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Chiari I Malformations; Clinical Presentations, Diagnosis and Management.

KEYWORDS: Chiari malformation, disruption of CSF flow, scoliosis, posterior cranial fossa decompression, syringomyelia, duroplasty.

Dr. Ramanjulu Mala	Dr. Rama Mohan Naik Ramachandra	Dr. Ananth Mugi Lakshmi
M.Ch. Associate professor, Department of Neurosurgery, Kurnool Medical College Kurnool, Andhra Pradesh, India	M.Ch.Assistant professor, Department of Neurosurgery, Kurnool Medical College Kurnool, Andhra Pradesh, India	M.Ch. Assistant professor, Department of Neurosurgery, Kurnool Medical College Kurnool, Andhra Pradesh, India

Dr.Nagaraju Bugude Nagireddy	Dr.Ram Balaji Naik Gonavath	Dr. Venkateswara Rao Chiniga
M.Ch. Assistant professor, Department of Neurosurgery, Kurnool Medical College Kurnool, Andhra Pradesh, India	M.Ch.,Assistant professor, Department of Neurosurgery, Kurnool Medical College Kurnool, Andhra Pradesh, India	M.S., Tutor, Department of Neurosurgery, Kurnool Medical College Kurnool, Andhra Pradesh, India

ABSTRACT *Background: In this study we have evaluated the clinical presentations, diagnosis, intraoperative findings and clinical outcome in patients with Chiari I malformations undergoing posterior cranial fossa decompression*

Materials and Methods: Outcome was assessed by Thirty patients with different age groups with Chiari I malformations who underwent suboccipital craniectomy and wide duroplasty .The clinical evaluation of postoperative signs and symptoms and magnetic resonance imaging of the craniovertebral junction were analyzed.

Results: Headache in 24(80%) patients and neck pain in18 (54%) were the most common symptoms. Syringomyelia was present in 21 patients (21%).scoliosis in 4 patients (12%), basilar invagination in 2 patients (6%). Female predominance of the malformation was observed, with a female: male ratio of 3:2.The mean age of onset was 27 years and 7 months. No reported positive family history. All patients had descent of cerebellar tonsils. The cerebellar tonsils were found to lie at C1 and C2 in 24 and 6 patients respectively. All patients underwent suboccipital decompresssion and autologous duroplasty using fascia lata in 18 patients and G patch (prolene) in 12 patients. The immediate postoperative period,24(91 %) of patients showed improved symptomatology and post operative magnetic resonance imaging revealed favourable findings comprising syrinx collapse or reduction of syrinx diameter in 85 % of patients. The mean follow-up period was 3 years and 4months. There was no mortality associated with the procedure. Cerebrospinal fluid leak (CSF) and meningitis was present in 7(21%) cases using G patch for duroplasty and 2 cases(6%) using fascia lata for duroplasty.

Conclusions: The ACM type I can vary in presentation throughout in different age groups, Suboccipital decompression and duroplasty in selected cases are effective treatment for most patients with Chiari I malformations.

INTRODUCTION

The Chiari I malformation is a disorder of mesoderm characterized by herniation of cerebellar tonsils through the foramen magnum. The malformation typically presents with neurological symptoms during early adulthood. Patients can be asymptomatic or can have a variety of neurological symptoms including headache, neck pain, visual disturbances, vertigo and ataxia [1]. Chiari I may lead to the development of syringomyelia , which can lead to additional neurological deficits .Since surgical intervention can improve existing symptoms as well as prevent further neurological deterioration from syringomyelia , there may be a benefit to early identification of these patients. Symptomatic patients usually demonstrate at least 5mm of tonsillar hernlation below foramen magnum.

MATERIALS AND METHODS

The medical records of patients aged between 9 to 40 years undergoing surgery with a diagnosis of Chiari I malformations year 2003 to 2014, were retrospectively evaluated. The data was analyzed for type of presentation, radiological features, indications for surgery, and results in follow-up. Mean follow-up period was 3 years and 5 months. Outcome was assessed by thirty patients with different age groups with Chiari I malformations who underwent suboccipital craniectomy (Figure; 1)

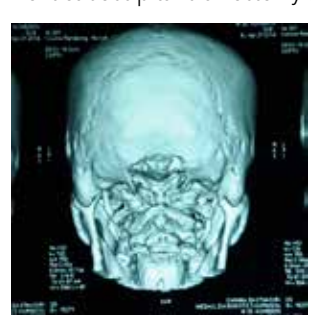

Figure 1; CT scan of post operative patient shows the extent of suboccipital craniectomy and removal of CI

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arch.and wide duroplasty .The clinical evaluation of postoperative signs and symptoms and magnetic resonance imaging of the craniovertebral junction was done.

RESULTS

Headache in 24(80%) patients and neck pain in18 (54%) were the most common symptoms. Syringomyelia was present in 21 patients (21%) (Figure; 2)

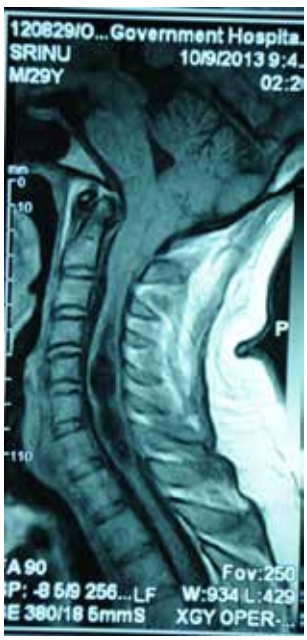

Figure 2; cervicomedullary magnetic resonance imaging showing a type I Chiari malformation,syringomyelia.

scoliosis in 4 patients (Figure; 3) (12%), basilar invagination in 2 patients (6%). Female predominance of the malformation was observed, with a female: male ratio of 3:2.

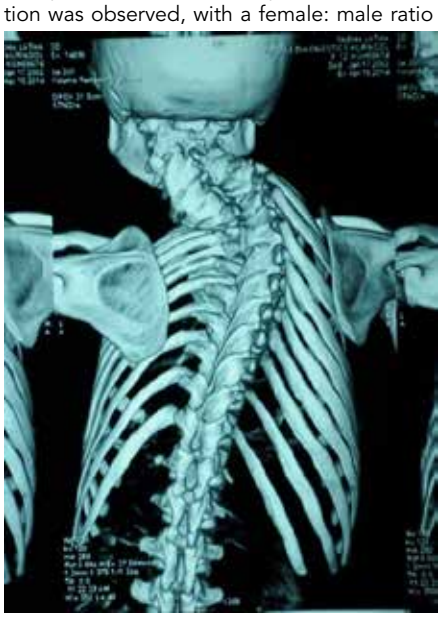

Figure 3; CT scan of patient with Chiari I malformation showing scoliosis of dorsal spine.

The age of onset was 27 years and 7 months. No reported positive family history. All patients had descent of cerebellar tonsils (Figure; 4, 5).

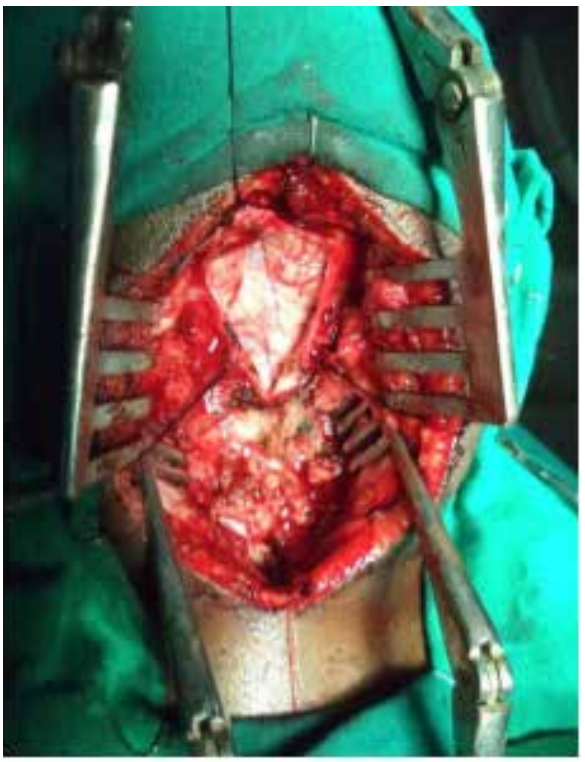

Figure 4; Intraoperativephotograph of Chiari I malformation showing descent of cerebellar tonsils well below the foramen magnum.

Figure 5; MRI scan showing herniation of the cerebellar tonsils more than 10 mm below the foramen magnum into the cervical spinal canal.

The cerebellar tonsils were found to lie at C1 and C2 in 24 and 6 patients respectively. All patients underwent decompresssion and autologous duroplasty using fascia lata in 18 patients and G patch (prolene) in 12 patients. The immediate postoperative period,24(91 %) of patients showed improved symptomatology and post operative magnetic resonance imaging revealed favourable findings comprising formation of neo-cistern behind the tonsils and the tonsils are round in shape(Figure 6),

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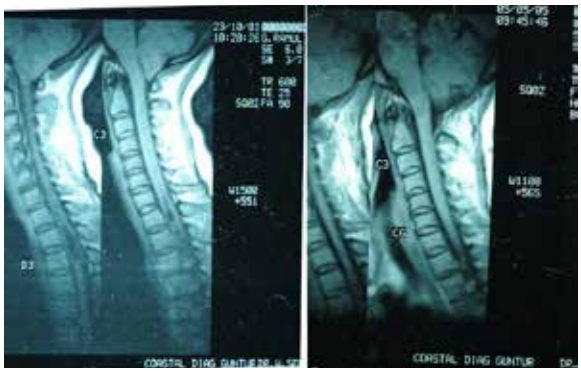

Figure6; Image Left,sagittal MRI scan showing the cerebellar tonsils shifted downwards through the foramen magnum into the upper cervical canal. Image Right, post operative showing cerebellar tonsils shifted upwards and the tonsils are rounded in shape

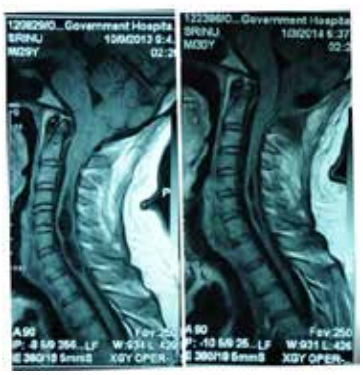

Figure7;Image Left,sagittal MRI scan showing the cerebellar tonsils shifted downwards with syringomyelia. Image Right, post operative showing cerebellar tonsils shifted slightly upwards and the resolution of syrinx.

Table I: clinical and radiographic presentation and management outcome in 30 patients with Chiari I malformation.

Clinical and radiographic findings	No. of cases	Management outcome Mean follow-up=3 years and 5months	
		Improved	unchanged
Head ache	24(80%)	22(91%)	2(8%)
Neck pain	18(54%)	16(88%)	2(12%)
Vertigo	16 (53%)	14(87%)	2(13%)
Ataxia	10(30%)	7(70%)	3(30%)
Paraesthesia of upper limbs	14 (42%)	8(57%)	6(43%)
Syringomyelia	21 (70%)	18(85%)	3(15%)
Scoliosis	4(12%)	_	4(100%)

syrinx collapse(Figure;7, 8) in 85 % of patients(Table I) There was no mortality associated with the procedure. Cerebrospinal fluid leak (CSF) and meningitis was present in 7(21%) cases using G patch for duroplasty(Figure 9) and 2 cases(6%) using fascia lata for duroplasty. The clinical presentations and surgical outcome is similar to the results reported by Milhorat et al, Park et al, Klekamp et al, and Fischer series. Our observations are similar to these series.

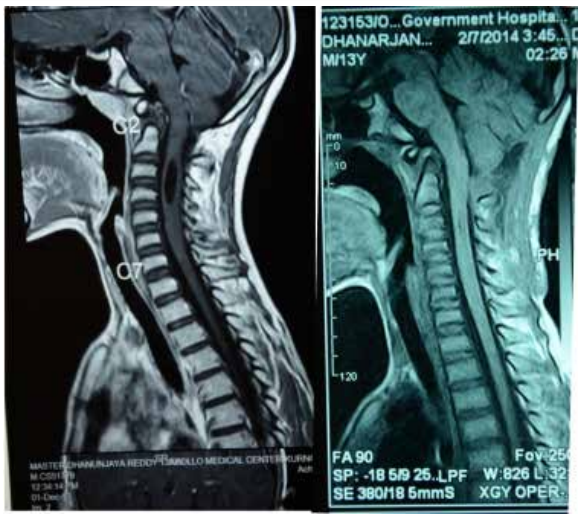

Figure8;Image Left,sagittal MRI scan showing the cerebellar tonsils shifted downwards with syringomyelia. Image Right, post operative showing cerebellar tonsils shifted slightly upwards and the resolution of syrinx.

Figure 9;A dural decompression graft (G-patch)has been sutured in place creating larger foramen magnum region and relieving pressure on the medulla and spinal cord.

DISCUSSION

Although Cleland described the first cases of malformations in 1883, the disorder is named after Hans Chiari, an Australian pathologist who, between 1891 and 1896, described various anomalies of the caudal cerebellum and brainstem from post mortem studies. Chiari colleague, Julius Arnold made additional contributions to the definition of Chiari malformation[2, 3] .In his honour, the students of Dr. Arnold later named the type II malformation as Arnold-Chiari malformation. Chiari classified the hind brain malformations into types I, ii, III, and then in his latter publication added the type IV malformation. Chari 0 and Chiari 1.5 is an interesting nomenclature introduced by Iskander and Oakes to the more subtle forms of malformations [4, 5]. Chiari I malformation is most common, peg-like cerebellar tonsils displaced into the upper cervical canal through foramen magnum. Chiari II malformation in which there is displacement of the medulla, fourth ventricle and cerebellum through the foramen magnum, usually associated with a lumbosacral myelomeningocele. Chiari III malformation, features similar to Chiari but with an occipital and /or high cervical encephalocele, Chairi IV malformation, severe cerebellar hpoplasia without displacement of cerebellum through foramen magnum. Chiari Zero, the case of differ-

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ential pressures exerting influence across the cervicomedullary junction is exemplified by presentation of certain group of people with minimal or no hindbrain herniation and syringomyelia. Chiari 1.5specifically address patients with tonsillar herniation but without brainstem elongation or fouth ventricle deformation.

The prevalence of Chiari I malformations in the general population has been estimated one in 1000 with a slight female predominance. Based on analysis of family aggregation a genetic basis for Chiari I have been suggested. Recent studies suggest linkage to chromosomes 9 and 15[6]. It is hypothesized that Chiari type I originates as a disorder of Para-axial mesoderm, which subsequently results in formation of a small posterior fossa. The development of cerebellum within this small compartment results in overcrowding of the posterior fossa, herniation of the cerebellar tonsils, and impaction of the foramen magnum. This theory is consistent with the observed association of Chiari I and other hereditary mesodermal connective tissue disorders [7].

Patients with Chiari I malformation usually remain asymptomatic until late childhood or early adulthood [1].Symptoms of Chiari I develop as a result of pathophysiological consequences of the disordered anatomy resulting in compression of medulla and upper spinal cord, compression of cerebellum, and disruption of CSF flow through foramen magnum. Compression of the cord and medulla may result in myelopathy and lower cranial nerve and nuclear dysfunction. Compression of cerebellum may result in ataxia, dysmetria, nystagmus, and disequilibrium. Disruption of CSF flow through foramen magnum probably accounts for the most common symptom pain. Patients can be asymptomatic or can have a variety of neurological symptoms including headache, neck pain, visual disturbances, vertigo and ataxia. Chiari I may lead to the development of syringomyelia or spinal cord cavitation which leads to additional neurological deficits[8].

Headache and neck pain in Chiari I are often exacerbated by cough and Valsalva manoeuvre. Hydrocephalus occurs less frequently. The disordered flow of CSF through the foramen magnum may result in formation of syringomyelia and central cord symptoms such as hand weakness and dissociated sensory loss. These symptoms are usually asymmetrical, as a syrinx has a tendency to develop in the side of the spinal cord that is more significantly affected by tonsillar ectopia.

Anomalies of the base skull and spine are seen in 30-40% of patients with the Chiari I malformation .These anomalies include basilar impression, atlanto-occipital fusion, Klippel-Feil deformity, cervical spina bifida occulta and scoliosis[9]. There may be brainstem dysfunction in some who have a Chiari I malformation(10-40%) resulting in drop attacks, postural and cough headaches, visual disturbances from nystagmus, spasticity, sensorimotor deficits, ataxia, dysarthria, dysphagia.

The diagnosis of ACM is often delayed. The interval from clinical presentation to diagnosis usually is 5 years. The diagnostic imaging modality of choice is MRI. Abnormal tonsillar descent with 3-5 mm is chosen by most radiologists as the threshold value [10]. Other findings encountered include compression of the posterior fossa subarachnoid space, overcrowding in the posterior fossa, peg-shaped tonsils, and increased slope of the tentorium, medullary kinking and basilar impression. Cine MRI CSF flow study shows obstruction of movement of CSF caused by the peg -like tonsils, caudal systolic motion of tonsils, and loss of flow behind the tonsils or anterior to the brainstem [11, 12].A more frequent observation is syringomyelia ,reported in 50-75%of cases[13]. Cervical and skull radiographs are of limited diagnostic value and usually reveal bony anomalies. Computed tomography is useful to delineate any associated bony anomalies.

Some of the entities to be entertained in the differential diagnosis are multiple sclerosis, fibromyalgia, psychogenic disorder, migraine, idiopathic intracranial hypertension and spinal cord tumours.

Treatment of Chiari I malformations and syringomyelia depend upon the exact type of malformation, as well as progression in anatomy changes or symptoms. The Chiari symptomatology is brought on by a lack of cerebrospinal fluid flow through the foramen magnum because of crowding of the tonsils, restoration of foramen magnum CSF flow becomes the goal of surgical intervention. The current practice involves suboccipital craniectomy including foramen magnum and removal of the laminar arch of C I, followed by augmentation duroplasty facilitating CSF flow .Small sub-occipital craniectomy is performed, exposing the dura and the occipital venous sinus while excision of the posterior arch of CI facilitates the dural exposure [5, 14, 15, 16,].The dura is opened vertically in the midline and tacked laterally to the muscles. Very often, the dural bands opposite CI, thickened arachnoid membrane and septations bridging the cerebellar tonsils are seen in the midline. The tonsils are usually tongue like and smooth, very often pulling the inferior cerebellar artery along with them into the spinal canal. Pericranium from the occipital region or the dural substitute can be used to perform duroplasty[17]. Fischer reported resolution of syrinx in 93% of cases on a literature survey[18].Park et al., had improvement in all the cases over the long term follow up [15, 19.] The early and timely treatment of a Chiari I malformation in a child with progressive scoliosis can yield a good result, in terms of halting the progression of the spine deformity. A CSF flow study in patients with Chiari malformation who undergone posterior cranial fossa decompression using spatial modulation of magnetization [20].Patients presenting with a clear anatomic Chiari malformation but no demonstrable clinical syndrome are followed expectantly, a yearly follow-up that includes questions regarding historical changes consistent with Chiari symptoms and a neurological examination. Medical management can put off surgery for some time, allowing ongoing discussion and better understanding of the impact of the symptoms on life and lifestyle.

Postoperative complications include problems with wound healing, infection, muscle spasm, pain in the suboccipital region and the risk of CSF leakage and meningitis.

Prognosis depends on severity of malformation, associated neurological abnormalities and complications.

CONCLUSIONS

CM I is a disorder of mesoderm, the anomaly occurs sporadically but can be transmitted genetically in some families. Patients with Chiari malformation type I are often asymptomatic. Presence of anatomic Chiari I malformation or compelling clinical Chiari syndrome should lead to evaluation by a neurologist or neurosurgeon experienced with the syndrome and their treatment. Surgical treatment for symptomatic Chiari I malformation is usually very successful when treated promptly[21].

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