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ORIGINAL ARTICLE

A RETROSPECTIVE ANALYSIS OF CLINICAL AND RADIOLOGICAL FEATURES OF SYMPTOMATIC CHIARI I MALFORMATIONS

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ABSTRACT

Background: To study the clinical and radiological features of symptomatic Chiari I malformations. **Materials and methods:** This study was a retrospective study conducted in Government medical college, Thrissur, which is a tertiary care government medical college in Kerala, India. The patients admitted with diagnosis of symptomatic Chiari I malformations during the period of 2015 to 2019 were selected for the study. **Results:** The mean age was 23.3 years. Females were 66.7% and males were 33.3%. Mean duration of symptoms (onset of first symptom to the time of presentation) was 16.75 months. Occurrence of symptoms include - sensory symptoms 14(58.3%), walking difficulty 10(41.7%), headache 8(33.3%), neck pain 6(25%), vertigo 5(20.8%), weakness of limbs 4(16.7%), spine deformity 2(8.3%), seizures 2(8.3%) diplopia 1(4.2%) , hoarseness of voice 1(4.2%), swallowing difficulty 1(4.2%) and signs (table3&figur3) include gait disturbance 12(50%), hyperreflexia 8(33.3%), nystagmus 4(16.7%), hypertension 4(16.7%), cape sensory loss 4(16.7%), hand atrophy 3(12.5%), scoliosis 2(8.3%). Mean tonsillar descend from the level of foramen magnum was 11.5 mms. Syringomyelia was present in 14(58.3%) cases. **Discussion:** Patients with Chiari I malformation usually do not become symptomatic until adulthood hence it is also known as adult Chiari malformation. In our study, mean age is 23.5 years. The anomaly is more common among females and our study also females are affected more(66.7%).The most common presenting symptom is usually occipital and upper cervical pain(60% to 70%), often induced by sneezing or coughing according to most of the studies.[5,6,7] Sensory symptoms 14(58.3%) are the commonst symptoms in our study. Magnetic resonance imaging is the imaging modality of choice and it is helpful in differentiating intramedullary spinal cord tumours from syrinx.[13,14,15,16] The radiological definition of Chiari I malformation has been reported as tonsillar herniation of at least 3 mm[17] or at least 5 mm[18] below the foramen magnum. We considered tonsillar herniation 5mm as the cut off for the diagnosis of Chiari I malformation. Mean tonsillar descend from the level of foramen magnum was 11.5 mm as per our study. Syringomyelia was present in 58.3% of cases in our study. **Conclusions:** Chiari I Malformations are more common in adults and incidence is slightly higher in females. Various symptoms include sensory symptoms,walking difficulty, headache , neck pain, vertigo, weakness of limbs, spine deformity, seizures, diplopia, hoarseness of voice, swallowing difficulty and signs include include gait disturbance, hyperreflexia, nystagmus, hypertension, cape sensory loss, hand atrophy, scoliosis in their order of frequency occurrence. Syringomyelia is commonly associated with Chiari I Malformations.

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INTRODUCTION

The Chiari malformation is a condition characterized by herniation of the posterior fossa contents below the level of the foramen magnum and the Chiari I Malformations has been defined as the 5-mm or more downward herniation of the cerebellar tonsils through the foramen magnum (Chiari, 1891; Dyste, 1989; Menezes *et al*., 1990). The etiologies may be congenital or acquired (Menezes, 1990). Chiari I Malformations commonly associated with syringomyelia and it may occurs in association with bony abnormalities at the craniovertebral junction (Menezes, 1990). Patients with Chiari I malformation do not become symptomatic until adulthood hence it is also known as adult Chiari malformation. Patients may present with a variety of symptoms and signs (Chiari, 1891; Dyste, 1989; Menezes *et al*., 1990). The purpose of this study was to study the various clinical and radiological features of Chiari I Malformations.

MATERIALS AND METHODS

Aim: To study the clinical and radiological features of symptomatic Chiari I malformations.

Methods

This study was a retrospective study conducted in Government medical college, Thrissur, which is a tertiary care government medical college in Kerala, India. The patients admitted with diagnosis of symptomatic Chiari I malformations during the period of 2015 to 2019 were selected for the study. Asymptomatic cases of Chiari I malformations and those patients who are not willing to participate in this study were excluded from the study. Consent was taken from all patients. Data collected from medical records. A total of 24 patients with diagnosis of symptomatic Chiari I malformations included in this study. All clinical and important radiological features were recorded and analysed.

RESULTS

SPSS software was used for analyzing data. The mean age was 23.3 years. Females were 66.7% and males were 33.3%. Mean duration of symptoms (onset of first symptom to the time of presentation) was 16.75 months. Occurrence of symptoms include (Table 2 & Figure 2) - sensory symptoms 14(58.3%), walking difficulty 10(41.7%), headache 8(33.3%), neck pain 6(25%), vertigo 5(20.8%), weakness of limbs 4(16.7%), spine deformity 2(8.3%), seizures 2(8.3%) diplopia 1(4.2%) , hoarseness of voice 1(4.2%), swallowing difficulty 1(4.2%) and signs (Table 3 & Figure 3) include gait disturbance 12(50%), hyperreflexia 8(33.3%), nystagmus 4(16.7%), hypertension 4(16.7%), cape sensory loss 4(16.7%), hand atrophy 3(12.5%), scoliosis 2(8.3%). Mean tonsillar descend from the level of foramen magnum was 11.5 mms. Syringomyelia was present in 14(58.3%) cases (Figure 4 & Figure 4)

Table 1.

Age	Mean: 23.5 SD: 14.06
Sex	Male: 8(33.3%) Female: 16(66.7%)
Duration of symptoms	Mean: 16.75 months SD: 11.804

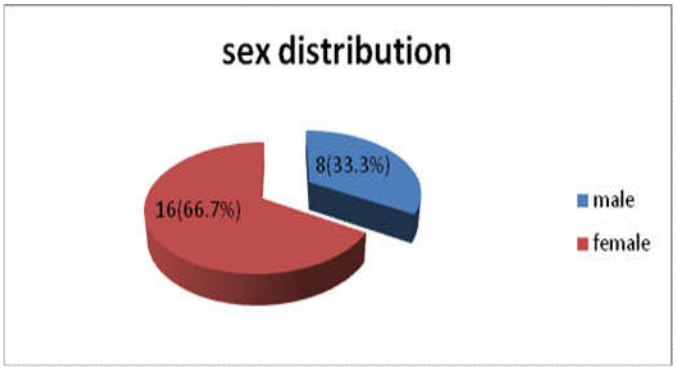


Table 2. Analysis of symptoms

Symptoms	Percentage of occurrence(n=24)
Sensory symptoms	14(58.3%)
Walking difficulty	10(41.7%)
Headache	8(33.3%)
Neck pain	6(25%)
Vertigo	5(20.8%)
Weakness of limbs	4(16.7%)
Spine deformity	2(8.3%)
Seizures	2(8.3%)
Diplopia	1(4.2%)
Hoarseness of voice	1(4.2%)
Swallowing difficulty	1(4.2%)

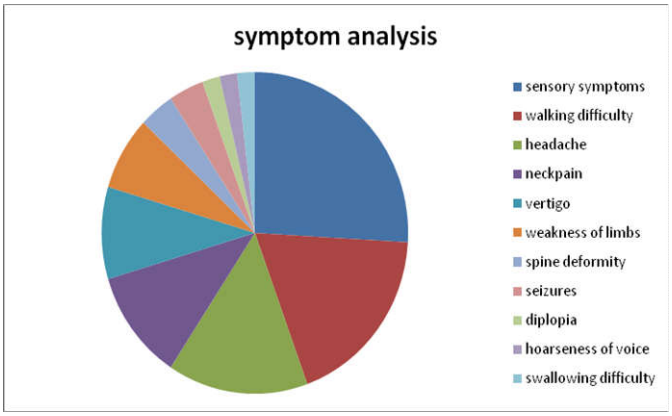


Table 3. Analysis of signs

Signs	Percentage of occurrence(n=24)
Gait disturbance	12(50%)
Hyperreflexia	8(33.3%)
Nystagmus	4(16.7%)
Hypertension	4(16.7%)
Cape sensory loss	4(16.7%)
Hand atrophy	3(12.5%)
Scoliosis	2(8.3%)

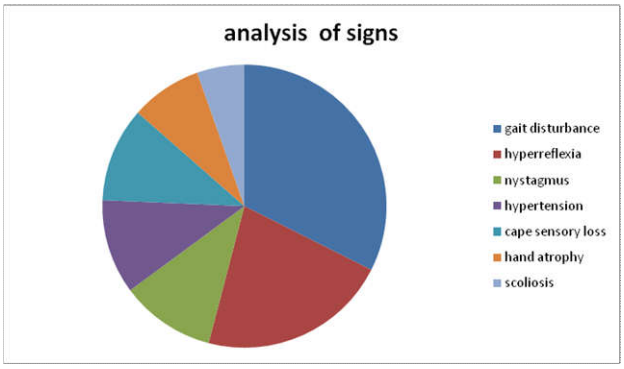
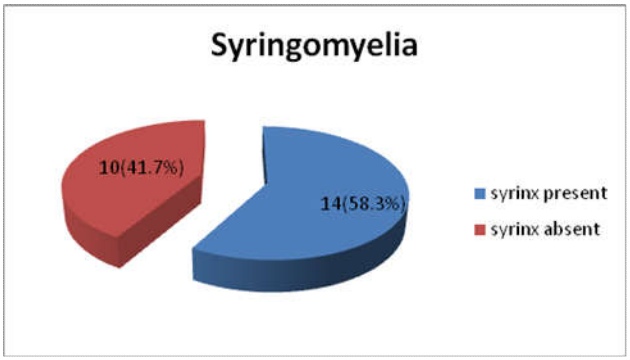


Table 4. Analysis of radiological findings

Tonsillar descend(level below foramen magnum)	Mean: 11.5 mm SD: 4.54
Occurrence of Syringomyelia	14(58.3%)



DISCUSSION

Chiari I malformation (CIM) refers to the downward herniation of cerebellar tonsils through the foramen magnum (FM) (Chiari, 1891; Dyste, 1989; Menezes *et al*., 1990). Patients with Chiari I malformation usually do not become symptomatic until adulthood hence it is also known as adult Chiari malformation. In our study, mean age was 23.5 years. The anomaly is defined as a rare disorder and more common among females (National Organization of Rare Disorder, 1997; Levy, 1983) and females are affected more (66.7%) our study also. The most common presenting symptom was usually occipital and upper cervical pain (60% to 70%), often induced by sneezing or coughing according to most of the studies (Levy, 1983; Paul, 1983; Nohria, 1990). Sensory symptoms 14(58.3%) are the commonst symptoms in our study followed by walking difficulty 10(41.7%), headache 8(33.3%), neck pain 6(25%), vertigo 5(20.8%), weakness of limbs 4(16.7%), spine deformity 2(8.3%), seizures 2(8.3%) diplopia 1(4.2%), hoarseness of voice 1(4.2%), swallowing difficulty 1(4.2%). Gait disturbance 12(50%), hyperreflexia 8(33.3%), nystagmus 4(16.7%), hypertension 4(16.7%), cape sensory loss 4(16.7%),

hand atrophy 3(12.5%), scoliosis 2(8.3%). Headaches may occur in association with ocular, otoneurological, brain stem, and spinal Cord disturbances (Pascual, 1992; Maroun, 1975; Da Silva, 1992). Blurring of vision, nystagmus, extraocular muscle palsies, diplopia, and visual feld deficits are the various ophthalmologic symptoms associated with Chiari I malformations (Bronstein, 1987; Gingold, 1991). Magnetic resonance imaging is the imaging modality of choice and it is helpful in differentiating intramedullary spinal cord tumours from syrinx (Lee, 1985; Spinos, 1985; Lee, 1985; Pojunas, 1984). The radiological definition of Chiari I malformation has been reported as tonsillar herniation of at least 3 mm (Banerji, 1974) or at least 5 mm (Gripp *et al*., 1997) below the foramen magnum. We considered tonsillar herniation 5mm as the cut off for the diagnosis of Chiari I malformation. Mean tonsillar descend from the level of foramen magnum was 11.5 mm as per our study. The pathophysiology of the Chiari malformations and syringomyelia has been already described in the literature, Chiari I malformation commonly associated with Syringomyelia (30% to 70%) (List, 1941; Hankinson, 1978; Banerji, 1974; Williams, 1978; Cahan, 1982). Syringomyelia was present in 58.3% of cases in our study.

Conclusion

Chiari I Malformations are more common in adults and incidence is slightly higher in females. Various symptoms include sensory symptoms, walking difficulty, headache, neck pain, vertigo, weakness of limbs, spine deformity, seizures, diplopia, hoarseness of voice, swallowing difficulty and signs include include gait disturbance, hyperreflexia, nystagmus, hypertension, cape sensory loss, hand atrophy, scoliosis in their order of frequency occurrence. Syringomyelia is commonly associated with Chiari I Malformations.

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