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SYRINGOMYELIA

Okan TÜRK[1]
Şahin YÜCELİ[2]

[1]*İstanbul Training and Research Hospital, Department of Neurosurgery, İstanbul*
[2]*Neon Hospital, Neurosurgery Clinic, Erzincan*

Orcid Numbers:
Okan TÜRK:
0000-0002-0074-2835
Şahin YÜCELİ:
0000-0002-9471-3575

Corresponding author: Okan TÜRK
Address: İstanbul Eğitim ve Araştırma Hastanesi, Fatih-İstanbul
E-mail: drokanturk@gmail.com
Phone: +90 532 781 49 61
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ABSTRACT

Objective: The cavities imbued with glia cells which may develop around the central canal of spinal cord are called syringomyelia. The pathophysiology of syringomyelia is yet to be defined in full. The aim of our study is to investigate the patients with syringomyelia.

Material and Method: The patients who applied to our clinics between 2007-2017 and were diagnosed with syringomyelia in consequence of neurological and radiological examinations were included in the assessment from the automation system of our hospital.

***Results*:** This study was carried out on 208 cases, in total, of which 35.1 % (n=73) were male while 54.9 % (n=135) were female. The ages of the cases included in this study varied between 9 and 81, and the average of ages was 42.52 ± 16.30 years. Chiari malformation were the most frequent etiology among these (28.4 %). The rate of cervical engagement of Chiari Malformation and Disc Pathology were significantly higher compared to the rates of the lumbar engagement of spinal congenital anomaly and the thoracic engagement of trauma.

***Conclusion*:** In consequence of this study, only 43.5 % of the patients with syringomyelia demonstrated etiological causes. On the other hand, a small part of the cases arose due to spinal mass, trauma and discopathy.

Key words: Syringomyelia, spinal cord, syrinx

Level of Evidence: Retrospective clinical study, Level III.

INTRODUCTION

Syringomyelia (SM) emerges due to the deteriorations in the cycle of cerebrospinal fluid (CSF), and it is widely believed in the recent literature even though the pathophysiology cannot be explained to the fullest extent that it follows the accumulation of extracellular fluids in spinal cord due to the difference of intracerebral pressure and the pressure of surrounding CSF [17].

While the etiology cannot be elaborated completely, the causes can be classified under three main titles which are congenital causes, acquired diseases and the deterioration of the flow dynamics of cerebrospinal fluid [6,17].

This study aims to contribute to the literature by assessing the epidemiological characteristics of 208 patients who were diagnosed with syringomyelia.

MATERIAL AND METHOD

The patients who applied to our clinics between 2007-2017 and were diagnosed with syringomyelia in consequence of neurological and radiological examinations were included in the assessment from the automation system of our hospital. Retrospectively, the spinal magnetic resonance images (MRIs) of the patients which were taken of cervical, thoracic, lumbar and more than one anatomical regions of spinal columns were analyzed. The patients included in this study were evaluated according to their age, sex, anatomical region, etiological cause and dimension of syringomyelia.

Statistical Analysis

NCSS (Number Cruncher Statistical System) 2007 (Kaysville, Utah, USA) program was used for the statistical analyses. Descriptive statistical methods (average, standard deviation, median, first quartile, third quartile, minimum and maximum) were used while evaluating the data of the study. The compatibility of quantitative data with normal distribution was tested with Shapiro-Wilk Test and graphical analyses. Kruskal-Wallis and Dunn-Bunferroni tests were availed of in the comparison of more than two groups in which quantitative variables did not show normal distribution. On the other hand, Fisher-Freeman-Halton Exact Test was used for the comparison of qualitative data. Statistical significance was accepted to be $p<0.05$.

RESULTS

This study was carried out on 208 cases, in total, of which 35.1 % (n=73) were male while 54.9 % (n=135) were female patients who had applied to Istanbul Training and Research Hospital between 2007 and 2017. The ages of the cases included in this study varied between 9 and 81, and the average of ages was 42.52 ± 16.30 years (Table-1, Figure-1).

Table-1. The Distribution of Demographic Characteristics

Age (years)	*Min-Max (Median)*	9 – 81 (44)
	Ave±Sd	45,52±16,30
Gender n(%)	**Male**	73 (35,1)
	Female	135 (64,9)

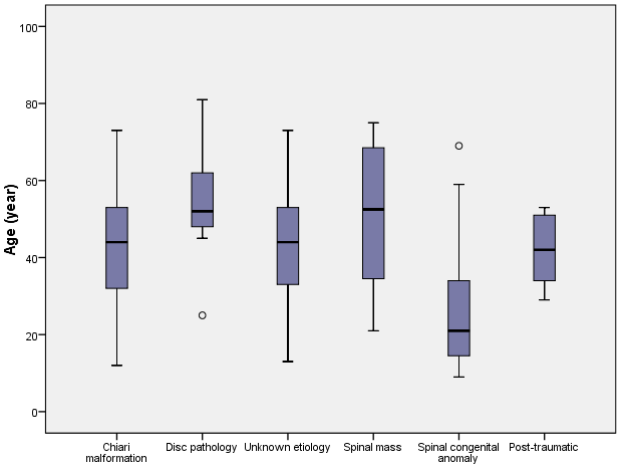

Figure-1. The Distribution of Ages According to Causes

Syringomyelia was found to be in cervical, thoracic, lumbar and more than one region of 61.5 % (n=128), 61.5 % (n=49), 8.2 % (n=17) and 6.7 % (n=14) of the patients, respectively. 51.4 % (n=107) cases showed milimetric syrinx while the other 48.6 % (n=101) demonstrated other dimensions (Table-2).

Table-2. Evaluation of Ages According to Causes

Cause	N	Age (years)			
		Average	SD	Median	Min-Min
Chiari Malformation	59	42.54	14.319	44.00	12-73
Disc Pathology	15	55.00	13.480	52.00	25-81
Unknown Etiology	97	43.69	14.923	44.00	13-73
Spinal Mass	8	50.88	20.420	52.50	21-75
Spinal Congenital Anomaly	23	26.70	17.844	21.00	9-69
Post-traumatic	6	41.83	9.847	42.00	29-53
Total	**208**	**42.52**	**16.302**	**44.00**	**9-81**

Kruskal Wallis test; p=0,001; p<0,01

The causes of syringomyelia were found to be Chiari malformation, disc pathology, unclear etiology, spinal mass, spinal congenital anomaly and post-traumatic in 28.4 % (n=59), 7.2 % (n=15), 46.5 (n=97), 3.8% (n=8), 11.1% (n=23) and 2.9% (n=6) of the patients, respectively (Table-3).

Table-3. Evaluation of Ages According to Regions

Regions	N	Age(year)			
		Average	SD	Median	Min-Min
Cervical	128	44,78	14,158	46,00	9-81
Thoracic	49	38,31	18,260	34,00	9-75
Lumbar	17	36,35	21,374	27,00	9-69
More than one region	14	44,14	17,615	48,50	12-67

Kruskal Wallis test; p=0,039; p<0,05

There was a statistically significant difference in terms of the distribution of ages according to the causes (p<0,01). In consequence of the Dunn-Bonferroni tests carried out in order to distinguish from which group the significance arose, the ages of spinal congenital anomaly cases were found to be significantly lower than those of Chiari Malformation, Disc Pathology, Unclear Etiology and Spinal Mass cases (p=0,006; p=0,001; p=0,001; p=0,020, respectively). No statistical significance was found among the distribution of the ages of other groups (p>0,05).

There was a statistically significant difference in terms of the distribution of ages according to the regions (p<0,01). In consequence of the Dunn-Bonferroni tests carried out in order to distinguish from which group the significance arose, it was inferred that the ages of cervical cases were significantly higher compared to the thoracic cases (p=0,041). No statistical significance was found among the distribution of the ages of other regions (p>0,05)(Figure-2).

There was a statistically significant difference in terms of the dimensions according to the causes of syringomyelia (p<0,01). The investigation of the group that caused significance showed that the determination rates of spinal congenital anomaly and unclear etiology cases were significantly higher than the others (Table-4).

There was a statistically significant difference in terms of the regions according to the causes of syringomyelia (p<0,01). Similarly, the analysis of the group that led to significance indicated that the rate of cervical engagement of Chiari Malformation and Disc Pathology were significantly higher compared to the rates of the lumbar engagement of spinal congenital anomaly and the thoracic engagement of trauma.

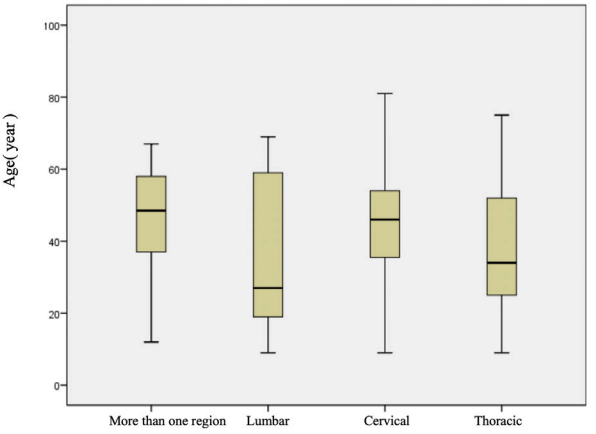

Figure-2. Distribution of ages according to regions

Table-4. Evaluation of dimensions and regions according to syringomyelia causes

	Chiari Malformation	Disc Pathology	Unclear etiology	Spinal Mass	Spinal congenital anomaly	Post-traumatic	p
Dimension							
Other	36(61,0)	10(66,7)	41(42,3)	6(75,0)	5(21,7)	3(50,0)	***0,005***
Millimetric	23(39,0)	5(33,3)	56(57,7)	2(25,0)	18(78,3)	3(50,0)	
Regions							
Cervical	48(81,4)	14(93,3)	59(60,8)	4(50,0)	2(8,7)	1(16,7)	
Thoracic	7(11,9)	1(6,7)	27(27,8)	4(50,0)	6(26,1)	4(66,7)	***0,001***
Lumbar	0(0)	0(0)	4(4,1)	0(0)	13(56,5)	0(0)	
More than one region	4(6,8)	0(0)	7(7,2)	0(0)	2(8,7)	1(16,7)	

Fisher's Freman Halton test **p<0,01

DISCUSSION

Syringomyelia is generally characterized with the loss of pain heat sensation and the conservation of the sense of touch due to the exposure of surrounding spinothalamic tracts after the expansion of central canal. Pyramidal findings, weakness in extremities and muscle atrophies can also be seen. It appears with weakness and muscle atrophy in the lower extremities, neck, upper back and all other extremities. Most of syringomyelia cases are also associated with severe dysesthetic pain in neck, shoulders and back [2].

There was a distinct increase in the incidence of SM upon the emergence of MRI. Because, small sub-clinic lesions can be diagnosed with MRIs which were undiagnosed previously [14,17].

Though there have been numerous studies that have been offered until today, etiologic causes cannot be determined in most syringomyelia patients. The patients with an acquired disease or congenital anomaly are known to have the most frequent etiologic causes [6,13]. In consequence of this study, only 43.5 % of the patients with syringomyelia demonstrated etiological causes. Chiari malformation were the most frequent factor among these (28.4 %). On the other hand, the remaining part of the cases were associated with spinal mass, trauma and discopathy.

However, there has not been a consensus on the surgical treatment or follow-up of syringomyelia patients with unknown etiology. The exact pathogenesis and development of syringomyelia are unknown, and the natural course of it is variable [9,10,16,18]. Furthermore, there is little information on syringomyelia since there are very few studies on syringomyelia with unknown etiology. It does not have any kind of predisposing pathology like chiari malformation or spinal cord tumor. In this study, syringomyelia with unclear etiological cause corresponds to 46.5 % of all the syringomyelia cases. Further examinations that were carried out did not produce etiological factors (Figure-3).

Many studies revealed that there were cases of syringomyelia that developed as secondary to chiari malformation. Chiari malformation was considered to be responsible for almost half of secondary syringomyelia cases [5]. In this study, 28.4 % of the cases showed syringomyelia secondary to chiari malformation. Besides, in most cases, it was observed to have statistically significantly higher possibility to occur in cervical region compared to the other regions (Figure-4).

The exact pathophysiology of post-traumatic syringomyelia is now known clearly. However; mechanical spinal cord compression, spinal cord inflammation, hematoma, secretion of intracellular lysosomal enzymes, ischemia and arterial and venous occlusions are among the factors that are believed to occur prior to the initial formation of the cavity in spinal cord [1]. All of such factors are considered to contribute to the occurrence of syringomyelia [7,12]. In this study, it was observed that post-traumatic syringomyelia made up 2,9% of the cases and that these were most frequently found in thoracic region. Additionally, women were found to have post-traumatic syringomyelia twice as much compared to men. Certain studies indicated that advanced age (with spinal cord damage), the severity of trauma and the cervical localization of such damage have a high potential to develop syringomyelia within five years [11-12]. This study had quite a low number of post-traumatic syringomyelia cases and, particularly, they were generally observed in thoracic region.

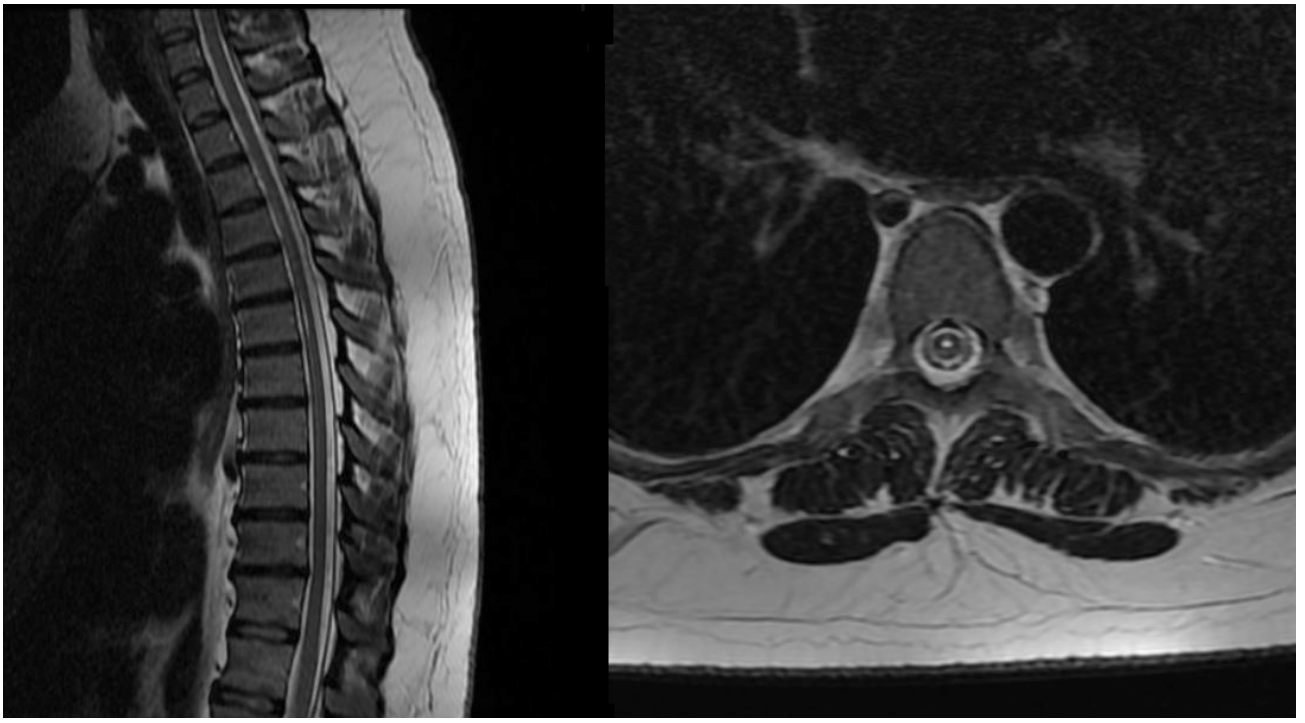

Figure-3. 50 years old female,T3-6 syringomyelia patients with unknown etiology

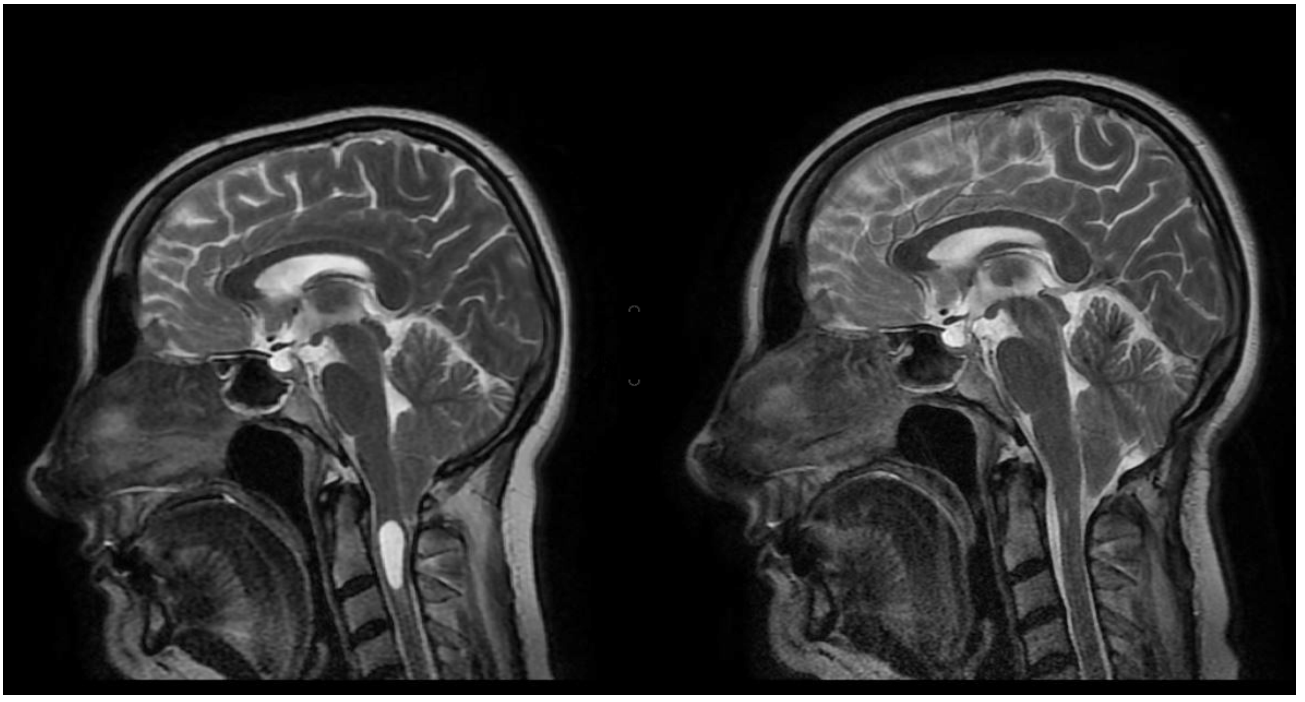

Figure-4. 50 years old female patient preoperative and postoperative chiari malformation

The study carried out by Ramnarayan et al. asserts and provides examples from the literature that childhood syringomyelias can regress without surgical treatment [15]. In this study, the dimensions of shrinks in spinal congenital anomaly and unclear etiology cases were smaller, and no statistically significant difference was found (p<0,01). The fact that milimetric syringomyelia was seen with spinal congenital anomaly in this study and that it was frequently observed in cases with unknown etiology leads to the consideration that this may be the continuation of regressed childhood syringomyelia.

The literature reports syringomyelia due to cervical spondylosis [3-4]. In this study, the rate of syringomyelia due to discopathy was 7.2 %, and 14 of these were in the cervical region while 1 was seen in thoracic region.

Spinal congenital anomalies are observed in lumbar region more compared to other regions. In this study, syringomyelia as secondary to spinal congenital anomaly was also found in lumbar region statistically significantly higher than the other regions (p<0,01).

The treatment of syringomyelia requires, first of all, research on etiology. Because, surgical plan in case of secondary syringomyelia is basically made in consideration of etiology. During follow-up period, shunting surgery may also be planned if there is no resorption [8].

Conclusion

Though there have been numerous studies that have been offered until today, etiologic causes cannot be determined in most syringomyelia patients. The patients with an acquired disease or congenital anomaly are known to have the most frequent etiologic causes [6,13]. In this study, only 43.5 % of the syringomyelia patients produced etiological causes. The most frequent etiological cause was chiari malformation (28.4 %) compared to the others. On the other hand, a small part of the cases arose due to spinal mass, trauma and discopathy.

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