Association of HTLV-I with Arnold Chiari Syndrome and Syringomyelia

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HTLV-I is associated with a broad spectrum of manifestations, including tropical spastic paraparesis and adult T-cell leukemia/lymphoma. Arnold Chiari syndrome is a condition characterized by herniation of the cerebellar tonsils through the foramen magnum. This condition should be suspected in all patients with headache and impaired motor coordination. Syringomyelia is a developmental anomaly that leads to the formation of an intramedullary cavity. Its clinical presentation is classically characterized by syringomyelic dissociation of sensation, with suspended distribution in the proximal portion of the trunk and upper limbs and preservation in other regions. We report here a case of association of the three diseases, which is rare in clinical practice, illustrating the difficulty in the diagnosis and therapeutic management of these conditions.

Key-Words: HTLV-I, Arnold Chiari syndrome, syringomyelia.

Human T-cell lymphotropic virus I (HTLV-I) is a type C virus of the family *Retroviridae*, subfamily Oncovirus, which contains double-stranded, enveloped RNA. Approximately 10 to 20 million people worldwide are infected with HTLV-I, with the infection thus being considered a global epidemic [1]. The virus is associated with a broad spectrum of manifestations, including tropical spastic paraparesis, a disease characterized by the slow and progressive development of neuromyelopathy, and adult T-cell leukemia/lymphoma, a lymphoproliferative disorder of rapid fatal progression which is characterized by its incurability and damage caused to the patient [2-4].

Infection with HTLV-I is endemic in different regions of the world, with high prevalence rates being observed in the south of Japan, Caribbean, Africa, southeastern Italy, Papua New Guinea, the Middle East, Australia and South America [5]. The prevalence increases with age and in the presence of a history of blood transfusion, and is two times higher in women older than 20-30 years than in men, especially among patients of low socioeconomic conditions [6,7]. This difference between genders after 20-30 years of age probably reflects more effective viral transmission from men to women during the sexually active years.

The most common routes of HTLV-I transmission are perinatal transmission (especially due to prolonged breast-feeding), parenteral exposure (blood transfusion and contaminated syringes) and sexual transmission [8]. Intrauterine transmission is rare, but postnatal infection plays an important role in the transmission from mother to infant. Among children born to infected mothers but who are not breast-fed, only 5.7% contract the infection, whereas the chance of vertical transmission ranges from 18% to 30% [9]. Blood transfusion is probably the most effective route of viral transmission, with a 40%-60% chance of seroconversion after the use contaminated blood [10].

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Arnold Chiari syndrome is a condition characterized by herniation of the cerebellar tonsils through the foramen magnum and is classified into three types according to the degree of herniation [11]. Although more than 100 years have elapsed since the identification of this disease, anatomopathological and embryological studies are still scarce [12]. The condition should be suspected in all patients with signs and symptoms involving the cerebrocerebellar system, particularly when they coexist with signs and symptoms of headache and impaired motor coordination.

Syringomyelia is a developmental anomaly that leads to the formation of an intramedullary cavity. The lower cervical segments are the most affected, but cranial extension to the brainstem (syringobulbia) or extension to the medullary cone might be observed. The clinical presentation is classically characterized by syringomyelic dissociation of sensation, with suspended distribution in the proximal portion of the trunk and upper limbs and preservation in other regions [8]. Symptoms include burning pain irradiating to the upper limbs and hypotrophy and weakness of the intrinsic hand and scapular muscles due to involvement of the anterior horn at this level of the spine. Later, signs of involvement of long tracts (lateral corticospinal tract) may arise, mainly pyramidal liberation [13].

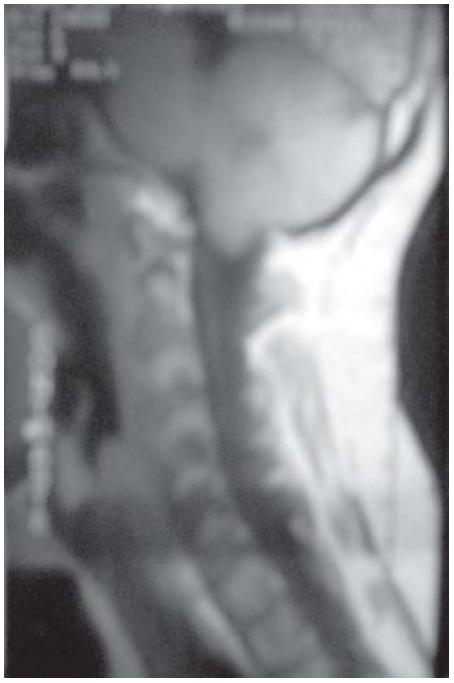
We report here a rare case of association of the three diseases, illustrating the difficulty in the diagnosis and therapeutic management of these conditions.

Case Report

A 44-year-old woman, a housewife, from the Northeast of Brazil, had been submitted 18 years earlier to neurosurgery (she did not know the type of surgery) for the treatment of progressive atrophy and a mild loss of muscle force in the left arm. Discrete loss of balance and gait alterations were observed during the immediate postoperative period. One month after surgery, radiologic investigation led to the diagnosis of Arnold Chiari malformation and syringomyelia (Figure 1). Ten years after surgery, the patient was no longer able to walk on her own, moving around only with the help of two persons, and had started treatment at a motor rehabilitation hospital where neurogenic

bladder and intestinal disorders were diagnosed. In addition, infection with HTLV was diagnosed by ELISA and Western blotting serology. From that time on the patient used a walker and has been using a wheelchair for locomotion for one year. The clinical course over the last 8 years was characterized by neuropathic pain, spasticity and lower limb contractions and the use of symptomatic medication. Imaging follow-up (computed tomography and magnetic resonance) did not show any increase in the syringomyelia cavities or in the extent of these lesions.

Figure 1. Syringomyelia associated with Chiari malformation. The intramedullary cavity is characterized by a homogenous hypointense and well-defined lesion, increasing the medullary diameter.



Discussion

The association of syringomyelia and Arnold Chiari syndrome has been commonly reported in the literature [13-17]; however, there are no reports regarding the association of these two diseases with HTLV-I infection.

The present case agrees with the age and gender most frequently affected by HTLV [18,19] and has occurred in an area of high prevalence of this infection [18]. In view of the clinical evolution of the patient, it is likely that she already presented Arnold-Chiari syndrome and syringomyelia at the time of surgery. In addition, although decompression surgery of the cerebellar tonsils is indicated as an effective treatment of the syringomyelia-Chiari I complex, resulting in the regression of symptoms during the immediate postoperative period or clinical improvement in 78% of cases [20], in the present study surgery did not result in clinical improvement and aggravation of the symptoms was observed.

Since imaging exams showed no atrophy of the thoracic spine nor an increase in the syringomyelia cavities or the extent of these lesions, the current presentation was attributed to infection with HTLV-I which explained the occurrence of intense pain, lower limb contraction, marked spasticity and greater walking difficulty, signs compatible with tropical spastic paraparesis/HTLV-I-associated myelopathy.

We emphasize the difficulty in establishing an early diagnosis in patients with neurological symptoms and especially in establishing therapeutic management aimed at improving the quality of life of these patients who are in their productive and socially active years. The aim of the present case report was not to describe a new association of diseases, but to alert colleagues about the possbility of association between these neurological diseases.

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