Syringomyelia: Retrospective Analysis of 30 Surgically Treated Patients from the Northeast of Brazil

Siringomielia: Análise retrospectiva de 30 casos operados no nordeste brasileiro

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Arq Bras Neurocir 2024;43(1):e1–e6.

Abstract

Introduction Syringomyelia is a chronic disease characterized by the presence of intramedullary cavity. Chiari malformation (CM) and basilar impression (BI) are conditions usually associated with syringomyelia. Its prevalence has wide geographical variation, being higher in the Northeast of Brazil, making it relevant to study the subject in this region.

Objective To analyze the frequency of signs, symptoms, and surgical aspects observed in patients undergoing decompressive treatment.

Methods We performed a retrospective analysis of the medical records of patients diagnosed with syringomyelia who received decompressive surgical treatment in various hospitals in João Pessoa, Paraíba, between 1994 and 2021.

Results Thirty patients were analyzed. Twenty-nine (96.7%) presented CM and 27 (90.0%) also presented BI. A wide variety of symptoms was found, with significant prevalence of muscle weakness, neck pain, and headache. Brevicollis, a finding considered typical of the Northeastern region and associated with craniocervical junction malformations, was present in 66.7%. The surgical technique used in 90% of patients was similar. Fourteen (46.7%) patients presented difficult craniocervical junction and 4 (13.3%) had occipital bone assimilation. Eighteen (60.0%) presented

Keywords
- craniocervical junction malformations
- syringomyelia
- Chiari malformation
- basilar impression
- surgery

received
August 9, 2022
accepted
March 7, 2023
article published online
October 18, 2023

DOI https://doi.org/10.1055/s-0043-1774746.
ISSN 0103-5355.

Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

thickening of the arachnoid membrane. Postoperatively, there was clinical improvement in 21 patients (70%).

Conclusions The sample majorly had CM and BI associated with syringomyelia. High prevalence of signs and symptoms related to the Brazilian northeastern phenotype was also found. Syringomyelia, therefore, has peculiarities in the population of the Northeast of Brazil that, when described, allow better understanding of the pathology in this group.

Resumo

Introdução Siringomielia é uma enfermidade crônica caracterizada pela presença de cavidade intramedular. Costuma vir associada a condições como malformação de Chiari (MC) e impressão basilar (IB). Sua prevalência tem grande variação geográfica, sendo maior no nordeste brasileiro, o que torna relevantes estudos sobre o tema nessa região.

Objetivo Analisar a frequência de sinais, sintomas e aspectos cirúrgicos observados em pacientes submetidos a tratamento descompressivo.

Método Foi realizada análise retrospectiva das informações contidas nos prontuários de pacientes diagnosticados com siringomielia submetidos a tratamento cirúrgico descompressivo em vários hospitais de João Pessoa, Paraíba, entre 1994 e 2021.

Resultados Foram analisados 30 pacientes, dos quais 29 (96,7%) apresentaram MC associada e 27 (90,0%) também tinham IB. Houve grande variedade de sintomas, com importante prevalência de fraqueza muscular, cervicalgia e cefaleia. Brevicolia, um achado considerado típico do nordestino e associado a malformações da junção craniocervical, esteve presente em 66,7% dos pacientes. A técnica cirúrgica utilizada em 90% dos pacientes foi semelhante. Quatorze (46,7%) pacientes apresentaram junção crânio-cervical difícil; e quatro (13,3%) apresentaram assimilação do osso occipital. Dezoito (60,0%) apresentaram espessamento da membrana aracnoide. No pós-operatório, houve melhora clínica em 21 pacientes (70%).

Conclusão A expressiva maioria da amostra possuía MC e IB associadas à siringomielia. Nota-se também grande prevalência de sinais e sintomas relacionados ao fenótipo nordestino. A siringomielia, portanto, possui peculiaridades na população do Nordeste brasileiro que, ao serem descritas, permitem a melhor compreensão da patologia nesse perfil de pacientes.

Palavras-chave
- malformações da junção craniocervical
- siringomielia
- malformação de Chiari
- impressão basilar
- cirurgia

Introduction

Syringomyelia (SM) can be defined as the presence of a cavity in the spinal cord.[1] Its etiology can be classified as idiopathic when it is not possible to identify a cause; or as secondary to cerebrospinal fluid (CSF) obstruction. Syringomyelia is commonly associated with Chiari malformation (CM) and basilar invagination (BI), but it can also be attributed to inflammation, trauma, tumor, or local compression.[2]

The prevalence of SM varies from 0.9 to 8.4 per 100,000 individuals, according to ethnicity and geographic variation.[3,4] The Brazilian Northeast is considered one of the regions in the world with the highest number of cases of craniocervical junction malformation (CJM).[5,6] Local studies conducted by Silva et al.[6] and Alves et al.[7] relate this fact to the great presence of brachycephaly in the area, which is so easily noted that it became popular knowledge, with people from the Northeast of Brazil, sometimes being referred to, pejoratively, as "flat head" or "short neck."[5] Associated with this craniometric profile, there is a marked prevalence of platybasia and BI.[5] This propensity toward flattening of the skull base and its correlation with SM may explain its high frequency.

Considering that the pathogenesis of symptomatic SM in this particular population located in the state of Paraíba may be different from others, it is important to study the clinical and surgical aspects from this region, to provide personalized treatment and better comprehension of the disease.

Material and Methods

We performed a retrospective analysis of data contained in the medical records of patients diagnosed with SM by magnetic resonance imaging (MRI) undergoing decompressive surgical treatment in several hospitals of João Pessoa, PB, Brazil, between 1994 and 2021. The identification of other

malformations of the CJM was performed as follows: CM was diagnosed using MRI or during the surgical procedure; BI, according to MRI results or other imaging examination, or by measuring the lines of Chamberlain and McGregor. Patients who were lacking the necessary data in their medical records were excluded from the study.

The research was based on secondary data, ensuring privacy, confidentiality, and the non-use of information to the detriment of the subject, in accordance with Resolution 580/18, of the National Health Council, which establishes the ethical principles of research in human beings. The study was approved by the ethics committee of Hospital Universitário Lauro Wanderley.

Results

For this study, 30 patients (15 women, 15 men) with SM were analyzed, 29 (96.7%) of whom had associated CM and 27 (90.0%) also had BI. Clinically, 6 (20.0%) had syringomyelic pain. The average age was 38.2 years, ranging from 8 to 72 years old.

The period between the onset of symptoms and surgery was over 3 years in 14 patients (46.7%); from 2 to 3 years in 2 of them (6.7%); from 1 to 2 years in 6 (20%); and less than 1 year in 8 (26.7%).

Table 1 Clinical symptoms observed in 30 cases of syringomyelia

Symptoms	Cases number	%
Neck pain	20	66.7
Muscular weakness	20	66.7
Numbness of limbs	20	66.7
Headache	16	53.3
Rhinolalia	15	50.0
Dysphagia	12	40.0
Sexual potency disturbances	11	36.7
Stiffness of neck	10	33.3
Vertigo	10	33.3
Nasal reflux	6	20.0
Hearing loss	5	16.7
Apnea	5	16.7
Libido disturbances	4	13.3
Constipation	4	13.3
Menstrual disorders	4	13.3
Vicious position of the head	3	10.0
Dysarthria	3	10.0
Numbness of face	2	6.7
Ataxic gait	2	6.7
Decreased visual acuity	1	3.3
Drop attack	1	3.3
Difficult urination	1	3.3
Anhidrosis	1	3.3

Table 2 Clinical findings in 30 cases of syringomyelia

Findings	Number of cases	%
Brevicollis	20	66.7
Brachycephaly	19	63.3
Low hair implantation	12	40.0
Alteration of the posterior fossa	9	30.0
Hypertelorism	8	26.7
Painful limitation of the neck	7	23.3
Scoliosis	6	20.0
Prognathism	5	16.7
Craniofacial asymmetry	4	13.3
Accentuated lumbar lordosis	1	3.3
Equinovaro feet	1	3.3

The symptoms observed in the preoperative period are shown in ►**Table 1**; the clinical findings are in ►**Table 2**; and the signs found on physical examination in ►**Table 3**.

Regarding the surgical treatment of SM, 27 (90%) of the patients were operated in the concord position (prone position with the head flexed forward), and the other 3 (10%) in a sitting position. All were submitted to bone, dural, and neural decompression, characterized by posterior fossa craniectomy and high cervical laminectomy (C1 and C2) according to the degree of tonsil herniation in the CM (►**Table 4**). In the 29 cases (96.7%) in which CM was present, Y-shaped opening of the 4th ventricle dura and subpial aspiration of the cerebellar tonsils was performed; in many cases, a small membrane that occluded the foramen of Magendie was found. After opening, the tonsils were fixed laterally, and duraplasty with a synthetic dural substitute was performed (100% of cases); thus, the cisterna magna was remodeled, allowing a more physiological circulation of the cerebrospinal fluid (►**Fig. 1**).

About half of the sample—14 patients (46.7%)—had a difficult craniocervical junction, and 4 (13.3%) had occipital bone assimilation. Eighteen (60.0%) presented thickening of the arachnoid membrane.

With surgical treatment, there was clinical improvement in 21 patients (70%) and 9 (30%) did not suffer changes; there was no worsening in any of the cases studied.

Discussion

Rodrigues et al.[8] stated that SM is associated with CM (or other anomalies that occlude the foramen of Magendie) in ¾ of the cases.[8] In the present study, CM was present in 96.7% of the cases, which reinforces CM and other CJM as major factors for the development of SM in our population.

When characterizing our sample, sex parity was perceived, similar to some previous analyses, but different from others in which there is a predominance in females.[9]

Table 3 Clinical signs observed in 30 cases of syringomyelia

Signs	Number of cases	%
Hyperreflexia	28	93.3
Syringomyelic dissociation	24	80.0
Hypoesthesia	23	76.7
Hoffmann sign	17	56.7
Abolition of gag reflex	16	53.3
Abolition of palatal reflex	15	50.0
Nystagmus	14	46.7
Hypertonia	14	46.7
Paresis	14	46.7
Clonus	12	40.0
Babinski sign	9	30.0
Rossolimo sign	9	30.0
Paresis of soft palate	8	26.7
Exophthalmos	6	20.0
Hypoacusis	6	20.0
Atrophies	6	20.0
Abolition of abdominal reflexes	4	13.3
Lesion of the V nerve	3	10.0
Vestibular disturbances	3	10.0
Hypertelorism	2	6.7
Lesion of the spinal nerve	2	6.7
Lesion of the XII nerve	2	6.7
Cerebellar disturbances	2	6.7
Unsteady gait	2	6.7
Fasciculations	2	6.7
Lesion of the VII nerve	1	3.3

Table 4 Surgical findings in 30 patients with syringomyelia

Findings	Number of cases	%
Difficult atlanto-occipital joint	14	46.7
Thinning of the occipital bone	12	40.0
Thickening of the occipital bone	17	56.7
Dural adhesion	11	36.7
Arachnoid thickening	18	60.0
Degree of cerebellar tonsillar herniation		
0	1	3.3
C1	15	50.0
C2	14	46.7
C3	0	0.0
Tonsillar symmetry		
Symmetrical	13	43.3
Right > Left	3	10.0
Left > Right	13	43.3

Regarding age, the population analyzed was similar to or older in relation to others.[9–11] Regarding the period between the onset of symptoms and surgical approach, there was a great variety in our sample, as well as among previous studies.[9,10]

With regard to the clinical aspects, as in other studies with SM, a wide variety of symptoms could be noted among patients, with higher frequency of complaints of muscle weakness, pain, and headache.[9,10] Paresthesia, neck pain and cranial nerve involvement were also observed in a proportion similar to the one described in the literature.[10] Rare findings in our sample have also been observed in some patients in other studies, such as ataxic gait and vestibular disorders.[9]

Some findings, however, were markedly more significant in our population. Brevicollis, for example—which is a finding considered typical of the Brazilian Northeast and associated with CJM—, was present in 66.7% of our patients and only in 22% of patients by Mariani et al. (1991). In the same study, a prevalence of 74% of CM and 28% of BI was reported among cases of SM.[10] Bogdanov et al. (2002) reports presence of BI in 46% of patients with SM.[3] In our sample, however, 96.7% had CM, and 90% BI.

Alzate et al.[9] demonstrated statistically that, among patients with type I CM, the presence of scoliosis is a risk factor for the development of SM. In our sample, the prevalence of scoliosis was 20%, which is similar to or smaller than observed in previous studies.[3,9–11]

One of the most widely reported symptoms among authors is headache, which also had a high frequency in our sample. Headache associated with SM tends to respond very well to decompressive treatment.[9] This characteristic can have an important influence on the rate of clinical improvement obtained with surgery.

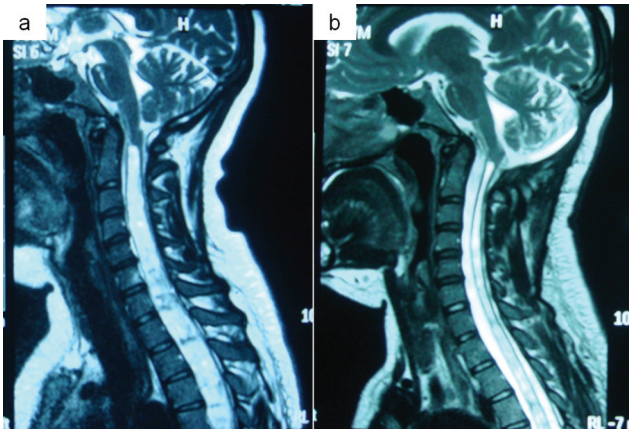

Fig. 1 (a) Preoperative magnetic resonance imaging showing extensive syringomyelia compressing the spinal cord. (b) Postoperative magnetic resonance imaging showing decompression of the spinal cord after the physiological return of the cerebrospinal fluid.

Generally, considering signs and symptoms of patients with SM, there is usually significant improvement in 50 to 90% of the cases with surgical treatment according to most studies.[9,12] In our sample, 70% of all patients showed improvement, and there was no worsening in any of the cases analyzed.

Considering the surgical outcomes, the study of Silva et al.[6] with 260 operated cases, in a population similar to ours, is certainly an important source of comparison for the obtained results. In it, 29.6% of patients had thin and 15% thickened occipital bone; similar to the results of Gonçalves et al. (2003), however, different from ours (40% and 56.7%, respectively).[6,12] Such variations are usually associated with greater technical difficulty in performing the craniectomy.[6]

Thickening of the arachnoid membrane was found in 60% of patients; similar to that observed by Silva et al. (1994), with 67.3%, and Gonçalves da Silva et al.,[12] with 67.9%.[6,12] In case series, this change was associated with an increased surgical risk.[10] It is also considered the most important factor for the development of neurological symptoms.[6] Thus, it is a frequent finding and has a great impact on the clinical picture of CJM. As for the topography of cerebellar tonsils, in our patients, there was symmetry in 43.3% of cases. This proportion approaches the one found in the literature.[6,13]

In our sample, there was clinical improvement in 70% of the operated patients, and outcome that was similar to the one found by Mariani et al.[10]

During the operative act, since the 1970s, duraplasty has been performed to increase the anatomical space of the posterior fossa, prevent cerebrospinal fluid leak, reduce arachnoiditis by preventing the penetration of blood in the manipulated region, recompose the pachymeninge, and protect the posterior fossa structures.[6,11,12] In the experience of Silva et al. (1994), duraplasty reduced the number of postoperative complications, especially mortality and the incidence of respiratory disorders.[6] Munshi et al. (2000) demonstrated that decompression of the posterior fossa with C1 laminectomy and dural graft caused hydromyelia to regress more quickly than with the same procedure without the use of dural graft.[11,12] In our sample, all patients treated with the decompressive procedure underwent duraplasty with a synthetic dural substitute. Another aspect of the surgical approach adopted in this sample was the aspiration of the herniated cerebellar tonsils. This procedure also had promising results presented by Silva et al.[6]

Note that our findings were similar to those of local studies but differed significantly from those of international studies. This can be related to the ethnic and geographic differences in SM incidence, which are associated with genetic and environmental differences.[3,4]

The average tonsillar position in asymptomatic individuals is another difference found between populations. Studies in normal patients have shown that the tonsillar position is usually higher in the Japanese population when compared with Euro-Americans: in these, more than 5% have tonsils between 1 and 5 mm below the FM, while in Japan, this prevalence is of only 0.24%.[13] This characteristic is especially relevant when it is pointed out that Japanese people are affected less frequently than Europeans with CM-related obstruction in the CSF pathways in FM, which explains the lower prevalence of SM in Japan (1.9 per 100,000) than in England or New Zealand (8.4 and 8.2 per 100,000, respectively).[13]

Among the regions identified globally as CM and SM clusters, there is the Brazilian Northeast, as already mentioned, some states in India, an area of Germany, and regions of Russia with a large proportion of Tatar.[13]

When studying the Tatar population, Bogdanov et al.[13] considered recommendation for surgery based on less objective criteria, as in groups with less than 5 mm descent of the tonsils, absence of SM, and predominantly subjective symptoms; but stressed that it is inappropriate to apply these criteria to populations in which the spread of CJM is not considered high.[13]

Furthermore, some studies suggest that cultural aspects present greater impact on the development of CJM than genetic and environmental. In the sample studied by Bogdanov et al.,[13] for example, 88% of symptomatic CM patients were male manual workers, who have done agricultural work all their lives. From this, the authors indicated that an occupation associated with physical tension imitating the Valsalva maneuver can lead to the development of SM in those who already had a posterior fossa underdeveloped.[13] Another fact that suggests that CM may be acquired, not congenital, is that the abnormal shape and position of the cerebellar tonsils disappears after simple extra-arachnoid decompressive surgery.[14]

Conclusions

There is great prevalence of signs and symptoms related do the Brazilian northeastern phenotype in patients with SM. This shows that regional determinants have an important role in the presentation of SM, thus requiring changes in treatment and prognosis.

Because it is usually associated with CM, in an even more expressive way in individuals from the Northeast of Brazil, surgical treatment of SM, in most cases, consists of suboccipital craniectomy, C1 laminectomy, and duraplasty.

Therefore, there are peculiarities in the Northeast of Brazil that, once described, allow for better understanding of SM.

Ethics Standards

This research was based on secondary data, ensuring privacy, confidentiality, and the non-use of information to the detriment of the subject, in accordance with Resolution 580/18 of the National Health Council, which establishes the ethical principles of research in human beings. The study was approved by the ethics committee of Hospital Universitário Lauro Wanderley.

Conflict of Interests

The authors have no conflict of interests to declare.

Acknowledgments

The authors would like to thank the research group on the study of craniocervical junction malformations.

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