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Familial Adhesive Arachnoiditis Associated with Syringomyelia

V. Pasoglou, N. Janin, M. Tebache, T.J. Tegos, J.D. Born, and L. Collignon



ABSTRACT

SUMMARY: Adhesive arachnoiditis is a rare condition, often complicated by syringomyelia. This pathologic entity is usually associated with prior spinal surgery, spinal inflammation or infection, and hemorrhage. The usual symptoms of arachnoiditis are pain, paresthesia, and weakness of the low extremities due to the nerve entrapment. A few cases have had no obvious etiology. Previous studies have reported one family with multiple cases of adhesive arachnoiditis. We report a second family of Belgian origin with multiple cases of arachnoiditis and secondary syringomyelia in the affected individuals.

Adhesive arachnoiditis is a relatively scarce condition. This pathology refers to an inflammation of the arachnoid matter, which becomes thick and adherent and is often associated with syringomyelia. Common etiologies are spinal surgery, inflammation and/or infection such as tuberculous meningitis, hemorrhage, trauma, and injection of anesthetic agents. During the past decades, this entity (syndrome) has been particularly related to an oil-based contrast agent used for myelographic studies. However, a few cases with no obvious cause have also been reported in the literature. In this regard, adhesive arachnoiditis is a sporadic condition, except for a single family of Japanese origin comprising 9 affected individuals. Part of this family immigrated to Canada and was reported by Duke and Hashimoto in 1974.¹ Adding new cases from part of the same family in Japan, Nagai et al² published an update in 2000 and drew a pedigree showing an apparent vertical transmission of the disease over 3 generations. We report here a second family with multiple cases of arachnoiditis and secondary syringomyelia in the affected individuals.

CASE SERIES

The pedigree of the Belgian family is shown in Fig 1.

Case 1

In 1979, a 35-year-old woman without a history of trauma, infection, and spinal operation presented to a local emergency department with flaccid paraparesis and complete sensory loss of all modalities below the level T6 (Fig 1, II.7). A myelographic examination was performed and revealed a blockage at the level of the dorsal spine. Surgery revealed the presence of attenuated arachnoidal adhesions at the levels T5–T6. The patient was treated by lysis of the adherent tissue. Postoperatively, the patient reported a slight amelioration of the symptoms, but 1 year later, she presented with walking difficulty, a right-sided Babinski sign, and complete sensory loss below the T6 level. CT of the brain and myelographic findings were normal; however, her symptoms worsened with time. In 1985, a routine MR imaging examination of the cervical and dorsal spine was performed and revealed a syringomyelic cavity from the C5 to T5 levels, just above where the surgeon had found the arachnoid adhesions.

Case 2

In the early 80s, the brother of case 1, a 50-year-old man, was diagnosed with syringomyelia (Fig 1, II.1). The patient underwent surgery for this condition but died due to perioperative complications. This information was disclosed by the family because no official documentation was available.

Case 3

The son of case 2 and brother of case 4 was neurologically impaired and presented with walking difficulties. No official diagnosis or documentation was available (Fig 1, III.6).

Case 4

A 49-year-old woman (daughter of case 2, sister of case 3, cousin of cases 5 and 6) without a history of trauma, infection, or spinal

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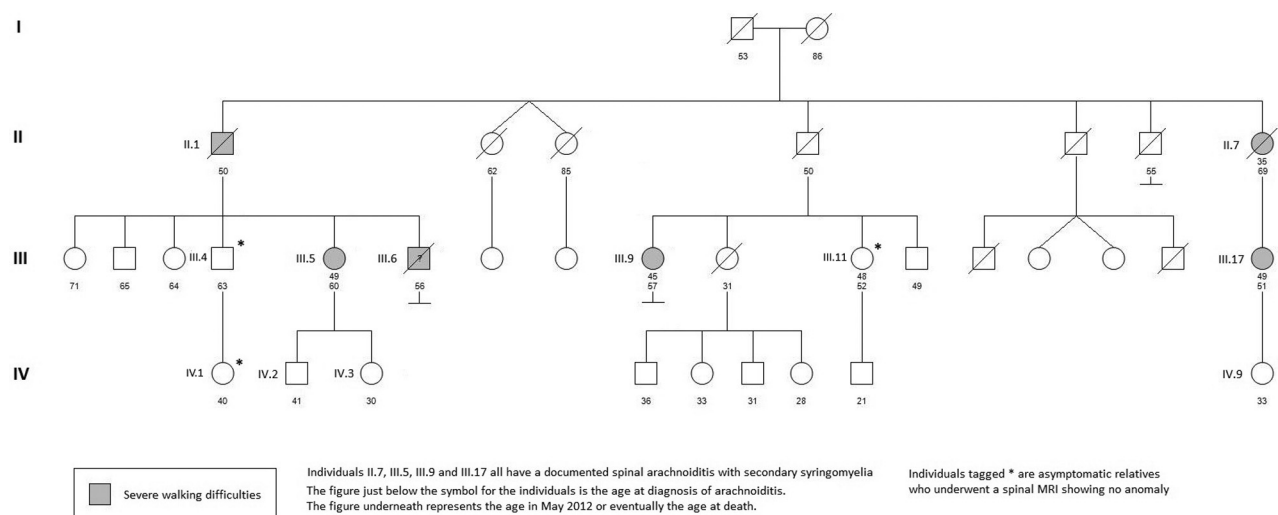


FIG 1. The pedigree of the Belgian family with spinal arachnoiditis and secondary syringomyelia. Squares and circles represent males and females, respectively. Solid symbols represent the affected family members.

operation presented in 2001 with walking difficulty and instability (Fig 1, III.5). The symptoms first appeared in 2000 when she reported paresthesia and bilateral dysesthesias below the knee, as well as chronic back pain. During the present neurologic examination, the patient could hardly lift her legs (flaccid paralysis). The examination revealed loss of pain and hot-cold sensation below the T3 level on both sides. The light-touch sensation and the vibration sensation in both medial malleoli were preserved. The Babinski sign was absent bilaterally. The routine serologic study findings were normal, including C-reactive protein levels. The CSF contained <5 white blood cells/mm³. CT of the brain revealed hydrocephalus. MR imaging using sagittal T2-weighted turbo spin-echo, T1-weighted TSE, a myelographic sequence, and a gadolinium-enhanced T1-weighted turbo spin-echo sequence of the whole spine, completed by a transverse T2-weighted TSE at the level of the detected anomalies, was performed and revealed the presence of a syringomyelic cavity at the T3–T8 levels. No arachnoid adhesions were detected by the examination. There was no anomaly in the rest of the spine, and the administration of contrast material confirmed the absence of a causative tumoral lesion. The neurosurgeon mentioned the presence of thick and adherent arachnoid matter just below the level of the tubular syringomyelic cavity; nevertheless, MR imaging did not depict this arachnoid web. The pathologic examination of the extracted specimen confirmed the fibrous nature of the meningeal tissue sample with meningioepithelial cells and lymphocyte infiltration. The diagnosis of adhesive arachnoiditis was made. Follow-up MR imaging examinations showed the regression of the transverse size of the spinal cord and syrinx.

Case 5

A 45-year-old woman (cousin of cases 3, 4, and 6) presented in 2007 with dorsal pain, walking difficulty, and paresthesias at the thoracic dermatomes (Fig 1, III.17). The neurologic examination revealed hyperesthesia at the T5 level. The routine serologic study findings were normal, including the C-reactive protein level. The CSF contained 1 white blood cell/mm³. All other CSF data were within normal ranges. The MR imaging examination included

the same sequences as those used in case 4 and demonstrated the presence of a syringomyelic cavity extending from the T2 to T7 levels and the presence of low-signal arachnoid adhesions at the T3–T4 level (On-line Fig 1). The rest of the spine was normal. After administration of contrast material, the arachnoid webs did not enhance and the presence of a causative tumoral lesion was excluded. The neurosurgeon confirmed the presence of thick and adherent arachnoid matter. The pathologic examination findings alluded to an adhesive arachnoiditis without signs of tumoral infiltration.

Case 6

In 2010, a 49-year-old woman (cousin of cases 3, 4, and 5) without any predisposing factors for arachnoiditis presented with left-sided numbness from level T4 to T10 (Fig 1, III.9). Light-touch sensation was decreased between the T4 and T8 levels, but the vibration sensation was normal. The deep tendon reflexes were normal in the upper and lower limbs. The Babinski sign was absent bilaterally. She also had walking difficulty and urinary incontinence. The routine serologic study findings were normal, including C-reactive protein. The CSF contained <5 white blood cells/mm³. CT of the brain revealed hydrocephalus. MR imaging examination of the spine included a sagittal T2 (TR/TE, 1900/100 ms), sagittal T1 (TR/TE, 330/7.4 ms), sagittal short-TI inversion recovery (TE/TR, 60/2500 ms and inversion recovery delay, 170 ms) of the whole spine completed by axial balanced turbo-field echo (TR/TE: 6.2/3.1 ms) at the level of the detected anomalies. We also used a midline sagittal cardiac-gated phase-contrast MR imaging (10-mm-thick sagittal section, a 250 × 250 mm FOV, acquisition matrix of 252 × 185, TR of 21 ms, TE of 6.4 ms, phase-contrast velocity set at 10 cm/s). Finally, a gadolinium-enhanced T1-weighted turbo spin-echo encompassing the whole spine was performed.

We observed the presence of a syringomyelic cavity extending from T4 to T8 with thickening of the meninges at the T5–T6 levels (Fig 2). The rest of the spine was normal, and the administration of contrast material confirmed the absence of a causative tumoral lesion. The arachnoid adhesion did not enhance. In our case, cine



FIG 2. Sagittal T2-weighted MR image. Note enlargement of the spinal cord with a syringomyelic cavity extending from levels T4 to T8 (arrow). At the T5–T6 levels, posterior to the cord, a band-like low-signal-intensity structure represents a fibrous thickening of the arachnoid matter (arrowhead). The superior half of the cavity, just above the arachnoid thickening, is much more dilated than the inferior half.

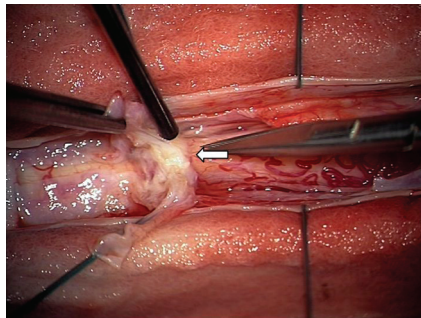


FIG 3. Intraoperative photograph confirming the presence of a thick and adherent arachnoid web (arrow), depicted by MR imaging.

MR imaging proved to be of little diagnostic value because the motion of the heart and great vessels produced significant artifacts in the thoracic spine (it did not help in identifying the site of CSF blockage). During surgery, the neurosurgeon confirmed the presence of thick and fibrous arachnoid matter (Fig 3 and On-line Video). The pathologic examination findings of the resected tissue confirmed the presence of adhesive arachnoiditis (Fig 4). The follow-up MR imaging examination showed that the transverse size of the spinal cord had regressed and the syrinx was less expansive.

Asymptomatic Members of the Family

All the living first-degree relatives of the affected individuals are asymptomatic. An MR imaging of the whole spine was recommended to exclude the possibility of an asymptomatic arachnoid-

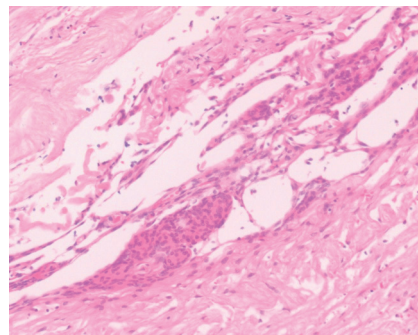


FIG 4. The pathologic examination demonstrated a thick membrane, rich in collagen limited by meningeoepithelial cells. There is no evidence of inflammation, infection, or old hemorrhage (H&E stain).

itis in these family members. The MR imaging examination included sagittal T2- and T1-weighted TSE sequences of the whole spine and a transverse balanced turbo-field echo sequence of the lower cervical and upper dorsal spine. The MR imaging showed no anomaly in the following individuals: III.4, III.11, IV.1, and IV.9.

DISCUSSION

Spinal adhesive arachnoiditis is an inflammation inside the dura, affecting the arachnoid layer of the meninges, resulting in fibrosis. As a consequence, the arachnoid becomes firm, adherent, and thick and gets attached to the pia and dura mater.³ The causes of spinal arachnoiditis include trauma, tumors, infections, spinal surgery, and spinal injection of substances such as anesthetics or oil-based myelographic contrast materials, which are no longer used clinically.^{4,5}

Adhesive arachnoiditis is a chronic inflammatory condition causing neurologic impairment. It usually presents with diverse symptoms such as persistent back pain, weakness, and dissociated sensory loss. This condition is notably underdiagnosed, even though there is a high and well-known association between adhesive arachnoiditis and failed back surgery syndrome. On the other hand, there have been reports suggesting that spinal arachnoiditis is a coincidental finding rather than a disease.⁶ Quencer et al⁷ described a number of patients in whom arachnoiditis was totally asymptomatic.

The complications of adhesive arachnoiditis include the formation of arachnoid cysts, spinal cord damage (ischemia possibly due to obliterative angiopathy), and syringomyelia.^{7–10} Jenik et al¹¹ highlighted, for the first time, the possible relationship between adhesive arachnoiditis and syringomyelia. Syringomyelia is an eccentric cavity within the spinal cord, which contains extracellular fluid (identical or similar to CSF). The most widely accepted theory regarding the etiology of syringomyelia is that the perturbation of CSF flow in the subarachnoid space modifies the fluid velocity, leading to a reduction of the fluid pressure and a passive distension of the spinal cord.^{12,13}

The pathophysiology of syringomyelia is still unknown despite numerous and thorough efforts.^{13–15} Older studies reported that CSF enters the syrinx via the fourth ventricle through the central canal.¹⁶ On the other hand, Ball and Dayan¹⁷ proposed that CSF enters the syrinx through the spinal perivascular space of Virchow-Robin. The studies of Heiss et al¹⁸ and Stoodley et al¹⁹ used

this hypothesis as a basis for explaining the pathophysiology of syringomyelia. According to Chang and Nakagawa,^{20,21} the formation of the syrinx is caused by CSF pressure gradients inside and outside the spinal cord. This hypothesis could explain the association of syringomyelia with both Chiari malformation and adhesive arachnoiditis. Concerning the relationship between arachnoiditis and syrinx formation, the authors support a pressure drop in the subarachnoid space distal to the blockage. As a result, the pressure inside the spinal cord becomes superior to the perimedullary pressure. The cyclic pressure gradient at each CSF pulse leads to the formation of a syrinx.

The symptoms of syringomyelia are usually progressive and worsen during a long period. The most usual clinical manifestation of syringomyelia is a dissociated suspended sensory loss (impaired pain and temperature sensation but preserved light touch, vibration, and position sense), motor deficit, dysesthesias, and pain. When occurring outside of the context of arachnoiditis, syringomyelia is commonly associated with extramedullary malformations, such as Chiari or Dandy-Walker malformation. Isolated syringomyelia, with no history of Chiari malformation, trauma, or infection, can also occur, but it should always raise the suspicion of an intramedullary tumor. When a syrinx has been diagnosed, a contrast-enhanced MR imaging study is, therefore, mandatory to exclude an associated expansive intramedullary lesion.

To the best of our knowledge, reports about familial cases of arachnoiditis in humans are limited to the articles of Duke and Hashimoto¹ and Nagai et al,² which deal with the same family of Japanese origin. There was no relation between the Japanese family and the family of native Belgians described here. Furthermore, all family members involved in the case study live in the same area of Belgium within a 50-km range. Nothing in our medical records or in our knowledge suggests the exposure of the family to toxic substances, endemic organisms, and so forth. Available data from our cases and the Japanese family suggest that arachnoiditis may sometimes be very strongly influenced by a very rare inherited genetic anomaly. A major gene with incomplete penetrance could explain the family data. Of course, multigenic inheritance is also a possibility. A genetic study of this family will be undertaken to look for a major gene.

Regarding syringomyelia, in 2002 a review by Yabe et al²² reported 21 cases of familial syringomyelia since 1899. Two more cases of familial syringomyelia were published since 2002.^{23,24} Almost all the published cases of familial syringomyelia (whether associated or not with Chiari malformation) entail only 2 affected individuals who often belong to the same sibship, but they sometimes stand in a parent-child relationship or in a more remote degree of relationship. Although a highly penetrant Mendelian transmission seems very unlikely, these published cases are consistent with the hypothesis that genetic factors might play a role in the development of sporadic syringomyelia and/or the development of the Chiari malformation. Consistent with this hypothesis, Newman et al²⁵ reported, 30 years ago, an association between human leukocyte antigen A9 and syringomyelia in a series of 53 cases (40 of which were associated with a Chiari malformation).

The CSF pulsates in the craniocaudal direction due to the cerebral blood volume variations during the cardiac cycle. Normal and pathologic CSF pressure pulse has been extensively studied

and recorded as waveforms.^{26,27} In their study, Enzmann et al²⁸ demonstrated that the most significant and consistent finding within preoperative syrinx cavities is the presence of fluid that pulsates in a similar way to subarachnoid CSF. The larger syringes demonstrated more important pulsations than the smaller ones. Nonpulsating syringes were usually the smaller ones. The postoperative studies of the syrinx showed a complete elimination of the pulsations in 3 of 5 patients. Furthermore, when narrowing of the subarachnoid space was observed, there were no CSF pulsations. There was no pulsation in patients with cystic formation associated with a tumor. More recent studies confirmed the pulsatility of the cyst and of the pericystic subarachnoid space.²⁹ Additionally, the occurrence of the systolic peak is earlier in the cyst than in the pericystic subarachnoid space. In patients with Chiari I malformation associated with syringomyelia, Koç et al³⁰ demonstrated that at the level of the foramen magnum, the flow pattern demonstrates a heterogeneous pattern, which was ameliorated after surgical decompression. Regarding the syrinx, the preoperative flow pattern was heterogeneous, but after the operation for decompression, there was an impressive decrease in CSF flow.

Mauer et al³¹ showed that cardiac-gated phase-contrast CSF flow studies are more reliable than invasive conventional myelography to detect the site of spinal CSF blockage in idiopathic syringomyelia. Concerning the imaging of adhesive arachnoiditis associated with syringomyelia, according to Inoue et al,³² the most striking MR imaging findings are the deformity of the cord at the level of the arachnoid thickening, blurring of a part of a syrinx wall, and the focal signal void within the syrinx.

For imaging of the subarachnoid space and arachnoid pathology, T2-weighted imaging is the method of choice. Nevertheless, there are some inconveniences arising from the loss of signal due to CSF flow provoking a loss of contrast within the arachnoid space. According to Roser et al,³³ 3D constructive interference in steady state MR imaging is an ideal method for the diagnosis of subarachnoid space disorders and syringomyelia due to the degradation of the flow-void artifacts compared with T2-weighted imaging and the amelioration of contrast within the subarachnoid space. Moreover, this method provides the possibility of producing multiplanar reconstruction images. Nevertheless, this sequence is very sensitive to motion caused by swallowing and breathing.³⁴ Gottschalk et al³⁴ suggested that cardiac-gated cine steady-state free precession MR imaging has a certain value in the evaluation of idiopathic syringomyelia and the detection of the arachnoid membranes.

For the imaging of future patients, we would suggest the use of sagittal T2, T2*, T1, and postgadolinium T1-weighted sequences from C0 to the cauda equina, a midline sagittal cardiac-gated phase-contrast MR imaging completed by axial balanced turbo-field echo at the level of the anomalies.

Regarding the treatment of adhesive arachnoiditis associated with syringomyelia, the first aim should be lysis of the adhesions, to resolve the pathologic factor. Drainage of the syrinx by myelotomy or by shunting is performed. Nonetheless, Klekamp et al³⁵ reported that recurrence of the syrinx after shunting occurs in 92% of cases for focal and 100% for extensive arachnoid scarring. The same authors reported that for long-term management of the

syrinx, microsurgical lysis of the arachnoid adhesions and decompression of the subarachnoid space with a fascia lata can lead to an improved outcome.

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