

ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXVIII | No. 2

June 2024

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DOI: 10.33962/roneuro-2024-040



Spinal arachnoid cysts. A case series and review of literature with subgroup comparison of intradural and extradural arachnoid cysts

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ABSTRACT

Objectives: Spinal arachnoid cysts (SAC) are rare lesions. There are many uncertainties regarding details about Intradural and Extradural SAC. We present a series of 12 cases of SAC along with a comprehensive review of the literature. In this review, we discussed the differences between Intradural and Extradural SAC pertaining to demography, pathogenesis, surgical procedures and outcomes.

Methods: We retrospectively collected the data of 12 patients of SAC treated at our Institute from 2012 to 2023. The age, gender, clinical, radiological, surgical data and outcome were noted. An extensive review of the literature was done to analyse and note the differences between Intradural and extradural SAC and their surgical management.

Results: In our series of 12 patients, 7 had Extradural SAC(58.3%), 3 had Intradural SAC(25%) and 2 had Intramedullary SAC(16.7%). . Of the 12 patients, 4 were males and 8 were females (1:2). Age range was 9 to 64 years and the mean age was 34.42 ± 17.71 years. There were four paediatric patients in the series. The most common symptoms reported at presentation are weakness of limbs(81.9%),back pain(36.4%), sensory symptoms(36.4%), radicular pain (18.2%), and bladder disturbances(9.1%). Out of the 12 patients, surgery was done in 11 patients and one patient was managed conservatively.

Conclusions: From the review of the literature, intradural SAC was twice as common as extradural SAC. Ventrally located SACs are more common in Intradural locations as compared to Extradural. The intradural SACs were more commonly located in the thoracic and cervical regions when compared to extradural SACs which were located commonly in the thoracolumbar, thoracic and lumbar regions. Extradural SACs were mostly Primary and the mainstay of treatment is complete or partial excision of the cyst with identification and ligation of the communicating pedicle. Selective laminectomy, partial excision of the cyst and wide fenestration to sustain CSF flow is the recommended treatment in Intradural SAC.

INTRODUCTION

Spinal Arachnoid cysts (SAC) are rare lesions which can cause myelopathy or radiculopathy.^{1,2}Spinal arachnoid cysts were classified into three major types by Nabors et al.: Extradural cysts without spinal

Keywords
spinal arachnoid,
cysts,
intradural,
extradural



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ISSN online 2344-4959
© Romanian Society of
Neurosurgery



First published
June 2024 by
London Academic Publishing
www.lapub.co.uk

nerve root fibers (Type I), Extradural cysts with spinal nerve root fibers (Type II), and Intradural cysts (Type III).³ Type I was further subdivided into two groups: extradural arachnoid cysts (type Ia) and sacral meningocele (type Ib).³ We describe here a series of 12 Spinal arachnoid cysts treated at our Institute between 2012 to 2023.

There were many uncertainties regarding details about intradural and extradural SAC. There was a difference in the opinion of authors regarding the incidence of intradural vs extradural SAC. Some authors claimed that intradural SAC⁴ were common whereas others claimed extradural SACs were more common.⁵ We wanted to clarify this ambiguity along with some other queries regarding SAC. We have done a comprehensive review of literature to find out the demographic, clinical and surgical details and differences between Intradural and Extradural arachnoid cysts.

METHODS

We retrospectively collected the data of patients treated for Spinal arachnoid cysts at our Institute between the years 2012 to 2023. All the demographic, clinical, radiological, surgical data and outcome was collected and analysed. The surgical technique and outcome were described in detail.

Comprehensive review of literature and detailed subgroup analysis of Intradural and Extradural SAC's was done.

We have defined Arachnoid cysts as Primary when there is no identifiable cause (Idiopathic). Secondary arachnoid cysts are defined as cysts secondary to inflammatory reactions related to trauma, subarachnoid haemorrhage, meningitis, post intradural surgery, after CT myelography or post lumbar puncture or epidural anaesthesia in few instances.⁶

Outcome analysis

Odom's criteria were used for evaluation of the surgical results and outcome: Excellent (no pain and no neurological deficit); Good (occasional, mild pain and no neurological deficit and no change in work status); Fair (frequent, persistent pain and slightly improved neurological deficit and some change in work status); Poor (disabling pain and unchanged neurological deficit and unable to work).

Statistics

IBM spss25 software was used for statistical analysis.

OUR CASE SERIES

We describe the results of our series of 12 patients of SAC.

Demography and clinical features

Twelve patients of Spinal arachnoid cysts were treated at our centre during the study period. Details of all the patients are described in **Table 1**. Of the 12 patients, 4 were males and 8 were females (1:2). Age range was 9 to 64 years and mean age was 34.42 ± 17.71 years. There were four paediatric patients in the series. The most common symptoms reported at presentation are weakness of limbs (81.9%), back pain (36.4%), sensory symptoms (36.4%), radicular pain (18.2%), and bladder disturbances (9.1%). Signs of myelopathy are present in 7 out of 12 (58.3%) patients. The duration of symptoms was ranging from 2 days to 6 months.

Imaging features

All the patients underwent MRI spine for diagnosis. Out of the 12 patients, 7 had Extradural SAC (58.3%), 3 had Intradural SAC (25%) and 2 had Intramedullary SAC (16.7%).

Location of the Arachnoid cysts was thoracic in 5 (45.5%) patients, cervical in 3 (27.3%) patients, thoracolumbar in 2 (18.2%) patients and lumbar in 1 (9.1%) patient. All the Extradural SAC were dorsal to the spinal cord/ thecal sac. Both the Intradural SACs were ventral to the cord. In one patient (Case 2) there were associated cervical degenerative disc disease and cord atrophy with myelomalacia changes. CT myelography was done in one patient but we could not delineate the level of communication.

Surgical procedure

Out of the 12 patients, surgery was done in 11 patients and one patient with D12-L2 extradural arachnoid cyst (case 10) was managed conservatively.

Laminectomy and excision of the cyst was done in all the six patients with Extradural cysts. Total excision of an extradural SAC (case 1) was demonstrated in Figure 1. Intradural communication was identified in 5 patients which was ligated and closed. The site of intradural communication was the junction of thecal sac and dural sleeve of nerve root in 4 patients and posterior midline of the thecal sac in 1 patient.

In two patients with Intradural cysts, Laminectomy, fenestration and partial excision of the cyst wall was done. In the third patient (case 12) with intradural cyst, complete excision of the cyst was done. Preoperative MRI images of an intradural SAC(case 9) were shown in Figure 2.

In one patient with intramedullary cyst, myelotomy and partial excision of cyst wall was done where as in other subpial intramedullary cyst, laminectomy and fenestration of cyst was done (Figure 3).

Table 1. Our case series of 12 cases of SAC

S. no	Age/ M/F	Symptoms	Duration	Spinal level (no. of)	Horizontal level (Dorsal/ ventral)	Surgery	Complications	Outcome	Follow up
1	14 /M	Spastic paraparesis	45 days	D8-L1 (6)	Extradural (Dorsal)	D8-D12 Laminectomy and complete excision -Communication seen and closed	NIL	Improved in power Excellent	-6years -No recurrence
2	43 /F	-Band like sensation in lower limbs - Weakness of both upper and lower limbs, difficulty in walking	2 mts. 2 mts	C3-C6 (4)	Extradural (Dorsal)	C3-C6 Laminectomy and complete excision -Communication seen and closed	Patient deteriorated and developed Right hemiparesis (Power-2/5)	-Patient improved to 4-/5 power in 3 months Poor	- 3years& 6 months -No recurrence
3	47 /F	-LBA Left L1,L2 radicular pain -Left LL paraesthesia	15 days	D10-L2 (5)	Extradural (Dorsal)	D11-L1 Laminectomy and complete excision -Communication at L1 closed	NIL	-Pain relieved Excellent	-3 years& 6 months -No recurrence
4	44 /F	Right LL weakness, difficulty in walking -Paraesthesia both LL's k/c/o Pulmonary TB & spinal TB arachnoiditis on ATT since 6 months	3 mts	D9-D11 (3)	Intradural extramedullary (Ventral)	D9-D11 Laminectomy, Partial excision of cyst wall	NIL	Improved in power Fair	-3years& 4 months -No recurrence
5	50 /F	LBA Weakness of both LL's	6 mts 2 mts	D9D10 (2)	Intramedullary	D8-D10 Laminectomy, myelotomy, partial excision of cyst wall	NIL	-Improved in power Good	-3 years -No recurrence
6	64 /F	Upper back pain	10 yrs.	D2-D4 (3)	Extradural (Dorsal)	D2- D4 Laminectomy and complete excision	NIL	Improved in power	-8 years& 2 months

		Numbness both LL's Difficulty in walking, Urinary urgency	6 mts			-communication not seen		Good	-No recurrence
7	9/ F	Paraplegia	2 days	D3- D6 (4)	Extradural (Dorsal)	D3- D6 Laminectomy and complete excision - Communication seen and closed	NIL	-No improvement of power Poor	-9 years & 6 months -No recurrence
8	16 /M	Spastic paraparesis	3 wks.	D4- D6 (3)	Extradural (Dorsal)	D4- D6 Laminectomy and complete excision - Communication seen and closed	NIL	-Improved in power Excellent	-10 months No recurrence
9	30 /F	Spastic quadripareisis, Right upper limb radiculopathy	10 days	C1- C3 (3)	Intradural extramedullary (ventral)	Laminectomy, subtotal excision of cyst wall & communication to subarachnoid space	NIL	Improved in power Excellent	-9 months -No recurrence
10	53 /F	LBA	1 mt.	D12- L2 (3)	Extradural (Dorsal)	Conservative	-	-LBA decreased -Repeat MRI showed no increase in size of lesion	3 years - No progression of symptoms
11	11 /M	quadripareisis	2 mt.	C6 C7 (2)	Intramedullary	C6, C7 Laminectomy and fenestration	NIL	-Power improved Good	-10 years & 6 months -No recurrence
12	32 /M	Back pain, difficulty in walking	1 year	D6 D7 (2)	Intradural extramedullary (dorsal)	D6 -D8 laminectomy and complete excision of cyst	NIL	-Back pain improved Excellent	- 9 months - No recurrence

Complications and outcome

Outcome was excellent in 5 patients, good in 3 patients and fair in 1 patient. Poor outcome was noted in 2 patients. One patient (case 2) developed right hemiparesis postoperatively (grade 2/5). This patient gradually improved in power to preoperative status (4-/5) on follow up over 3 months period. The other patient (case 7) with poor outcome presented with paraplegia did not improve after surgery. There was no CSF leak or wound infections in the series.

Follow up

The average follow up of our patients was 52 months. There was no reported recurrence or onset of new symptoms till the last follow up.

DISCUSSION

Spinal arachnoid cysts are rare and may account for only about 1-3 % of Primary spinal lesions.^{1,2} Spinal arachnoid cysts were described as Spinal Meningeal cysts by Nabors et al. and were classified into three types.

In our literature review of large case series⁵⁻¹³ of Spinal arachnoid cysts it was evident that Intradural arachnoid cysts are two times commoner than Extradural arachnoid cysts and Intramedullary arachnoid cysts are very rare.¹⁴

Ventrally located cysts in relation to the spinal cord were more common in Intradural arachnoid cysts. Ventral location of cysts is seen in about 15-20% of all Intradural arachnoid cysts.^{8,7,12,15}

Extradural ventral cysts are very rare.^{5,11,12,16,17} Similar findings were seen in our present series.

Intradural SACs were more commonly seen in the Thoracic, cervical and cervicothoracic and thoracic levels^{9,6,7,18} whereas extradural SACs were commonly seen in the Thoracolumbar, Thoracic, lumbar and lumbosacral levels.^{11,16,17}

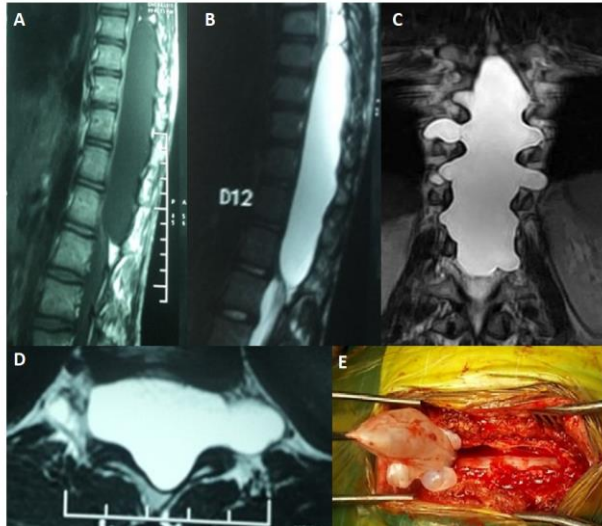


Figure 1. Extradural arachnoid cyst (case 1) extending from D8-L1. Sagittal T1 & T2 weighted (A, B) and coronal, axial T2W MRI images (C, D) demonstrating extension into neural foramina. Intra operative image (E) showing arachnoid cyst being excised and easily separable from underlying dura.

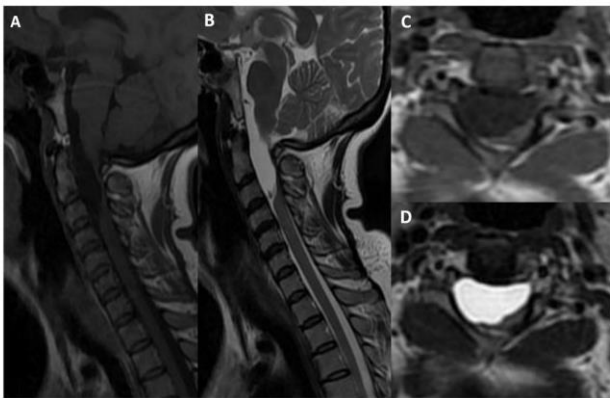


Figure 2. Sagittal T1 and T2 weighted (A, B) and axial T1 and T2W (C, D) MRI images demonstrating an Intradural arachnoid cyst (case 9) at C1 to C3 level with marked thinning of the cord.

PATHOGENESIS

The origin of Extradural arachnoid cysts in most cases is due to herniation of arachnoid through a congenital dural defect or could be a congenital diverticulum of the dura.¹⁹ The dural defect is located most often at the junction of thecal sac and dural

sleeve of the root or in the region of the dural sleeve of the root and less commonly dorsal midline of the thecal sac.¹⁹ Extradural secondary arachnoid cysts are very rare and causes are post traumatic or post surgical dural defects.²⁰ Various theories were proposed for expansion of Extradural cyst and one suggested theory is ball-valve mechanism in the communicating pedicle associated with pulsatile CSF dynamics and one way CSF flow which leads to cyst expansion.²¹ Other proposed mechanisms for cyst expansion are active secretion by cyst wall, Osmosis of water into cyst due to hyperosmolar cyst content.¹

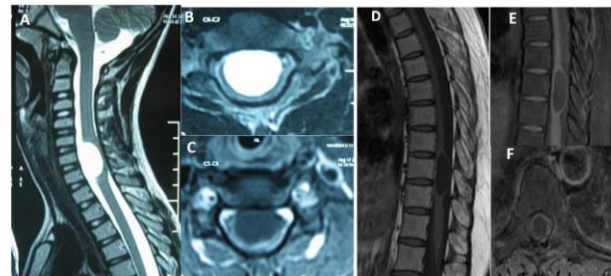


Figure 3. Sagittal and axial T2 weighted (A, B) and Axial T1 weighted (C) images of Case 11 demonstrating a Subpial intramedullary arachnoid cyst. T1 weighted sagittal (D), Sagittal T1 post contrast (E), Axial T1 post contrast (F) images of Case 5 showing a D9, D10 intramedullary arachnoid cyst with expansion of cord.

The pathogenesis of Primary (congenital) Intradural arachnoid cysts includes various theories and Perret et al.²² proposed that they arise from or within the septum posticum. Septum posticum is a midline membrane dividing the posterior cervical and thoracic subarachnoid space longitudinally. In our analysis most of the Intradural arachnoid cysts are seen in the thoracic and cervical location. But this theory does not explain the origin of ventral cysts. Another theory proposes that congenital arachnoid cysts originate from deviations in arachnoid trabeculations during the embryonic period.⁴⁵ The etiology of Secondary Intradural arachnoid cysts could be post traumatic, SAH, post surgical, lumbar puncture and infective causes.^{6,7,18}

Secondary arachnoid cysts are most commonly Intradural and Extradural secondary arachnoid cysts are very rare. Klekamp et al.⁶ described 130 cases of Intradural SAC's out of which 109 were primary and 21 were secondary. They have highlighted certain differences between Primary and Secondary Arachnoid cysts. Primary arachnoid cysts were mostly seen in the thoracic level (94%) whereas

Secondary Arachnoid cysts were seen in the thoracic spine, the cervicothoracic, or the thoracolumbar junction. Secondary Arachnoid cysts were located in a more lateral or even ventral in relation to the spinal cord but only one Primary Arachnoid cyst was located ventrally. They also observed that progression-free survival for 10 years following surgery was 83% for primary compared to 15% for secondary arachnoid cysts. Associated syrinx was more common in Secondary arachnoid cysts. 18 patients with secondary arachnoid cysts (86%) showed a syrinx and 50 patients with primary (46%) demonstrated a syrinx. Baig Mirza et al.⁹ also described in their series that a syrinx being present in 5 out of their 11 secondary SAC cases.

RADIOLOGY

MRI is the diagnostic imaging of choice as it can delineate exact location of the cyst (extra or intradural), extent and relationship to the cord (dorsal vs ventral)²⁴. Arachnoid cysts are similar to that of CSF signal intensity on T1 and T2 weighted sequences.

Extradural cysts show epidural fat capping of the lesion at the superior and inferior poles. The lesions cause spinal cord or thecal sac compression and vertebral body scalloping, expansion of the neural foramina bilaterally, thinning of pedicles which suggest longstanding mass effect from the cyst.²⁵

Neo et al.²⁶ have used Cine-MRI sequence for successful identification of the communication site(dural defect) which shows a pulsating turbulent flow void in an Extradural arachnoid cyst. Funao et al¹⁷ have used Myelography, CT myelography (CTM), and cine MRI to delineate the communication site between the spinal subarachnoid space and the arachnoid cyst cavity. Pooling of the contrast medium at the early phase in Myelography and CT myelography revealed the communication site between the spinal subarachnoid space and the cyst in 7 out of 12 cases in their series. Whereas Cine MRI revealed the communication site in 2 of these 7 patients.

Morizaneetal.²⁷have done CT Myelography(CTM) and Cine MRI to detect the communication site in 6 of their 12 Extradural SAC's. They also suggested to do CTM first and to do cine MRI if the communication site could not be detected by CTM.

Nakagawa et al.²⁸ have used 3D constructive interference in steady state MRI (3D CISS MRI) for visualizing the communication site.

MRI with CISS-3D sequences is also very precise in detection of intracystic septae, trabeculae, and intradural cystoid formations as compared to T2W imaging.²⁹ The differential diagnosis of Intradural cysts includes neurenteric cysts, neuroepithelial cysts, epidermoid cysts, dermoid cysts, cystic schwannomas, and parasitic cysts.⁶

SURGERY

In Extradural arachnoid cysts, the mainstay of management is complete excision of the cyst with closure of dural defect and obliteration of communicating pedicle.^{2,3,10} In case of dense adhesions of cyst wall with dura, partial or subtotal removal of the cyst wall, with ligation of the communicating pedicle is recommended.^{3,10} Selective laminectomy, incision of the cyst wall and closure of the dural communication has been done by few authors with good neurological outcomes and cyst resolution.

Selective laminectomy or hemilaminectomy, incision of the outer cyst wall and closure of the dural defect microscopically was done by Xu et al.¹⁶ in their series of 10 patients. There was shrinkage or even complete disappearance of residual cysts in this series with an average follow up of 13.2 months.They have not identified any recurrence of cyst with this technique.

Funao et al¹⁷ reported 12 cases of Extradural SAC. Laminectomy and total resection of the cyst was done in 7 patients and closure of the dural defect without cyst resection was done in 5 patients. In the later group, preoperative identification of the communication site was done by CT myelogram and Cine MRI . They performed a selective laminectomy at that level, opening of the dorsal cyst wall, and closure of dural defect with the aid of a flexible neuroendoscope. They observed no difference in functional recovery between the two groups but the mean postoperative kyphotic angle was more (statistically significant) in the patients treated by total resection of the cyst.

Morizane et al.²⁷described 12 patients of Extradural SAC out of which 6 patients underwent massive laminectomy along with total resection of the cysts. In the remaining 6 patients in whom dural defects could be detected preoperatively, partial

laminectomy and closure of the dural defects without resection of the cysts was done. Selective laminectomy or hemilaminectomy with closure of dural defect is recommended as compared to massive laminectomy and total cyst excision in view of similar results, reduced operative time, less bleeding, less muscle damage, and less incidence of postoperative kyphosis.

If the communication site is not detectable preoperatively, they advocated selective laminectomy and closure of the defect at the mid-level vertebrae affected by the cyst or at the T12-L1 level in patients with a cyst at the thoracolumbar level.

Hatashita et al³⁰ proposed that closure of the communication site is not important if total removal of the cyst could be done. Hence total Excision of the cyst can be done in cases where communication site cannot be detected either by preoperative imaging or intraoperatively.

The goals of surgery in Intradural SAC are decompression of the cord, establishing sustained flow of CSF and preventing postoperative adhesions⁶. Selective laminectomy, partial excision of cyst and wide fenestration to sustain CSF flow is the recommended treatment in Intradural SAC.^{6,8} Klekamp et al.⁶ and Schmutzer et al.⁸ recommended resection of upper pole or upper half of the cyst with fenestration. Partial excision or fenestration was also recommended in ventrally located cysts or adherent cysts.^{3,13} Duraplasty with graft was recommended by few authors^{6,7} to prevent post operative adhesions and also achieve the goal of unobstructed CSF flow.

Complete excision of the Intradural SAC was recommended by few authors to prevent recurrence.^{3,7} Complete excision of the Intradural SAC can be done wherever possible in case of short segment cysts extending not more than 3 vertebral levels.⁶

Schmutzer et al.⁸ observed that there was no significant difference in terms of size reduction of the cyst or clinical outcome between the two groups of total cyst excision vs partial excision and fenestration. Cystoperitoneal shunt was a treatment option in few cases of recurrent intradural cysts.^{8,13}

Endo T et al¹⁸ have done Endoscopic assisted surgery in 6 of their 11 cases of Intradural SAC's. In all these six patients, 2 to 3 level hemilaminectomy or laminectomy and microsurgical removal of part of the cyst wall was done. Then a flexible endoscope of

outer diameter 2.5 mm was introduced to inspect the cranial and caudal end of the cyst and further fenestration was done. The endoscope was passed forward to exit from the cyst cavity into the subarachnoid space. Endoscope was also used to inspect the area ventral to the cord and to look for adhesions. They concluded that the number of levels of laminectomy were significantly lower and operative times were shorter in the endoscopically treated group.

Limited laminectomy or Laminoplasty whenever possible may decrease the incidence of post operative deformity. Lesions requiring laminectomy of five or more vertebral segments in the cervical and thoracic region may result in kyphosis of the spine.^{1,2}

CONCLUSIONS

From the review of literature, Intradural SAC was almost twice as common than extradural SAC. Ventrally located SAC are more common in Intradural location and Extradural ventral cysts are very rare. Intradural SAC were more commonly located in the thoracic and cervical region when compared to extradural SAC which were located commonly in the thoracolumbar, thoracic and lumbar regions.

Extradural SACs were mostly Primary and mainstay of treatment is complete or partial excision of the cyst with identification and ligation of the communicating pedicle. After preoperative imaging for detection of communication site, Selective laminectomy or hemilaminectomy with closure of dural defect is recommended in Extradural SACs. Total excision can be reserved to cases where dural communication site could not be detected by preoperative imaging or intraoperatively.

Selective laminectomy, partial excision of cyst and wide fenestration to sustain CSF flow is the recommended treatment in Intradural SAC. Total excision can be done if cyst is not more than 3 vertebral segments and not adherent to cord. Cystoperitoneal shunt can be done in few cases of recurrent intradural cysts.

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