Spinal arachnoid cysts

Report of six cases

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Six cases of symptomatic spinal arachnoid cysts are presented. All lesions but one were intradural. Although these cysts are frequently indistinguishable clinically from spinal cord tumors, their correct early diagnosis by myelography and treatment by excision or marsupialization gives gratifying results. The authors emphasize that precise diagnosis and definitive treatment are sometimes delayed because of intermittent symptoms.

KEY WORDS • spinal arachnoid cyst • spinal cord • myelopathy

Symptomatic spinal arachnoid cysts, particularly in the cervical region, are rare; extradural lesions occur more often. Of the six we are presenting, five were intradural and one extradural. Two of the intradural lesions were located in the cervical region. They were all treated at the Radcliffe Infirmary, Oxford, England, between 1938 and 1970.

Case Reports

Case 1

This 19-year-old girl was admitted to the Wingfield Morris Orthopaedic Hospital on May 24, 1938, because of headache followed by a stiff neck 3 weeks previously and progressive difficulty in micturition plus weakness, numbness, and tingling of all limbs for 3 days before admission. When examined, the patient exhibited marked spastic quadriplegia, increased deep tendon reflexes, and extensor plantar responses. Pinprick sensation was lost in the left leg, but light touch and position sense were intact; recognition of temperature was absent on the neck and below both knees. She had urinary retention. A lumbar puncture revealed a pressure of 130 mm H₂O; jugular compression elicited a maximum rise of 30 mm; cerebrospinal fluid (CSF) protein content was 110 mg%, and there was one cell. The following morning she was drowsy and almost totally quadriplegic, including most respiratory muscles. There was marked neck stiffness. Cervical spine films were negative.

While being examined by a neurosurgical consultant (H. Cairns), the patient stopped breathing. Artificial respiration was instituted followed by mechanical support, and she was transferred to the Radcliffe Infirmary. A cystic lesion of the foramen magnum was suspected. Within a day she made a remarkable spontaneous recovery.

Examination. Repeat lumbar puncture revealed a pressure of 140 mm H₂O with normal rise in response to jugular compression;
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the protein count was down to 50 mg%. These changes were attributed to a spontaneous resolution of a CSF pathway block. She continued to improve during the following 2 weeks. A third lumbar puncture was performed. The pressure was too low to register, but the Queckenstedt test was normal. The next day recurrence of pain in the neck was followed by increased weakness of the limbs, more marked on the left. Coarse lateral nystagmus and a relative hypalgesia were noted in the C2-4 distribution. During the subsequent 2 weeks, neck pain and stiffness increased. A progression of motor and sensory deficits indicated deterioration of cervical cord function.

Operation. On June 30, 1938, a cervical laminectomy from C1-4 inclusive was performed (H. Cairns). The dura felt firm and appeared opaque over an area 1.5 cm wide and immediately below the foramen magnum. When the dura was opened, the cord seemed swollen and filled almost the entire canal. When the dura had been reflected, a large cyst was noted opposite the first three cervical segments, to the right and in front of the cord. The cyst had a thin transparent wall and contained about 10 cc of a milky white fluid, which was suggestive of a hydatid cyst but on histological examination proved to be arachnoidal in origin. After the dentate ligament and upper two dorsal roots had been divided, the cyst was entirely removed.

Postoperative Course. The patient made an uneventful recovery. The discharge examination 1 month after operation showed hypalgesia in the right C-2 distribution, slight left hemiparesis with brisker deep tendon reflexes, and equivocal plantar responses, normal sensation, and no disturbance of sphincter control. She returned to normal activity including work within 3 months. She moved to the United States but a medical report received 11 years later revealed no complaints referable to the previous cervical lesion.

Case 2

This 3-year-old boy was admitted to the Radcliffe Infirmary on January 19, 1970, because of stiff neck and abdominal pain for the 2 weeks since a febrile upper respiratory infection.

First Examination. The patient was alert but very fretful. The temperature was 37.5°C. Both ear drums were dull, inflamed, and indrawn. The neck was stiff and exhibited slight resistance to flexion, although Kernig’s sign was negative. Neurological examination was otherwise unremarkable. A lumbar puncture yielded clear CSF without cells, a normal sugar content, and a protein count of 40 mg%. Cervical spine films were normal. Ten days after admission some weakness was detected in both arms. The lumbar puncture was repeated and showed a rise in protein to 120 mg%; on the day following this procedure the patient became markedly quadriparetic with only slight movement detectable in the right leg, absent reflexes, and extensor plantars. A myelogram showed an obstruction at C2-3.

First Operation. On February 2, 1970, a cervical laminectomy was performed (J. B. Pennybacker). The upper part of the cervical cord was enlarged. Probing on either side and in front of the cord failed to reveal any abnormality. Aspiration through the midline raphe at C1-2 yielded 3 cc of clear colorless fluid and resulted in collapse of the cord and the appearance of normal pulsation. It was, therefore, thought that a syrinx had been evacuated, and nothing further was done pending observation of the clinical outcome from this decompression.

The patient made a remarkable recovery. When examined 1 month later he had resumed normal play and except for slight spasticity of the legs and equivocal plantar responses he was neurologically intact. On February 21, 1970, he started to wake up at night complaining of severe neck pain which was followed by decerebrate posturing and variable periods of flaccidity of both arms. There was no weakness of the legs or sphincter disturbance. He seemed unresponsive during these episodes, which lasted about 30 seconds; he returned to normal following them. He was readmitted to Radcliffe Infirmary 2 weeks after onset.

Second Examination. There was no muscle wasting or weakness; muscle tone in the legs was slightly increased. The deep tendon reflexes were absent in the arms but hyperactive in the legs and both plantar reflexes were extensor. Sensory testing within the limits of cooperation was considered near normal. A cervical myelogram demonstrated a complete block at C-4, thought to represent a reaccumulation of the syrinx.
Second Operation. On September 17, 1970, the cervical cord was reexplored from C1–5. The theca was pulsating at the level of the atlas but not below it. The cord looked normal at C-5 but from C-4 upward it was bound down by numerous adhesions. A fine needle was again introduced in the midline at C1–2, and 2 cc of clear, colorless fluid were aspirated, resulting once again in collapse of the upper cervical cord. A midline incision measuring 3 cm was made at that level but failed to reveal a cyst large enough to accommodate a tube. A small wick of gutta percha was fashioned and introduced into the opening in the hope of providing drainage. The patient again made an uneventful recovery and resumed normal childhood activities. On October 21, 1970, he was re-admitted because of recurrence of identical symptoms; they were nocturnal at first, but then also occurred during the day.

Third Examination. The gait was spastic. Only slight weakness was noted in the left arm. The left biceps jerk and right ankle jerk were decreased, and both plantar reflexes were extensor. There seemed to be no sensory deficit. After 3 days of observation during which he was normally active, he was discharged. He was readmitted 2 days later because first the right arm and then the left had become progressively weaker. Pain was not a prominent feature. He had had no further decerebrate episodes. He was uncooperative and walked with a broad-based gait. Both arms were very flaccid. He was still able to grasp with the right hand but had only a flicker of finger movements in the left. Both legs were strong but hypertonic. The deep tendon reflexes were symmetrically active and the plantar reflexes were extensor. No clear sensory level could be detected although a sweat level was noted at the cervicothoracic junction. During a 5-day observation period his legs became progressively weaker and he developed urinary incontinence.

Third Operation. On November 2, 1970, reexploration of the cervical wound and posterior fossa decompression were undertaken but discontinued after infected subcutaneous stitches were found. The wound was drained, and the patient treated with antibiotics. Over the following 2 weeks the quadriparesis progressed; intercostal paralysis precluded satisfactory pulmonary function. He became pyrexic. Another lumbar puncture released sterile fluid under normal pressure, but the protein was 770 mg%. He died on the following day. At autopsy a large intradural, arachnoid cyst was found anterior to the upper cervical cord (Fig. 1).

Case 3

This 15-year-old girl was admitted to the Radcliffe Infirmary on July 21, 1942. Five months earlier she had fallen backward against a wall and sustained a minor back injury and a sprained ankle. She went to work the next day but experienced a pain in the right leg and progressive difficulty walking. Bed rest resulted in slight improvement, but 5 weeks later her legs became “stiffer” and 2 weeks before admission she developed urinary retention and decreased sensation from the waist down. She had no back pain.

Examination. Spastic paraparesis was noted, most marked on the right, and severe enough to prevent standing. The deep tendon reflexes in the legs were increased with a
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sustained ankle clonus on the right and un-
sustained clonus on the left. Both plantar
reflexes were extensor. The lower abdominal
reflexes were absent. A sensory level was
observed at T8–9 with sacral sparing,
although the anal wink was diminished on the
right. Spine films revealed wedge-shaped mid-
dorsal vertebrae.

Operation. A laminectomy was performed
from T4–10. At T-7 there was an extradural
arachnoid cyst 6 x 2.5 cm containing clear
CSF. It had a thin, firm wall that was
adherent to the dura by a pedicle; a 3 mm
hole communicated with the subarachnoid
space. The hole was closed and the cyst
removed.

The patient made an uneventful recovery.
At follow-up 17 years later she was married
and living a normal life. Examination
revealed only a middorsal kyphosis.

Case 4

This 25-year-old woman was admitted to
the Radcliffe Infirmary on March 21, 1958.
Ten days earlier she had experienced, after
gardening, a low thoracic back pain which
became localized to the left T7–8 root dis-
tribution. Five days before admission she ex-
perienced paresthesias in both legs, progress-
ing to a sensory anesthesia below T-8, and 2
days later developed progressive paraparesis.

Examination. The patient had a profound
but incomplete paraparesis, affecting the left
leg more than the right, with increased reflex-
es and extensor plantar responses. There was
a sensory level at T-8. Tenderness to percus-
sion could be elicited at T6–7. Lumbar punc-
ture revealed a complete block, and examina-
tion of the CSF showed 90 mg% protein and
12 cells. A myelogram demonstrated a com-
plete block at T-8.

Operation. A laminectomy extending from
T7–9 was performed. There was no epidural
fat at the T-8 level. When the dura was
opened, the cord looked normal but was dis-
placed backward by an anteriorly placed sub-
arachnoid cyst measuring 1 cm in diameter
and containing grayish fluid. During the
course of retraction the cyst ruptured and the
cord returned to its normal position.

The patient made an uneventful recovery.
She showed immediate sensory improvement
and by the time of discharge she had regained
some power in both legs. She continued to ex-
perience intermittent nocturia. Her neuro-
logical function gradually improved and she
was able to lead a normal life. She gave birth
to two children; there was some uterine iner-
tia and forceps delivery was needed in both
cases.

Five years after laminectomy she fell downstair,
after which she noted back stiffness and leg weakness. She had two other falls, after which a greater paraparesis developed and she was readmitted on July 22, 1964.

Second Examination. The patient now had
weakness of both legs, the left more than the
right, with normal tone, bilaterally increased
reflexes, and extensor plantar responses. Hypesthesia to pin prick and touch were
present below the right T-8 dermatome. A
lumbar puncture revealed no block, and CSF
examination showed a protein content of 70
mg% with 5 lymphocytes and 1400 red cells.
A myelogram revealed a free flow of contrast
material with only a slight deformity at the
site of the previous operation. She was dis-
charged to continue with physiotherapy.

She was readmitted to the Neurology
Department at the Churchill Hospital on
May 26, 1965, for reassessment. Examination
then demonstrated weakness of the left leg
and normal strength in the right leg. Sensa-
tion was still diminished on the right below
T-8 and was normal on the left. A myelo-
gram now showed almost complete obstruc-
tion at the T-9 level. The previous operative
site was reopened. Adhesions were found at
the level of the block but there was no reac-
cumulation of cyst fluid. Postoperatively the
left leg became stronger but the sensory level
remained at about the T-10 dermatome on
the right without sacral sparing. At the last
follow-up 3 years after reexploration she was
coping with her household duties.

Case 5

This 40-year-old man was admitted to the
Radcliffe Infirmary on April 7, 1965. He
related a 16-year history of difficulty walking
because of weakness of both legs which had
started on the left and gradually progressed to
the right leg. Fourteen years earlier he had
undergone bilateral Achilles tendon lengthen-
ing, and 9 years earlier a diagnosis of multiple sclerosis was made. At that time a lumbar
puncture revealed a normal pressure and the
CSF contained 50 mg% protein and 2 cells. Six months prior to admission he had become unable to walk, and examination revealed a spastic paraparesis with a T-9 sensory level. He was admitted 3 months later to another institution where a myelogram revealed three fairly large arachnoid diverticula, two at the level of T-6 and a large one at T-10 (Fig. 2).

Examination. Upon admission he exhibited a spastic paraparesis, on the left more than the right, with increased reflexes, bilateral sustained clonus, and extensor plantar responses. A sensory level was detected at T-6 with sacral sparing. Abdominal reflexes were absent. Repetition of the myelogram revealed the presence of two more cysts at the first and twelfth thoracic levels; both filled when the patient was standing and emptied when the head was lowered.

Operation. A laminectomy from T5–7 was performed; epidural fat was absent, and the dura was bulging. When the dura was opened a translucent arachnoid cyst was seen covering the posterior part of the cord and pushing it to the right side. It extended the length of three vertebrae. Aspiration of the cyst yielded a clear colorless fluid. After the cyst was marsupialized the cord resumed a normal position and shape. Histological examination of the cyst wall showed thickening by an acellular increase of connective tissue but no evidence of inflammation. The patient made a satisfactory recovery. The superficial sensory deficiency became less apparent, and motor strength returned in his legs.

Case 6

This 14-year-old girl was admitted to the Radcliffe Infirmary on July 19, 1945. Since childhood she had a tendency to “throw” her left leg as she walked. Five months prior to admission she began to be “flat-footed” and slap her feet when she walked. She then began to trip during games. Five weeks prior to admission she fell 4 feet during physiotherapy, and 5 days later she was unable to raise her left foot sufficiently to walk upstairs. At the same time she noticed diminished feeling in both legs, especially on the left. She also complained of intermittent right-sided dorsal pain. By the time of admission she was totally unable to walk and was incontinent of urine and feces.

Examination. The patient had a marked spastic paraparesis with increased deep tendon reflexes and extensor plantar responses. Complete anesthesia was present below the T-11 dermatome. A lumbar puncture showed a pressure of 40 mm H₂O, and the Queckenstedt maneuver suggested a block. Examination of the CSF showed a protein content of 140 mg% and 7 lymphocytes. Spine films revealed kyphoscoliosis of the lower dorsal spine and scalloping of the posterior aspect of the vertebral bodies from the midthoracic region to L-3 (Fig. 3).

Operation. A laminectomy extending from T7–10 was performed. The bone was thin and friable; there was no extradural fat, and the dura was thin and translucent. The arachnoid was opaque and contained a large cyst divided into three compartments. The central cyst extended from T6–10. Fluid from the cyst contained 60 mg% protein and 21 cells.

Fig. 2. Thoracic myelogram demonstrating two small arachnoid diverticula at T-6 and a large one at T-10. Left: Anteroposterior view. Right: Oblique lateral view.

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Histological examination of the cyst wall revealed collagenous tissue covered by flattened meningothelial cells. The septa separating the cyst from the general sub-arachnoid space were divided to accomplish free circulation of CSF. The dura was left open.

Follow-up examination 3 years after the operation showed normal power, tone, and sensation. There was, however, a marked kyphosis.

Discussion

The clinical signs and symptoms of spinal arachnoid cysts are frequently indistinguishable from tumors in similar locations. They most commonly become evident in adolescence or early adulthood. In a review of 99 Mayo Clinic cases of intraspinal tumors in childhood, only one cyst was found. Svien, et al., reported two cases among 41 children under the age of 15 with spinal cord tumors. Spiller, et al., presented one such patient in addition to 11 cases of spinal tumors. Asymptomatic arachnoid pockets in various locations along the spinal axis are much more commonly demonstrated at the time of myelography.

Pain, paresthesias, varying degrees of spastic weakness, and sphincteric disturbances are the most common presenting complaints. Remissions and exacerbations may occur, and lead to the erroneous diagnosis of multiple sclerosis. This was the case with one of our patients, who carried this diagnosis for 9 years before the correct cause of his illness was ascertained. One patient was referred with the diagnosis of Guillain-Barre syndrome, and our second case was thought to have a cervical syrinx. A lumbar disc exploration was performed in one case prior to definitive treatment. The intermittent nature of the symptoms has been explained on the basis of an emptying and filling of the cyst which could occur with changes in position. With the patient standing, filling of the cyst could result not only in local compression, but also traction due to gravitational force. The presence of an unexplained kyphosis in childhood or adolescence should arouse suspicion of an arachnoid extradural cyst.

Various etiologies have been stressed. Some cases clearly seem to be congenital in origin. Our second case probably falls into this category. Elsberg, et al., also stressed the congenital origin of extradural cysts and postulated two mechanisms, namely, a congenital diverticulum of the dura mater, or a herniation of the arachnoid through a congenital defect in the dura. It seems unlikely that these cysts arise from cell rests as suggested by Hyndman and Gerber, since frequently a communication exists between the cysts and the subarachnoid space. In other cases trauma plays a role. Adhesive arachnoiditis secondary to various inflammatory processes has been cited as a potential cause. Skoog felt that most arachnoid cysts had a parasitic etiology. Perrett, et al., postulated an origin from the septum pocusum. The hypothesis does not explain cysts that appear on the anterior aspect of the cord, as with Hoffman’s patient and our Cases 1, 2, and 4. Teng and Papatheodorou saw no evidence of adhesive arachnoiditis in their cases and on the basis of operative findings suggested that the cysts might be the
result of "faults" in the distribution of arachnoidal trabeculae.

When the lesions occur in the thoracic region, radiographic studies sometimes reveal a kyphosis with signs of longstanding pressure in the form of scalloping of the posterior surfaces of the vertebral bodies, thinning of the pedicles, and widening of the canal. A myelogram may show a block or actually demonstrate a filling of the cyst, particularly if efforts are made to study the patient first in the supine position and then erect. The CSF protein is usually elevated under these circumstances.

The majority of spinal arachnoid cysts are found on the posterior aspect of the thoracic spinal cord. Two cervical cases have been described; one extradural and the other intradural. Spiller, et al., described a lumbar intradural cyst. Our two cervical cases are, therefore, quite rare. The second case is particularly unusual in that exploration of the lateral gutters at the time of operation failed to reveal the true nature of the lesion, which was thought to be a syrinx until autopsy showed the anterior cervical arachnoid cyst. Our fourth case is also unusual in that the cyst was anterior to the thoracic spinal cord. Histological studies of the cysts usually reveal a lining of meningothelial cells on a thin fibrous base. At times thickening and round cell infiltration suggests an inflammatory response. The cysts usually contain fluid indistinguishable from CSF. In our Case 1 the fluid was described as "milky" and suggestive of a hydatid cyst, but on histological examination it proved to be arachnoidal in origin. In Case 4 the fluid was described as "grayish." The arachnoid was opaque in Case 6, and the cyst was divided into three compartments.

The operative treatment most commonly advocated has been total removal of the cyst. However, removal may be hazardous and entail the sacrifice of nerve roots, particularly if the cyst is in an anterior location. For this reason marsupialization has been recommended as a sufficient treatment to provide a lasting cure. Teng and Papatheodorou recommended that when symptoms were not disabling a trial of frequent rest periods in recumbent position usually provided temporary relief. They reserved surgical removal of the diverticula for patients disabled by persistent pain or weakness. The extent and rapidity of neurological recovery depend largely on the length and severity of preoperative cord compression.

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**References**

9. Pennybacker JB: Personal communication
13. Spiller WG, Musser JH, Martin E: A case of intradural spinal cyst with operation and recovery; with a brief report of eleven cases of tumor of spinal cord or spinal column. Trans Stud Coll Physicians Philad 25:1-18, 1903
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