Scholars Journal of Applied Medical Sciences (SJAMS) **ISSN 2320-6691 (Online)**
Sch. J. App. Med. Sci., 2014; 2(2C):740-743 **ISSN 2347-954X (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjams.2014.v02i02.055

Research Article

A Study of Clinical Features and Surgical Outcome of Chiari Type-I Malformation Associated with Syringomyelia

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Abstract: The Chiari malformation type 1 (CM1) has been subject to a recent examination anatomically and clinically where caudal displacement of the hindbrain through the foramenmagnum more than 5mm. The advent of modern neuroimaging has increased not only the detection of the malformation but also our understanding of it. The aim of this study was to attempt and to understand the clinical presentation of the disease with respect to morphology on MRI. The study consisted of 36 patients who presented to our institute Andhra Medical College between January 2008 and December 2011, in whom a diagnosis of CM1 with syringomyelia. Incidence of CM1 with syringomyelia in the present study was observed in youngest patient were 15years old and the oldest being 45 years with an average of 30 years and the sex distribution in the present study was female predominance. Examination by CT-scan showed no hydrocephalous, Magnetic resonance imaging (MRI) showed CM1 with syringomeylia in all 36 patients. The location of the syrinx was cervical in 14 cases and 22 cases had cervico dorsal. The symptoms observed in the present study were ranging from 3 months to 2 years with an average period of 13.5 months. All the patients in this study population underwent small sub occipital craniectomy and foramenmagnum decompression. The findings from the present study conclude that, clinical presentations of patients with CM1 with or without syringomyelia can be extremely diverse and heterogenous. The advent of magnetic resonance imaging has revolution the mainly, chari1 in our understanding of disease pathogenesis and may help to elucidate the different presentations of the disease. This would help to elucidate the nature and pathogenesis of atypical presentations.
Keywords: Chiari malformation type 1, syringomyelia, Charctos joints

INTRODUCTION

Chiari type 1 malformation (CM1) encompasses a spectrum of congenital hindbrain herniation syndromes which was first described by the Austrian pathologist, Hans Chiari in the year 1890. CM1 occurs in the region where the brain and the spinal cord join. Generally, the presenting symptom of CM1 includes head pain, high cervical or occipital pain or dysesthesia, upper limbs weakness or atrophy, lower limbs spasticity, scoliosis and Hemiparesis [1]. In addition, some sort of neurologic deficit is also common. Whereas, scoliosis or other symptoms related to spinal cord dysfunction are occasionally observed [2-4]. Often, individuals with CM1 are also diagnosed with syringomyelia, which is characterized by a fluid-filled cyst in the spinal cord due to an injury or may also present at birth. Several theories has been proposed to explain the CM1 involving hindbrain herniation, but none of them currently been accepted.

To rule out CM1 the choice of diagnostic method is the MR scan in which the tonsillar herniation is best seen on sagittal imaging [5-7]. Syrinx, which occurs most commonly in the cervical spinal cord in association with Chiari [4], can be investigated by spine MRI scan survey [5].

The classic symptoms of a syrinx within the spinal cord include dissociated sensory loss, amyotrophy, and spastic paraparesis and lower motor neuron symptoms in the upper limbs [8]. Weakness and sensory loss, particularly in the hands is typical which may be asymmetrical inspite of a centrally placed syrinx [9]. In later stages and with larger syrinxes the deficits may become bilateral, although in early stages there may be involvement of only one modality and the deficit may be unilateral [8]. Damage to the nerve bodies innervating the lumbricals leads to a claw-hand deformity. In advanced stages of the disease, significant wasting of the intrinsic hand muscles, marked sensory

changes, and even development of a Charcot Joint, which may occur in upto 25% of patients [8].

The present study was a prospective study conducted at Andhra Medical College from 2008 January to December 2011 with the prior approval from the Institutional Ethics Committee. The objective of the present study was to elucidate the clinical presentation of patients with CM1 with syringomyelia and to compare the clinical data, clinical improvement after surgical treatment.

MATERIALS AND METHODS

The study consisted of 36 patients who presented to Andhra Medical College between January 2008 and December 2011, in whom a diagnosis of CM1 with syringomyelia was made, and who underwent surgery. Patients with co existing other anomalies at the cranio-vertebral junction that were also responsible for symptoms and signs were specifically excluded from this study. A protocol was developed to permit the organization of clinical data from the case records of patients treated during the aforesaid period. Clinical data such as age, sex, chief complaints, presenting complaints, symptoms, related to syringomeylia were recorded. Further, a detailed analysis of signs was done and recorded. All available imaging, including X – rays were reviewed and the findings were recorded. MRI scans were reviewed with respect to the presence of CM1, presence and extent of syrinx, and presence of other anomalies.

RESULTS

Incidence of CM1 with syringomyelia in the present study was observed in youngest patient were 15years old and the oldest being 45 years with an average of 30 years. Maximum number of cases was found to be in the age group 35–45 years (48%) followed by 25-35years (36%) and 15-25 years (22%) respectively (Table 1).

Table 1: Distribution of Cases According to Age Distribution

Age Distribution	No. of Patients	Percentage(%)
15-25	08	22
25-35	13	36
35-45	15	42

The sex distribution in the present study was female predominance, There were 15 males (42%) and 21 females (58%) mean age of presentation is 28.4 years. The age distribution is given in the table 2.

Table 2: Distribution of Cases According to Sex Distribution

Sex Distribution	No of Patients	Percentage (%)
Males	15	42
Females	21	58

In the present study, symptom duration ranged from 3 months to 24 months with an average duration of symptoms of 13.5 months. The commonest presenting symptoms were neck pain in 32 patients (21%), weakness in 31 patients (19%), sensory symptoms in 26 patients (16%), unusual presenting symptoms like charcots joints in 2 patients (1%) (Table 3).

Table 3: Distribution of Cases According to Symptoms

Symptoms	No. of Patients	Percentage (%)
Sub occipital & High cervical pain	32	21
Upper limb weakness	31	19
Lower limb weakness & Spastisity	25	15
Tingling and Numbness in upper and lower limbs	26	16
Small muscle wasting	20	12
Bladder involvement	10	06
Swaying	16	10
Charctos joints	02	01

Basic clinical signs data of the patients were as shown in Table 4. The commonest physical sign was dissociated sensory loss and upper limb weakness with diminished reflexes, which was present in 25 and 26 patients respectively. Followed by In coordination signs in 18 patients. Other signs were short neck, posterior column disturbances, wasting of small muscles hand, XI Nerve involvement, nystagmus and down beat.

Cases were studied with emphasis on elucidation of atypical presentations. Atypical presentation was deemed to be present when the overall presentation of the patient did not fit into the classical pattern of presentation. There were 02 patients in all, who had some atypical feature in their presentation of Charcoats Joints with sensory loss (Table 4).

Table 4: Distribution of Cases According to Signs

Study	No. of Patients	Percentage (%)
Short neck	02	01
Upper limb weakness with diminished reflexes	25	16
Lower limb weakness with Babinski's positive	15	09
Dissociated sensory loss	26	17
Posterior column disturbances	15	09
In coordination	18	11
Wasting of small muscles hand	16	10
Fasciculation's	06	04
Horner's syndrome	03	02
XI Nerve involvement	04	02
Nystagmus	15	09
Horizontal	13	08
Down beat	02	01
Charcots joints	02	01

In the present study the radiological examination by CT-scan showed no Hydrocephalous, Magnetic resonance imaging (MRI) showed CM1 with syringomeylia in all 36 patients. The location of the syrinx was cervical in 14 cases and 22 cases had cervico dorsal. Plain X – Rays of the cervical spine were available in 36 patients, loss of funneling / increased canal diameter was observed in 15 cases.

All the patients in this study population underwent small sub occipital craniectomy and foramenmagnum decompression. Removal of Posterior arch of C1 and Duraplasty with fascialata graft. Clinical improvement after surgical procedure was shown in table 5.

Table 5: Distribution of Cases According to Treatment

Symptom/sign	Preoperative	Improvement
Suboccipital & High Cervical pain	32	28
Weakness	31	25
Sensory Symptoms	26	18

DISCUSSION

The clinical presentation of patients with Chiari malformation with or without syringomyelia is complex and heterogenous, and the number of studies aimed at elucidating the presentation and attempts at classification bear testimony this fact. In the days before modern neuroimaging, diagnosis relied on clinical findings; pnuemoencephalography, myelography and intrathecal contrast CT Scanning. However with the advent of MRI there is an evolution towards the better understanding of the pathophysiology of this complex problem and also a better understanding of the correlation between clinical signs and imaging data. The aim of this study was to attempt and to understand the clinical presentation of the disease with respect to morphology on MRI. Atypical presentations were also studied which were presented in the form of Charcots joints with sensory loss.

In the present study, symptom duration ranged from 3 months to 24 months with an average duration of symptoms of 13.5 months. As in the order series, the symptoms like sub occipital & cervical pain 32 (89%), limb weakness 31 (86%) sensory loss 26 (72%) small muscle wasting 20 (56%) nystagmus 15 (42%) was observed. Horner's syndrome was another unusual finding in most studies, our study showed 1% of the cases. Whereas, Dyste *et al*. [10] observed 4% of patients, Levy *et al*. [11] found 6% of cases. Sphincter disturbances were encountered in 6% of patients in this study compared to 4% in the study by Saez *et al*. ([12], nil in the study by Levy *et al*. [11] and 4% in the study by Dyste *et al*. [10]. In the present study, youngest patient was 15years old and the oldest being 45 years with an average of 30 years. Whereas, earlier studies showed that the mean average of the age at onset were in agreement with our present study [10-12].

The symptoms observed in the present study were ranging from 3 months to 2 years with an average period of 13.5 months. The present study was comparable with earlier study by Pillary *et al*. [13] who reported 18 months of average period for duration of symptoms. The observations from the present study also had shown that males outnumbered females. Banerji and Millar [14], Saez *et al*. [12], Levy *et al*. [11] reported similar type of sex incidence in males and females

Based on clinical presentation, sub occipital and cervical pain was the commonest symptom and was present in 89% of cases. The symptoms of weakness and Sensory occurred in 86%, 72% of the cases respectively. The present study was comparable with Dyste *et al*. [10], Pillary *et al*. [13], Verasriet *et al*. [15] study who reported pain abdomen in 73%, 85% and 60% of the cases. As in the order series, weakness is the commonest symptom which was seen in 86% of cases, followed by Nystagmus (42%), wasting (56%) and sensory loss (2%). Similar observations were also observed by Saez *et al*. [12] studies; Sensory loss occurred in 63.3% cases Dyste *et al*. [10] and co-workers observed Scoliosis was presenting symptom in 42% of cases, followed by Nystagmus (24%).

The studies by Pillay *et al*. [13], Versari *et al*. [15], Stevens *et al*. [16], and Bindal *et al*. [17] grouped patients into the various "syndromic" presentations rather than symptoms-wise. In most studies, the predominant complaints were related to foramen magnum compression syndrome, such as pain, which is probably reflective of the difference in the patient populations. Most studies have included patients with CM1 without syringomyelia, whereas in the present study the population is a fairly homogenous group of patients with CM1 and syringomyelia. To conclude, clinical presentations of patients with CM1 with or without syringomyelia can be extremely diverse and heterogenous. The understanding of the disease and its pathogenesis is still evolving. The advent of magnetic resonance imaging has revolution the mainly, chari1 in our understanding of disease pathogenesis and may help to elucidate the different presentations of the disease. This would help to elucidate the nature and pathogenesis of atypical presentations.

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