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Arnold-Chiari Type I malformation: a look at two cases in the adult

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This paper reviews the varied presentations of Arnold-Chiari Malformation (ACM) in terms of anatomy, clinical presentation, diagnostic workup, and therapeutic considerations. Emphasis is placed on the Type I condition as it appears in the adult. ACM is a developmental anomaly in which the cerebellar tonsils and portions of the posterior fossa structures herniate through the foramen magnum. It is frequently accompanied by a broad spectrum of additional anatomical variations and can present clinically in a variety of ways. Two cases of ACM Type I in the adult are presented. Both patients were treated initially by conservative measures before MRI could be obtained to establish the diagnosis. Neither one of these patients showed improvement with such a therapeutic regimen. It is important for practitioners to be aware of the varied presentations of ACM in the adult.
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KEY WORDS: Arnold-Chiari Malformation, syringohydromyelia, cerebellar tonsils, posterior fossa, chiropractic.

Cet article se penche sur les diverses présentations de la malformation d'Arnold Chiari (ACM) en termes d'anatomie, de tableau clinique, d'établissement du diagnostic et de considérations thérapeutiques. Nous nous intéressons plus particulièrement à la condition de Type I telle qu'elle se manifeste chez l'adulte. L'ACM est une anomalie congénitale au cours de laquelle les amygdales cérébelleuses et des portions des structures de la fosse postérieure traversent le trou occipital. Cette anomalie s'accompagne souvent d'une vaste gamme de variations anatomiques additionnelles et peut se manifester en clinique sous diverses formes. Nous présentons deux cas de ACM de Type I chez l'adulte. Les deux patients ont d'abord reçu des soins conventionnels avant que l'on puisse obtenir l'IRM pour confirmer le diagnostic. Aucun de ces deux patients n'a démontré d'amélioration à l'issue de ce traitement. Il est important pour les praticiens de connaître les diverses présentations de l'ACM chez l'adulte.
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MOTS-CLÉS : Malformation d'Arnold Chiari, syringohydromyèlie, amygdales cérébelleuse, fosse postérieure, chiropractie.

Introduction

In the adult, Arnold-Chiari Malformation (ACM) can manifest itself in a variety of ways. Not only can it be totally asymptomatic, but it can also be very dramatic in its presentation. ACM can also be associated with other conditions, one of which is syringomyelia. This assocation may add further dimensions to the clinical presentation. These patients sometimes provide a diagnostic challenge to the health care practitioner. Two cases of ACM Type I are presented, one of which had an associated syringomyelia. A discussion follows in which the clinical, pathological and therapeutic considerations are reviewed.

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Case reports

Case 1. A 43-year-old white female had been a chiropractic patient for the past ten years. She had been treated for a variety of back and neck complaints including occasional episodes of suboccipital pain which had always responded well to chiropractic care. February 15, 1985 she fell on some ice and hit the back of her head. Since that time she complained of lightheadedness, dizziness, photophobia, diminished smell and taste, neck soreness, diminished mobility of the head and neck. She also described a peculiar suboccipital pain that she likened to the sensation that one gets after eating ice cream too fast. This sensation would come in brief paroxysms lasting approximately ten seconds. As time went on she would experience them more frequently, up to 40 episodes per day. These sensations appeared to be triggered by activities that increased CSF pressure such as straining, laughing, coughing. Maneuvers that increased axial pressure on the spine or those that extended the cervical

spine, as well as those that increased the cerebral spinal fluid (CSF) pressure such as the Valsalva maneuvre, also evoked the peculiar suboccipital sensation.

Plain film radiographs of the cervical spine and skull were normal. Magnetic resonance (MR) imaging of the cervical spine and skull base (Figure 1) revealed herniation of the cerebellar tonsils through the foramen magnum as well as caudal displacement of portions of the cerebellar hemispheres, elongation of the medulla and thinning of the cisterna magna.

A diagnosis of Arnold-Chiari Malformation Type 1 was made. This patient was admitted for surgery and underwent a posterior fossa craniectomy and a complete laminectomy of the arch of C1. The dura was subsequently opened and microdissection was performed to remove tonsillar adhesions. It was found that the Foramen of Magendie was occluded with choroid plexus. This was gradually teased away to allow the efflux of CSF across the floor of the fourth ventricle.

This patient has recovered fully from her surgery and reports complete relief of her symptoms. She continues to be monitored by her chiropractor who provides periodic care as indicated for other spinal conditions.

Case 2. A 50-year-old white female presented complaining of paresthesia in the left arm, forearm and hand, as well as mild left sided facial paresthesia in the region of the tempromandibular joint. She described tightness involving the left anterolateral aspect of the neck and in the left supraclavicular and infraclavicular areas. There was tenderness over the sternoclavicular and acromioclavicular articulations. She also reported mild weakness of the left thigh and leg noticeable only when she was climbing a number of stairs.

The patient first noticed an insidious onset of mild paresthesia in the left 4th and 5th fingers 6 years earlier. At the time, her physician diagnosed a C8 radiculopathy and prescribed a course of physical therapy. This proved largely ineffective. Her symptoms were modest and she simply tolerated them. About 2 years later, her symptoms became worse, eventually involving the entire left hand.

While waiting for MR imaging she presented to a chiropractor in an attempt to obtain symptomatic relief. Examination revealed superficial and deep tendon reflexes to be normal. Sensitivity and differentiation between sharp and dull and hot and cold were normal. She was unable to differentiate subtle changes in texture with the fingers of the left hand. She reported previous nerve conduction tests were normal and orthopedic testing was normal. Cervical mobility was essentially full, but mildly limited by hypertonic musculature. Testing the thoracic outlet produced some diminution of the radial pulse with the costoclavicular test. Plain film radiographs of the neck and the chest were normal.

A trial of chiropractic manipulation to the cervical and thoracic spine involving twice-weekly treatments over a period of four weeks was ineffective. Efforts to reduce the hypertonicity of the cervical musculature, particularly the scalenus anticus, sternocleidomastoid and other regional musculature by spray and stretch, massage, microcurrent and TENS were ineffective. Manipulation of the shoulder, acromioclavicular and sternoclavicular articulations also failed to change the symptoms.

MR imaging of the cervical spine and skull base (Figure 3) revealed ACM Type I and syringomyelia.

Discussion

ACM is a congenital anomaly involving protrusion through the foramen magnum of the cerebellum, the medulla, the fourth ventricle and other posterior fossa structures.[1,2,3,4,5] This process usually begins during the third week of embryonic life at the time of closure of the neural groove. This anomaly may be associated with other ancillary findings. It is generally classified into three types.[1,4]

Type I: The cerebellar tonsils herniate through the foramen magnum. The inferior lobes of the cerebellum are displaced caudally. The medulla tends to become elongated, and the cisterna magna is thinned or obliterated.[2,3,4] It is associated with syringomyelia in about 40% of the cases.[4,6]

Type II: Type II is the most common type of ACM.[4] In addition to the changes of Type I, the inferior vermis, lower pons, and medulla are elongated and displaced caudally and herniate through the foramen magnum. The medulla remains in correct alignment with the cord in 30% of cases or buckles behind the cord in 70% of the cases creating a kink in the cervical cord.[3] The fourth ventricle is elongated and may descend into the cord.[2,3,6] Shortening of the cervical cord, upward angulation of the cervical nerve roots, hydromyelia and other cord malformations can occur. It is nearly always accompanied by meningomyelocele and hyrocephalus.[2,3,4,5] It occurs in about 2 per 1,000 births.[2]

Type III In addition to the findings of Type II ACM, herniation of practically the entire cerebellum and medulla into a cervical meningocele occurs.[1,6] The fourth ventricle typically empties into an external sac or hydroencephalocele. Type III ACM is rare.[6]

In recent years, however certain authors have restricted their classification of ACM to Types I and II only. They make the distinction only of a cerebellomedullary malformation with and without a meningomyelocele.[2,3,8]

ACM may present with a number of coexisting findings. Associated skeletal anomalies are present in about 23–45% of patients.[6] Osseous manifestations include scoliosis, Kleippel Feil Syndrome, and various skull abnormalities such as enlargement of the foramen magnum, flattening of the posterior fossa, and basilar impression.[1,3,6,9] If the protrusion is severe enough to cause obstruction of the Aqueduct of Sylvius and the 4th ventricle, internal hydrocephalus may occur.[1] Compression of the anterior spinal and the vertebral arteries at the foramen magnum is seen in severe cases.[1] Syringomyelia is seen in about 30–56% of the Type I cases.[6]

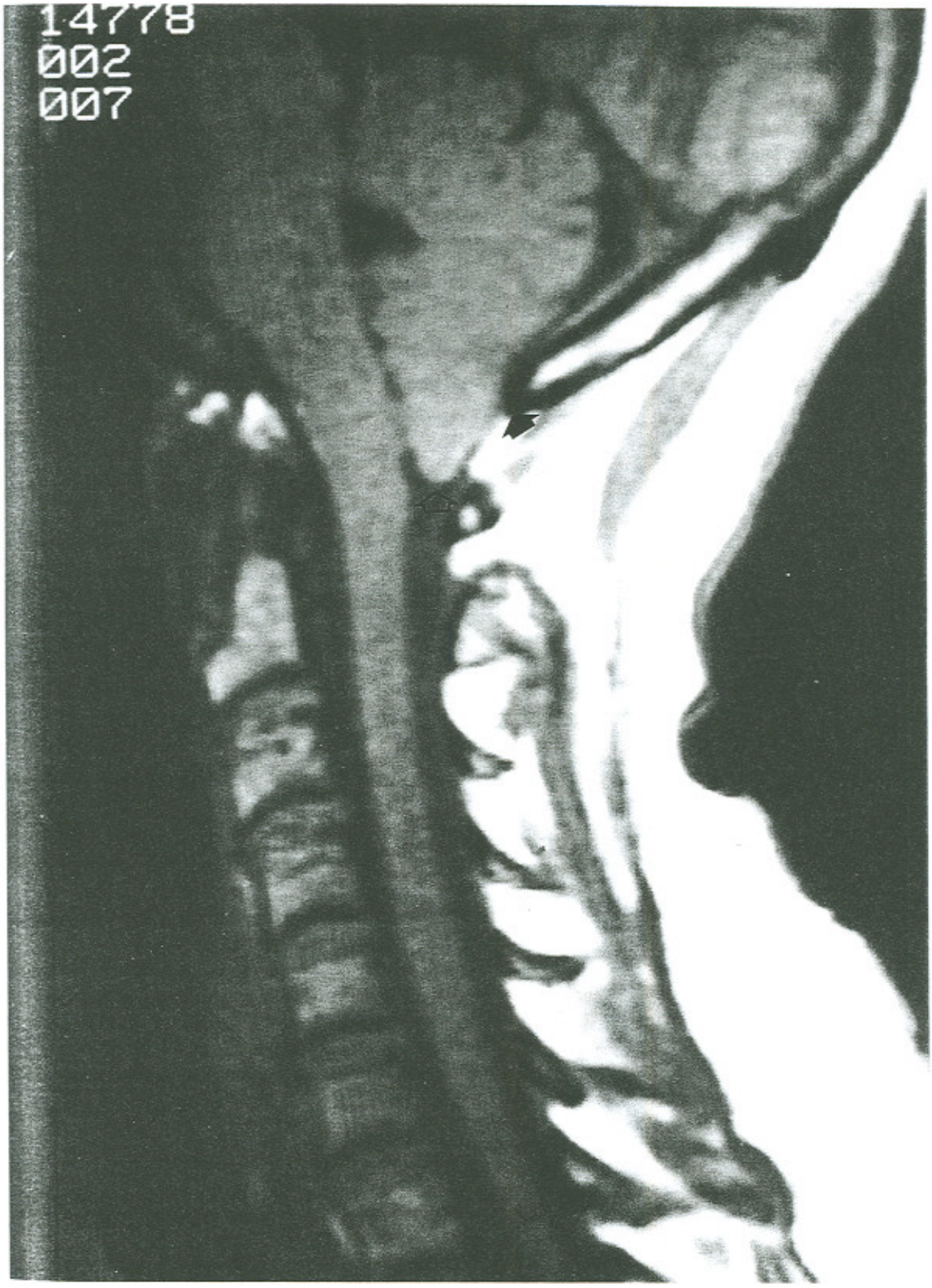

Figure 1 Sagittal T1 weighted MR image. There is tonsillar herniation to the level of the C1 posterior arches (open arrow). The medulla oblongata is elongated and the cisterna magna is thinned. There is caudal displacement of the cerebellar hemispheres. Solid arrow indicates the level of the foramen magnum.

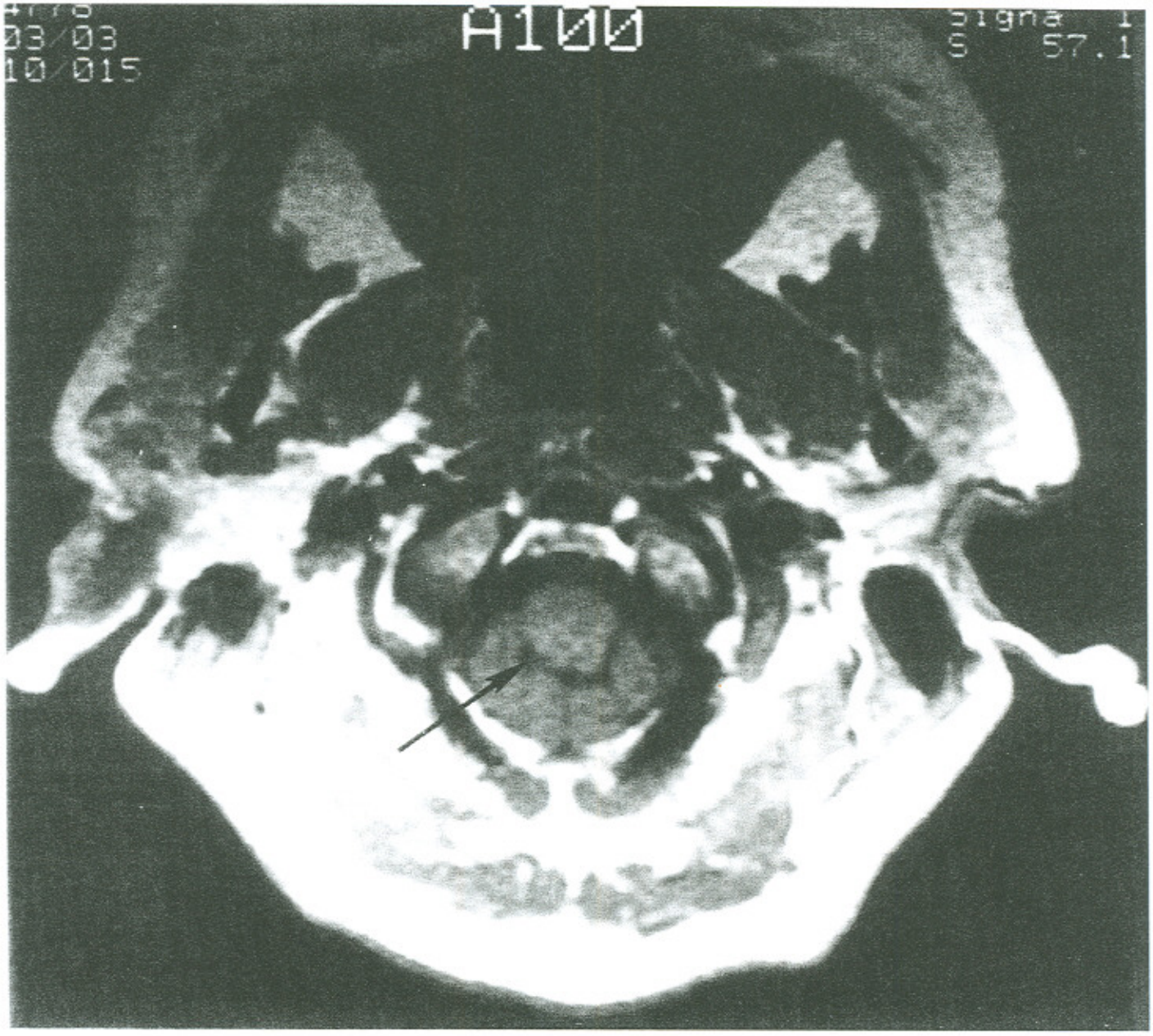

Figure 2 Axial T1 weighted MR image. The medulla is slightly compressed (short arrow) and there is thinning of the cisterna magna (long arrow).

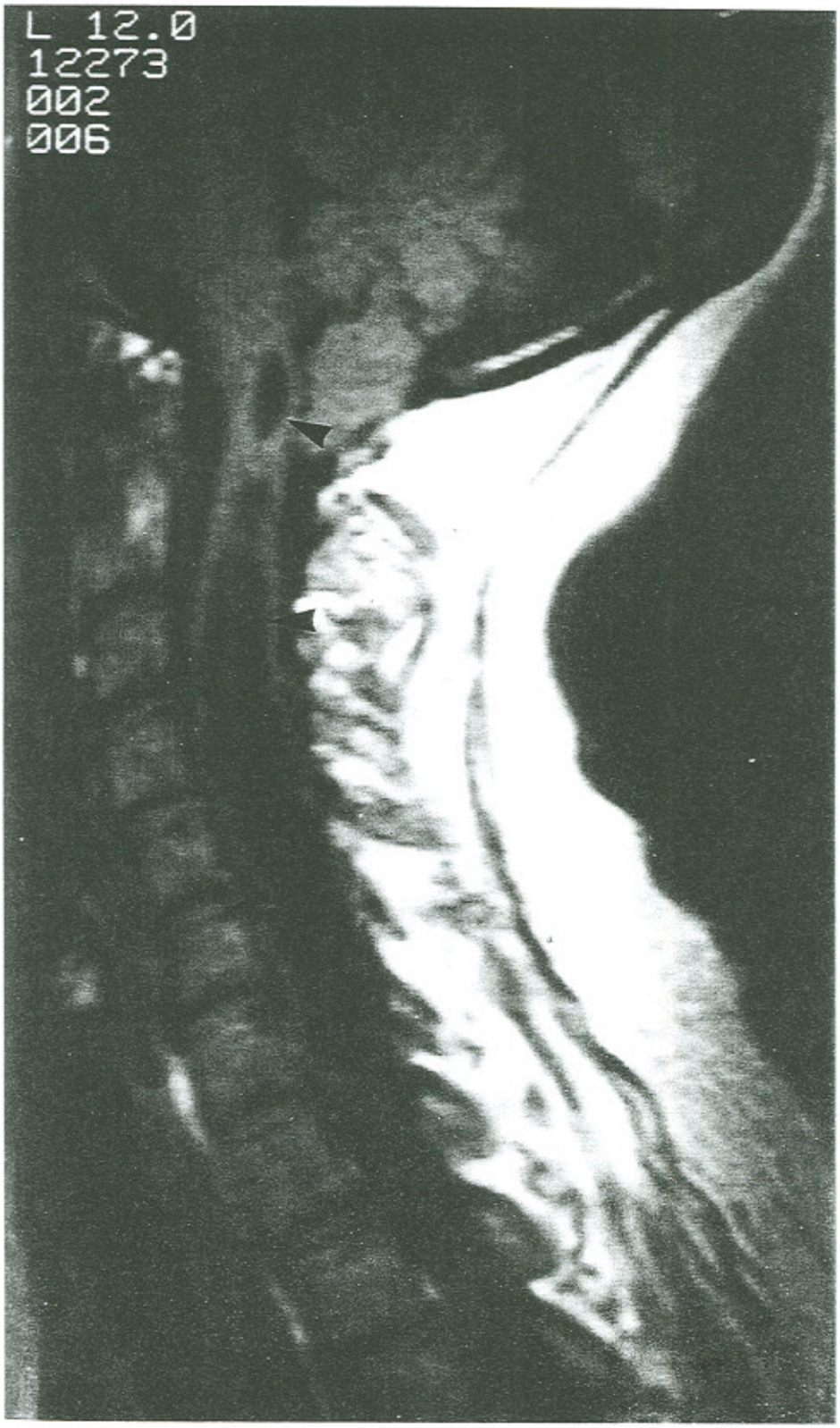

Figure 3 Sagittal T1 weighted MR image. There is herniation of the cerebellar tonsils to the level of the arches of C1. There is caudal displacement of the cerebellar hemispheres and elongation of the medulla. The cisterna magna is thinned. A small syrinx is present at the level of C1 and extends into the medulla (arrow). A large syrinx is present extending from C2 to C7 (arrow).

ACM Type II and III malformations are suspected in infancy because of associated meningoceles.[6] ACM I, however frequently remain undetected until late childhood or adulthood because they are often relatively asymptomatic. With the advent of MR imaging many ACM I are found as incidental findings on studies performed for other reasons.[6]

Clinical features of the ACM I, without syringomyelia, include symptoms and signs relating to hindbrain compression.[1,4,6,8] These include headache, neckache, weakness, numbness, or symptoms suggestive of a cerebellar tumor such as ataxia, cranial nerve palsies (nystagmus, oscillating vision, dysphagia, dysphasia), apnea, sudden death, persisting crying, and incontinence.[6] The most common symptom is pain in the occiput and/or neck due to compression of the upper cervical nerve roots. Symptoms can be precipitated by laughing, coughing, and running.[4] These episodic exacerbations often simulate an electric shock. The most common signs are motor and sensory deficits and changes in deep tendon reflexes.[4] Hydrocephalus and papilledema are the predominant findings when CSF circulation is impaired due to 4th ventricle outlet obstruction.[1]

Syringomyelia is a slowly progressive condition in which one or more eccentric fluid filled cysts are present in the spinal cord and the medulla.[10] Most cases are congenital in origin relating to maldevelopment of the cervicomedullary junction. Other cases are caused by arachnoiditis, intraspinal tumor or trauma and are referred to as secondary syringomyelia. An incidence of 8.4 per 100,000 births has been reported.[10] The most common type is the communicating syringomyelia or hydromyelia, in which the fourth ventricle communicates with the spinal cord cavity. This is thought to represent a dilated remnant of the fetal central canal and 60% of these patients have a coexisting ACM.[8] Noncommunicating syringomyelia develops in a segment of the cord rendered abnormal by an intramedullary tumor, spinal arachnoiditis, or severe traumatic injury. The two conditions are more appropriately combined into a single category called syringohydromyelia.[5]

A communicating syringohydromyelia typically is a slowly progressive destructive process in the cervical and thoracic portions of the spinal cord.[1,6,8,9] It occupies the central grey matter of the cervical portion of the cord where pressure on the crossing spinothalamic tract fibres causes a dissociated sensory loss with impaired perception of pain and temperature in the neck, arms, and upper trunk. Light touch perception and proprioception usually is preserved. As the syrinx expands it involves the anterior horn cells causing weakness and wasting of the muscles of the hands and arms, diminished reflexes in these areas, and often scoliosis due to denervation of the paraspinal muscles. Spastic weakness in the lower extremities is often found due to involvement of the white matter of the corticospinal tracts.[8,10] The precise clinical picture at any given point in time depends upon the cross-sectional and longitudinal extent of the syrinx.[8] Symptoms usually begin between 25 and 40 years of age and advance relentlessly for decades, although ⅓ of the patients have long periods of stability.[10] The deficits may worsen suddenly after a fall or after coughing or sneezing. Ten per cent of patients develop a painless neuroarthropathy (Charcot joint) of the shoulder, elbow, or hand.[10] Pain, however, can be present in ⅓ to ½ of patients. It is usually a unilateral or asymmetric burning or aching sensation present at the border of areas of sensory impairment.[8]

Patients with ACM Type I and a syrinx nearly always present with symptoms referable to the syrinx rather than the hindbrain compression.[6] When the syrinx extends into the medulla, (usually in the vicinity of the descending tract of cranial nerve V) however, bulbar symptoms will predominate.[1,6,10] Signs of syringobulbia include analgesia and thermoanesthesia of the face, wasting and weakness of the tongue, and palatal and vocal cord paralysis.[8] Other possible symptoms include nystagmus, 5th nerve palsy, mixed nerve palsy, as well as 12th nerve involvement.[6] Some patients may have hydrocephalus or cerebellar signs if they have an associated Chiari malformation.

Exceptions to the classical presentation described above do exist. In unusual cases, motor function may be spared and segmental dissociated sensory loss and/or pain may be the only signs of the disease. In a few cases, especially those with associated ACM, upper extremity deep tendon reflexes are preserved or even hyperactive. The shoulder muscles can be atrophic and the hands spastic. Occasionally the characteristic bilateral sensory loss may be only unilateral, or the patient will present with unilateral amyotrophy. Finally, while tactile sensation is classically preserved, there are cases where it is impaired.[8]

Syringomyelia can be classified into three types:[8]

I Syringomyelia with obstruction of the foramen magnum and dilation of the central cord:
   a) with ACM Type I,
   b) with other obstructive lesions of the foramen.
II Syringomyelia without obstruction of the foramen magnum (idiopathic type).
III Syringomyelia with other diseases of the spinal cord:
   a) spinal cord tumors,
   b) traumatic myelopathy,
   c) spinal arachnoiditis and pachymeningitis.
IV Pure hydromyelia with or without hydrocephalus.

Diagnostic imaging

Radiography may offer early clues that a congenital anomaly exists. Findings which raise suspicion for ACM include a small posterior fossa, enlarged foramen magnum with a convex posterior border, and enlarged diameter of the spinal canal which may be associated with scalloping of the posterior margins of the vertebra or widened interpedicular distance.[1] Other important findings include Kleippel Feil Syndrome, basilar invagination, platybasia, atlanto-occipital assimilation, spina bifida, fused cervical vertebra or fused ribs, and scoliosis.[6,9] A rapidly progressive scoliosis especially a left convex thoracic curve

should alert the practitioner toward the possibility of underlying neurologic disease.[4,18]

Conventional myelography, computerized tomography (CT) and computerized tomography with myelography (CTM) are useful imaging procedures in assessing ACM and syringomyelia, however MR imaging is the procedure of choice.[2,5,6] Sagittal T1-weighted images of the cervical spine and skull base would reveal the extent of involvement of the posterior fossa structures in ACM. To determine the extent of herniation of these structures a line representing the plane of the foramen magnum is constructed extending from the basion to the opisthion according to the method established by Aboulezz.[6] The degree of herniation is measured along a perpendicular line extending from this baseline. Coronal images may reveal asymmetry in tonsillar herniation, however this is not usually regarded as being significant.[6] Sagittal T1-weighted images also will reveal the presence of myelomeningeocele or hydrocephalus.

Elster and Chen,[6] use the following criteria to diagnose ACM Type I:

a) herniation of at least one cerebellar tonsil 5 mm or more below the plane of the foramen magnum;
b) herniation of both cerebellar tonsils 3–5 mm below the foramen magnum if accompanied by other definite feature consistent with a ACM Type I (i.e. syrinx, cervicomedullary kinking, or elongation of the fourth ventricle);
c) no clinical history of myelomeningocele or radiologic evidence of a ACM Type II;
d) no prior cranial or cervical spinal surgery; and
e) tonsillar herniation appearing to be a primary dysplasia and not secondary to a cerebellar mass lesion.

Syrinx cavities associated with ACM will be readily apparent as a well defined cavity within the central cord, having the same signal characteristics as CSF.[2,5] No capsule is identified on any imaging modality. In one study the most common level for a syrinx was between C4 and C6.[6] The thoracic cord was involved in 20 of 27 patients and only one syrinx began below C4. Skip areas of syrinx were encountered in 3 of these patients.[6] When a syrinx is detected that is apparently not related to ACM, the decision has to be made whether it is a primary syrinx (congenital) or a secondary syrinx (neoplastic, inflammatory, or traumatic). In the majority of cases the diagnosis of primary syrinx may be used on thinning of the parenchyma adjacent to the expanded cavity. The internal margins are smooth, but may be interrupted by septations that can be continuous or incomplete.[2] Intravenous injection of gadolinium during MR imaging helps to distinguish neoplasms from a primary syrinx. Following contrast administration if no enhancing nodule is seen, a cord neoplasm is unlikely.[2]

Examination of the entire cord with T1-weighted sagittal images is suggested. Failure to examine the entire cord may result in missing cavities that are separated from the primary or larger cavity. Long segments of normal appearing cord may intervene between a cervical cavity and one located in the conus.[5]

On comparing the volume of the cyst detected by MR imaging with clinical symptoms, signs, and disability, Grant[11,12] recognized that there was no direct relationship between the imaging dimension of a syrinx and the associated neurological disability. Patients with a normal or atrophied spinal cord lesion were just as severely affected as those with an expanded cord. According to Modic[5] patients with congenital cavities tend to be less symptomatic than patients with acquired cysts. This may be because congenital cavities are not associated with as much focal myelomalacia as traumatic cysts. This implies that a more gradual growth of a cyst in an otherwise normal cord may be less symptomatic and that the cord may accommodate a cyst if the changes are gradual.

Treatment considerations

Arnold-Chiari malformations. Surgical treatment of ACM Type I without associated syrinx involves a posterior fossa craniectomy and usually a C1 laminectomy. The dura may be opened and adhesions may be divided. According to Nohria and Oakes[4] this type of procedure produces greater improvement in symptoms than signs. Surgery is much more likely to improve the presenting complaints, while the improvement in the objective examination may be less impressive, particularly concerning motor deficits. They go on to conclude that the longer the duration and the greater the severity of the symptoms and signs the less likely any resolution will occur.[4]

Syringohydromyelia. The utility of surgical procedures for dealing with syrinx cavities, either associated with ACM or independent of ACM is not clear. The overall impression is that there is little difference in effectiveness between the various surgical procedures[11,16,17] and that although improvement occurs in ⅓ of patients it persists only in 18% after 4 years.

Munari[11] concluded that there was no advantage to surgery for syringomyelia when neurologic damage is severe as the majority of cases worsened anyway. Surgery may improve the status of patients whose neurologic deficits are only segmental, however these cases should be operated on early. It should be remembered nevertheless that in about half of all the cases of syringomyelia the disease is not progressive. The presence of a ACM did not seem to affect the course of the disease nor influence surgical considerations.

Conclusion

It is essential for practitioners to be aware of the varied presentations of ACM Type 1 in the adult. Two cases are presented which have widely differing clinical manifestations. Case 1 was one of sudden onset with very marked symptomatology requiring immediate surgery. Case 2 exhibits a similar degree of tonsillar herniation, but also had a very large cervical syrinx and a smaller cervicomedullary syrinx. Clinically, this case pro-

gressed very insidiously. Her symptoms were comparatively mild and they were somewhat atypical in presentation. Her neurologist and her neurosurgeon have elected to monitor this individual and observe its progression.

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