

## CASE REPORT

# Arnold–Chiari malformation with syringomyelia in an elderly woman

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## Abstract

**Presentation:** a 76-year-old woman, complaining of leg pain and unsteady gait for 3 years, presented with a spastic paraparetic gait, severe spasticity and touch, thermal and pain sensory loss limited to arms, lower thorax and upper abdomen. Brain and spinal cord magnetic resonance imaging showed a large loculated syrinx. Cerebellar tonsillar herniation into the foramen magnum was also seen (Arnold–Chiari malformation, type D).

**Outcome:** the patient had successful cervico-spinal surgical decompression which resulted in marked reduction in hypertonia and weakness, normal gait and normal joint movement at 6 months.

**Conclusion:** this unusual, late clinical presentation of a congenital disease underlines the importance of a comprehensive diagnostic work-up in the elderly patients with complex neurological signs.

**Keywords:** *Arnold–Chiari malformation, elderly people, syringomyelia*

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## Introduction

Arnold–Chiari malformation is characterized by the herniation of the cerebellar tonsils into the foramen magnum, often associated with syringomyelia and therefore presenting with features of a spinal cord lesion. It is congenital in all cases associated with spina bifida (type II Arnold–Chiari malformation) and in many cases without spina bifida (type I malformation). Acquired forms have also been described, and may be due to an abnormality of cephalo-cranial development in some cases and to a dissociation of pressures in the cranial and spinal compartments in others [1–3]. While patients with tonsillar herniations greater than 12 mm are usually symptomatic, 30% of patients with tonsils herniating 5–10 mm below the foramen magnum are asymptomatic [4].

Syringomyelia as a complication of Arnold–Chiari malformation is usually apparent in early or middle life [5, 6]. We report case of late-onset syringomyelia in a 76-year-old woman with Arnold–Chiari malformation.

## Case report

A 76-year-old woman came to our day hospital with a progressive gait disturbance. Fifteen years earlier, cancer of the left breast had been diagnosed and treated by radical excision and radiotherapy (caesium, 4500 rads over 40 days). Over the past 3 years, she had developed leg pain and an unsteady gait, which slowly worsened. Eight months before examination, her legs became spastic with progressive worsening of leg pain, gait disturbance and functional impairment. Three months earlier, she became aware of thermal sensory loss in the hands and forearms.

Neurological examination showed a spastic paraparetic gait, severe spasticity, more marked on the left side and in the arms, and bilateral weakness. There was touch, thermal and pain sensory loss limited to the arms, lower thorax and upper abdomen (transversal syndrome). Tendon reflexes were normal in the arms, exaggerated in the legs. Hoffmann and Babinski signs were elicited bilaterally. Cranial nerves were spared and cerebellar signs absent. Brain and spinal cord magnetic resonance imaging showed a large loculated



Figure 1. Brain and spinal cord magnetic resonance showing a large loculated syrinx, spreading from the bulbar-spinal junction to a mid-dorsal level. The syrinx is more expanded at the cervico-dorsal junction level, causing fusiform enlargement of the spinal cord, with obliteration of the surrounding cerebro-spinal space. Cerebellar tonsillar herniation into the foramen magnum is also seen, with restriction of the outlet of the fourth ventricle and of the cerebello-medullar cistern (Arnold-Chiari malformation, type D).

syrinx, spreading from the bulbar-spinal junction to a mid-dorsal level (Figure 1), with cerebellar tonsillar herniation (about 8 mm) into the foramen magnum (Arnold-Chiari malformation type I) that had normal dimensions (30 × 33 mm).

The patient was referred to the neurosurgical unit, where she underwent cervico-spinal surgical decompression. No signs of chronic arachnoiditis were found at operation. Seven days later, cervico-dorsal spinal cord magnetic resonance showed good decompression. On neurological examination, spasticity was milder, being restricted to the legs, and movement had improved a little. She experienced less effort while walking. Six months and 2 years later, she had further neurological examinations, which revealed marked reduction in spasticity and weakness, normal gait and normal joint movements. Tendon reflexes remained exaggerated in the legs, the Hoffmann sign was still bilaterally elicited, but the plantar reflex was now flexor. Touch, thermal and pain sensory loss on the arms, thorax, and upper abdomen were still present on clinical examination, but the patient reported much subjective improvement.

## Discussion

An unusual feature of this case is the age of the patient at onset of the first symptoms (before the age of 74 the

patient had been asymptomatic). Syringomyelia is a common complication in type I Arnold-Chiari malformation [5], but in late adulthood and in old age it is more frequently caused by acquired factors. In particular, adhesive arachnoiditis of any cause (infective, ischaemic, traumatic) can cause a block of the cerebrospinal fluid dynamics in the spinal subarachnoid space. This can initiate the extension of the syringomyelic cavity [7, 8]. The only major medical event in the history was the mastectomy followed by caesium radiotherapy. Before neurosurgery, we had considered that the prolonged radiation therapy to the left breast might have caused chronic arachnoiditis, with adherence of the cord to the meninges, and downward traction of the cerebellum and brainstem. However, no signs of arachnoiditis were found. Therefore, the only explanation is of congenital disease with late expression: Arnold-Chiari malformation in this subject was a 'malformation', which had been asymptomatic until her eighth decade.

This case illustrates that in elderly patients neurological symptoms can be caused by unusual, even congenital, conditions. This underlines the importance of thorough diagnostic investigation, even in old age, in selected patients.

## Key points

- A 76-year-old woman presented with Arnold-Chiari malformation with syringomyelia, which had been asymptomatic for 74 years.
- Neurological symptoms in elderly patients can be caused by unusual, even congenital, conditions.

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