Who is the most appropriate specialist to assess and treat arachnoiditis? The short answer is, someone who has some experience of the condition.

Practically speaking, this may mean that a local specialist who is familiar with arachnoiditis is hard to find.

However, neurologists or rheumatologists are both likely to be able to make a comprehensive assessment.

Surgical specialists are less likely to be consulted unless a surgically treatable problem comes to light. (one must bear in mind the original underlying spinal pathology present in the majority of cases).

Arachnoiditis does not present with a specific clinical picture of motor, sensory and reflex abnormalities: diagnosis tends to rest on further investigations such as MRI scans.

There are, however, still some centres which perform myelograms as diagnostic procedures for arachnoiditis.

Taking into consideration the fact that any foreign agent introduced into the spine has the potential to cause arachnoiditis, the rationale behind this type of testing seems questionable.

Burton ([1]) points out that whilst the incidence of arachnoiditis following non-ionic water-soluble myelographic agents is "quite small", if the

" wrong water-soluble agents, or the wrong concentrations of agents, are administered there can be serious consequences such as permanent neurological injury or death. "

Moreover, he goes on to state that

" It is important to understand that myelography has never really been a great diagnostic study...a poor means of demonstrating many important entities such as pathology in the foraminal zone of the vertebral canal."

The current investigation of choice is a T2 weighted, fat suppressed, high resolution MRI scan, including axial views.

(Delamarter et al ([2]) suggested that T2-weighted images rendered the nerve root changes more visible than T1-weighted images.)

Ideally, this should be read by a neuroradiologist experienced with the appearance of arachnoiditis.

Whilst MRI scan CAN be used in patients who have ?hardware' in their spine (pedicle screws, plates, rods etc.), the presence of metal may cause significant image artefacts which make interpretation difficult. Titanium implants produce fewer artefacts.

MRI scans can be performed on patients with a ?pump' (intraspinal delivery of drugs). However, the scanner will temporarily halt the rotors of the pump.

MRI scans may not be possible in patients with a spinal cord stimulator.

MRI cannot be used in the following circumstances:

In patients with:

- a heart pacemaker
- a metallic foreign body in the eye
- metallic vascular aneurysm clips
- a cochlear implant
- on life support systems which use ferromagnetic materials

Gadolinium has been used extensively to facilitate diagnosis of patients with Failed Back Surgery Syndrome, as it enhances scar tissue but not disc tissue, which therefore allows distinction to be made between recurrent disc herniation (treatable by surgery) and scar tissue (fibrosis), which tends not to be amenable to surgery.

However, more recently, more high resolution scanning techniques have rendered the use of gadolinium less necessary for this purpose. 3D imaging allows for high resolution images in a reasonably short time sequence (other techniques may sacrifice resolution in a bid to achieve ultrafast MRI images.

A routine cervical spine MRI would include:

- Sagittal T1-weighted (T1W)
- Sagittal T2W
- Axial T2W

Routine lumbar spine MRI includes:

- Sagittal T1W
- Sagittal T2W
- Axial T1W and/or T2W

Simmons et al ([3]) note:

" It is critical when performing an MRI that a complete study be done. This includes sequential sagittal and axial high-resolution images to optimally evaluate all components of the spine. "

Nelson recommends that MRI should include the thoracic region as this may well be a site for constrictive arachnoiditis.

Massie et al. ([4]) gave a poster session at the 2001 Annual Meeting of the American Orthopaedic Research Society on the assessment of post-laminectomy scar formation.

The goal of their animal study was to

"correlate the histology results with MRI results and to show if the MRI can be used for assessing evidence of scar as new anti-inflammatory agents are approved for clinical trials."

They found that there was a

"pattern towards a correlation between the MRI (with an injection of gadolinium) results and the histophomotery results."

They therefore concluded that

"The MRI can be a useful tool in assessing the presence and amount of scar",

although they did remark that when the defect was small or there was minimal enhancement of the tissue between the osseous defect of the laminectomy site, that there was a discrepancy between MRI and histological findings.

ARACHNOIDITIS ON MRI

Nerve roots normally appear like strands of spaghetti, lying adjacent to each other, but in adhesive arachnoiditis the nerve roots are adherent to each other and thus distorted, resembling overcooked and matted spaghetti.

This nerve root ?clumping' is one of the cardinal signs of arachnoiditis on radiographic images. As arachnoiditis progresses, the nerve roots are drawn out to the edge of the dural (thecal) sac, thereby producing an appearance known as the ?empty sac'. This may cause a ?string of pearls' appearance on axial views.

Various authors have described an intrathecal soft tissue mass (matted cauda equina) with a broad dural base, which can cause obstruction to CSF flow. Focal or diffuse constriction and thickening of the thecal walls may also be noted.

Use of gadolinium contrast agent (see below) may show enhancement of thickened intradural (intrathecal) scar tissue.

Aldrete included the following arachnoiditis-related lesions that may be diagnosed by MRI scan:

- nerve root clumping, unilaterally or bilaterally
- adherence of nerve roots to the dural sac wall
- ?empty sac'
- arachnoid cyst with scarring
- Swelling or thickening of cauda equina
- Partial or complete obstruction of dural sac
- Compartmentalisation of dural sac
- Tethering of spinal cord
- Atrophy of spinal cord
- Filling defect of spinal cord
- Syrinx
- Subdural calcification

In terms of severity, Aldrete describes MRI findings as follows:

- Absent: normal distribution of nerve roots
- Minimal: 2 clumped nerve roots (unilateral) or thickened dural sheath
- Mild: 3-5 clumped roots, 1 or more adherent to dural sac

- **Moderate:** clumped nerve roots, bilateral, adhered to spinal cord, partial compartments within dural sac

- Severe: deformity of dural sac, pseudomeningocele, partial/complete obliteration

- **Extreme:** calcification of dural sac; pachymeningitis, nerve roots adherent to dura and/or cord, syringomyelia

Note:

False arachnoiditis: pseudo-clumping seen with spinal stenosis

lophendylate residue may be seen as localised deposits, typically encapsulated and in some cases calcified (these may exert a localised mass effect) or as a thin film, which resembles a layer of fat on either T1 or T2 images.

MRI in specific situations:

Intraspinal TB:

Gupta et al. ([5]) in India, looked at 20 patients. TB leptomeningitis was characterised by CSF loculation, nerve root thickening and clumping (in the lumbar region) or complete obliteration of the subarachnoid space.

Gadolinium enhancement showed linear enhancement of the surface of the spinal cord and nerve roots or plaque-like enhancement of the dura-arachnoid complex. Intramedullary features included tuberculoma, cord oedema and cavitation.

Imaging Arachnoiditis Ossificans

Frizzell et al. ([6]) looked at MRI findings in arachnoiditis ossificans. They noted linear or mass-like intrathecal lesions with hypersensitivity on T1-weighted sequences and hyper- or hypo-intense on T2 weighted images within a setting of arachnoiditis.

However, MR is limited in demonstrating small areas of calcification or ossification.

Faure et al. ([7]) stated:

"MR imaging is a poor examination for this disease because it fails to show intracanal osseous formations."

Braz et al. ([8]) recently noted "On MRI imaging the manifestations could be minimal and variable."

CT scan shows a typical picture depending on the type; a circumferential high-density structure following the contours of the spinal cord or cauda equina is path gnomonic of arachnoid ossification.

Revilla et al ([9]) described the ability of helical computed tomography (CT) in diagnosing calcified plaques; multiplanar reconstruction can be helpful in pinpointing location for the neurosurgeon?s reference.

Arachnoid cysts:

Yu et al. ([10]) have studied arachnoid cysts using phase-contrast cine MRI, which allowed them to measure brain motion and CSF flow during the cardiac cycle.

They showed that brain motion was due to the volume difference between arterial and venous blood flow during a cardiac cycle, which thus drives CSF pulsation.

Arachnoid cysts and subarachnoid space enlargement carried different curve patterns, thereby demonstrating that phase-contrast MRI and flow quantification can be a useful for non-invasive evaluation of brain motion and CSF flow and differentiation between arachnoid cysts and subarachnoid space enlargement.

Syringomyelia: MR features

Inoue et al. ([11]) looked at MRI features in 7 patients with syringomyelia associated with surgically proven spinal arachnoiditis, of which 5 were thoracic, 1 cervicothoracic and the remaining one extended from C4 to L1.

All cases showed cord deformity due to adhesion or displacement due to an associated arachnoid cyst. This was best imaged on axial views. Flow voids could be seen in all cases on T2 weighted images.

How Useful is the MRI?

Kenneth Light, MD, a spine surgeon who has himself experienced severe back pain, has worked extensively with patients who have Failed Back Surgery Syndrome (i.e. have failed to get better after a back operation.

He wrote in his article, " When the MRI lies" ([12]):

" It is up to the clinician, not the radiologist, nor the MRI scanner, to decide whether the anatomic lesion discovered by the test is clinically significant. "

He concludes:

"This test, like the myelogram and CT scan before it, is not a substitute for a careful history and physical examination."

One must also bear in mind that, as Deyo wrote in the New England Journal of Medicine ([13]) in 1994,

" interpretation of MRI findings can vary substantially, so that the results may be equivocal despite the technique's aura of infallibility. "

Deyo goes on to comment:

"This variation...creates further opportunities for erroneous clinical decisions."

Sadly, at present, as Dr. Charles Burton of the Institute of Low Back and Neck Care in Minnesota remarks ([14]):

"most clinicians and radiologist are presently uninformed regarding quality studies and their interpretation", but, as he points out, this does not necessarily invalidate MRI scans themselves.

The NZHTA Report noted in its conclusions:

"Dependence on MRI or CT alone to detect abnormalities could result in inappropriate clinical evaluation and intervention."

It is important that treatable causes of Failed Back Surgery Syndrome (FBSS) such as recurrent disc herniation, disc fragments or spinal stenosis be excluded.

Functional MRI (fMRI) and PET scans have recently been developed: these are able to demonstrate the cerebral features of centralised pain.

New imaging techniques such as MTI may help in the future, as yet being untried with regard to arachnoiditis.

Ramli et al. reported recently ([15]) on a new imaging technique known as CISS (constructive interference in steady state), a type of MRI scan refinement. CISS is useful in

"intraaxial and extraaxial cystic abnormalities, dysraphic malformations and disturbances of cerebrospinal fluid circulation, including post-traumatic and post-surgical scarring."

One must bear in mind that MRI demonstrates gross anatomical abnormalities, but cannot image physiological deficit, any more than a chest X-ray can show heart rhythm.

This brings us to electromyography (EMG) and nerve conduction studies (NCS).

These tests are, of course, designed to elicit evidence of physiological impairment.

However, as an undetectable level of nerve impairment can cause severe pain, these tests have limited value unless they are being used to assess the level of functional impairment.

The Guidelines for Clinical Practice and Facility Standards facilitated by the College of Physicians and Surgeons of Ontario in 2001 ([16]), clearly state:

"Studies may be falsely negative if they are performed either too early and/or too late in the course of radiculopathy. Electrodiagnostic studies cannot be used to "exclude" a radiculopathy.

A clinically significant root lesion, particularly a chronic one, causing solely sensory complaints may be associated with a normal needle examination.

"..."Needle examination has a number of limitations. It assesses only motor fibres and detects primarily motor axonal loss."

The authors conclude:

"Few studies have been done to assess evidence based medicine and specifically sensitivity and specificity as it relates to radiculopathy. Much of the evidence is based on experience and expert opinion."

Fibrescopes:

Warnke et al. ([17]) recently published the results of the caloscopy performed on patients who had suspected arachnoiditis but no evidence on MR. The authors noted:

"The ?gold standard' did not establish any treatable diagnosis in these patients."

Their results showed pathomorphological evidence of thickening of the arachnoid membrane in all the patients treated, and rootlets suggested by clinical examination were confirmed as showing signs of arachnoiditis. Isolated arachnoiditis was observed on single levels and one side.

A further interesting finding was local raised CSF pressure, which caused CSF to pour out under pressure during the procedure.

They suggested that this is related to CSF circulatory disturbance which may also dilate the thecal sac. Warnke also remarked on cases in which a ?large theca' was observed on MR, and the patients who experienced symptomatic relief after lumbar puncture due to temporary reduction in CSF pressure.

"Thecaloscopy confirmed the suspected diagnosis in 100% of cases...We feel that the thecaloscope at least provides us with a safe diagnostic tool."

In the 9 patients with arachnoiditis, there were no serious adverse events.

In September 2003, Tobita et al. ([18]) reported on the use of ultrafine flexible fibrescopes to view spinal epidural and subarachnoid spaces in patients with chronic pain.

The authors assert that

"Fine flexible fiberscopes make it possible to visualize the entire length of the spinal subarachnoid space without major complications."

Their findings were: in 12 patients, new diagnoses were made, of which arachnoiditis 9, subarachnoid cyst 2, old subdural hematoma 1.

These had not been detected by MRI or CT.

Additionally, chronic arachnoiditis was found in 2 patients with spinal trauma.

Previously diagnosed pathologic changes were confirmed by fibrescopic examination in 16 patients of which arachnoiditis 11, spinal trauma 2, arteriovenous malformation 2, subarachnoid cyst 1.

The authors concluded:

"These fine flexible fiberscopes may provide new diagnostic and interventional tools for spinal canal diseases, provided skilled techniques are applied."

Whilst minimally invasive, this type of investigation nevertheless carries a potential risk of exacerbating any existing arachnoiditis, although it may be of help in diagnosing the condition prior to proposed major invasive therapies (e.g. unavoidable spinal surgery), so that measures may be undertaken to minimize the negative impact of such treatment.

Thermography to image pain: Infrared thermal imaging (ITI) has a role in the assessment of neuropathic pain. Neurovascular autonomic tests have been performed in studies of patients with complex pain and ITI has been found to successfully differentiate between mechanical causes of back and neck pain and nerve-generated (neuropathic/neurogenic) pain.

Hooshmand ([19]), an expert in Reflex Sympathetic Dystrophy (CRPS Type 1) has stated

"ITI identifies hyperthermic foci of permanent sympathetic system damage sparing the patient from further damage by trauma of sympathetic nerve blocks."

A significant number of arachnoiditis patients have features strongly suggestive of RSD-type damage and thus this test might be of help in ascertaining the source of their pain.

Lumbar puncture (LP): (Invasive: NOT recommended); in infective cases: chronic meningitis, which can be associated with arachnoiditis will show as: increased CSF pressure, possibly decreased glucose, increased protein, evidence of mononuclear cells. LP may be suggested if clinicians suspect Multiple Sclerosis.

Bladder dysfunction:

For bladder dysfunction, urological assessment and possibly urodynamic studies may be required. The Urodynamic test is designed to test the pressure in the bladder. In order to test the pressure a small catheter is inserted into the bladder. There is also a small rectal catheter and two small patches placed by the rectum. The bladder is then filled with water. When the bladder feels full, the patient urinates.

Cystometrogram (CMG) measures the bladder pressure as it fills, as well as testing bladder sensation, capacity and ability to expel urine.

Urethral pressure profile (UPP) measures the pressure along the length of the urethra as a catheter is being slowly removed. Uroflowometry (VFR) tests the force of the urine flow, measuring the amount and time taken voiding (into a special toilet)

Pressure flow study combines the CMG, VFR and electromyography (EMG). It measures both bladder pressure and urine flow. The EMG measures the activity of the pelvic floor muscles.

Multi-channel video urodynamics combine a pressure flow study with X-ray imaging of the bladder, to study the appearance of the bladder during filling, straining, coughing and voiding.

Post-voiding residual urine volume may be measured using ultrasound.

Urologists can also help to identify and treat sexual dysfunction. Gastroenterologists may be helpful in assessing bowel problems or difficulties in swallowing.

Clinical Investigations of ANS dysfunction.

By virtue of its many and complex functions, a complete assessment of the ANS is, of necessity, extremely complex. ANS testing has been used most often by cardiologists, gastroenterologists, urologists, and endocrinologists, with neurologists and pain specialists only recently developing ways to evaluate patients with autonomic dysfunction.

The autonomic nervous system cannot be tested directly by conventional neurophysiologic techniques, i.e., nerve conduction studies and electromyography.

The only way to assess function is indirect, by evaluating the response elicited reflexly by appropriate stimuli.

Until very recently, autonomic tests were available only in a few specialised centres.

Autonomic Reflex Screen (ARS)

Tilt table test Deep breathing Valsalva manoeuvre QSART Adrenergic vasomotor function Cardiovagal Cardiovagal and adrenergic vasomotor Postganglionic cholinergic sudomotor

CRPS Screen

Temperature measurements RSO* QSART* TST Index of sympathetic vasomotor tone Sudomotor and partially vasomotor Postganglionic sudomotor (stimulated) Thermoregulatory sudomotor pathways

*Performed simultaneously in bilateral, symmetrical sites.

(Source: Testing the Autonomic Nervous System, Paola Sandroni, MD, PhD *IASP Newsletter,* November/December 1998)

Investigations of Pelvic Pain

Perineal electrophysiological investigations (detection of neurogenic muscles of the perineal floor, increased sacral latency).

These constitute: bulbocavernosus muscle EMG, measurements of bulbocavernosus reflex latencies (BCRLs) somatosensory evoked potentials of the pudendal nerve (SEPPNs) and pudendal nerve motor latencies (PNTMLs).

Cystometry and urethral pressure profile may be investigated if there are urinary symptoms.

Note:

"the presence of pudendal neuralgia should prompt a search for an underlying cause, and that this severe neuropathic pain syndrome is effectively managed with adjuvant analgesics." ([20])

In the Canadian study, 6 patients with pudendal neuralgia all had diabetes and/or neoplastic conditions.

NOTE: 25 percent of lumbosacral plexopathies are metastatic ([21]). Pain is usually felt in the lower abdomen, buttock, and leg. Infiltration of the sacral plexus may produce perineal and perirectal pain, which is exacerbated by sitting and lying prone.

Pain typically precedes, by weeks or even months, the neurological signs of weakness, sensory loss, or urinary incontinence. Abdominal and pelvic CT or MRI may provide the diagnosis.

MRI of the epidural space is also required to exclude epidural disease of the cauda equine or leptomeningeal mass which may produce a clinical syndrome similar to lumbosacral plexopathy([22]).

ASSESSMENT: OF OSTEOPOROSIS

Osteoporosis risk-factor questionnaire

DEXA study (radio-imaging with low radiation): bone scan of spine, hip and wrist checks bone mineral density.

If there are osteophytes (bony spurs) and/or crush fracture, then an X-ray is also required. Note that X-ray alone only detects 30% or greater bone density loss.

Risk of fracture may be assessed using a Quantitative Ultrasound (QUS)

Investigations for Hydrocephalus:

CT, MRI, Lumbar Puncture, Intracranial pressure (ICP) monitoring; measurement of CSF outflow resistance; isotopic Cisternography.

Psychological assessment:

Chronic, intractable conditions such as arachnoiditis present the sufferers with a wide range of challenges day by day; in addition to the physical effects, it is important to acknowledge the inevitable psychological toll of unremitting pain and loss of function.

Haythornthwaite and Benrud-Larson ([23]) reviewed studies on the psychological assessment and treatment of neuropathic pain conditions, including postherpetic neuralgia (PHN), diabetic neuropathy, complex regional pain syndrome, post spinal cord injury, post amputation, and AIDS-related neuropathy.

They noted:

" the assessment of neuropathic pain needs to include measurement of multiple dimensions of quality of life. "

They included mood, physical and social functioning, and pain-coping strategies as important aspects of the patient's overall wellbeing.

TREATMENT OPTIONS

Generally speaking, this complex neurogenic pain syndrome is best treated at a specialist pain clinic, with a multidisciplinary approach.

As yet, a curative treatment is unavailable, so management revolves around palliative care principles.

Alert: http://www.fda.gov/cdrh/safety/neurostim.html

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