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Original Article

Chiari malformation and types of basilar invagination with/without syringomyelia

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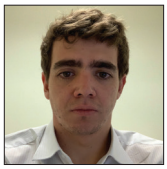

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ABSTRACT

Background: Craniometric studies document different subtypes of craniocervical junction malformations (CCJM). Here, we identified the different types and global signs and symptoms (SS) that correlated with these malformations while further evaluating the impact of syringomyelia.

Methods: Prospective data concerning SS and types of CCJM were evaluated in 89 patients between September 2002 and April 2014 using Bindal's scale.

Results: The mean Bindal's scores of each type of CCJM were Chiari malformation (CM) = 74.6, basilar invagination Type 1 (BI1) = 78.5, and BI Type 2 (BI2) = 78. Swallowing impairment and nystagmus were more frequently present in the BI patients. Symptomatic burdens were higher in patients with syringomyelia and included weakness, extremity numbness, neck pain, dissociated sensory loss, and atrophy.

Conclusion: There were no statistically significant differences in SS between the different CCJM types. BI patients had more swallowing and nystagmus complaints versus CM patients, but there were no significant differences in clinical SS between BI1 and BI2 patients. Notably, those with attendant syringomyelia had a higher SS burden.

Keywords: Arnold–Chiari malformation, Basilar impression, Neurologic manifestations, Platybasia, Signs and symptoms

INTRODUCTION

Chiari malformations (CMs) and basilar invagination (BI – BI1 and BI2) are the most common craniocervical junction malformations (CCJM) seen in adults; both may also be associated with syringomyelia.[1,3,4,6,7]

BI1 is associated with craniocervical instability, odontoid insinuation toward the foramen magnum, and often anterior arch of the atlas assimilation.[2-4] BI Type 2 (BI2) is not associated with instability, but there is an exacerbation of the clivus canal angle and craniocervical kyphosis resulting in ventral compression of the brainstem and cranial nerves.[2-4] Syringomyelia is frequently encountered in all of these three types of CCJM and can be correlated with a variety of increased signs and symptoms (SS).[1] Here, we analyzed the different frequencies of SS, between these three CCJM subtypes with/without syringomyelia.

METHODS

Study design

This study represented a cross-sectional evaluation of prospectively collected data conducted between September 2002 and April 2014 using Bindal's scale. All patients filled out informed consent to enter in this study. The research protocol was approved by the research ethics committee (CAAE 32361614.6.0000.5551).

There were 89 consecutive symptomatic patients with CCJM; 46 (52%) had CM, 43 (48%) had BI; among those with BI: 13 (30%) had BI1, and 30 (70%) had BI2. Females represented 57% of the patients with CCJM (51 patients), and they averaged 47.7 years of age (range: 11–77 years). All exhibited varying degrees of SS attributed to neural structure/posterior fossa compression and/or CSF blockage. Syringomyelia was present in 44 (49%) of all CCJM (CM and BI) patients.

Definitions and diagnostic criteria of CM, B11, and B12 malformations [Figure 1]

CM

Herniation (variable) of the cerebellar tonsils below the foramen magnum results in compression of the tonsils in the cisterna magna without odontoid insinuation toward the skull base.[3,4]

BI

BI1: Odontoid insinuation toward the foramen magnum, violating the McRae line, associated with the anterior arch of the atlas assimilation, and craniocervical instability.[2-4,7]

BI2: Odontoid axis process above the Chamberlain line, without violation of the foramen magnum.[2-4,7]

Syringomyelia

Syringomyelia constitutes a fluid accumulation around the spinal cord central canal, identified as intramedullary hypersignals on T2-weighted magnetic resonance imaging (MRI).[1,4]

Assessment of SS of CCJM

Bindal's standardized scale longitudinally evaluated CCJM patients' clinical manifestations and provided the following score for follow-up comparisons; each sign or symptom received a score of 10, and the score (symptomatic burden) was defined as the sum of the SS presented by the patients [Table 1].[1]

Statistical evaluation

Bindal's scores for the three types of malformations were compared using ANOVA (the Kruskal–Wallis test).

SS were described by percentages and were compared between types using the Chi-squared test; this was similarly applied to patients with/without syringomyelia [Graphs 1-5 and Table 2].

Table 1: Bindal's scale.

	Brainstem compression	Syringomyelia
Vertigo		
Diplopia		
Hoarseness		
Swallowing difficulty		
Tinnitus		
Persistent cough		
Hearing loss		
Nystagmus		
Sleep apnea		
Ataxia		
Shoulder pain		
Weakness (sign)		
Weakness (symptoms)		
Spasticity		
Extremity numbness		
Neck pain		
Headache		
Atrophy		
Dissociated sensory loss		
Scoliosis		
Extremity pain		

Based on: Bindal AK, Dunsker SB; Tew JM. (1995) Chiari I Malformation: Classification and Management. Neurosurgery

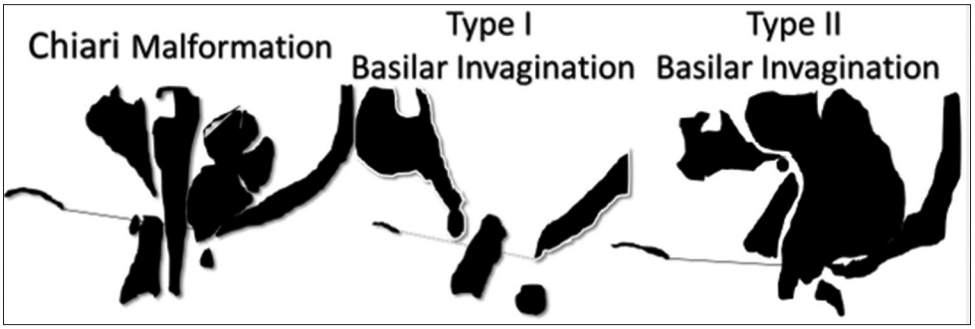
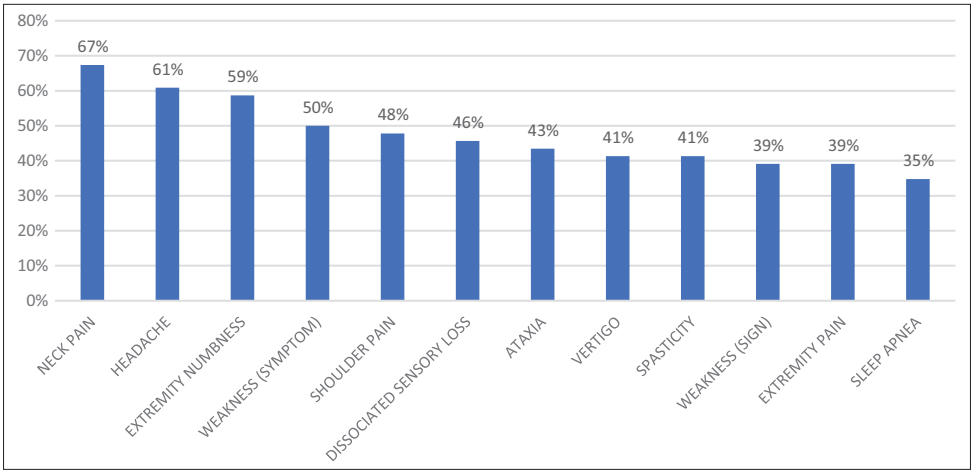
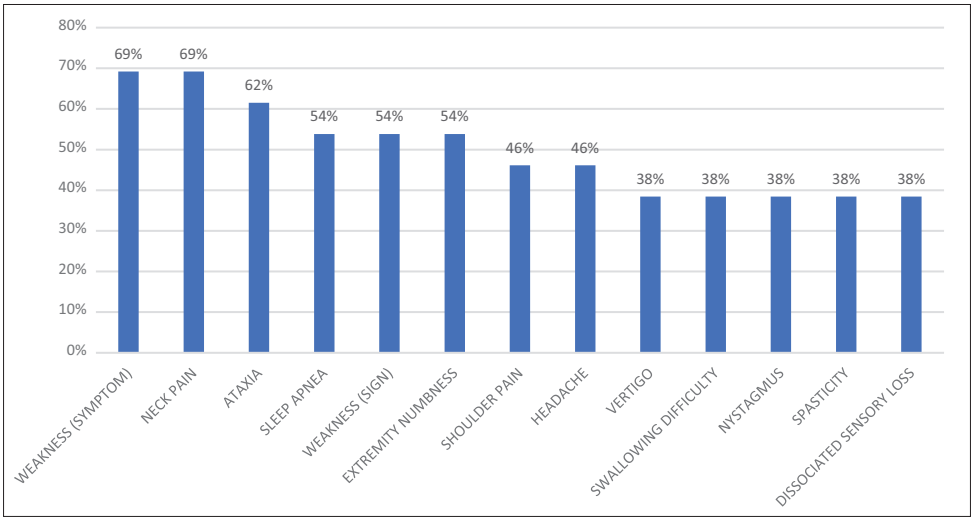


Figure 1: Craniocervical junction malformations.



Graph 1: Prevalence of Chiari malformation signs and symptoms.



Graph 2: Prevalence of basilar invagination Type 1 signs and symptoms.

RESULTS

Prevalent SS among three CCJM malformations

Predominant SS for patients with these three malformations (CCJM, B11, and B12) included neck pain, ataxia, numbness, and weakness of the extremities (e.g., with added headaches for CM patients and sleep apnea for BI patients). For CM patients, SS in descending order included neck pain, headache, and numbness of extremities followed by weakness [Graph 1]. For patients with B11, SS in descending order included weakness and neck pain, ataxia, sleep apnea, and extremity numbness [Graph 2]. For BI2 patients, SS included (descending order) weakness, ataxia, nystagmus, sleep apnea, extremity numbness, neck pain, and headache [Graph 3].

The mean Bindal's score for each type of CCJM was 74.6 for CM, 78.5 for BI1, and 78 for BI2 (Kruskal–Wallis test, $P = 0.9$) [Graph 4]. There were significant differences in SS, swallowing difficulty ($P = 0.03$) and nystagmus ($P = 0.007$) for the three groups [Table 2].

SS for patients with/without syringomyelia

The mean Bindal's score was 69.3 for the patients without syringomyelia and 83.4 for the syringomyelic patients (Mann–Whitney U-test; $P = 0.054$) [Graph 5]. Weakness (SS), extremity numbness, neck pain, dissociated sensory loss, and atrophy were significantly more frequent ($P < 0.05$) in the patients with syringomyelia.

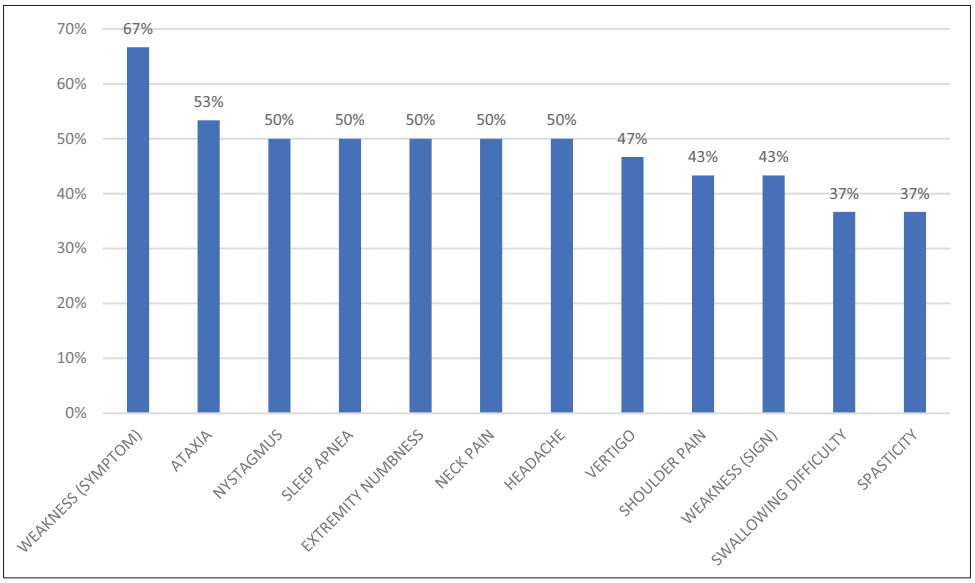
DISCUSSION

Our understanding of CCJM (e.g., CM, B11, and B12) has evolved with the introduction of computed tomography, MRI, linear and angular craniometric studies, and genetic

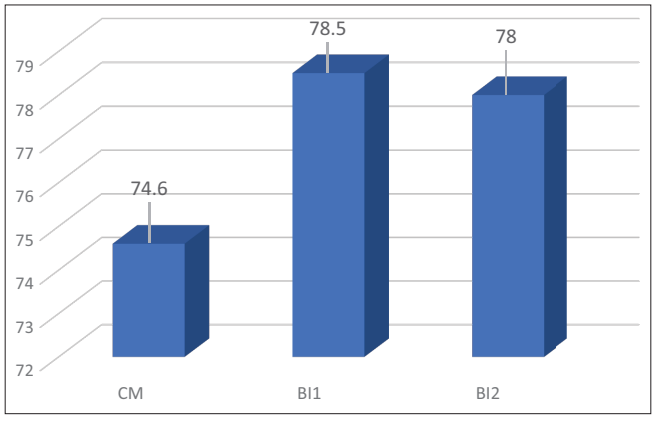
Table 2: Prevalence of signs and symptoms of craniocervical junction malformations as measured using the Chi-squared test.

	Chiari malformation (%)	IB1 (%)	IB2 (%)	$X^2(p)^a$
Vertigo	41	38	47	0.85
Swallowing difficulty	13	38	37	0.03
Nystagmus	20	38	50	0.007
Sleep apnea	35	54	50	0.29
Ataxia	43	62	53	0.45
Weakness (sign)	39	54	43	0.64
Spasticity	41	46	37	0.92
Extremity numbness	59	54	50	0.75
Neck pain	67	69	50	0.26
Headache	61	46	50	0.51
Dissociated sensory loss	46	38	23	0.14
Extremity pain	39	23	30	0.48

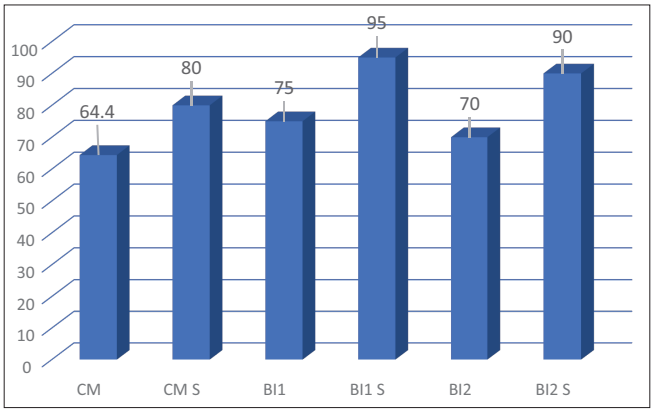
[a]Chi-square test



Graph 3: Prevalence of basilar invagination Type 2 signs and symptoms.



Graph 4: The mean Bindal's score for each type of craniocervical junction malformations.



Graph 5: The mean Bindal's score for each craniocervical junction malformation type with and without syringomyelia.

and embryological studies.[2-4] Notably, SS for each of these three entities differ. Although the current study found no statistically significant differences in the incidence of sleep apnea between the CCJM types, others indicated that sleep apnea was more frequent and intense in BI patients.[5] In 43 patients with atlantoaxial dislocation and CCJM, Shuhui *et al*. found the following SS to be prevalent (in descending order): weakness (93%), cervical motor limitations (88%), limb paresthesia (98%), neck and shoulder pain (58%), ataxia (42%), vertigo (23%), and respiratory difficulties (7%).[8]

Goel showed that the following symptoms were more predominant in BI1 patients: paresthesias (55%), spinothalamic dysfunction (36%), cervicalgia (77%), and torticollis (41%). The study also reported that BI2 physiopathology was the same as for CM, largely attributable to the diminished cranial fossa volume contributing to SS.[4] Our results showed that the only SS differences in the three subtypes of CCJM were swallowing difficulties ($P = 0.03$) and nystagmus ($P = 0.007$) in the BI patients.

Bindals' scale

Bindal's scale was created to quantify SS, and in this study, no differences were found in the SS burden between the three types of CCJM, but SS burdens were uniformly increased in all patients with accompanying syringomyelia.[1,4]

CONCLUSION

For the three types of CCJM, swallowing difficulties and nystagmus were more prevalent in BI versus CM, while all CCJM patients with syringomyelia had higher frequencies of weakness, extremity numbness, neck pain, dissociated sensory loss, and atrophy.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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