal muscles ➢ Tissue over lumbar spine indents or "caves in" ➢ Arms can't extend
Sitting/Standing Ability Impaired	➢ Can't sit or stand in one position very long
Intractable Pain Syndrome	➢ Constant ("24/7") pain ➢ Cardiovascular, metabolic, and endocrine dysfunctions
Neuropathic Symptoms	➢ Burning feet ➢ Shooting pains into buttocks or legs ➢ Radiating type pain
Impaired Immunity	➢ Sepsis (infection) ➢ Premature death
Loss of Bladder, Bowel, and Sexual Function	➢ Urgency, incontinence, hesitancy, or paralysis ➢ Bloating, abdominal pain, alternating constipation, and diarrhea ➢ Sex organs disabled, loss of libido
Leg and Foot Paralysis	➢ Weakness ➢ Inability to stand or walk ➢ Foot drop
Bizarre Neurologic Symptoms	➢ Sensation of water or insects on legs ➢ Burning of feet or buttocks ➢ Leg jerking, spasms, "restless legs"
Dietary Dysfunction	➢ Loss of appetite for proteins ➢ Excess sugar intake ➢ Malnutrition/weight loss ➢ Anorexia
Hormonal Deficiencies	➢ Cortisol ➢ Pregnenolone ➢ Dehydroepiandrosterone (DHEA) ➢ Testosterone

TABLE 4.
Categories and stages of AA

Stage One – Mild ➢ Extremities: full range of motion, strength, extension ➢ No urinary or central symptoms* ➢ Normal ambulation ➢ Intermittent pain: non-opioid management is sufficient
Stage Two – Moderate ➢ Extremities: full range of motion, strength, extension ➢ Some urinary, gastrointestinal tract, and/or central symptoms* ➢ Normal ambulation ➢ Constant pain: non-opioid management is sufficient
Stage Three – Severe ➢ Extremities: some deficiency in range of motion, strength, extension ➢ Significant urinary, gastrointestinal tract, and/or central symptoms* ➢ Ambulates with assistance ➢ Severe, constant pain that requires daily opioids
Stage Four – Catastrophic ➢ Extremities: significant deficiency in range of motion, strength, extension ➢ Significant urinary, gastrointestinal tract, and/or central symptoms* ➢ Bed bound part of each day ➢ Ambulation requires assistance ➢ Severe, intractable pain that requires palliative care
Notes on Interpretation ✓ Central refers to headaches, and eye/ear/nasal symptoms such as blurred vision, tinnitus, vertigo, or nasal dripping ✓ Ambulation assistance means cane, walker, wheelchair ✓ MRI fmdings do not necessarily correlate with staging although the severe and catastrophic categories usually show one or more of these findings: dense scruring of nerve root clumps, multiple clumps, lower spinal canal distension (''empty sac"), peripheralization of nerve roots, calcification.

Note: Categories can overlap. Mild and moderate categories have the best potential for recovery.