



Review Article

Review/Perspective On the Diagnosis and Surgical Management of Spinal Arachnoid Cysts

Nancy E. Epstein, M.D.

Clinical Professor of Neurosurgery, School of Medicine, State University of NY at Stony Brook, and c/o Dr. Marc Agulnick, 1122 Franklin Avenue Suite 106, Garden City, NY, 11530, USA.

E-mail: *Nancy E. Epstein - nancy.epsteinmd@gmail.com



***Corresponding author:**

Nancy E. Epstein, MD,
Clinical Professor of
Neurosurgery, School of
Medicine, State University
of NY at Stony Brook, and
c/o Dr. Marc Agulnick, 1122
Franklin Avenue Suite 106,
Garden City, NY 11530, USA.

nancy.epsteinmd@gmail.com

Received : 07 February 2022

Accepted : 08 February 2022

Published : 25 March 2022

DOI

10.25259/SNI_153_2022

Quick Response Code:



ABSTRACT

Background: Spinal arachnoid cysts (SAC) are typically congenital, spontaneous, traumatic (i.e., including iatrogenic/surgical), or inflammatory in origin. In descending order, they occur in the thoracic, lumbar, and cervical spine, and originate from focal entrapment of the arachnoid membrane. Arachnoid cysts represent 1–2% of all cystic spinal masses/tumors. The majority are extradural arachnoid cysts (EDAC) while 10% of all arachnoid cysts are intradural (IDAC) including subarachnoid, or extra-arachnoidal/subdural. Only rarely are they intramedullary in location. The clinical symptoms/signs of IDAC/EDAC include; intracranial hypotension (i.e., due to continued cerebrospinal fluid drainage), radiculopathy, and/or myelopathy.

Methods: Magnetic Resonance Images (MR) and Myelo-Computed Tomography (Myelo-CT) studies classically document the predominant dorsal location of IDAC/EDAC. They also show their extent and severity contributing to root, cord, and/or cauda equina compression. In the cervical/thoracic spine, MR/Myelo-CT studies classically show the “double cord” or “windsock” signs, while the “fake arachnoiditis sign” may be seen in the lumbar spine. The latter sign signals the presence of a circumferential extra-arachnoidal-subdural cyst that centrally “traps” the cauda equina. Note, that this resembles and is often misinterpreted as adhesive arachnoiditis.

Results: Patients with significant SAC-related neurological deficits typically warrant early surgery. That surgery includes; partial/total resection/fenestration of cyst walls, and occlusion of communicating fistulas with or without accompanying shunts.

Conclusion: It is critical to recognize the clinical (i.e., intracranial hypotension, radiculopathy, and/or myelopathy) and radiographic MR/Myelo-CT signs (i.e., “double cord,” “windsock signs,” or “fake arachnoiditis sign”) of IDAC, EDAC, or intramedullary spinal arachnoid cysts to appropriately offer treatment. For those with significant neurological deficits, early surgery (i.e. optimally 0- < 24 hours after the onset of symptoms/signs consisting of laminectomies, partial/total cyst resection/fenestration, and ligation/occlusion of the subarachnoid-cyst fistula with or without shunt placement), is essential to avoid significant permanent neurological sequelae.

Keywords: Diagnosis/Surgery; Spinal Arachnoid Cysts; Intradural/Intramedullary; Intradural/Extramedullary; Extradural; Marsupialize, Spinal, Subdural Cyst, Cyst Resection; Double Cord/Windsock Signs; Fake Arachnoiditis Sign

INTRODUCTION

Spinal arachnoid cysts (SAC) may be congenital, spontaneous, traumatic (i.e. including iatrogenic/surgical), or due to inflammation [Table 1].^[1-23] SAC develop as outpouchings/entrapments of the arachnoid membrane resulting in a fistulous ball-valve communication/

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Table 1: Clinical, radiographic, and surgical management of intradural extramedullary/subdural arachnoid cysts, epidural arachnoid cysts, and intramedullary arachnoid cysts.

Author ref date	subject Symptoms (Sx)	etiology Location	Type of AC symptoms-signs-deficit	MR Myelo-CT	Surgical treatment timing surgery
Choi <i>et al.</i> ^[4] 2006	EDAC Review+Case Study 59 yo F	Unknown	Spinal Cord Comp; 6 mos LBP Rad BLE	MR ED AC Diagnosed	Surgery Curative
Filho <i>et al.</i> ^[7] 2009	Case Giant ID EM AC 28 yo Thoracic	Congenital Thoracic ID AC 12 Levels	10 mos Pain Paresthesia Paraparesis Ataxia Myelop	MR Dorsal T3-T6 ID EM AC	T3-T6 LAM Durotomy- Posterior Wall Cyst Resected Fenestrated Shunted to SA Space
Nottmeier <i>et al.</i> ^[15] 2009	2 Cases Iatrogenic ID AC	Iatrogenic- Surgical Durotomy	Postop Comp Myelop CES	Postop MR Comp CES ID AC	Lumbar Intradural Exploration Fenestration of AC
Tureyen <i>et al.</i> ^[21] 2009	ED AC Case 25 yo M	Idiopathic Congenital	1 yr LBP Progressive Myelop BLE Neurogenic Claudication	MR Dorsal ED AC T11-L2 Postop MR Cyst Resolved	T11-T12 Laminotomy Total Resect Cyst-Repair Dural Defect @ T12+Fibrin Sealant-LOP No Postop Deficit
Gomez <i>et al.</i> ^[8] 2011	ID AC 4 Cases	Thoracic Spine 3 Dorsolateral 1 Ventral	Myelop	MR Best Study 1 Septum 3 Multiple Septations Cysts@1-8 Levels	3 LAM/Cyst Fenestration (1 Died Preop) 1 Reoperation for Syrinx/ Cyst-SA Shunt
Kong <i>et al.</i> ^[11] 2013	ED AC Case Paraparesis Voiding Difficulty	Post Traumatic	Myelop Paraparesis Neurogenic Bladder	MR ED AC Dorsal T11-L3 Cord Comp	Microsurgery Repair Right Dorsolateral Dural Defect Between Root Sleeves T11/T12
Rahimizadeh and Sharifi ^[18] 2013	2 Anterior Cervical ID AC<20 yo	Congenital	Neck Pain Paraparesis	MR ID AC Cyst	LAM LOP-Wide Fenestrated Cyst Wall-Subtotal Excision Good Result
Choi <i>et al.</i> ^[5] 2013	ED AC 2 Cases 59 yo M 51 yo F	T12-L3 ED AC T1-L2 ED AC	59 yo M Myelop Bilateral Rad x 4 Yrs 51 yo F Unilateral Rad/Buttock Pain 3 years	59 yo M MR ED AC T12-L3 51 yo F MR ED AC T1-L2 Myelo-CT Dural Com	59-yo LAM T12-L3/Cyst Fenestration Dural Defect Repaired 51 yo F T1-L2 ED AC Fenestration Primary Dural Repair Sx and MR Findings Resolved 1-2 mos
Deutsch ^[6] 2014	Thoracic SAC Review	Spontaneous Iatrogenic Traumatic	Pain Myelop	MR-Focal Displaced Thoracic Cord	Thoracic LAM Resect SAC Wall
Novegno <i>et al.</i> ^[16] 2014	IM AC Case 31 yo F	14 Cases in Literature	7 Years+3 mos Worse Myelop Paraparesis Urinary Incont Abd Pain	MR Dorsal IM AC T11-T12 No Enhance	T11-T12 LAM Fenestrated Cyst 6 mos Walk No Incont 2 Years MR No Cyst
Min and Kim ^[14] 2015	Case ID AC C6-T12 43 yo F	Spontaneous Extensive ID AC+Double Cord Sign	Left HP Cervical Myelop, GD Hypes, Rad	MR ID EM AC Multiple Septations C6-T12 Double Cord Sign Block CSF Flow Cord Comp	1 st LAM/Cyst Wall Resection T5-T7- Site Max Comp (Postop Worse Right C8T1 Deficit-New) 2 nd : Surgery Right C6-T12 LAM Resect AC Wall

(Contd...)

Table 1: (Continued).

Author ref date	subject Symptoms (Sx)	etiology Location	Type of AC symptoms-signs-deficit	MR Myelo-CT	Surgical treatment timing surgery
Habibi <i>et al.</i> ^[9] 2016	7 Sacral ED AC with SCM Average Age 56.7 mos Rg 18–119	Congenital SCM with Anomalies	Myelop Paraparesis Deformity Neurogenic Bladder	MR 7 Tethered Cords 6 Syrinxes 7 Sacral EDAC	Surgery Resect Sacral ED AC+SCM LAM/Fenestrate Cyst Wall/7 Obstruct Com
Menezes <i>et al.</i> ^[13] 2017	ED AC Case 14 yo M	Congenital Mother had L2-L4 ED AC and Disc L34	14 yo M Myelop Slowly Progressive	MR T8-L2 Dorsal ED AC Cord Comp	Laminectomy Fenestration Resection of Cyst
Shrestha <i>et al.</i> ^[19] 2017	Anterior Cervical ID AC 20 yo C34	Most IID AC Posterior Surgery	Myelop Spastic Quadriparesis Pain Neurological Sx	MR ID AC C34 Anterior Approach MR “Scalpel Sign” Indicates SAW	C34 Anterior Corpectomy Fusion ACF; Cyst Resection T3-T4 LAM Fenestrate SAW Resolution of Sx
Ali <i>et al.</i> ^[11] 2018	SAW Case 70 yo M T3-T4	Arachnoid Membrane SAW Displace Cord+Syrinx			
Himes <i>et al.</i> ^[10] 2019	Case ID AC T3-T7 60 yo F Multiple Comorbidities	Idiopathic Pain Between Shoulders	Pain Relieved Supine Postural Hypotension	T3-T7 Dorsal ID AC Postop MR 7 mos IC AC Resolved	LOP T3/T7 Levels Excise Limits Cyst-Fenestrated Passed Lumbar Drain T3-7 No Shunt-Not Resect Full Cyst
Ozdemir <i>et al.</i> ^[17] 2019	ED AC 1% Spine Tumors Thoracic or Thor-Lum-3% All EDAC Cervical	Arachnoid Membrane Herniates Through Dura	Myelop Reflects Case Cerv-Thor Jnct	MR Best Shows ED AC Cerv-Thor Jnct May Enlarge	Surgical Resect Recommend Early Surgery Prevent Worsening
Lee <i>et al.</i> ^[12] 2019	ID AC: ISD Placed 10 year Ago; Now 50 yo with ID AE/Fx ISD	ID AC=10% of all AC Rare Iatrogenic Here Postop Surgery	LBP S1 Rad RLE (10 years Postop)	X-rays: Broken ISD L45 MR Multiple ID AC Cystic Tumors L1-S1 MR ED AC Lumbar Spine	Resect ISC LAM L1-S1 Resection Cysts/Septations ID AC/Fenestration Resect ED AC Fenestrate (Scar) Repair Dural Defect-Grafts or Lumbar Drains - Shunts
Boody <i>et al.</i> ^[2] 2019	ED AC	Multiple Congenital Spontaneous Traumatic Infectious	Back Pain Rad, Myelop CES		11 (84.6%) Total AC Resect; 2 (15.3%) Partial AC Resect All 13 Closed ID Com/Fistula
Singh <i>et al.</i> ^[20] 2019	Sx ED AC<2% All Spinal Tumors 13 Surgery 2006-16	Congenital All 13 ED AC Com ID Space Mean 52.2 mos (Rg. 1-160)	Rad Myelop	MR-Most ID EM/SD AC Case ED AC	
Bowman and Edwards ^[3] 2020	Case ED AC L3-L5 38 yo F	Congenital Lumbar ED AC	Left Quad Wk Dyses	ED AC-X-rays MR/Myelo-CT CT: L4 Erosion VBody/Pedicle Identify Cyst-Dural Fistula- MR ID AC Cord Comp	L3-5 LAM/Fusion Resect ED AC No Comp No Residual Deficit
Zapata <i>et al.</i> ^[23] 2020	Traumatic ID Case AC Cord Comp	Congenital Spontaneous Trauma	Myelop		Surgical Options

(Contd...)

Table 1: (Continued).

Author ref date	subject Symptoms (Sx)	etiology Location	Type of AC symptoms-signs-deficit	MR Myelo-CT	Surgical treatment timing surgery
Zanon <i>et al.</i> ^[22] 2021	ID AC Thoracic Dural Tear	Surgical Trauma Thoracic Lumbar Cervical	ID AC Thoracic Myelop	Diagnosis MR Thoracic ID AC	Cyst Resect Drain Timing Surgery Best Immediate

AC: Arachnoid Cyst, TR: Traumatic, CC: Cord Compression, CES: Cauda Equina Compression, ID: Intradural, EM: Extradural, SD: Subdural, ED: Epidural/Extradural, Cong: Congenital, Iatro: Iatrogenic, Surg: Surgical, Inflamm: Inflammatory, Rad: Radiculopathy, Resect: Resection, Drain: Drainage, Fenestrate, CSF: Cerebrospinal Fluid, LAM: Laminectomy, LOP: Laminoplasty, yo: Years Old, mos: Months, SA: Subarachnoid Space, Com: Communicated, Rg: Range, F: Female, M: Male, Wk: Weakness, Dyes: Dysesthesias, Vert: Vertebral, Hem: Hemorrhage, ISD: Interspinous Device, CSDH: Chronic Subdural Hematoma, AE: Adverse Event, LBP: Low Back Pain, RLE: Right Lower Extremity, Thor-Lum: Thoraco-Lumbar, LE: Lower Extremities Comp: Compression, Jnct: Junction, Cer-Thor: Cervico-Thoracic, Preop: Preoperative, HP: Hemiparesis, GD: Gait Disturbance, Max: Maximal, Hypes: Hypesthesias, Spont: Spontaneous, Sx: Symptomatic, SAW: Spinal Arachnoid Web, SA: Subarachnoid, IM: Intramedullary, DL: Dorsolateral, SCM: Split Cord Malformation, CE: Cauda Equina, , CES: Cauda Equina Syndrome, ACF: Anterior Corpectomy/Fusion, BLE: Bilateral Lower Extremities, Myelop: Myelopathy, Incont: Incontinence, Abd: Abdominal, Fx: Fractured, VBody: Vertebral Body

continued cerebrospinal fluid (CSF) leak into loculated intradural/subdural (i.e. intradural arachnoid cysts [IDAC] constitute 10% of all arachnoid cysts), or extradural cysts (i.e. extradural arachnoid cysts [EDAC] represent 1-2% of all spinal tumors) [Table 1].^[12,17,20] Clinical syndromes include intracranial hypotension (i.e. intermittent chronic headaches frequently with a postural component due to continued drainage of CSF into cysts), radiculopathy and/or myelopathy. Cervical or thoracic Magnetic Resonance Images (MR)/Myelo-Computed Tomography (Myelo-CT) studies typically demonstrate the “double cord” and/or “windsock/intussusception” signs, while those with lumbar circumferential subdural SAC may demonstrate the “fake arachnoiditis sign”.^[15,22] Patients with neurological deficits should undergo early surgery (i.e. optimally 0-< 24 hrs after the onset of symptoms/signs). Surgery for IDAC/EDAC usually includes a laminectomy with partial/total resection or marsupialization/fenestration of the cyst wall, obliteration of the fistulous subarachnoid communication, with or without placement of a shunt.^[2,3,6,18,20,22]

EDAC AND IDAC VERSUS SPINAL SUBARACHNOID WEB (SAW)

MR followed by Myelo-CT studies document that the majority of IDAC or EDAC are dorsally located, resulting in severe nerve root, cervical/thoracic cord, and/or cauda equina compression [Table 1].^[2-23] In the cervical/thoracic regions they often demonstrate the “double cord” or “windsock signs”.^[14] Alternatively, in the lumbar spine they may contribute to the “fake arachnoiditis sign,” signaling the presence of a circumferential subdural arachnoid cyst that centrally traps the cauda equina, making it appear as if adhesive arachnoiditis (AA) is present; in fact, AA is absent. Further, for IDAC some of these studies will document multiple intra-arachnoid septations.^[8] Notably, the Myelo-

CT examination is the study of choice to most directly/best document the fistulous tracts communicating between the subarachnoid space and either the IDAC or EDAC.^[3,5]

Additionally, IDAC and EDAC must be differentiated from spinal arachnoid webs (i.e. SAW). These occur when abnormal arachnoid membranes within the subarachnoid space contribute to cord/root “displacement” and/or obstruction of CSF flow.^[1]

INTRAOPERATIVE DURAL INJURY RESULTING IN POSTOPERATIVE IATROGENIC ARACHNOID CYSTS

Postoperative traumatic/iatrogenic arachnoid cysts are rare. Notably, establishing the diagnosis of these lesions utilizing MR and/or Myelo-CT studies is critical so that they can be effectively treated early (i.e. optimally 0-< 24 hours after the onset of symptoms/signs), prior to the onset of a fixed neurological deficit [Table 1].^[1-23] Surgical management usually includes partial/total cyst resection/removal, or marsupialization/occlusion of the communication between the IDAC or EDAC. To attain a water-tight arachnoid IDAC/EDAC fistulous closure often requires a primary sutured repair (i.e., typically using 7-0 Gore-Tex sutures) with or without attendant microdural staples, muscle patch grafts, adjunctive patches (i.e. sheep pericardium), and/or the addition of microfibrillar collagen, and a fibrin sealant. Clearly, a watertight closure should be confirmed immediately after the sutured-repair, not just after the adjunctive microfibrillar collagen/fibrin sealants have been applied. Rarely, additional cyst/subarachnoid, cyst/pleural, cyst/peritoneal shunts, or lumboperitoneal shunts may be warranted to divert residual loculated fluid collections within these SAC.^[2,6]

ETIOLOGY OF INTRADURAL EXTRAMEDULLARY/SUBARACHNOID/SUBDURAL ARACHNOID CYSTS (IDAC)

Intradural/extramedullary or subdural/extra-arachnoid arachnoid cysts (IDAC) account for 10%^[12] of all arachnoid cysts [Table 1].^[8,12,14,15,19,23] Many authors warn that IDAC should be strongly considered in patients who present with the chronic, subacute, or acute onset of a progressive cervical quadriplegia, thoracic paraparesis, or lumbar cauda equina syndrome.^[7,8,10,12,14,15,19] IDAC lesions occur due to the disruption of the arachnoid membrane with/without a dural laceration, either on a congenital, spontaneous, traumatic (i.e. including iatrogenic/surgical), or infectious basis. This results in arachnoid out-pouching into the subarachnoid or subdural spaces (exclusive of the extradural compartment) where they can loculate and continue to fill via a ball-valve mechanism.

Posterior surgical management of congenital IDAC

Two studies involving congenital IDAC warranted surgical resection, and fenestration; one patient required shunting to the subarachnoid space [Table 1].^[7,18] Filho *et al.* (2009) diagnosed a congenital giant dorsal T1-T12 intradural/extramedullary arachnoid cyst (IDAC); a focal T3-T6 laminectomy with durotomy, including wide resection of the posterior IDAC wall plus fenestration and shunting to the subarachnoid space, resulted in significant postoperative improvement within the 1st postoperative year.^[7] In Rahimizadeh *et al.* (2013), two patients <20 years of age presented with neck pain/upper extremity weakness due to an MR-documented anterior cervical IDAC (i.e. presumed congenital).^[18] They successfully underwent laminectomy and laminoplasty respectively, allowing for “wide” cyst-fenestration/marsupialization, and cyst wall removal.

Anterior cervical corpectomy/fusion for managing a congenital IDAC

One study uniquely utilized a direct anterior cervical approach (i.e. anterior corpectomy/fusion ACF) to resect a C3-C4 IDAC [Table 1].^[19] Shrestha *et al.*'s (2017) 20-year-old presented with a spastic quadriplegia attributed to an MR-documented anterior C3-C4 IDAC; following a C3-C4 ACF involving resection/marsupialization of the cyst wall, the patient neurologically improved.^[19]

Posterior surgical management of spontaneous or traumatic (non-surgical) IDAC

Three studies, involving 6 patients were variously attributed to spontaneous or non-surgical/non-traumatic IDAC [Table 1].^[8,10,14] Gomez *et al.* (2011) studied 4 patients with

compressive thoracic dorsolateral (3 patients) or ventral (1 patient) IDAC; surgery, performed in 3 cases (i.e. one died of cardiovascular disease prior to surgery) consisted of laminectomies involving cyst fenestration.^[8] Although two patients developed recurrent postoperative MR findings of residual IDAC, only one with clinical worsening required a reoperation consisting of arachnoid cyst/subarachnoid shunt placement. As of 2015, Min *et al.* evaluated a 43 year old female with a left hemiparesis attributed to an MR-documented massive C6-T12 IDAC.^[14] The MR clearly showed IDAC's classical “double cord sign” along with demonstrating multiple intradural cyst septations resulting in marked cord compression.^[14] After the initial T5-T7 laminectomy for resection of the cyst wall (i.e. level of maximal cord compromise), the patient developed a new right-sided C8/T1 deficit. This warranted a second operation consisting of a right C6-T1 laminectomy for extensive cyst resection/marsupialization; it resulted in significant neurological improvement. In Himes *et al.*, (2019) a 60 year old female presented with a one year history of increased pain between the shoulder blades uniquely relieved when supine (i.e. postural hypotension).^[10] When the MR documented a dorsal T3-T7 IDAC, she underwent “selective laminoplasties” at the T3 and T7 levels for excision of the extremes of the cyst. At surgery, they passed a lumboperitoneal shunt catheter between the two ends of the cyst to confirm communication; the catheter was then removed. Seven months later, the MR showed “near complete resolution of the cyst,” and the patient was asymptomatic.

Posterior surgical management of traumatic surgical durotomies resulting in IDAC

In three studies, traumatic surgical durotomies resulted in IDAC requiring surgical resection, marsupialization, and cyst wall fenestration [Table 1].^[12,15,23] In 2009, Nottmeier *et al.* had two patients sustaining incidental durotomies during lumbar surgical procedures resulting in MR-documented IDAC with cauda equina syndromes; both of their symptoms improved following intradural exploration and fenestration of the IDAC cyst wall.^[15] In Lee *et al.* (2019) a 40 year old male had originally undergone a L4-L5 placement of an interspinous device (ISD).^[12] Ten years later, he presented with low back and right lower extremity pain in the S1 distribution. His complaints were attributed to an X-ray-documented broken ISC. Further, the MR showed multiple IDAC “loculated cysts” filling the lumbar canal from L1-S1. After a laminectomy and excision/fenestration of the multiple loculated arachnoid cysts, the patient neurologically improved. In Zapata *et al.* (2020), a patient with a traumatic IDAC was successfully treated with surgical decompression, cyst removal, and fenestration.^[23]

ETIOLOGY OF EDAC

EDAC are rare, and may be due to a congenital anomaly, appear spontaneously, or occur secondary to infection

[Table 1].^[1-23] Ozdemir *et al.* (2019) noted that EDAC represent 1% of all spinal tumors, and likely arise following herniation of the arachnoid membrane through a typically small arachnoid/dural defect (i.e. likely the result of extension of arachnoid into the epidural compartment).^[17] EDAC fill and enlarge based upon a one-way, ball-valve mechanism of the fistulous communication between the subarachnoid and epidural spaces; in short, CSF flows into the cyst, but becomes entrapped, and cannot exit.

Posterior surgical management of presumed post-traumatic EDAC

EDAC comprise 1% of all spinal tumors, and are typically found at the thoracic/thoraco-lumbar regions, but only rarely (3%) occur in the cervical spine [Table 1].^[17] EDAC are typically found dorsal to the cervical/thoracic cord or cauda equina, and may be post-traumatic in origin [Table 1].^[5,11,17] In 2013, Choi *et al.* had two patients, ages 59 and 51, who presented with myelopathy attributed to EDAC located at the T12-L3 and T1-L3 levels respectively; both patients did well following “focal laminectomies” with cyst fenestrations, and primary dural-epidural fistulous repairs (i.e. the latter patient’s Myelo-CT confirmed the fistulous site).^[5] They emphasized the benefits of performing more limited decompressions directly over the EDAC cysts and dural-epidural fistulous sites rather than performing extensive laminectomies covering the entire expanse of the EDAC. When Kong *et al.* (2013) myelopathic patient with a post-traumatic dorsolateral-right sided T11-L3 EDAC underwent focal occlusion of the dural fistula located between the T11 and T12 nerve root sleeves, the patient improved.^[11] Ozdemir *et al.* (2019) reported a patient with an EDAC at the cervicothoracic junction that resulted in cord compression/myelopathy that favorably responded to early surgical intervention.^[17]

Posterior surgical management of congenital EDAC

Congenital EDAC typically appear in younger patients, and may occur in conjunction with other vertebral anomalies [Table 1].^[3,9,13,20,21] Tureyen *et al.* (2009) had a 25 yo male with 1 year of low back/bilateral leg pain attributed to an MR-documented dorsal T12-L2 EDAC; he did well following a focal T11-L2 laminotomy/laminoplasty accompanied by total resection of the cyst wall, and suturing/application of fibrin sealant to the T12 fistula site.^[21] Habibi *et al.* (2016) presented 7 patients, averaging 56.7 mos of age (range 18–119 months), who presented with paraparesis/urological dysfunction attributed to congenital sacral EDAC with congenital split cord malformations/tethered cords, and 6 syrinxes; fistulas were successfully sutured/ligated (6 patients) or occluded/repared (1 patient).^[9] In 2017, Menezes *et al.* presented a 14 year old male with thoracic myelopathy and a dorsal T7-L2 EDAC whose mother had previously had surgery for a dorsal L2-L4 EDAC and L3/4 disc; following decompression/

fenestration, this 14 year-old did well.^[13] Singh *et al.* (2019) 13 congenital EDAC patients averaging 52.2 mos of age were all successfully treated with total (11 patients: 84.6%) or partial (2 patients: 15.3%) cyst wall removals, and complete occlusion of their fistulous tracts [Table 1].^[20] Bowman *et al.* (2020) treated a 38-year-old female with an “atypical” congenital EDAC at the L3-L5 levels.^[3] The MR documented multiple lumbar loculated EDAC cysts along with chronic erosion/scalloping of the L4 vertebral body and pedicle [Table 1]. Here, the Myelo-CT best documented the specific location of the arachnoid-epidural fistula responsible for filling the cyst which was successfully resected along with removed of the cyst itself during the surgery.

INTRAMEDULLARY SPINAL ARACHNOID CYST (IMAC)

Only very rarely are intramedullary SAC reported.^[16] In 2014, Novegno *et al.* (2014) observed just 14 cases of IMAC reported in the literature, and added their 31 year old female to this list [Table 1].^[16] Their patient had a 7 year history of a progressive myelopathy that had worsened over 3 months; she presented paraparetic with urinary incontinence due to a T11-T12 IMAC [Table 1]. She underwent a T11-T12 laminectomy for fenestration of the cyst. Six months postoperatively, she could ambulate without assistance, and her urological symptoms resolved. The MR 2 years later, showed resolution of the lesion. Here the authors strongly recommended early surgery for IMAC due to their increased risk of permanent significant neurological sequelae with delayed surgery.

DIFFERENTIATING SAW FROM IMAC, IDAC, AND EDAC

SAW may be traumatic or atraumatic in origin, and are due to the formation of abnormal arachnoid membranes within the subarachnoid space [Table 1].^[1] Classical MR findings may include extramedullary transverse bands extending to the dorsal cord resulting in the “Positive Scalpel Sign” (i.e. dorsal cord compression attributed to a loculated CSF collection that looks like a dorsally-pointing scalpel blade). As SAW can contribute to the obstruction of normal CSF flow, they can result in root, cord, or cauda equina compression, and/or syrinxes. Their management usually includes surgical excision of the thick arachnoid membrane. Ali *et al.* (2018) reported 31 SAW cases from the literature, 13 of which were surgically confirmed; to this they added their own case of a 70 year old with the acute onset of left leg weakness resulting from a fall.^[1] When the 70-year-old patient’s MR showed the “Positive Scalpel Sign” at the T3-T4 level resulting in dorsal to ventral cord compression, the patient successfully underwent a laminectomy for removal/fenestration of the intra-arachnoidal web with resolution of his complaints.

SHUNTING OPTIONS FOR IDAC, EDAC, AND IMAC

Several articles mention placing different types of shunts for IDAC and EDAC [Table 1].^[2,7,8] In Filho's case report, the patient had a giant T3-T6 IDAC that required a laminectomy/decompression, fenestration of the cyst, plus the primary placement of an IDAC-subarachnoid shunt.^[7] One of Gomez's 3 patients with IDAC who initially underwent a thoracic laminectomy with cyst fenestration, developed a postoperative syrinx that warranted placement of a syrinx-subarachnoid shunt.^[8] In Boody *et al.* the patient presented with a lumbar EDAC and cauda equina syndrome; following the initial cyst resection with occlusion/repair of the cyst-dural fistula, the patient eventually required a lumboperitoneal shunt.^[2]

CONCLUSION

IDAC, EDAC, and IMAC occur due to outpouchings/entrapment of the arachnoid membrane fed by one-way fistulous tracts between the subarachnoid space and these cysts [Table 1].^[1-23] The location, extent, and/or severity of neural/root/cord/cauda equina compression documented on MR/Myelo-CT studies, determines their clinical presentation (i.e. with intracranial hypotension, radiculopathy, and/or myelopathy). Cervical or thoracic MR/Myelo-CT findings include the "double cord" or "windsock/intussusception" signs. In the lumbar spine the "fake arachnoiditis sign" may be seen with circumferential cauda equina IDAC (i.e. due to 360 degree herniation of the arachnoid membrane into the subdural space).^[15,22] Early surgery (i.e. optimally 0- < 24 hours after the onset of symptoms/signs) is typically advocated and includes a laminectomy, partial/total cyst resection/fenestration with ligation/occlusion of the subarachnoid-cyst fistula with or without shunt placement.^[2,3,6-8,18,20,22]

Declaration of patient consent

Patient's consent not required as there are no patients in this study.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ali BH, Hamilton P, Zygmunt S, Yakoub. Spinal arachnoid web-a review article. *J Spine Surg* 2018;4:446-50.
2. Boody B, Lucasti CJ, Schroeder GD, Heller JE, Vaccaro AR. Extradural arachnoid cyst excision. *Clin Spine Surg* 2019;32:E403-6.
3. Bowman JJ, Edwards CC. Extradural arachnoid cyst with bony erosion: A rare case report. *J Spine Surg* 2020;6:736-42.
4. Choi JY, Kim SH, Lee WS, Sung KH. Spinal extradural arachnoid cyst. *Acta Neurochir (Wien)* 2006;148:579-85.
5. Choi SW, Seong HY, Roh SW. Spinal extradural arachnoid cyst. *J Korean Neurosurg Soc* 2013;54:355-8.
6. Deutsch H. Thoracic arachnoid cyst resection. *Neurosurg Focus* 2014;37 Suppl 2:Video 4.
7. Filho SC, da Silva HB, de Albuquerque LA, de Almeida JP, Santos FP, Sciubba DM. Giant intradural extramedullary arachnoid cyst of the thoracic spine. *J Clin Neurosci* 2009;16:1369-71.
8. Gomez E, Quiles AM, Pedraza S. Spinal arachnoid cyst as an infrequent cause of spinal cord compression. *Neuroradiol J* 2011;24:535-45.
9. Habibi Z, Hanaei S, Nejat F. Sacral extradural arachnoid cyst in association with split cord malformation. *Spine J* 2016;16:1109-15.
10. Himes BT, Kerezoudis P, Rajjoub KR, Shepherd DS, Bydon M. Resection of an extensive thoracic arachnoid cyst via less-invasive targeted laminoplasties. *Int J Neurosci* 2019;129:397-400.
11. Kong WK, Cho KT, Hong SK. Spinal extradural arachnoid cyst: A case report. *Korean J Spine* 2013;10:32-4.
12. Lee HG, Kang MS, Na YC, Jin BH. Spinal intradural arachnoid cyst as a complication of insertion of an interspinous device. *Br J Neurosurg* 2019 Sept 24: 1-5.
13. Menezes AH, Hitchon PW, Blouhy BJ. Symptomatic spinal extradural arachnoid cyst with cord compression in a family: Case report. *J Neurosurg Spine* 2017;27:341-5.
14. Min WK, Kim JE. Extensive spinal intradural arachnoid cyst exhibiting a "double cord sign" on magnetic resonance imaging. *J Orthop* 2015;13:110-4.
15. Nottmeier EW, Wharen RE, Patel NP. Iatrogenic intradural spinal arachnoid cyst as a complication of lumbar spine surgery. *J Neurosurg Spine* 2009;11:344-6.
16. Novegno F, Umana G, Muro LC, Fraioli B, Fraioli MF. Spinal intramedullary arachnoid cyst: Case report and literature review. *Spine J* 2014;14:e9-15.
17. Ozdemir M, Kavak RP, Gulgonuf N. Spinal extradural arachnoid cyst in cervicothoracic junction. *Spinal Cord Ser Cases* 2019;5:45.
18. Rahimizadeh A, Sharifi G. Anterior cervical arachnoid cyst. *Asian Spine J* 2013;7:119-25.
19. Shrestha P, Shrestha P, Deykota UP. Excision of an anterior intradural arachnoid cyst of the cervical spine through central corpectomy approach. *Eur Spine J* 2017;26 Suppl 1:154-7.
20. Singh S, Bhaishora KS, Sardhara J, Kanti K, Attri G, Mehrotra A, *et al.* Symptomatic extradural spinal arachnoid cyst: More than a simple herniated sac. *J Craniovertebr Junction Spine* 2019;10:64-71.
21. Tureyen K, Senol N, Sahin B, Karahan N. Spinal extradural arachnoid cyst. *Spine J* 2009;9:e10-5.
22. Zanon BC, Kanas M, Joaquim MA, Martins, DE, Waichenberg M, Astur N. Posttraumatic arachnoid cyst in the thoracic spine with medullary compression: Case report. *Ver Bras Orthop (São Paulo)* 2021;56:114-7.
23. Zapata HD, Monsalvez EU, Arribas PJ, Carretero MD, Fernandez CS, Garcia AF, *et al.* Spinal cord compression

secondary to traumatic intradural arachnoid cyst. *Neurocirugia (Astur: Engl Ed)* 2020;31:195-200.

Commentary

Preventing Arachnoid Cyst Recurrence

Intradural spinal arachnoid cysts may be dormant for many years and go unnoticed until gait difficulties and other signs of cord compression and myelopathy develop. At some point CSF dynamics change and the cyst becomes larger and pressurized. At that point you have a typically slowly growing mass in a confined space, and the first thing that gets compressed due to the pressure differential is the spinal cord. Once you suspect cord compression the workup and treatment need to be expedited. MRI is usually the first choice of diagnostic studies given that it is noninvasive. But if MRI does not show you the top and bottom of the cyst clearly or the cyst is multiloculated, strong consideration should be given for obtaining a myelogram with CT. If there is delayed filling of the cyst post-myelogram, you potentially may need a delayed CT also.

It is important to look for such loculations that could make your fenestration surgery less effective. If there is a simple single cyst, knowing the top and bottom of the cyst can enable the surgeon to do fewer laminectomies by approaching the cyst at each end for fenestration, and resection of the cyst. Generous resection and removal of the cyst along the lateral wall away from the cord is safest, and still gives a low recurrence rate. You should be able to irrigate between the top and the bottom of the cyst resection. Passing a catheter between sites may break up loculations within the cyst, but will also yield a higher risk of cyst recurrence, and risk potential cord injury (i.e. depending on the type of catheter that is passed, and the number of passes needed to open the cyst fully). If you suspect the cyst is still loculated, it is recommended to perform additional levels of laminectomy to allow for better exposure, and a more thorough cyst resection. A watertight dural closure is essential. Neurologically patients typically have good outcomes if surgery is done early in the course of the cord compression.^[2, 3, 6] Radiographic follow-up should be performed if any signs of myelopathy recur or if there is concern for cerebral hypotension.

VENTRAL INTRADURAL CYSTS

Ventral Intradural cysts are thought to occur by a different mechanism and with resection, arachnoid elements are typically lacking. These cysts are thought to be formed from ventral dural defects that dissect the dura. These may also be associated with ventral cord herniation and are much harder to safely repair.^[4]

EXTRADURAL ARACHNOID CYSTS

Extradural spinal arachnoid cysts can also cause cord compression and typically require multilevel laminectomy to expose the extent of the cyst to determine the site(s) of CSF communication so that the dural defect can be secured and occluded to minimize the chance of cyst recurrence.

CONCLUSION

Although uncommon, treatment with resection or fenestration of the arachnoid cyst (both IDAC and EDAC) offer a good prognosis with a low recurrence rate.^[1,5]

REFERENCES

1. Alvisi C, Cerisoli M, Giulioni M, Guerra L. Long-term results of surgically treated congenital intradural spinal arachnoid cysts. *J Neurosurg* 1987;67:333-5.
2. Mirza AB, Bartram J, Sinha S, Gebreyohanes A, Boardman T, Vastani A, *et al.* Surgical management and outcomes in spinal intradural arachnoid cysts: The experience from two tertiary neurosurgical centres. *Acta Neurochir (Wien)* 2021 Oct 27 (ahead of print)
3. Mirza AB, Bartram J, Vastani A, Gebreyohanes A, Al Banna Q, Lavrador JP, *et al.* Systematic review of surgical management of spinal intradural arachnoid cysts. *World Neurosurg* 2021 Oct 30. (ahead of print)
4. Ball BG, Luetmer PH, Giannini C, Mokri B, Kumbar N, Piepgras DG. Ventral "spinal epidural meningeal cysts"-not epidural and not cysts? Case series and review of the literature. *Neurosurgery* 2012;70:320-8.
5. Fam MD, Woodroffe RW, Helland L, Noeller J, Dahdaleh NS, Menezes AH, *et al.* Spinal arachnoid cysts in adults: Diagnosis and management. A single-center experience. *J Neurosurg Spine* 2018;29:711-9.
6. Petridis AK, Doukas A, Barth H, Mehdorn HM. Spinal cord compression caused by idiopathic intradural arachnoid cysts of the spine: Review of the literature and illustrated case. *Eur Spine J* 2010;19 Suppl 2:S124-9.

Jamie Baisden, M.D

Department of Neurological Surgery, Professor of Neurological Surgery, Medical College of Wisconsin, Fellow of the American College of Surgeons (FACS), and Fellow of the American Association of Neurological Surgeons (FAANS)

E-mail: jbaisden@mcw.edu

How to cite this article: Epstein NE. Review/Perspective On the Diagnosis and Surgical Management of Spinal Arachnoid Cysts. *Surg Neurol Int* 2022;13:98.