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Study of Chiari Malformations in North Karnataka Region

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Abstract

The most common anomalies of cranio-vertebral junction are Chiari malformations. Chiari malformations constitute a variety of four main syndromes (I, II, III, and IV), which describe the protrusion of brain tissue into the spinal canal through the foramen magnum. These malformations frequently occur in combination with other pathological entities such as myelomeningocele, hydrocephalus, and/or hydrosyringomyelia.They involve both skeletal and neural structures. The incidence of Chiari Malformation was 1 in 1000 births but with the increased use of imaging techniques such as CT scans and MR imaging it is suggested that this condition is much more common than thought earlier to be. However it is very difficult to estimate the exact rate of occurrence as some of cases are asymptomatic or do not develop symptoms till adulthood. In the present study 100 MRI films of patients with symptoms pertaining to chiari malformations from North Karnataka region were studied and interpreted.

Keywords: Chiari Malformations; Hydrocephalus; Myelomeningocele; hydrosyringomyelia.

Introduction

Chiari malformations constitute a group of different clinicopathological entities with varying etiology, pathophysiology, and clinical features. They represent varying degrees of hindbrain herniation through the foramen magnum. In 1883, John Cleland described a case of hindbrain malformation found during autopsy. Hans Chiari, an Austrian pathologist, performed post-mortem examination of forty cases in 1891 and 1896 and gave a detailed description of hindbrain malformations [1]. Chiari described these malformations as congenital anomalies of the hindbrain characterized by downward elongation of the brain stem and cerebellum into the cervical portion of spinal cord [2]. In his initial description, Chiari classified the hindbrain malformations into type I, II and III and then latter added type IV malformation [3].

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Classification of Chiari Malformation

Chiari malformations were described to be of four types:

Type I

It is the most commonly observed Chiari malformation. In this type, there is tonsillar herniation through foramen magnum. It is often associated with syringomyelia but not hydrocephalus. This type of Chiari malformation is congenital as well as acquired. Radiologically, Type I is described as tonsillar decent of 5 mm below foramen magnum. Patients with Type I Chiari malformation may be asymptomatic or present with mixture of cerebellar and pyramidal tract signs associated with dysfunctioning of lower cranial nerves [4].

Type II

It is also called as classic Chiari malformation or Arnold- Chiari malformation. It is less common. In type II Chiari malformation there is caudal descent of cerebellar tonsils and the vermis into the spinal canal

along with brain stem and fourth ventricle. Type II Chiari malformation is usually accompanied by myelomeningocele. Hydrocelaphus is seen in 90% of cases. Symptoms arise from dysfunctioning of brain cells and lower cranial nerves. Myelomeningocele results in the partial or complete paralysis of area below the spinal opening. Due to the severity, Type II patients become symptomatic in infancy or early childhood [4].

Type III

It is the most serious form of Chiari malformations. There is occipital or cervical encephalocele alongwith intra cranial abnormalities seen in type II Chiari malformation and a wide foramen magnum. This defect is readily visible and palpable. Plain radiographs help to identify the scull or cranial defects while MR imagining identifies the herniated brain tissue [4].

Type IV

It is a very rare type. It is characterised by cerebellar hypoplasia or aplasia and tentorial hypoplasia. There is no hind brain herniation in this type.

Other types of Chiari malformations include Chiari 0 and Chiari 1.5 types. Chiari 0 includes minimal or no hind brain herniation but the headache and other symptoms of Chiari malformation are present. Chiari 1.5 includes patients with tonsillar herniation without brain stem elongation or fourth ventricle deformation [5].

The incidence of Chiari malformations has been increased with the evolution of CT and MRI scans and hence this study was taken up to give further more insight on Chiari malformations in North Karnataka region.

Materials and Methods

In the present study 100 MRI films from different scan centers in North Karnataka region were studied. The MRI films of patients presenting with Headache and problems in balance and cordinationwere selected for the study. The signs of chiari malformations like herniation of hind brain, syringomyelia and myelomeningocoeleswerelooked for in the MRI films and tabulated according to different parameters.The reliable morphological features leading to diagnosis of Chiari II malformation on MR imaging are downward herniation of the cerebellum, downward displacement of the medulla, pons and fourth ventricle, medullary kinking, abnormally shaped fourth ventricle, hypoplastic tentorium and breaking mesencephalic tectum [6]. Along with above signs, Myelomeningoceole and Hydrocephalus are very often associated with Chiari malformations [7,8]. Hence these signs on MRI were considered for diagnosis of Chiari malformations in present study.

Results

Out of the 100 cases studied with headache, imbalance and incoordination, 58 cases were of females and 42 cases were of males (Table 1). There were 78 cases with age less than 7 years. Out of all the MRI films studied Cerebellar herniation was seen in two cases and both the patients were female child. Hydrocephalus was seen in 6 cases. Myelomeningocoele was seen in 2 cases. The cases in which cerebellar herniations were seen also had syringomyelia.

Table 1: Shows the gender distribution of cases with headache, imbalance or incoordination

Gender	Male	Female
No. of Cases(n=100)	48	52

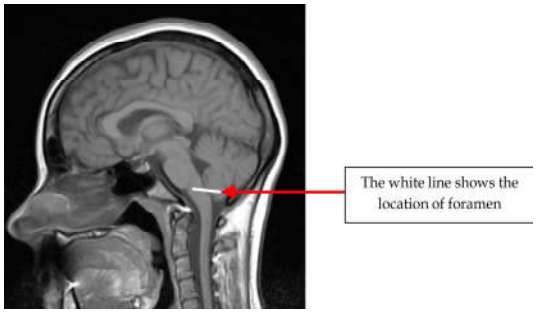

Fig. 1: Shows T1 weighted MRI image of normal sagittal section of brain

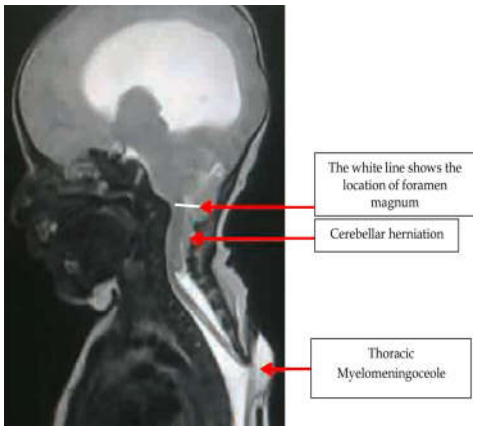

Fig. 2: Shows T2 weighted MRI image of sagittal section of brain with cerebellar herniation with Thoracic myelomeningoceole

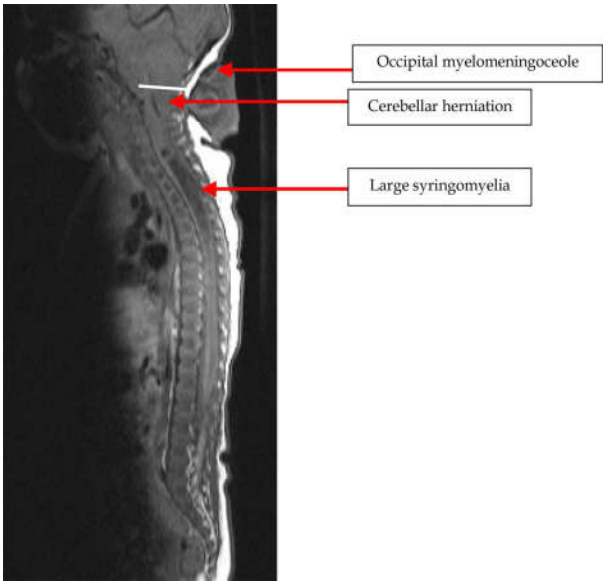

Fig. 3: Shows T1 weighted MRI image of sagittal section of brain with cerebellar herniation with Occipital myelomeningoceole and large syringomyelia

Case 1 (Fig. 2) had cerebellar herniation with thoracic myelomeningoceole with hydrocephalus and syringomyelia. Case 2 (Fig.3) had cerebellar herniation with occipital myelomeningoceole with large syringomyelia.

Discussion

Numbers of studies have been done on Chiari malformations. It has been found that the prevalence of Chiari I malformation is one per thousand in general population. With invent of the CT scans, MRI scans and newer imaging techniques, the diagnostic abilities have also improved.

Niel sGeerdinll et al carried out MR imaging study on 79 children and concluded that the reliable morphological features leading to diagnosis of Chiari II malformation on MR imaging are downward herniation of the cerebellum, downward displacement of the medulla, pons and fourth ventricle, medullary kinking, abnormally shaped fourth ventricle, hypoplastic tentorium and breaking mesoncephalic tectum [6]. Gammal T et al stated that myelomeningocoele is present with Chiari II malformation almost in all cases. However, the reverse is not true all the time [7]. According to Rauzzino M et al, Hydrocephalus is seen in 90% of the cases and ventricles are seen asymmetrically [8]. According to Stevenson KL, approximately 1/3rd of the patients with Chiari II malformation develop signs and symptoms of brain stem compression [9]. Curnese JT carried out a study on 33 patients with Chiari II malformation and found out that 36% of patients were symptomatic while 64% were asymptomatic [10]. Wolpert SM et al carried out a study to see the relation between the amount of brain stem herniation and neurological status of the children with Chiari II malformation and found out that the neurological status was not affected by either the amount of herniation of the characteristics of cervico-medullary and hence concluded that the breathing and swallowing difficulties experienced by children with Chiari II malformation is due to other factors like disorganization of brain stem nuclei [11]. Tsai T et al did a biometric analysis of 25 patients with myelomeningocele and Chiari II malformation and concluded that degree of vermion herniation and cervicomedullary junction herniation are independent variables in Chiari II malformation while the size of posterior cranial fossa is an important factor in explaining the variability of vermian herniation [12].

The initial description of CM type III was based on the description of a single case with a large dermal sac in the occipital region, containing herniated cerebellum.Type III is characterized by caudal displacement of the medulla and herniation of part of the cerebellum intoan occipitocervicalmeningocele. Sometimes, part of thehindbrain is also herniated. Hydrocephalus is present in 50% of these cases and is always of obstructive etiology, due to either aqueductal stenosis or an associated Dandy-Walker malformation. Chiari type III is a neuroectodermal malformation [13].

In the present study Arnold-chiari malformation was seen in one case and Chiari III malformation was seen in other. Invariably both the cases were having myelomeningoceoles and syringomyelia. It is would be appropriate to say that all chiari malformations cases are associated with myelomeningoceoles.

The embryological basis of above anatomical variation can be understood by different theories suggested by researchers. The theory of overgrowth suggested that the overgrowth of neural plate before neurulation prevents fusion of neural folds. According to hydrodynamic theory, imbalance between pulsating choroid plexus of forth and lateral ventricles result in Chiari malformation [14]. According to Jenning et al, Chiari malformation occurs because the normal zone of fusion at third and fourth somites is displaced caudally below the third to fifth somite pairs thus causing the displacement of the area of formation of cervicomedullary junction [15]. A theory was given by Daniel and Strich, which stated the developmental arrest, especially in the progression of pontine flexure

during 28th and 29th day of gestation as a cause of Chiari malformation [16].

Conclusion

Chiari malformations are not as rare as would be expected from the small number of reported cases but with the increased use of CT and MRI scans they can be much more common. The defect is almost always, but not invariably, associated with meningomyelocele or spina bifida occulta in lumbosacral region. Hydrocephalus is present in most cases. Other associated defects of development include craniolacunia, hydromyelia, syringomyelia, double cord, basilar impression [5]. Limitations of present study are the selection of target population, as mentioned in literature not all cases present with symptoms, No proper diagnostic facilities in rural areas and poor people cannot afford the cost of CT and MRI scans.

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