

ORIGINAL RESEARCH

Spinal arachnoid cysts: diagnosis and management: A single center experience

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ABSTRACT

Introduction: Spinal arachnoid cysts (SACs) also called arachnoid diverticula, are **uncommon** benign lesions that occur in the spinal axis which are indistinct etiology that present with pain or myelopathy. Diagnosis is made incidentally on imaging studies with varying degrees of specificity. In symptomatic cases treatment involves surgical exploration and relief of neural tissue compression. **Aim:** The aim of this study was to illustrate features of SACs, surgical management, and outcomes. **Methods:** We searched medical records for all SACs in adults in the 10-year period ending in December 2018. Radiology and pathology reports were reviewed to exclude spine cystic disorders like syrinx, hydromyelia, cystic tumors and recurrent or previously treated patients were excluded. Demographic variables (age, sex) and clinical presentation (symptoms, duration, history of infection or trauma, and examination findings) were extracted. Radiological features were collected and interpreted on imaging studies. Operative findings were reviewed to accurate description of the surgical technique. Finally, patient-reported outcomes were analysed at every clinic visit. **Results:** The authors' search identified 10 patients with SACs (mean age at presentation 31.5 years). Eight patients were male, representing an almost 4:1 (male:female) sex distribution. Symptoms comprised back pain (n = 6, 60%), weakness (n = 6, 60%), gait ataxia (n = 8, 80%), and sphincter dysfunction (n = 3, 30%). The mean duration of symptoms was 8 months. Four patients (40%) exhibited signs of myelopathy. All patients underwent preoperative MRI; SACs were located in the thoracic spine (n = 4, 40%), and less commonly in the sacral spine (n = 2, 20%) and cervical/cervico thoracic, thoracolumbar region (n = 1, 10%). Based on imaging findings, the cysts were interpreted as intradural SACs (n = 9, 90%), extradural SACs (n = 1, 10%), or ventral spinal cord herniation (n = 1, 10%); All patients underwent surgical treatment consisting of laminoplasty in addition to cyst resection (n = 6, 60%), ligation of the connecting pedicle (n = 1, 10%), or fenestration/marsupialization (n = 3, 30%). Postoperatively, patients were followed up for an average of 12.2 months (range 6-18 months). Postoperative MRI showed complete resolution of the SAC in 8 of 10 patients. Patient-reported outcomes showed improvement. One patient suffered a CSF leak with delayed wound infection which treated conservatively and one patient developed recurrent spinal arachnoid cyst underwent redo excision. **Conclusions:** Spinal arachnoid cysts are rare disease entities, which are idiopathic, incidentally diagnosed and most of them are asymptomatic needs close clinical observational management. In symptomatic patients with imaging findings suggestive of spinal arachnoid cyst with neural compression, surgical exploration and complete resection is the treatment of choice. Treatment is usually well tolerated, carries low risks, and provides the best chances for optimal recovery.

Key words: Arachnoid cysts, meningeal cysts, meningocele, spinal cysts, congenital

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INTRODUCTION

Spinal arachnoid cysts (SACs) in adults are rare pathological entities that often present with back pain or myelopathy. Theories on the pathogenesis of these cysts, suggesting that they arise from diverticula in the septum posticum or ectopic arachnoid granulations,

have not been substantiated. In most cases, no underlying cause for the SAC can be identified and is considered as an idiopathic SAC¹. In some cases, SACs can develop from arachnoid adhesions following trauma or inflammatory or infectious causes. Surgical etiologies of arachnoid cysts have

also been described after conducting various procedures, such as a lumbar myelography, laminectomy and vertebroplasty. Inflammatory adhesions within the arachnoid are known to form arachnoid webs that can result in direct cord compression and myelopathy. Such adhesions can also form a 1-way valve entrapping circulating CSF, ultimately resulting in the formation of an SAC. Diagnosis is usually suspected on MRI with focal displacement/buckling of the spinal cord and less frequently on direct visualization of a cystic structure within the spinal canal with signal characteristics similar to those of CSF². Better delineation can often be obtained through CT myelography, which, in addition to diagnosis, can gauge the degree of free-flowing CSF between the arachnoid cyst and the subarachnoid space. Management consists of surgical exploration and decompression via total cyst excision, marsupialization, fenestration, ligation of the communication site, or shunting, or a combination of these techniques. Limited series have been published on SACs, and little is known about their natural course and optimal management. The aim of this study was to illustrate features of spinal arachnoid cysts in adults including surgical management and outcomes^{3,4}.

METHODS

We searched medical records for all adult patients (>18 years) with a diagnosis of arachnoid cyst or spinal cyst in the 10-year period. Radiological reports were individually reviewed, and all patients with radiological reports including spinal arachnoid cysts of the spine were included. We excluded other cystic disorders of the spine, such as syrinx, hydromyelia, and cystic tumors. Patients with recurrent or previously treated arachnoid cysts were also excluded as well as patients who were lost to

follow-up. For all included patients, we extracted demographic variables (age at presentation, sex) and clinical presentation (medical comorbidities, presenting symptoms, duration of symptoms, and examination findings). All surgically treated patients underwent clinical follow-up at 6 weeks postoperatively, at 3 and 6 months, and then annually as dictated by clinical progress. Radiological features of the arachnoid cyst were collected from formal radiology report and direct interpretation of imaging studies by the senior author (P.W.H.). In patients who underwent surgical treatment, operative reports and intra operative photomicrographs were reviewed to accurately describe and confirm the surgical technique. Finally, subjective outcomes were collected at every clinical visit using the SF-36.30 Scores from the initial clinic visit and the last follow-up visit were available for 15 surgically treated patients and were used to elucidate patient-reported outcomes.

SURGICAL TREATMENT

Patients were selected for surgery based on the degree of symptoms and correlation to imaging findings (Figs. 1-A,B). Intra operative fluoroscopy was used in all cases to identify spinal level, and laminectomy was then undertaken under electrophysiological monitoring for somatosensory and motor evoked potentials. Exposure was tailored to expose the entire SAC whenever possible, exposing the caudal and rostral boundaries. Tack-up dural sutures were then placed, and the cyst wall was dissected free from the surrounding dura and cord and then resected (Fig. 1-C). In the case of cervico dorsal cysts (Fig. 2), these were also exposed via laminectomy and dissected free of their surroundings before resection of the cyst.

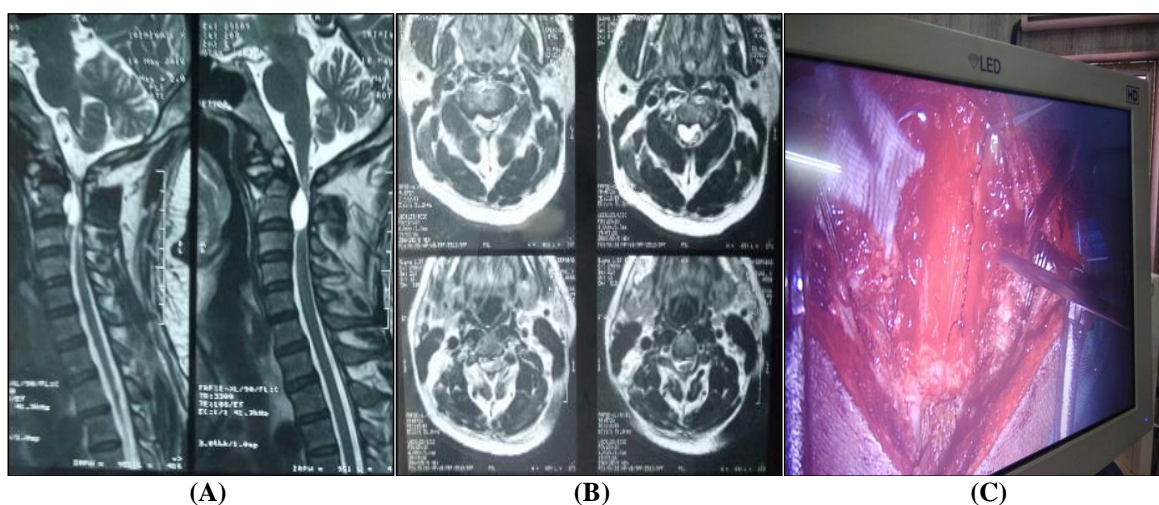


FIG 1: Case 8. A 42-year-old male presented with a 1/2-year history of progressive weakness of all 4 limbs and numbness Rt upper & lower limb with urinary incontinence since 6 months. Sagittal (A) and Axial (B) T2-weighted MR images showing dorsal displacement of spinal cord. Surgical exploration revealed a ventral arachnoid cyst at C2-C3 level, which was completely resected (C)

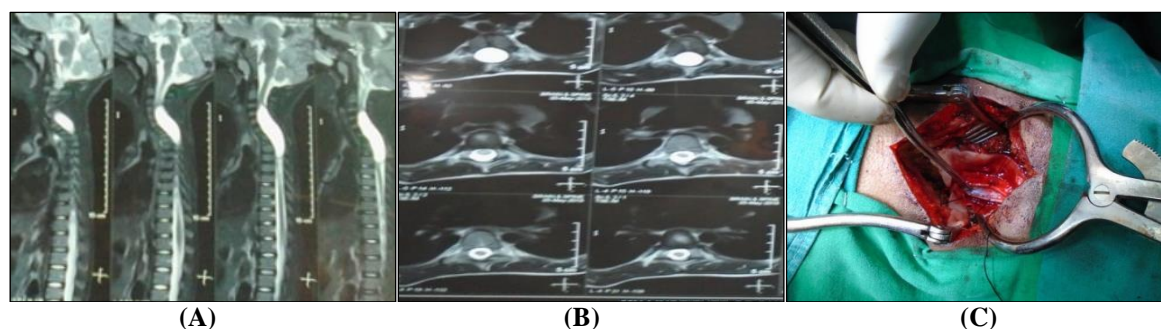


FIG 2: Case 9. A 7-year-old male child presented with a 6 months history of gait ataxia while playing and progressive weakness of all 4 limbs. Sagittal (A) and Axial (B) T2-weighted MR images showing dorsal compression of spinal cord. Surgical exploration revealed a spinal arachnoid cyst at C4-D4 level (cervico dorsal), which was completely resected (C)

RESULTS

DEMOGRAPHIC AND CLINICAL FEATURES

Ten previously untreated patients with SACs were identified from our department. The mean age at presentation was 31.5 years (range 7-60 years). 8 patients were male, demonstrating a sex distribution exceeding a 4:1 ratio. The commonest presenting complaint was recurrent falls (n = 8,80%), followed by back pain (n=6,60%) and lower-extremity paresthesia (n = 2,20%). Symptoms reported at the

initial clinical encounter comprised back pain (n = 6,60%), lower-extremity weakness (n = 6,60%), loss of coordination (n = 8,80%), and sphincter disorder (n = 3,30%). The mean duration of symptoms was 8 months (range 1-18 months). On clinical examination 4 patients (40% of all cases and 30% of cervical and thoracic cysts) exhibited clinical signs of cord compression, including lower-extremity stiffness, hyperactive reflexes, ankle clonus, and/or loss of proprioception (**Table 1**).

Table 1: Demographic and Clinical Features

SL.No.	Age/Sex	Clinical Present N	Durtn months	Pain	Wkns	Gait	Sphinctr	Signs of Cord chngs
1.	50Y/M	LBA	6	P	A	P	A	A
2.	22Y/M	FALL	18	A	P	A	A	P
3.	20Y/M	LBA	12	P	A	A	A	A
4.	60Y/M	LBA+Prsth	3	P	A	P	A	A
5.	50Y/M	FALL	6	P	P	P	P	A
6.	35Y/M	FALL	12	A	A	P	A	A
7.	13Y/M	FALL	1	P	P	P	P	P
8.	42Y/M	Prsth	18	P	P	P	P	P
9.	7Y/M	FALL	1	A	P	P	A	A
10.	16Y/M	FALL	2	A	P	P	A	P

RADIOLOGICAL FEATURES

All patients in our series underwent MRI at the time of presentation. MRI identified the abnormal focal displacement/buckling of the spinal cord or thecal sac. The distribution of these cysts was thoracic in 4 (40%), sacral in 2 (20%), and cervical/cervicothoracic in 2 (20%).

4 patients (45%) had associated intrinsic cord signal changes or myelomalacia. On MRI evaluation preoperative radiological diagnoses comprised intradural arachnoid cyst (n = 9,90%), extradural arachnoid cyst (n = 1, 10%), or ventral spinal cord herniation (VSCH; n = 1, 10%).

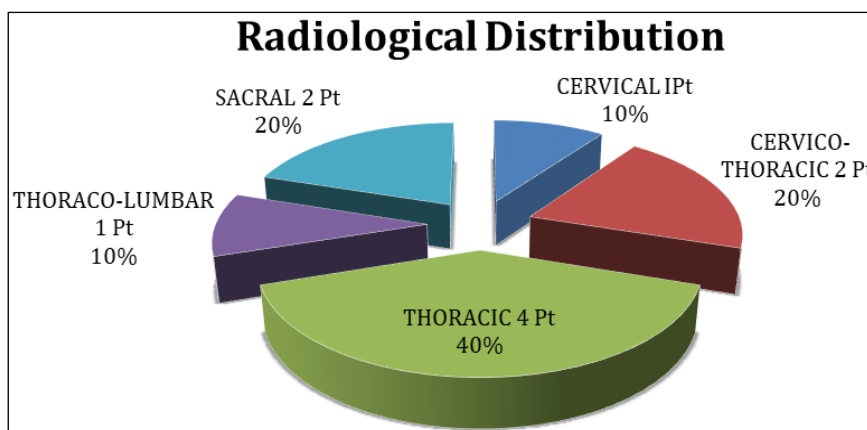


Fig 3: Radiological Features

SURGICAL TREATMENT

All patients in our study underwent surgical treatment (Table 2). All surgical procedures were performed by a single senior surgeon (P.W.H.). In all cases, the cyst was exposed via a posterior-approach laminectomy spanning the entire lesion for an average of 4 levels (range 2-7 levels). The cyst could then be identified using ultrasound and direct inspection after durotomy (Figs.1 & 2). If a dorsal arachnoid cyst could not be identified despite its suggestion on preoperative imaging, the same approach was then used to inspect the canal ventral to the cord in search of a ventral SAC or VSCH. Dorsal extradural cysts (n =1) were encountered immediately after reflection of the laminae. Those were carefully dissected and resected in their entirety. In all cases of extradural cysts, a transdural pedicle/conduit connecting the cyst to the main subarachnoid space could be identified

intraoperatively and was ligated. For dorsal intradural cysts (n =4), midline durotomy and tack-up sutures allowed identification of the cyst dorsal to the spinal cord. Blunt dissection of the cyst avoids violation of the cyst wall so that the cyst can be well visualized in its entirety. In most cases, arachnoid mater forming the cyst wall is thickened and milky white, allowing its identification and excision (Fig.2). The ventral cysts that were surgically treated (n = 1) required extension of the surgical approach via drilling of the medial portion of the ipsilateral facet joints. In the lumbar spine, retraction of the thecal sac allowed for complete resection of a limited extradural arachnoid cyst (case 1). Another 2 spinal arachnoid cysts in our series extended over multiple levels along the cervical and thoracic spine. Adequate exposure and dissection was therefore technically difficult, and both were fenestrated/marsupialized (cases 5 and 9).

Table 2: Outcome

SL.No.	Age/sex	Cli-presentation	Extension	Location	Treatment	Follow-up(Months)
1.	50Y/M	Paraparesis	D11-L3	Extradural	Resection	12
2.	22Y/M	Paraparesis	D3-D9	Intradural	Resection	8
3.	20Y/M	LBA	S1-S3	Intradural	Marsupulisation	12
4.	60Y/M	LBA	S1-S2	Intradural	Marsupulisation	12
5.	50Y/M	Quadriparesis	C7-D2	Intradural	Partial Resection	10
6.	35Y/M	Paraparesis	D7-D12	Intradural	Resection	6
7.	13Y/M	Paraparesis	D3-D8	Intradural	Resection	18
8.	42Y/M	Quadriparesis	C2-C3	Intradural	Resection	16
9.	7Y/M	Quadriparesis	C4-D4	Intradural	Resection	10
10.	16Y/M	Paraparesis	D4-D12	Intradural	Marsupulisation	18

OUTCOMES

Patients who underwent surgery in our series underwent follow-up at 6 weeks, at 3and 6 months, and then annually thereafter based on clinical outcome, with a mean follow-up of 12.2 months (range 6-8 months). Postoperative MRI scans were obtained in 10 patients. Postoperative imaging showed residual cyst in 1 patient withSAC that underwent marsupialization. In the remaining patients, complete resolution of the SAC and relaxation of the spinal cord was radiologically confirmed. Patient-reported outcomes were routinely collected at each clinical

encounter using the SF-36.30 In surgically treated patients, scores obtained at the last clinic follow-up visit were individually compared with those at presentation. Pre- and postoperative scores were available for 10 patients. Direct comparison showed improvement in SF-36 parameters across all domains: average increase in physical functioning (8.4 points, 23%), physical limitation (22.2 points, 30%), emotional limitation (2.2 points, 17%), energy/fatigue (4.2 points, 21%), emotional well-being (4.1 points, 9%), social functioning (10.7 points, 31%), pain (5.7 points, 26%), and general health (1.7 points, 2%).

Surgical complications were encountered in 1 patient with a delayed wound infection that was treated conservatively.

DISCUSSION

DEMOGRAPHIC AND CLINICAL FEATURES

Spinal arachnoid cysts represent a peculiar pathological entity in the differential diagnosis of spinal cystic lesions. The benign disorder is quite rare and has a poorly understood pathogenesis, especially in the absence of a history of trauma or infection. While cystic malformations in the spine are often encountered in the pediatric age group, especially in association with various congenital disorders, only a few reported series in the literature have attempted to characterize SACs. In our series only 1 patient had a remote history of significant trauma, none of the patients had a history of meningitis or prior spine surgery⁵. Also of interest in our series is that over males 4 times more commoner than female. While this could imply a sex predilection when taken in isolation, sex distribution in published series varied significantly. This marked variation could possibly be due to the small series but might actually refute an actual sex predilection (Table 1). The mean presenting age for our patients was 31.5 years (range 7-60 years), which is in keeping with that in other series, reflecting a more common presentation in the 3rd and 5th decades of life. A characteristic feature of SACs is their anatomical distribution, having a strong predilection to the dorsal thoracic spine, usually intradural. This location is invariably more common than all other locations combined. Less common locations are extradural, ventral, lumbar, and, least commonly, cervical (Table 2). Our results reflect those of other published reports (Table 3). The peculiar distribution led authors to suspect that ventral arachnoid cysts could represent a special subgroup with different pathogenesis. Bassiouni *et al.* noted that all patients with ventral SACs in their series were younger, had greater craniocaudal extension, and exhibited intracystic tough fibrous septae that were not encountered with primary dorsal lesions. This led to their hypothesis that ventral SACs represent a special subgroup arising after adhesive arachnoiditis due to traumatic subarachnoid hemorrhage. Except for the greater craniocaudal extension, the other features of ventral SACs were not seen in our series. Another previously reported feature of SACs that was not validated in our series is their association with syringomyelia⁶.

RADIOLOGICAL DIAGNOSIS

Diagnosis of an SAC represents the greatest challenge in the management of this disorder. In most practices, thin-slice MRI represents the imaging modality of choice for diagnosis and follow-up. MRI is the diagnostic test of choice because of its ability to accurately demonstrate the anatomical location and the extent and relationship of the arachnoid cyst to the

spinal cord. MRI also has the advantage over CT myelography of visualizing intrinsic cord changes and atrophy that can be used for prediction of neurological outcome¹¹. The classic scalpel sign commonly seen with SACs represents the abrupt buckling or change in contour of the spinal cord. Other less specific imaging findings like vertebral scalloping or widening of the vertebral pedicle have also been reported, especially with chronic, large SACs. SACs and arachnoid pathologies in general are also known to have a causative association with syringomyelia, which can aid in radiological diagnosis⁸. In our series, however, not a single patient associated with syrinx. Specificity of MRI in diagnosing SACs, however, is debated, especially because of a very similar radiological picture of an even rarer disorder, ventral spinal cord herniation. The modality also has some limitations with SACs given their thin cyst walls and identical signal characteristics of surrounding CSF. Improved visualization of arachnoid webs and cystic formations can be expected with novel high-resolution MRI sequences, such as constructive interference in steady states (CISS) and cine-mode steady-state free-precession imaging (SSFP) studies⁷. However, specificity and superiority to conventional imaging techniques are yet to be validated. Being an invasive imaging technique, CT myelography is usually reserved for diagnosis after inconclusive findings on MRI. The modality has the relative advantage in its ability to localize the connection between the SAC and the main subarachnoid space as well as quantifying the degree of free-flowing CSF between the 2 compartments. Myelography is not conclusive in SACs, as rapid filling of the cyst can result in loss of the interface between the 2 compartments and concealment of the cyst boundaries. Neither modality is therefore conclusive in diagnosing SAC. In our series, all patients were referred with MRI studies that invariably demonstrated the spinal cord or thecal sac displacement. All patients in our study underwent MRI radiological interpretation was conclusive for an SAC (100%). No patient in our series underwent CT myelography.

Radiological interpretation favored an SAC in all 10 patients which was confirmed on surgical exposure. Differentiation of disorders on radiographic imaging, Schultz *et al.* proposed relying on 2 indirect signs, namely the contour of dorsal cord indentation on sagittal plane, and presence or absence of CSF signal ventral to the cord²⁴. In our experience with applying those criteria, differentiating spinal arachnoid cysts from other spinal cystic diseases⁸.

TREATMENT AND OUTCOMES

The main lines of management of spinal arachnoid cysts comprise complete total resection, fenestration/marsupialization, disconnection from the main arachnoid space, cysto-subarachnoid/cystoperitoneal shunting, or a combination. **In our practice, laminoplasty is usually adopted to gain access to**

thoracic spine lesions in order to avoid delayed kyphosis seen after laminectomy. Exposure usually extends 1 extra level rostral and/or caudal to the cyst, allowing for adequate exposure. The goal of surgery is excision of the cyst with spinal cord or thecal sac relaxation and free-flowing CSF proximally and distally. To the best of our knowledge, no prior study has directly compared the success and recurrence rates among different surgical techniques. Recent studies reported sustained recovery after cyst deflation and disconnection from the main subarachnoid space through minimally invasive approaches to avoid complications of an extensive laminectomy. It is believed that cyst excision with complete cyst wall resection, when technically feasible, provides the least chances of recurrence. **In our experience, laminoplasty allows for complete cyst excision to minimize the chances of recurrence while avoiding delayed kyphosis encountered after conventional laminectomy.** All our patients had sustained improvement of symptoms except for 1 patient who underwent cyst fenestration due to the poorly accessible ventral location. In this patient, symptoms temporarily improved for 4 months, followed by relapse and reappearance of the cyst on follow-up imaging studies. Shunting of the cyst was recommended to the patient, but the patient declined. There is a strong association between SACs and syringomyelia due to aberrant CSF flow dynamics⁹. Holly and Batzdorf reported neurological improvement and reduction in syrinx cavity size after resection of dorsal thoracic SAC in 8 patients⁸. In our cohort, no patient presented with syrinx. In this study, we shed light on a rather rare spinal disorder and illustrate the pitfalls in diagnosis and surgical management. Our results reflect those of other series in the literature (Table 3). We also included patient-reported outcomes to reliably appraise the impact of surgery for symptomatic SAC alongside postoperative radiological changes. Limitations in our study include the small number of cases and short follow-up. The short follow-up also limited our ability to assess the recurrence rates in surgically treated SACs. While long-term clinical and radiological surveillance could ascertain the recurrence rates, in our practice, long-term follow-up was not possible, given continued symptomatic improvement in most patients with the exception of one with a ventral SAC. Our study was also limited in appraising patient-reported outcomes. While newer, more specific scores such as the Oswestry Disability Index and modified Japanese Orthopaedic Association (mJOA) scoring system are now routinely implemented in our practice, the SF-36 score, although relatively nonspecific, was consistently recorded for all patients in our cohort over the study period and was therefore used in our study. The small number of patients also precluded a meaningful statistical analysis of patient-reported outcomes. Finally, despite its benign nature, an SAC represents a diagnostic and therapeutic challenge. In

the absence of standard guidelines, management is largely dependent on surgeon's experience and institutional practice, which greatly influence the treatment and follow-up strategies. Our review provides insight into the presentation, management, and natural course of SAC at a single institution¹⁰.

CONCLUSIONS

Radiological findings, more helpful in diagnosis of SACs except where it cannot differentiate SACs from other spinal disorders such as VSCH. In symptomatic patients with clinical and imaging findings suggestive of an SAC, surgical exploration with complete resection is the treatment of choice. Treatment is usually well tolerated, carries a low risk of complications, and provides the best chance for optimal recovery.

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