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Original Article

Radiology Section

Role of MRI in Selection of Patients for Surgery and Assessing the Post Operative Outcome in Chiari 1 Malformation

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ABSTRACT

Introduction: Chiari malformation also known as Arnold-Chairi malformation is a malformation of the skull. They are of four types among them Chiari 1 malformation is most common. It is characterised by inferior herniation of cerebellar tonsil through foramen magnum for more than 5mm and leads to compression of cervicomedullary junction and interruption of normal flow of CSF.

Aim: To identify patients with Arnold-Chiari malformation type 1 who are ideal candidates for early surgical intervention.

Materials and Methods: In this prospective study, 20 patients with age ranging from 15 to 47 years with Arnold-Chiari type 1 malformation were included. All the patients were evaluated with MRI brain and spine. Length of tonsillar herniation from foramen magnum, presence or absence of syrinx and maximum diameter of syrinx were assessed. Post operative outcome were evaluated by serial six months follow-up for resolution, reduction and persistence of symptoms.

Results: The mean age of patients with Chiari 1 malformation was 28 years. Chiari 1 malformation was common in females in our study. The mean duration of symptoms was 5.2 years. Ten patients had associated syrinx. Preoperative MRI showed the length of herniation between 6 to 10mm in seven patients and more than 10mm in 13 patients. In patients with syrinx mean maximum diameter of syrinx was 7.8 mm. Evaluation of postoperative patients for clinical recovery were done by serial follow-up at 1st, 3rd and 6th months. After surgery 9 patients with Chiari 1 malformation without syrinx had complete recovery. One patient without syrinx had partial recovery. Three patients with syrinx had complete recovery. Four patients with syrinx had partial recovery and three patients with syrinx had persistence of symptoms.

Conclusion: MRI has important role in diagnosing the Chiari 1 malformation and its association. Presence of syringomyelia and large diameter of syrinx implies a less favorable response to surgery. Patients without syrinx have better results with decompression. So, MRI plays a very important role in selection of patients for early surgical intervention and assessing the post operative outcome.

Keywords: Brain, Recovery, Syringomyelia, Syrinx

INTRODUCTION

Chiari malformation is the malformation of craniovertebral junction. There are four types of Chiari malformations, them Chiari 1 malformation is most common one and mildest form of various congenital hindbrain malformations. It is characterised by inferior herniation of cerebellar tonsil through foramen magnum for more than 5 mm and leads to compression of cervicomedullary junction and interruption of normal flow of CSF. It is asymptomatic in the initial period and become symptomatic later on. It is the dynamic disease and symptoms are due to altered CSF flow dynamics. Neck pain and headache are the most common symptoms of Chiari 1 malformation. It is commonly associated with syrinx. Syrinx refers to abnormal collection of CSF within the spinal cord. Less commonly it is associated with osseous abnormalities like basilar invagination, scoliosis etc. MRI is the investigation of choice for diagnosing the Chiari 1 malformation and its associations [1]. Several prognostic factors can be studies with MRI in addition to diagnosis line presence of syrinx, maximum diameter of syrinx and length of tonsillar herniation. Treatment is not needed in asymptomatic patients. Surgical decompression is the treatment of choice for patients with symptomatic Chiari 1 malformation [2,3]. Long standing syrinx will destroy the fibres of spinothalamic tract and leads to poor

surgical results. Early identification of symptomatic patients with Chiari 1 malformation is very important for better surgical results.

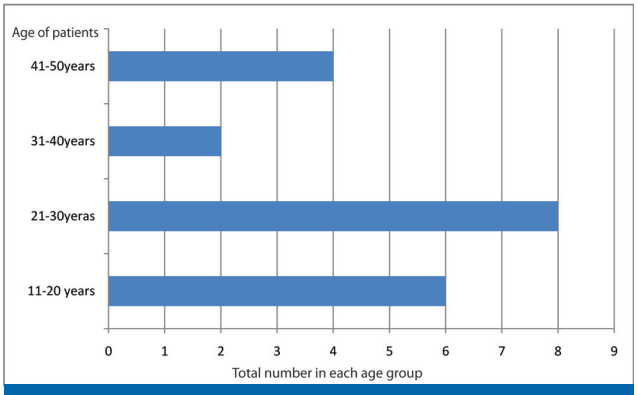
MATERIALS AND METHODS

This prospective study was conducted in KMCT Medical College, Calicut District of Kerala state, India during the period of January 2013 to January 2014 for the duration of 13 months.

Approval for this study was obtained from scientific and ethical committee in our college and written informed consent was obtained from all the patients.

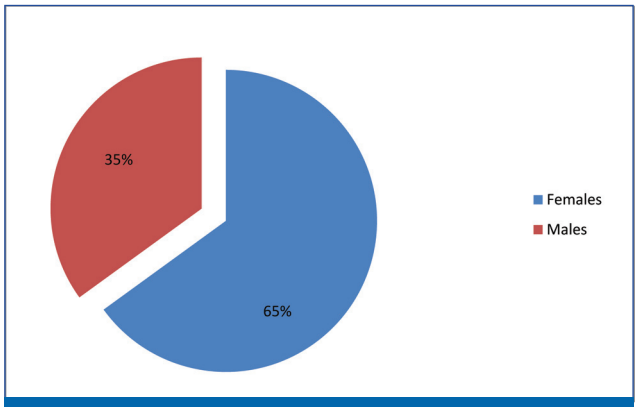
A total of twenty symptomatic patients of Chiari 1 malformation with an age range of 15-47 years (mean age 28 years) who underwent surgery in our medical college from January 2013 to January 2014 were included in this study [Table/Fig-1]. Claustrophic patients, patients with metallic implants and patient not willing to give consent were excluded from the study. Patients with associated scoliosis and other vertebral malformations were also excluded from the study. No dropouts noted in entire study.

There were 13 female (65%) and 7 male (35%) patients in this study [Table/Fig-2].



[Table/Fig-1]: Age distribution (in numbers).

Radiologists trained in musculoskeletal radiology compared the preoperative MRI findings and postoperative outcome. Several variables were studied including age at presentation, duration of symptoms and types of symptoms. All patients underwent preoperative MRI brain and spine with GE HDxt series 1.5 Tesla systems. Imaging sequences included conventional spin-echo T1- weighted images (500/14 ,TR/TE, 4mm thick, 256x256 matrix), fast spin-echo T2-weighted images (3000/105,TR/TE, 3mm thick, 256x256 matrix) obtained in the sagittal plane and T2-weighted images in axial plane for spine. T1 sagittal, T2 coronal and FLAIR axial were done for brain. Radiologists experienced in musculoskeletal MRI analysed the length of herniation, presence of syrinx in sagittal plane and syrinx size in axial plane. Post operative outcome was evaluated for resolution; reduction and persistence of symptoms with serial follow-up of patients for six months.



[Table/Fig-2]: Sex distribution (in percentage).

RESULTS

The mean age of patients with Chiari 1 malformation was 28 years. Chiari 1 malformation was common in females in our study. The mean duration of symptoms was 5.2 years. Ten patients had associated syrinx. Preoperative MRI showed the length of herniation between 6 to 10mm in 7 patients and more than 10mm in 13 patients. In patients with syrinx mean maximum diameter of syrinx was 7.8mm. Evaluation of postoperative patients for clinical recovery was done by serial follow up at 1st, 3rd and 6th months. After surgery nine patients with Chiari 1 malformation without syrinx had complete recovery. One patient without syrinx had partial recovery [Table/Fig-3].Three patients with syrinx had complete recovery. Four patients with syrinx had partial recovery and three patients with syrinx had persistence of symptoms [Table/Fig-4].

Age/Sex	Duration of Symptoms	Length of Herniation	Post-operative Outcome
24/M	3 years	8mm	Resolution
18/F	3 years	10mm	Resolution
18/F	2 years	28mm	Persistence
24/F	4 years	15mm	Resolution
44/F	6 years	8mm	Resolution
21/F	5 years	18mm	Resolution
32/M	4 years	14mm	Resolution
42/M	5 years	15mm	Resolution
30/M	6 years	14mm	Resolution
26/F	4 yeras	12mm	Resolution

[Table/Fig-3]: Post-operative outcome of patients without syrinx.

DISCUSSION

Chiari malformations are the group of hindbrain malformation characterised by disorganised relationship between the posterior fossa structures.

Age/ Sex	Duration of Symptoms	Maximum Diameter of Syrinx	Length of Herniation	Postoperative Outcome
16/F	2 years	6mm	10mm	Resolution
21/F	6 years		12mm	Reduction
15/F	2 years	10mm	10mm	Resolution
40/F	10 years	17mm	10mm	Persistence
45/F	10 years	17mm	12mm	Persistence
27/M	4 years	10mm	12mm	Resolution
47/F	7 years	6mm	10mm	Reduction
20/M	5 years	10mm	12mm	Persistence
30/M	9 years	10mm	15mm	Reduction
20/F	8 years	12mm	16mm	Reduction

[Table/Fig-4]: Postoperative outcome of patients with syrinx.

There are four types of Chiari malformation.

Chiari Type-1 malformation is characterised by inferior herniation of peglike cerebellar tonsil.

Chiari Type-2 malformations consist of inferior herniation of cerebellum, hindbrain, fourth ventricle, tectal beaking and associated abnormalities like myelomeningocele.

Chiari Type-3 malformation consists of inferior tonsillar herniation with cervical myelomeningocele and,

Chiari Type-4 malformation is characterised by cerebellar dysgenesis [4,5].

Chiari 1 malformation is most common malformation. It is characterised by inferior herniation of cerebellar tonsil through foramen magnum for more than 5mm and leads to compression of cervicomedullary junction and interruption of normal flow of CSF. Position of cerebellar tonsils with respect to foramen magnum will vary with respect to age.

Inferior tonsillar herniation below the basion – opisthion line is acceptable up to 3mm and is borderline or benign tonsillar ectopia between 3 and 5mm and in considered abnormal above 5mm [6].

In a study conducted by Barkovich et al., on Chiari malformation had sensitivity and specificity of 96% and 99.5% respectively by keeping the tonsillar herniation of 3mm from foramen magnum and sensitivity and specificity of 100% and 98.5% respectively with tonsillar herniation of 2mm. In their study they concluded that inferior tonsillar herniation of 2mm from the foramen magnum is not clinically significant unless associated with syrinx [7].

In a study conducted by Mikulis et al., concluded that the position of tonsils changes with age. According to their study the distance greater than two standard deviations beyond the normal values is abnormal for the patients. For the ages of 1st decade of life, 2nd and 3rd decades of life, 4th to 8th decades of life and 9th decade of life these distances correspond to 6mm, 5mm, 4mm and 3mm respectively [8].

Review of literatures indicates that Chiari 1 malformation is a dynamic disease and the problem is located at the level of foramen magnum .True incidence of Chiari 1 malformation is not known. It is common in females with a male-to-female ratio of 2:3.

In a study by Balsa et al., 17 patients with Chiari I malformation were included. There were 12 women and 5 men [9]. This correlates with our study in gender ratio. In our study there were 13 females and 7 males.

Syringomyelia is commonly associated with Chiari 1 malformation. It is seen in about 30-70% of patients with Chiari 1 malformation [10]. It is a condition in which a fluid-filled cavity develops inside the spinal cord. This cyst can enlarge over period of time, causing the spinal cord to expand and stretch the adjacent nerve fibres. Finally the syrinx can cause permanent damage to nerve fibres and paralysis. Presence of syrinx will destroy the spinothalamic tract and even after surgery patients will have persistence of symptoms.

It is asymptomatic initially, became symptomatic in early adolescent period. Headache and neck pain are the most common symptom (due to herniated tonsil causing pressure at the base of the brain and obstruction to the CSF flow). Other symptoms are unsteady gait, numbness and tingling of the hands and feet, muscle weakness etc. Flow obstruction at the craniovertebral junction is believed to be responsible for the signs and symptoms of this disease [11,12].

Other associations with Chiari 1 malformations are scoliosis, segmentation anomalies of spine, platybasia /basilar invagination, atlanto-occipital assimilation and sprengel deformity of shoulder [13].

Magnetic resonance imaging of cervical spine is the imaging procedure of choice to diagnose Chiari 1 malformation and its associated anomalies [Table/fig-5-7]. Recently phase contrast velocity encoded cine MRI is used in addition to routine sequences to assess flow dynamics and volumetric analysis.

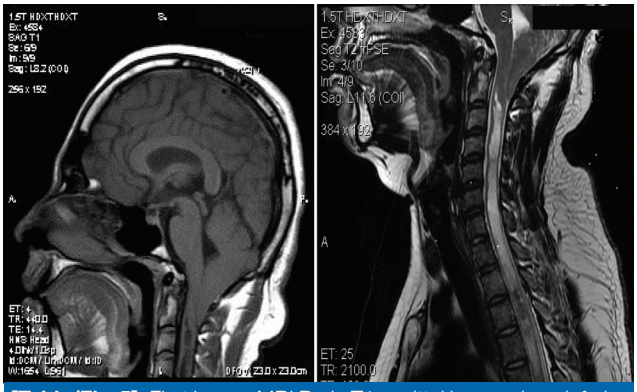
Balsa et al., studied 17 patients of Chiari 1 malformation and conducted post operative follow up for six months and found resolution of symptoms in seven patients, reduction of symptoms in eight patients and no change in symptoms in two patients [9].

In a study conducted by Pillay et al., had 35 patients with Chiari 1 malformation. In their study he divided the patients into two types. In type a twenty patients with syringomyelia were allotted and in type B fifteen patients without syringomyelia were included [14].

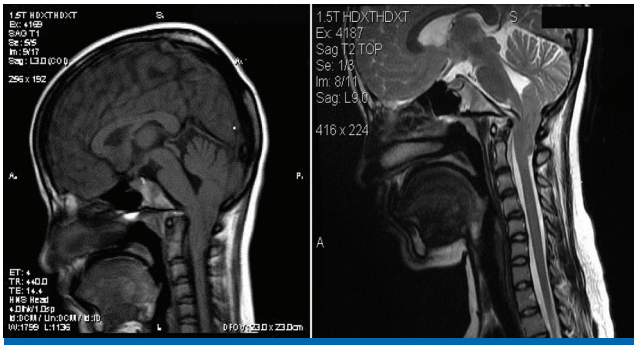
Significant change in clinical condition was noted only in 9 out of the 20 patients (45%) with syringomyelia. In contrast significant improvement occurred in 13 out of 15 patients (87%) without syringomyelia. Reduction in syrinx volume was

obtained in 11 of the 20 patients with syringomyelia in the post operative follow-up [14] [Table/Fig-8].

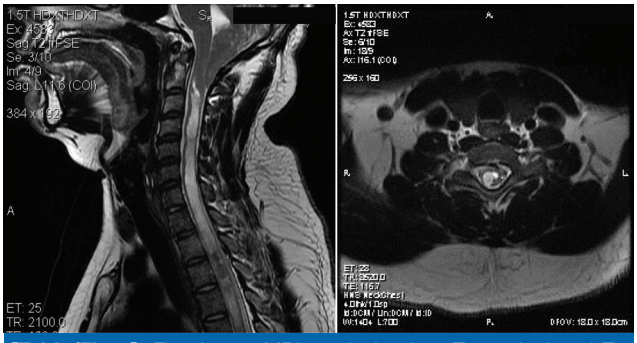
In our study, nine patients with Chiari 1 malformation without syrinx had complete recovery (90%). One patient without syrinx had partial recovery (10%).Three patients with syrinx had complete recovery (30%). Four patients with syrinx had partial recovery (40 %) and three patients with syrinx had persistence of symptoms (30%). Patient with persistence of symptoms had long duration of symptoms, inferior tonsillar herniation of more than 10mm and with large maximum diameter of syrinx.



[Table/Fig-5]: First image-MRI Brain T1 sagittal image show inferior herniation of cerebellar tonsil. Second image- MRI Cervical spine T2 sagittal show presence of syrinx.



[Table/Fig-6]: First image-MRI Brain T1 sagittal image show inferior herniation of cerebellar tonsil. Second image- MRI Cervical spine T2 sagittal show absence of syrinx.



[Table/Fig-7]: First image-MRI cervical spine T2 sagittal and T2 axial images with syrinx.

Study by Pillay et al., [14] (Total 35 patients)				Our Study (Total 20 patients)			
Patients without Syrinx (15 patients)		Patients with Syrinx (Total 20 Patients)		Patients without Syrinx (Total 10 Patients)		Patients with Syrinx (Total 10 Patients)	
Significant response	Partial/no response	Significant response	Partial/no response	Significant response	Partial/no response	Significant response	Partial/no response
13(87%)	2(13%)	9(45%)	11(55%)	9(90%)	1(10%)	3(30%)	7(70%)

[Table/Fig-8]: Comparison of our study with a similar study done by pillay et al.

LIMITATION

Sample size was small in our study was one of the limitations of our study and also we didn't include CSF flow study.

CONCLUSION

MRI has important role in diagnosing the Chiari 1 malformation and its association. Presence of syringomyelia and large diameter of syrinx implies a less favorable response to surgery. Patients without syrinx have better results with decompression. So, MRI plays a very important role in selection of patients for early surgical intervention and assessing the post operative outcome.

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