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CHIARI II MALFORMATION: COMPUTED TOMOGRAPHIC EVALUATION*

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In 1883, Clealand described the morbid findings in a full-term infant with spina bifida, craniolacunia, an elongated brainstem and herniated fourth ventricle through the foramen magnum. In 1891, Chiari reported the anomalies in the pontobulbocerebellar area and classified them into three types. An infant with fourth ventricular and cerebellar herniation was reported by Arnold in 1894. In 1896, Chiari added a fourth type of malformation to the three types of malformations he had previously described-the Chiari II or Arnold-Chiari malformation which is the most common of the four types[1-4].

The Chiari II malformation is characterized by the caudal elongation of the cerebellum and brainstem through an enlarged foramen magnum. There may also be accompanying midbrain, forebrain, and cranial bony deformities, as well as marked hydrocephalus. It must be differentiated from other types of hydrocephalus preoperatively, since the surgical management of the Chiari II malformation may differ from other types of hydrocephalus. The surgical management of the Chiari II malformation consists of cervicomedullary decompression in addition to ventricular shunting.

Several reports about the cranial computed tomography (CT) findings of this malformation have appeared in the literature. With the use of magnetic resonance imaging (MRI), some new features of the malformation, such as increased mamillopontine distance, have been reported[2].

In this article, we analyzed the CT findings in 28 patients with Chiari II malformation in the light of CT and the recently defined MRI characteristics.

Material and Methods

Twenty-eight patients with Chiari II malformation were evaluated. There were 15 males (53.6%), and 13 females (46.4%). The youngest of the patients was ten-days-old, and the oldest was 44-years-old, with the mean age being 7.8 years. The age distribution of the patients was as follows: 12 (42.9%) were less than

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one-year-old, six (21.4%) were 1-5 years-old, three (10.7%) were 6-10 years-old, four (14.3%) were 11-20 years old, and three (10.7%) were more than 21-years-old. In seven patients CT examinations were performed after a ventricular shunt was placed. Two patients had CT examinations both before and after shunting.

All the CT examinations were performed with the third generation Philips Tomoscan-350 scanners in the Radiology Department of the Hacettepe University Children's Hospital. All of the patients had CT scans in the transaxial plane parallel to the orbitomeatal line. Three patients had additional direct coronal and direct sagittal scans taken.

Results

The results of the study are categorized and summarized in Table I.

TABLE I: Computed Tomographic Abnormalities in Chiari II Malformation

Abnormalities grouped by location	Number of patients (Total 28)	Percentage (%)
Cranial bones		
Asymmetry of the cranial bones	16	57.1
Sagittally enlarged foramen magnum	23	82.1
Craniolacunia	18	64.3
Petrous/Clival scalloping	27	96.4
Rhombencephalon		
Fourth ventricle		
Position		
Low	15	53.6
Normal	10	35.7
Size		
Not seen	3	10.7
Small	22	78.6
Normal	3	10.7
Cerebellum		
Enveloping	22	78.6
Upward displacement	22	78.6
Orientation of vermian folias		
Not seen	13	46.4
Transversally	4	14.3
Oblique posteromedially/Sagittally	11	39.3
Mesencephalon		
Tectum		
Pointed	22	78.6
Flattened	6	21.4
Diencephalon		
Third ventricle		
Size		
Normal	6	21.4
Enlarged	22	78.6
Telencephalon		
Lateral ventricles	15	53.6
Normal		
Enlarged		
Asymmetry	2	7.1
Corpus callosum	26	92.9
Partial agenesis	24	85.7
Supracerebellar CSF containing space		
	9	32.1
Tentorium and Falx	8	28.6
Wide tentorial hiatus		
Falx hypoplasia/Interdigitation of gyri		
	17	60.7
	13	46.4

Discussion

The following is a discussion of our findings from 28 cases grouped according to location of abnormality:

Cranial bones

Enlarged foramen magnum: The reports about the size of the foramen magnum in cases with Chiari II malformation are controversial. By using conventional radiologic techniques, Kruyff and Seffs[5] reported enlarged foramen magnum in 71 percent of cases with Chiari II malformation. According to Naidich et al[6], it is difficult to assess the size of the foramen magnum by computed tomography unless the slices are taken in the coronal or sagittal planes[6]. We found that the sagittal diameter of the foramen magnum was larger than normal (more than 40 mm) in 23 (82.1%) of our cases, with the mean value being 42.2 mm.

Scalloping of the petrous bones and clivus: Since the posterior fossa is small in Chiari II malformation, as the cerebellum and hindbrain grow there is erosion of the clivus and the posterior aspects of the petrous bones due to pressure[2,6,7]. On axial CT scans, this is seen as a loss in the normal convexity of the posterior aspects of the petrous bones (flattening), or the concavity of the posterior borders of the petrous bones and clivus (scalloping)[6]. Naidich et al[6], using CT, and Wolpert et al[7], using MRI, reported flattening or scalloping of the petrous bones and clivus in 90% and 79% of their cases, respectively. We observed petrous and/or clival flattening or scalloping in 27 (96.4%) of our cases (Fig. 1).

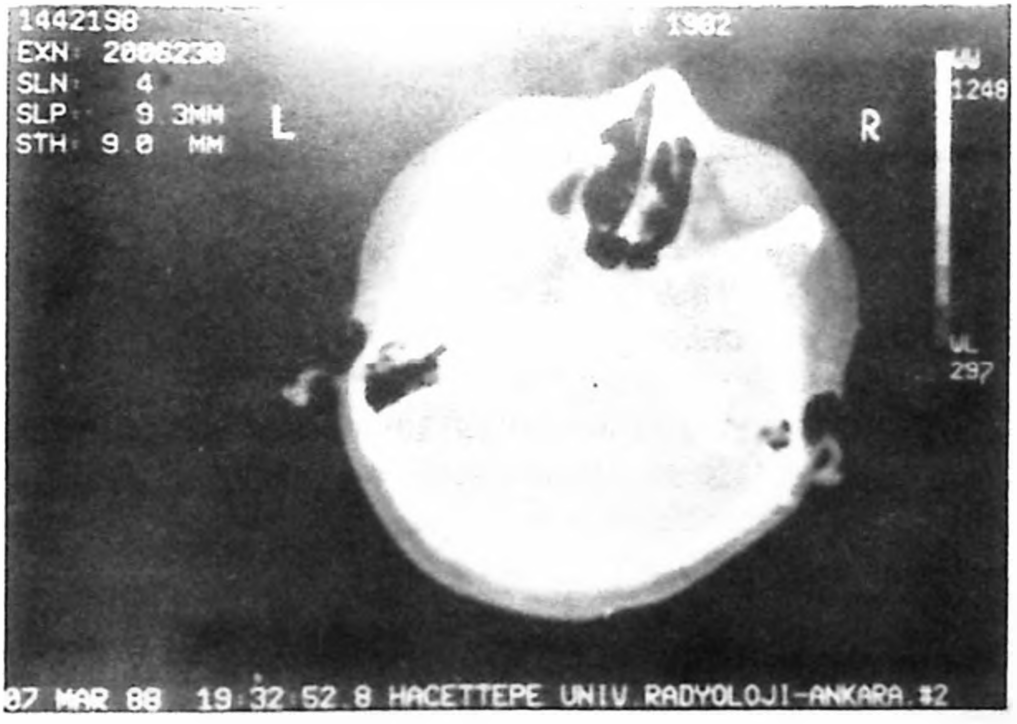

Fig. 1: Convex borders of the posterior aspects of the petrous bones and clivus are noted in this section through the posterior fossa.

Craniolacunia (Luckenschadel): Craniolacunia or lacunar skull, which is caused by calvarial mesodermal dysplasia, is seen as thinnings, pits, or fenestra predominantly involving the inner aspects of the frontal, parietal, or membranous occipital bones[6,7]. The lacunae mostly disappear after six months of age[1,6,7]. We found that craniolacunia was present in 18 (64.3%) of our cases, with seven of them being more than one-year-old (up to 18-years-old).

Asymmetry of the cranial bones: We found that the cranial bones were asymmetric in 16 (57.1%) of our patients. Asymmetric lateral ventricular dilatation was noted in all these 16 patients, and 13 of them also had craniolacunia. Asymmetry of the cranial bones due to asymmetric lateral ventricular dilatation has been previously reported[8]. We believe that the asymmetry of the cranial bones in patients with Chiari II malformation is due to asymmetric lateral ventricular dilatation, with craniolacunia being a contributing factor.

Rhombencephalon

Fourth ventricular size: Elongation and sagittal flattening of the fourth ventricle is the hallmark of the Chiari II malformation[2-4,7-10]. Naidich et al[10] have reported that the fourth ventricle was not seen in 70%, and was narrow in 25% of their cases. We found that the fourth ventricle was not seen in three of our cases (10.7%), and was small in 22 (78.6%). The fourth ventricle was of normal size in the remaining three cases. We were able to identify a small fourth ventricle in more cases than Naidich et al[10], maybe because a high resolution scanner was used on all of our patients which might cause reduced posterior fossa artefacts.

Fourth ventricular position: In the Chiari II malformation, along with the caudal displacement of the brainstem, the fourth ventricle may also be low in position[4]. According to Zimmerman et al[11], failure to demonstrate a fourth ventricle at or above the upper third level of the petrous pyramids indicates pathologic displacement. Elgammal et al[2] have reported that the fourth ventricle was stretched and low in position in 75% of their cases. The fourth ventricle was low in position in 15 of our 25 cases, in whom we managed to identify a small or normal sized fourth ventricle (Fig. 2). The caudal displacement of the fourth ventricle is better evaluated with sagittal MR sections[2,7]. The fourth ventricular size and position can also be assessed by intraoperative ultrasound, and the surgical approach is tailored to the specific abnormality for improving the safety of the procedure[12]. Three major types of fourth ventricular malformation have been identified in preoperative MRI and intraoperative ultrasound studies[7,12].

Cerebellar enveloping: Since the posterior fossa is small, the anterior parts of the cerebellar hemispheres migrate into the prepontine cistern causing envelopment of the brainstem[7,13]. Naidich et al[13], and Wolpert et al[7] reported that the cerebellar enveloping of the brainstem was present in 75% of their cases[7]. Cerebellar enveloping was identified in 22 (78.6%) of our patients (Fig. 3).

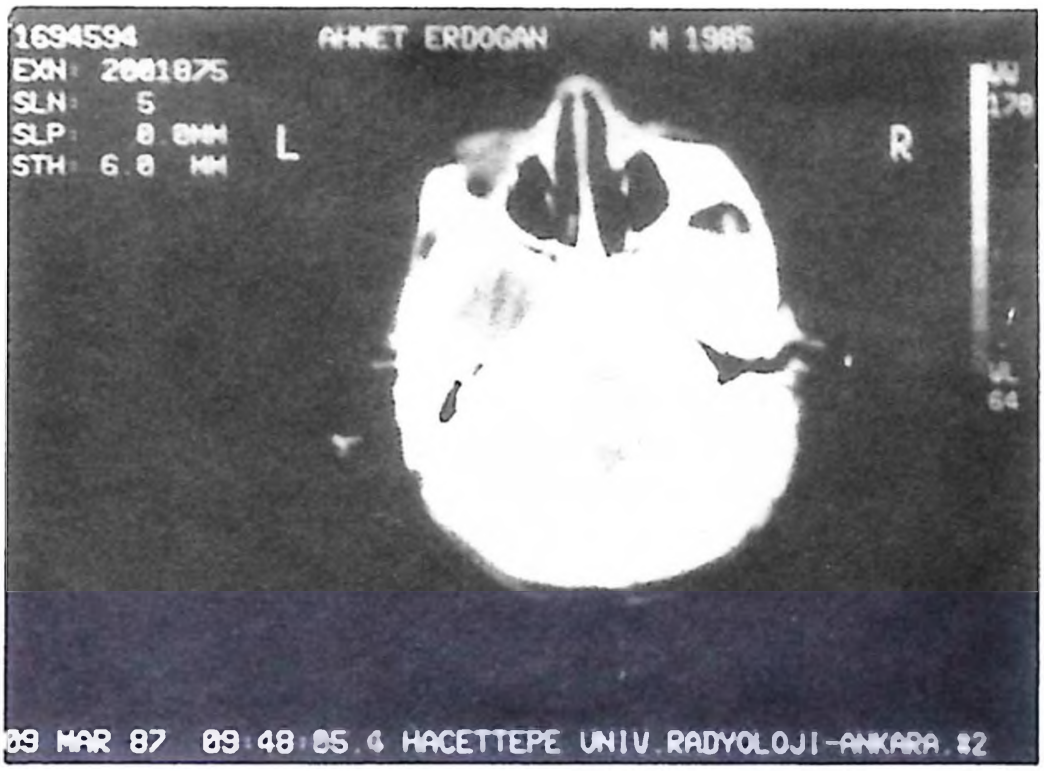

Fig. 2: A narrow fourth ventricle is seen at the level just above the foramen magnum, characteristic of Chiari II malformation.

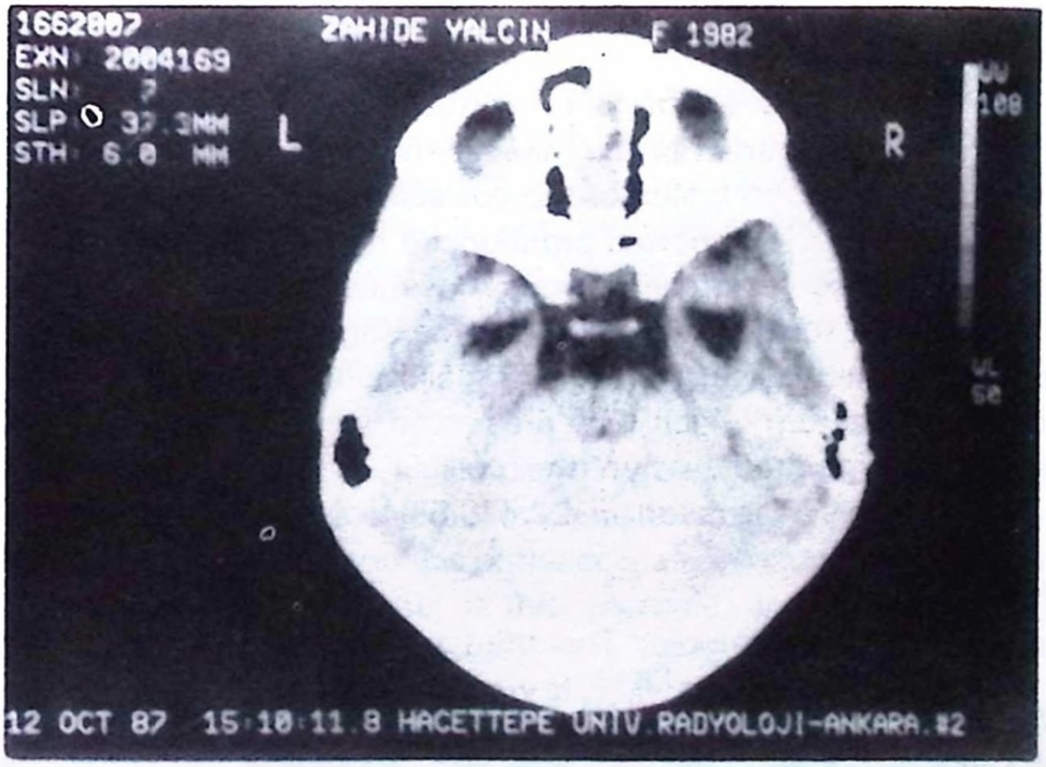

Fig. 3: The anterior cerebellar margins are sharply pointed and overlap the cerebral peduncles on both sides.

Cerebellar vermian folias: In patients with Chiari II malformation, some form of cerebellar dysplasia and disorders of migration were described by Gilbert et al[14]. Wolpert et al[7], mentioned the nonvisualization of orientation in the obliquely posteromedial direction of the vermian folias, and explained this feature by the dorsal angulation and displacement of primary and postlunate fissures. We found that the superior vermian folias were not seen in 13 (46.4%), and were directed obliquely posteramedially or sagittally in 11 (39.3%) of our cases.

Cerebellar displacement: The inability of the small posterior fossa to accommodate posterior fossa contents in Chiari II malformation causes upward herniation of the cerebellum through the wide tentorial incisura[2,7,11,13]. Zimmerman et al[11] reported the CT appearance as a "pseudotumor of the tentorium", and Naidich et al[13] described it as a "towering cerebellum". Transincisural growth of the cerebellum in 43% of Chiari II patients before shunting, and in 76% after shunting have been reported[13]. Upward cerebellar displacement was present in 22 (78.6%) of our cases, and also identified in all the seven patients having ventricular shunts. But recent MR studies indicate that these findings may be related more to the technique of the radiologic examination, than to true upward herniation of the cerebellum[2]. Caudal displacement of the medial posterior cerebrum through the wide tentorial hiatus may allow visualization of a centrally placed cerebellum and a peripherally placed supratentorial brain in axial CT scans[2,11].

Mesencephalon

Tectum: The midbrain is abnormal in virtually every patient with Chiari II malformation[13]. The anomalies of the mesencephalon range from a slight loss of the intercollicular groove, to fusion of the colliculi into an elongated beak-shaped structure[15]. Naidich et al[13], reported pronounced mesencephalic beaking in 81% of their cases. We observed an anomalous mesencephalic tectum in all of our 28 cases. In the mildest form of the anomaly, the sagittal length of the tectum is shortened and the intercollicular groove is shallow. We defined this from as the with a "flattened" tectum which was present in six (21.4%) of our cases. The tectum is termed "pointed" when the colliculi is fused into a beak-shaped structure (Fig. 4). This was seen in 22 (78.6%) of our patients.

Diencephalon

Third ventricle/massa intermedia: The third ventricle is reported to be mildly dilated in most Chiari II patients[7,8,10]. It was enlarged in 22 (78.6%), and was of normal size in (21.4%) of the patients. The massa intermedia is reported to be enlarged in 67-90% of Chiari II cases[7,10]. The enlarged massa intermedia was noted in 15 (53.6%) of our cases.

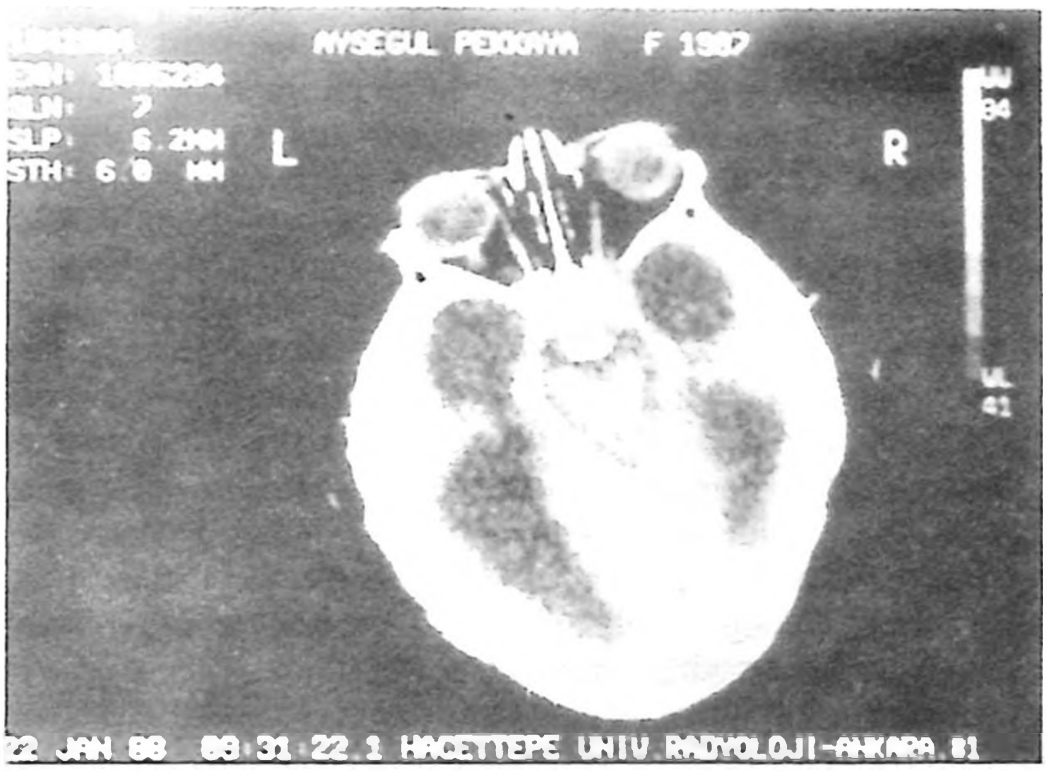

Fig. 4: In mesencephalon, fusion of the colliculi into a beak-shaped structure with posterior elongation is seen. Marked dilatation of the temporal and occipital horns of the lateral ventricles are also noted.

Telencephalon

Lateral ventricles: At necropsy, the lateral ventricles are dilated in 95% of Chiari II cases, and in most cases the dilatation is bilaterally asymmetric[16]. The atria and the occipital horns are more dilated than the frontal horns[16]. Dilatation of the lateral ventricles has been reported in 74% of cases with Chiari II malformation[10]. We observed dilated lateral ventricles in 26 (92.9%) of our cases, and in 24 of them the dilatation was asymmetric. Though the absence, or the fenestration of the septum pellucidum has been reported in necropsy studies, axial CT scans failed to demonstrate this anomaly in 50% of patients[10].

Corpus callosum: We have observed partial agenesis of the corpus callosum in nine (32.1%) of our 28 cases. We believe that the axial CT sections are not very reliable for assessing the dysgenesis of the corpus callosum. Sagittal MR sections better demonstrate the presence of this anomaly, and it was reported to be present in 33-83% of Chiari II cases[7,17].

Supracerebellar CSF containing spaces: A wide subarachnoid cistern above the cerebellum involving, to varying degrees, the quadrigeminal plate cistern, the cistern velum interpositum, the interhemispheric fissure, and the ambient cistern has been reported[2,7]. This may be due to parenchymal loss in the medial posterior aspects of the cerebral hemispheres resulting from pressure atrophy from

hydrocephalus[2]. A relationship between the size of the cistern and the degree of hydrocephalus has been noted. The spaces were compressed or smaller when hydrocephalus was present, and were larger when the ventricles were effectively shunted[2,7]. A large supracerebellar CSF containing space was identified in eight (28.6%) of our cases. We found that the size of the cistern was greater in five patients, in whom the lateral ventricles were compressed due to shunting. In cases with marked hydrocephalus, the cistern could not be identified.

Tentorium and Falx

Wide tentorial hiatus: Widening of the tentorium is seen in axial scans by the appearance of cerebellar tissue extending superiorly without the normal tapering caused by the cerebellar margins of the tentorium[7]. The tentorial margins may also be identified on contrast enhanced scans[6]. Peach[16] has reported the widening of the tentorial incisura, and its low insertion to the occipital bone in 95% of cases coming to necropsy. Wolpert et al[7] have reported the widening of the tentorial hiatus in 15 of their 24 cases. Widened tentorial hiatus was present in 17 (60.7%) of our cases.

Falx hypoplasia/Interdigitation of gyri: Partial absence or hypoplasia of the falx, and fenestrations were reported to be present in all Chiari II cases at necropsy[16]. Interdigitation of the gyri, which indicates falx fenestration has been significantly demonstrated by CT[6]. While Naidich et al[6] reported a 22% rate, Wolpert et al[7] reported that the interdigitation of the gyri was present in nine of their 24 cases. Gyral interdigitations were present in 13 (46.4%) of our cases (Fig. 5).

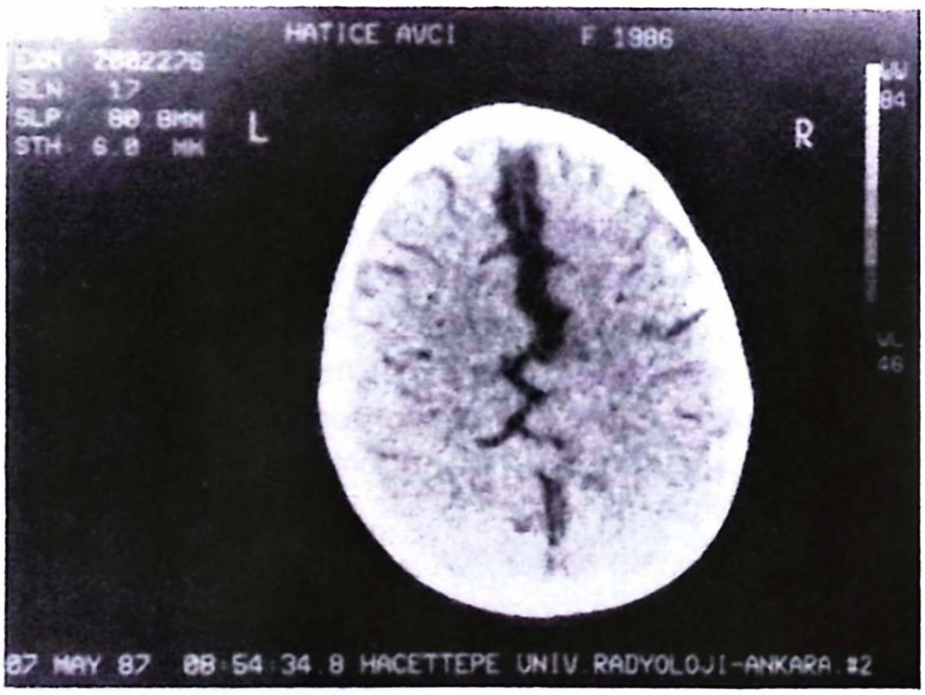

Fig. 5: The fenestration of the falx and the apposition between the gyri of the two hemispheres are noted.

Surgery

Respiratory or swallowing dysfunction associated with poor feeding and failure to thrive is considered an indication for surgery in an infant[12]. In the older child, progressive spasticity and upper extremity weakness are the most common indications for surgery[12]. The treatment of hydrocephalus in Chiari II malformation requires decompressive approaches in the suboccipital and cervical areas in addition to ventricular shunting[18]. Surgery has been designed to decompress the fourth ventricle and spinal cord promoting relief of pressure symptoms resulting from either internal expansion, or dorsal compression of the medulla and the cervical cord[12]. It is important to make a differential diagnosis between the Chiari II malformation and other types of hydrocephalus preoperatively. Although malformations have rich computed tomographic findings, none of them are pathognomonic for Chiari II[6,10,13]. Each may be seen infrequently in other patients with other conditions. But the presence of several of these anomalies together, helps to identify the patients with Chiari II malformation, thus differentiating them from other hydrocephalus cases (non-Chiari hydrocephalus)[6,10,13].

Summary

In this study cranial computed tomographic (CT) features of 28 cases with Chiari II malformation are presented. 26 of these cases had hydrocephalus (92.9%) and the fourth ventricle was either absent or small in 25 (89.3%) of the cases. Caudal displacement of the fourth ventricle was noted in 15 (53.6%) of the cases, and craniolacunia was present in 18 (64.3%) of the cases with seven of them being more than one-year-old (up to 18-years-old).

We found CT to be a safe, non-invasive and effective method of elucidating the types of abnormalities associated with this disorder. Preoperative computed tomographic evaluation of patients with Chiari II malformation helps to differentiatic it from other types of hydrocephalus, thus yielding a change in the operative approach.

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