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Case Report

Generalised spinal cord atrophy, Chiari-I malformation, and syringomyelia

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Objectives: To report the simultaneous occurrence of generalised spinal cord atrophy, Chiari-I malformation, and syringomyelia.
Setting: Vienna, Austria.
Case description: An 83-year old woman presented with predominantly lower limb and distal tetraparesis, diffuse wasting, spasticity of both lower limbs, contractures, and severe kyphoscoliosis. Radiographic examinations revealed generalised spinal cord atrophy, Chiari-I malformation, and syringomyelia between C5 and C7. The generalised spinal cord atrophy was assumed to be due to either secondary atrophy or spontaneous collapse of a formerly more extensive syringomyelia. All other causes for generalised spinal cord atrophy were excluded.
Conclusions: Generalised spinal cord atrophy in a patient with Chiari-I malformation and syringomyelia may be due to either secondary atrophy or spontaneous collapse of a formerly holocord syringomyelia. Myelography and MRI imaging of the spine are helpful in diagnosing this condition.
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Keywords: syringomyelia; Chiari-I malformation; generalised spinal cord atrophy; kyphoscoliosis; myelography; hydromyelia

Introduction

Generalised spinal cord atrophy is rare. Several causes can be found responsible for this entity.[1–6] One of these is Chiari-I malformation leading to secondary cord atrophy or holocord syringomyelia which may collapse spontaneously or be mechanically induced. The following report is the description of a patient with generalised spinal cord atrophy, attributed to either collapse of a formerly holocord syringomyelia or secondary compression of the cord due to Chiari-I malformation.

Case history

An 83-year-old woman was admitted and evaluated for painful muscle cramps and progressive weakness of the lower limbs, leading to inability to walk or even stand, since a fall 3 months earlier. The patient had a history including forceps cephalic delivery without asphyxia at birth. For the last 23 years type II diabetes was successfully treated with oral antidiabetic drugs. Since the age of 18, the patient has developed abnormal right finger position, slowly progressing weakness and wasting of the right lower arm and hand muscles. For 10 years, weakness has also developed in the left arm muscles and was followed by abnormal finger position. Over the last 7 years, weakness and wasting progressed to the abdominal muscles and lower limbs, leading to chronic constipation and gait disturbance with frequent falls. During the last 5 years, the patient has complained about recurrent dyspnoea, which has been attributed to chronic heart failure. Four years before admission, the patient was operated for a left-sided cataract. For the last 4 years, the patient could walk only with aid. Urine incontinence developed 2.5 years ago. Two years before admission, she underwent local resectioning of an intraductal carcinoma of the left breast and a basal cell carcinoma on the back. Repeated controls did not reveal relapses of either tumour. The father and oldest brother of the patient had gait disturbance of undetermined cause, but both had already died and no medical reports about their clinical presentation and instrumental findings were available. The other family members, including the

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patient's only daughter and two grandchildren, had none of the described symptoms.

On clinical neurologic examination, normal cognitive functions, palpebral fissures, wider on the left than the right side, unrounded pupil and weak reaction to light on the left side (attributed to cataract surgery) and oro-facial dyskinesia were found. The patient was tetraparetic (MRC grade 3–4) with distal, lower limb and right-sided predominance. Deep tendon reflexes were absent in the upper limbs and hyperactive in the lower limbs. Adductor and flexor spasticity was present. There was marked diffuse wasting of the upper and lower limbs with distal accentuation. Plantar responses were flexor bilaterally. Hips, knees and ankles were contracted in a flexor position. The patient was able to sit with support, but was unable to stand or walk. No foot deformities were found. There were sensory disturbances affecting all modalities of the right hand up to the wrist.

Routine laboratory tests were normal, except for anaemia and an increased C-reactive protein. Serum anti-HTLV-1 antibodies, immunological parameters, CSF examination, the carcino-embrional antigen, CA125, CA15-3, GM1 antibodies, non-esterified fatty acids and β-hexosaminidase levels were all normal. X-ray of the spinal column showed severe cervical hyperlordosis, gibbous and scoliosis without segmental instability. Additionally, spondylosis, spondlyarthrosis and osteoporosis were found. Myelography revealed severe spinal deformity and a band-like cord with a sagittal diameter of a few millimetres (Figure 1). Magnetic resonance imaging (MRI) of the spine also showed severe spinal deformity, Chiari-I malformation, and severe, generalised spinal cord atrophy with missing cervical and lumbar expansion. The sagittal diameter of the spinal cord was 2–3 mm and the subarachnoidal space was compensatorily enlarged. Between C5 and C7, two syringomyelinic cavities were found (Figure 2). Neither spinal stenosis nor dysraphism were present. A computed tomography (CT) scan of the brain showed moderate generalised cerebral atrophy and leucoaraiosis. The brainstem and cerebellum were normally sized. Excretory urography revealed bilateral hydronephrosis II. Nerve conduction studies of the left median and peroneal nerves showed reduced compound muscle action potentials and slowing of the nerve conduction. Orthodromic sural nerve stimulation failed to evoke a response. Electromyography of the left anterior tibial muscle was neurogenic. Pain due to cramps and spasticity were successfully relieved with tizanidine, tetrazepam, tramadol and botulinum toxin.

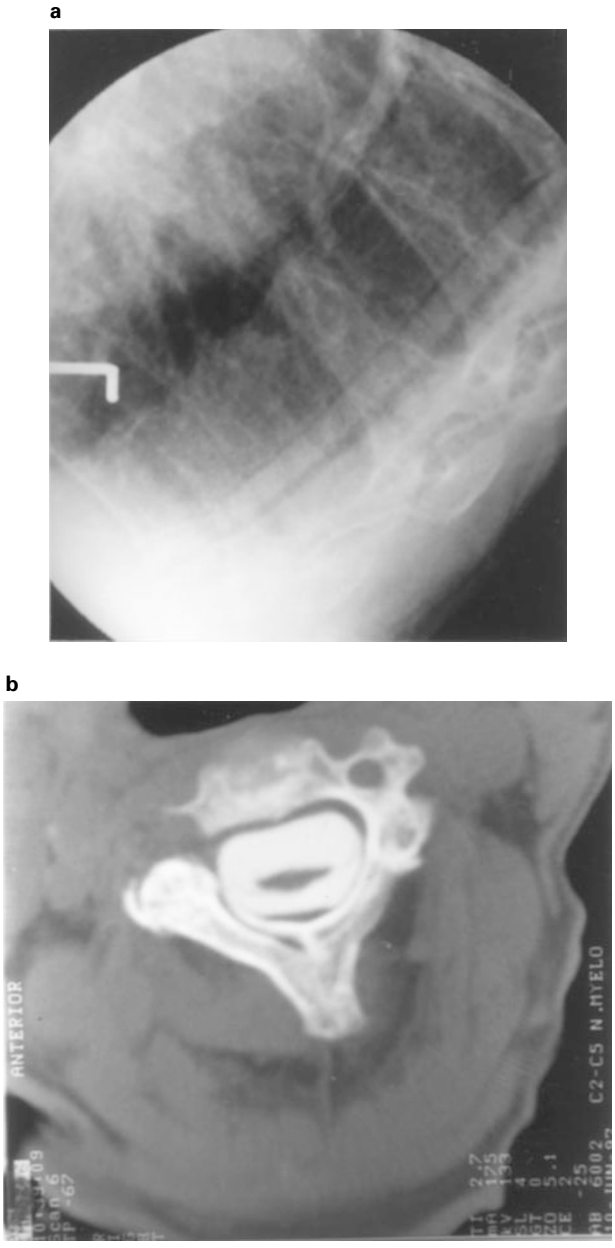

Figure 1 Myelography of the cervical and thoracic spine shows severe reduction of the sagittal spinal cord diameter (**A**). A CT-myelography scan of the cervical spine in a transverse plane shows flattened cross-sectional spinal cord area (**B**)

Discussion

The dominant feature in the presented patient was severe spinal cord atrophy. Generally, spinal cord atrophy may be focal or generalised. Focal spinal cord atrophy may be due to Chiari malformation, previous CNS infection, trauma, radiotherapy, spinal surgery, immunological disorder, vascular abnormality or CNS tumour.[1–3,6–8] Generalised spinal cord atrophy is rare and may be due to Chiari malformation, luetic CNS infection,[9] and hereditary disorders like arthrogryposis multiplex congenita,[10] Chediak-Higashi syndrome,[5] adrenoleucodystrophy,[4] Sjögren syndrome,[11] familial spastic paraparesis,[12] and hereditary motor and sensory neuropathy with pyramidal signs (HMSN-V).[6] Apart from these causes, generalised spinal cord atrophy may be attributed to a collapse of a formerly holocord syringomyelia, leading to reduction of the spinal cord

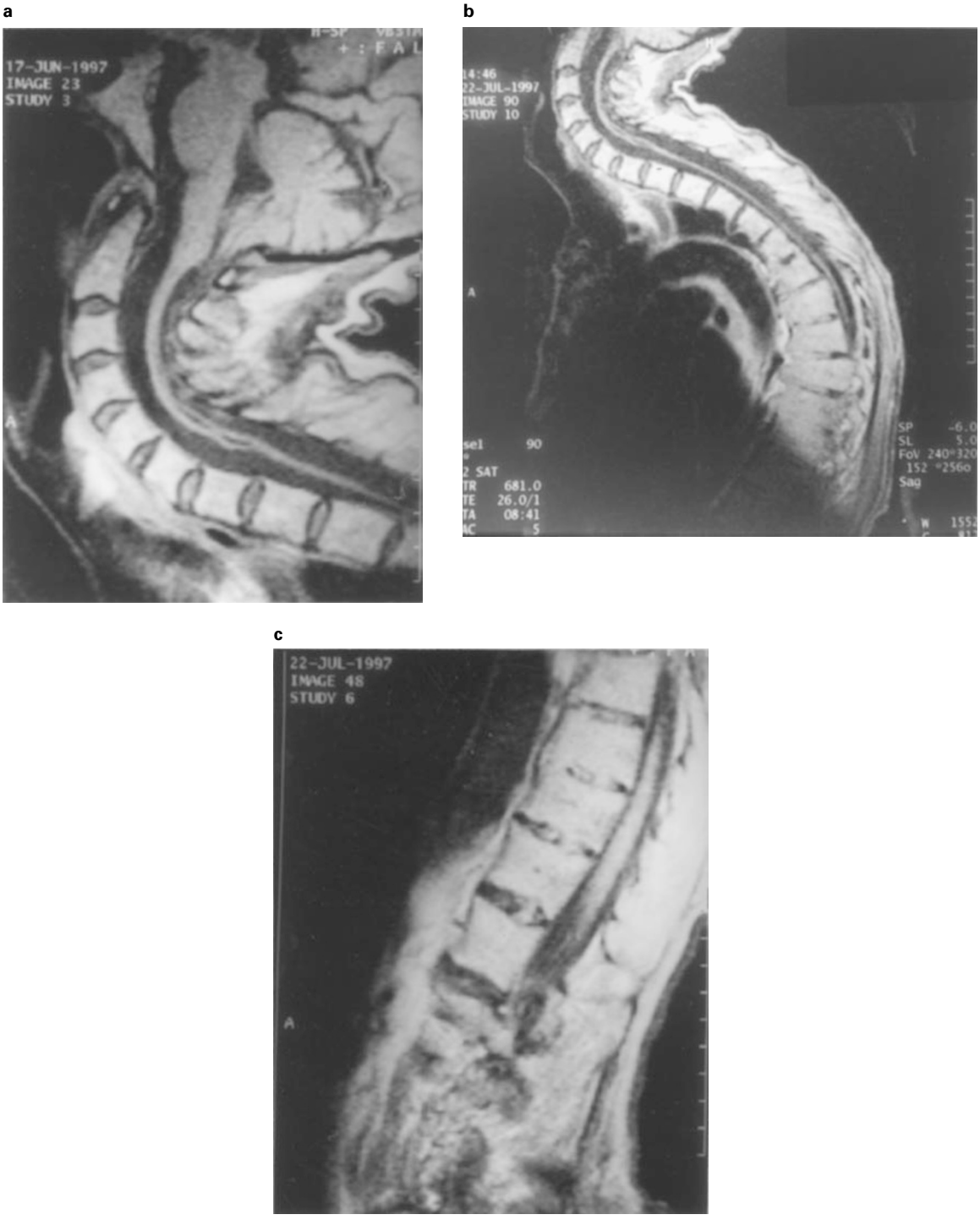

Figure 2 T1 weighted image of the cervical spine on a saggital plane shows Chiari-I malformation, cord atrophy with normal signal intensity and missing cervical expansion, compensatory enlargement of the subarachnoidal space and syringomyelinic cavities at C5–C7 (**A**). Cord atrophy extends to the thoracic and lumbar region (**B**, **C**). In addition severe spinal deformity can be seen

diameter, mimicking spinal cord atrophy. Except for Chiari-I malformation and collapse of a holocord syringomyelia, all other causes for generalised spinal cord atrophy were excluded in the described patient. Familial spastic paraparesis was excluded, because the onset of symptoms was in the upper limbs and there was tetraparesis instead of paraparesis.[13] HMSN-V was excluded, because the onset of symptoms was in the upper limbs and upper motor neurone signs were absent in the upper limbs.[14]

Chiari-I malformation, one possible cause for generalised spinal cord atrophy in the presented patient is defined as >5 mm herniation of the cerebellar tonsils through the foramen magnum, but only in the absence of dysraphism, craniocerebral dysgenesis, deformities of the foramen magnum (occipital encephalocele, multiple craniosynostosis, small skull base), fourth ventricle elongation and cervicomedullary kinking.[15,16] Chiari-I malformation may remain asymptomatic or may present with few clinical findings due to brain stem compression or cervical syringomyelia.[15] Usually, Chiari-I malformation is acquired, and rarely hereditary. Acquired Chiari-I malformation may be due to a cerebral mass lesion such as tumour, cerebral oedema and hydrocephalus, lumbar-peritoneal shunt or trauma. Except for birth trauma due to traction of the brain and cord during forceps delivery, all other causes for acquired Chiari-I malformation were excluded in the presented patient. Arguments against birth trauma as a possible cause of Chiari-I malformation are that the postpartum period was uneventful and that the initial symptoms did not occur before adulthood.[17] Most likely, Chiari-I malformation was hereditary in the presented patient. The malformation may have compressed the cord to such a degree that not only CSF circulation was disturbed, but also secondary cord atrophy due to direct cord compression developed.

Collapse of a formerly holocord syringomyelia may be another possible explanation for severe cord atrophy in the presented patient. Usually, syringomyelia is due to CSF circulation blockade with consecutive dilation of the central spinal canal. The degree of dilation relates to the degree of obstruction of the CSF circulation.[18] The most frequent cause of syringomyelia is Chiari malformation. All other causes were excluded in the presented patient, including meningeal inflammation, that could have left scars which later induced ischaemia and subsequent cavitation,[19] trauma or tumour that lead to ischaemia by compression, and myelomalacia and cavitation.[18] Whether the cervical syringeal cavities are remainders of a collapsed holocord syringomyelia remains unknown, since no delayed films were taken to look for central cord enhancement, and no axial MRIs were done to show small residual syringomyelias in the atrophic cord. The discrepancy between the comparatively small size of the cervical syrinx and the severity of the cord atrophy may favour the collapse theory. A further argument in favour of a pre-existing and more extensive syrinx is the appearance of an enlarged spinal canal on MRI. In view of the patient's symptoms, existing for decades, it is conceivable that the syrinx spontaneously decompressed at some point. Induction of this collapse by myelography is rather unlikely, since it is assumed to occur only with air and not with water-soluble contrast myelography, and since no acute deterioration could be observed during or after myelography. Particularly in long-standing syrinx cases, spontaneous decrease in syrinx size has been noted. A formerly widespread syringomyelia with almost normal spinal cord diameter can turn into limited residual cavitation and a severely wasted spinal cord.[20]

Kyphoscoliosis developed most likely secondary to the anterior horn cell damage, following spinal cord atrophy. Whether hydronephrosis was due to cord atrophy remains speculative. Though urologists found no cause for this abnormality, incontinence and recurring urine bladder infections could be indicators for a neurogenic ureter and urine bladder dysfunction, also suspected by urodynamic investigations. Cataract, oro-facial dyskinesia, and heart failure were interpreted to be senescent. Findings on nerve conduction studies and EMG, compatible with polyneuropathy, were attributed to the long-standing diabetes. Whether these findings were in part due to the spinal process remains speculative. No paraspinal EMGs were performed. Clinical implications of the presented case are that in patients with weakness and wasting of all four extremities, spasticity of both lower limbs, and severe skoliosis, Chiari-I malformation with syringomyelia and cord atrophy have to be ruled out by appropriate radiographic examinations. Early recognition of such abnormalities could prevent severe kyphoscoliosis by surgical stabilisation of the spine.

It is concluded that generalised spinal cord atrophy may be due to Chiari-I malformation, with either secondary spinal cord atrophy or spontaneous collapse of a formerly holocord syringomyelia. Myelography and MRI imaging of the spine are helpful in diagnosing this condition.

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