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COMPUTERIZED TOMOGRAPHY FINDINGS OF THE POSTERIOR FOSSA IN CHILDREN: ETIOLOGY AND CLINICAL CORRELATION*

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New developments in the field of radiodiagnostic procedures have brought changing clinical concepts with them. Since the advent of techniques like computerized tomography (CT) and nuclear magnetic resonance (NMR), detailed evaluations of cranial structures have become possible and the scope of observed morphological abnormalities has enlarged. The extent to which these findings are correlated with the clinical picture is a matter of curiosity.

In this study, we reviewed, retrospectively, children who had CT abnormalities confined to the posterior fossa and evaluated their clinical and neurological findings in relation to their appearance on tomography.

Material and Methods

Of the 1105 patients, two days to seventeen years of age, who were examined by cranial CT for various reasons at the Hacettepe University Children's Hospital between June 1986 - June 1988, 93 with posterior fossa abnormalities were reviewed. Twenty-five patients whose hospital records were incomplete, and nine with postoperative tissue defects, were not included. Table I shows the CT results of the 93 cases.

The CT examinations were routinely performed parallel to the orbito-meatal line, with nine mm slice thickness from the foramen magnum to the vertex. A second

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series of CT was performed with intravenous injection of contrast medium. The CT findings were grouped as follows: neoplasms of the brain stem or cerebellum, the Arnold-Chiari malformation, cerebellar hypoplasia, cerebellar atrophy, cerebral and cerebellar atrophy, cerebellar abcess, cerebellar hemorrhage and the Dandy-Walker malformation (Figs. 1-5).

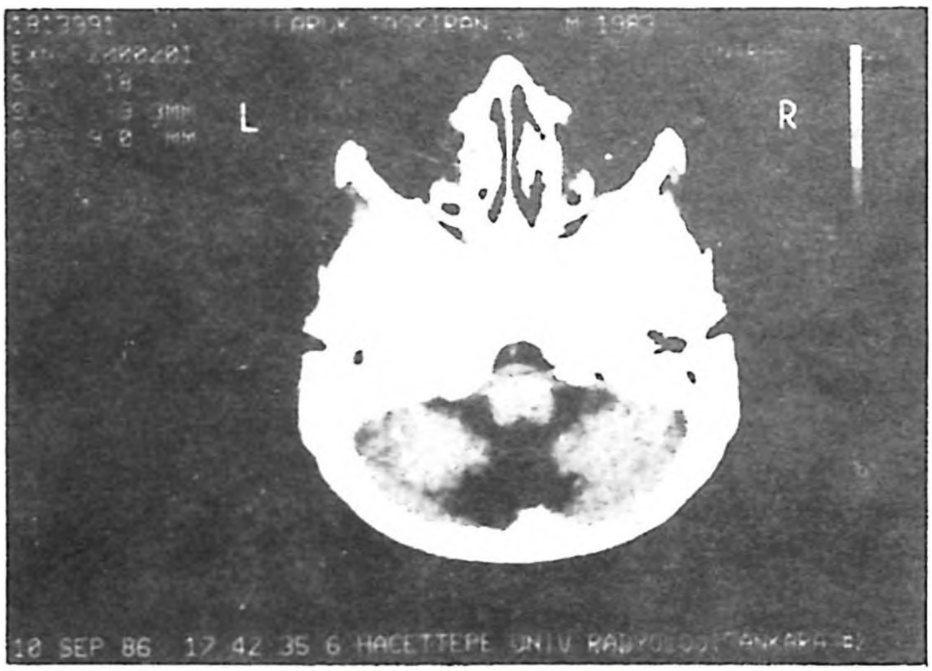

Fig. 1 (a): Cerebellar atrophy.

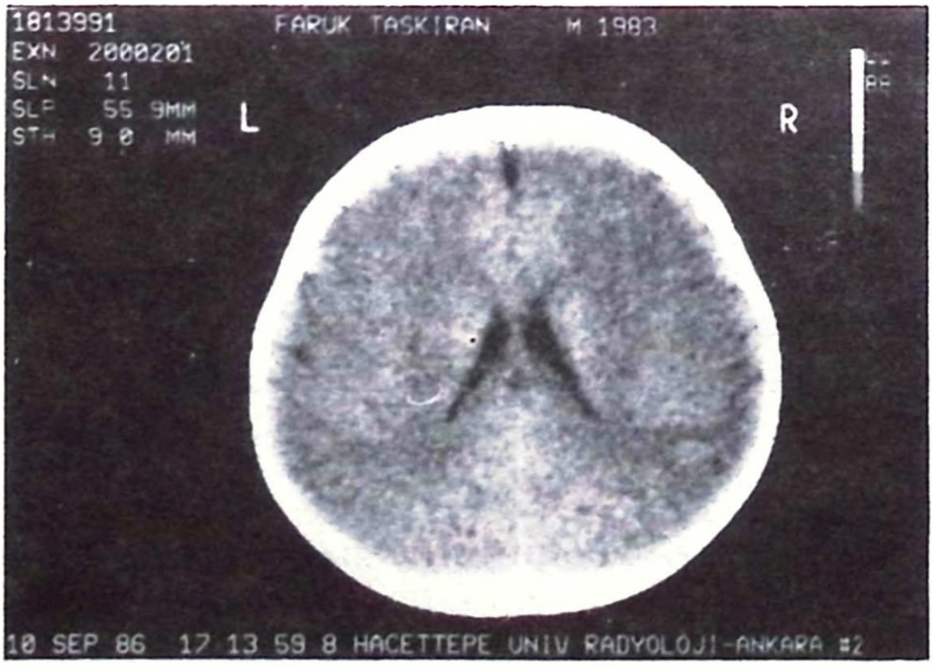

Fig. 1 (b): Compared to normal cerebellum.

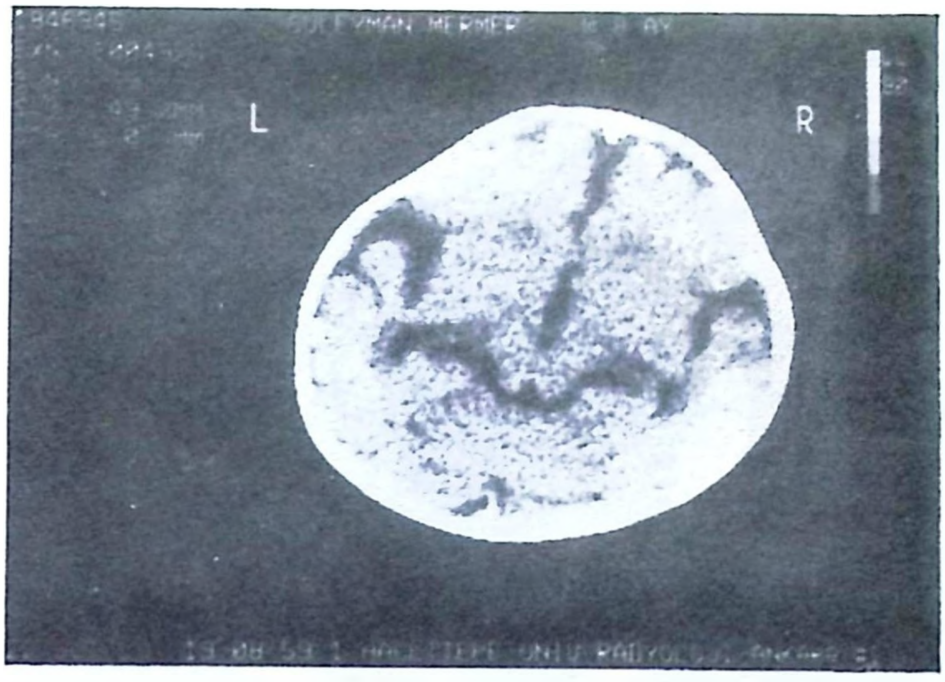

Fig. 2: Cerebral and cerebellar atrophy.

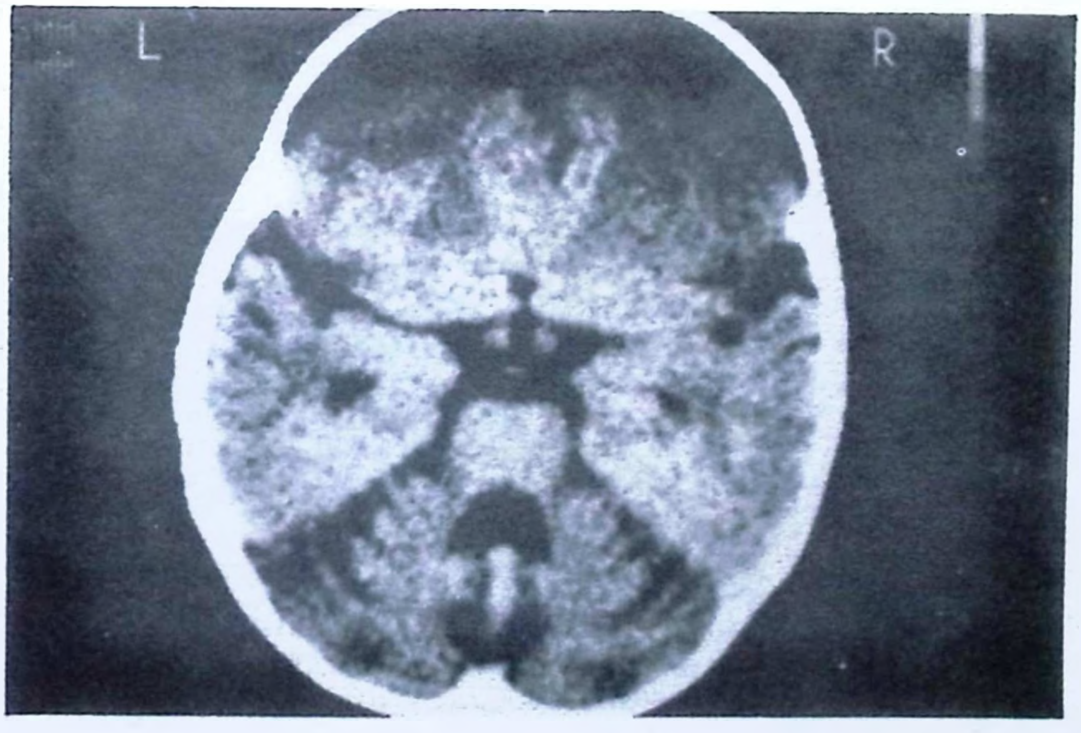

Fig. 3: Small posterior fossa and cerebellar hypoplasia.

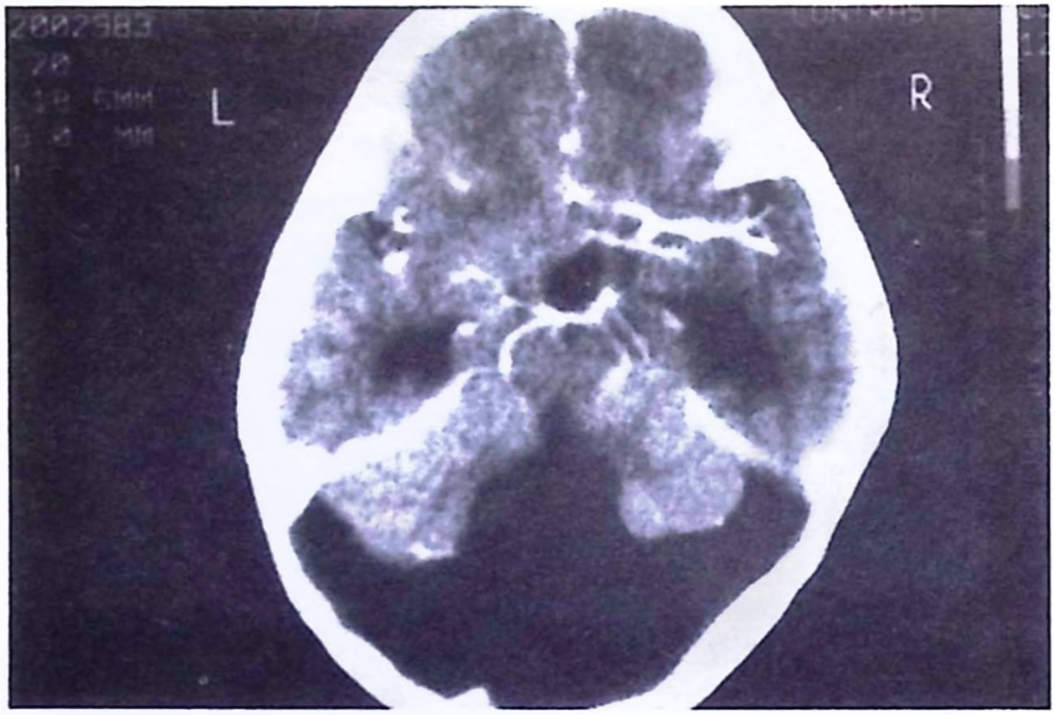

Fig. 4: Dandly-Walker malformation and partial cerebellar agenesis.

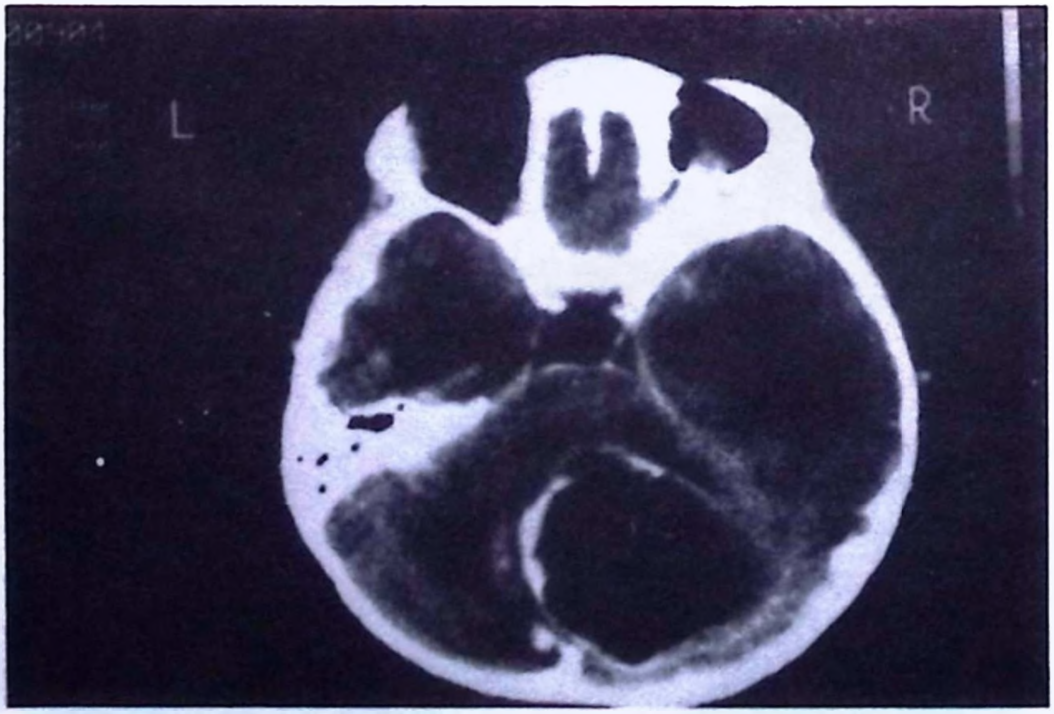

Fig. 5: Cystic astrocytoma in the right cerebellar hemisphere (left contrast substance injection).

TABLE I: CT Findings Related to the Posterior Fossa

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–Tumors	21
–brain stem-15	
–cerebellum 6	
–Cerebellar atrophy	13
–Cerebellar hypoplasia	6
–Cerebellar and cerebral atrophy	30
–Cerebellar abcess	1
–Cerebellar hemorrhage	2
–Arnold-Chiari malformation	20

A diagnosis of cerebellar atrophy was based on the visualization of the cerebellar sulci and the enlargement of the cerebellopontine and superior cerebellar cisterns. The term hypoplasia was used by the radiology department if the above findings were accompanied by other malformations on CT or if the clinical course was a non-progressive one starting from birth.

Symptoms, physical findings, developmental status and neurological signs of the patients were noted from hospital records. The patients were considered positive for cerebellar symptoms and signs if at least one of the following features was present: intentional tremor, ataxia, nystagmus, dysarthria, in addition to others (dysdiadochokinesia, etc.).

The clinical diagnoses and, if present, the suspected or definite cause for the patient's clinical condition was also recorded.

Results

The frequency of different CT findings, cerebellar and neurological findings and developmental status of the patients in each group are illustrated in Table II.

The radiological and clinical diagnoses were similar in 95% (20/21) of the tumor group. The CT results were considered unexpected if a discrepancy was present between the CT finding and the primary diagnosis or the reason for performing CT (i.e., if atrophy was observed in a patient investigated for malignant infiltration or cerebellar hypoplasia was found in a child with convulsions). The unexpected findings comprised 70% (30/43) cerebral and cerebellar atrophies, either alone or accompained by cerebral atrophy.

Cerebellar signs were present in 6/6 of the cerebellar tumors, 4/6 of the cases with cerebellar hypoplasia and 7/13 of the cases with cerebellar atrophy; they

TABLE II: Clinical and CT Findings

	Concordance with clinical diagnosis	Cerebellar signs	Other neurological signs	Delayed development
– Tumors				
– Brain stem	15/15	8/15	15/15	–
– Cerebellum	5/6	6/6	2/6	–
– Cerebellar atrophy hypoplasia	10/13	7/13	4(13	9/13
– Cerebellar hypoplasia	4/6	4/6	5/6	5/6
– Cerebral and cerebellar atrophy	3/30	3/30	26/30	19/30
– Cerebellar hemorrhage	1/2	0/2	2/2	–
– Cerebellar abcess	1/1	1/1	1/1	–
– Arnold-Chiari malformation	19/20	–	20/20	20/20

were less frequent in the group with cerebral and cerebellar atrophy (3/30; 10%). The later group presented with other neurological signs (26/30; 86%).

The frequency of delayed development was similar in cerebellar hypoplasia (5/6) or atrophies (9/13) and in cerebral and cerebellar atrophies (19/30).

The conditions leading to or accompanying the CT results are illustrated in Table III. No etiologic relationship between the CT finding and the patient's clinical condition was found in the two cases with hypoplasia, four with cerebellar atrophy and the seven with cerebellar and cerebral atrophy.

TABLE III: Etiology or Associated Condition

Hypoplasia	Cerebellar atrophy	Cerebral and cerebellar atrophy
Dwarfism (1)	Post infection (2)	Postinfection (2)
Arrested hydrocephaly (1)	Diphenylhydantoin (2)	Diphenylhydantoin (5)
Multiple congenital malformation (2)	Radiotherapy (1)	Degenerative (2)
	Ataxia telangiectasia (2)	Birth anoxia (2)
	Spinocerebellar (1) degeneration	Intrauterine infection (2)
	Birth anoxia (1)	Collagen disorder (3)
		Leukemia/lymphoma (3)
		Hepatic coma (1)
		Cyanotic heart disease (1)
		After pseudotumor (1)
		Mitochondrial myopathy (1)
		Unknown (7)

Discussion

Among the new, sometimes incidental findings which are being observed with the introduction of CT, those concerning the posterior fossa attracted our interest by their frequency (84/100 scans), and this observation became the starting point of our study. Similar studies have been done previously with particular emphasis on the enlargements of the cisterna magna[1,2] and the fourth ventricle[3]. It has been reported that solitary enlargements of these structures are not reliable predictors of clinical involvement because of the wide variation in size occurring in normal people, and that the mega cisterna magna and an enlarged 4th ventricle have a constant ratio on routine CT material. For this reason, we did not record these abnormalities when they were isolated but considered them as an indication of the other signs of cerebellar atrophy or hypoplasia.

The visualization of cerebellar sulci and the enlargement of the cerebellopontine and superior cerebellar cisterns may be due to cerebellar atrophy or hypoplasia. A definite discrimination between these diagnoses may be difficult on radiological

findings only. We used the term hypoplasia when other congenital anomalies (arachnoid cysts, hydrocephalus) were present or if the clinical course was consistent with a congenital malformation.

In contrast to the tumor cases, not all of the patients with cerebellar atrophy on CT had clinical signs of cerebellar dysfunction: 11/19 with atrophy or hypoplasia and only 3/30 with cerebral and cerebellar atrophy had cerebellar signs or symptoms. The cerebral and cerebellar atrophy group presented frequently with other neurological findings, particularly pyramidal signs and mental retardation which might have masked the cerebellar dysfunction. Our two cases with hemorrhagic infarcts did not give cerebellar signs and were investigated for altered consciousness or convulsions. The frequency of developmental delay was high among the hypoplasia, atrophy and the Arnold-Chiari groups (Table II).

Cerebellar atrophy may be due to a variety of causes which may be difficult to distinguish. Diphenylhydantoin (DPH) has been frequently reported as an etiological factor[4-6]. Some authors have pointed out that anoxia due to convulsions may be the real cause of, or a contributor to the atrophy[7]. Koller et al[6], who evaluated patients treated with DPH having cerebellar atrophy observed no signs of cerebellar dysfunction on CT. This result which is in contrast to other studies may indicate the state of preclinical DPH-induced cerebellar degeneration. Since our seven cases with atrophy, who had received DPH, had no cerebellar signs either, we can assume that in addition to the cerebellar syndrome associated with cerebellar degeneration, DPH may cause cerebellar atrophy without any clinical findings. Prospective studies indicating the clinical and CT evaluations of DPH-treated patients are needed to show the frequency of these conditions.

Malignant disorders may cause cerebellar atrophy by several mechanisms: the remote effect of cancer, the toxicity of chemotherapeutic agents or of radiotherapy[8-10]. One of our cases had received radiotherapy for a glial tumor of the pons; the other three cases with leukemia or lymphoma had received cytosine arabinoside or cyclophosphamide. However, we believe that chronic malnutrition and anemia, as demonstrated previously, are also important factors in patients with malignancies since cerebral atrophy frequently accompanied cerebellar atrophy in our patients[11].

A specific diagnosis of atrophy could be made in two of our cases with ataxia-telangiectasia, two with postinfectious cerebellar ataxia and in one with spinocerebellar degeneration. Cerebellar hypoplasia has been reported as a nonprogressive, isolated malformation of the brain of autosomal recessive or dominant inheritance or as a feature of some degenerative disorders of early onset[12,13]. Sarnat and Alcala[14] have reported that cerebellar hypoplasia is a clinical syndrome due to fetal exposure to some viruses, toxins, or radiation. In

most of our cases, other congenital malformations (micrognathia, clubfoot, etc.) accompanied the cerebellar hypoplasia.

The patients with cerebral and cerebellar atrophy were a heterogenous group presenting with many different underlying conditions (Table III).

In conclusion, cerebellar abnormalities have an incidence of 84 per 1000 CT scans in our hospital, and the atrophies are the most frequent (45%). Signs of cerebellar dysfunction are more likely to be present if cerebellar atrophy is the only finding; they are infrequent (10%) in patients with cerebral and cerebellar atrophy. A specific cause can be found in cases with cerebellar atrophy (9/13, 69%) or hypoplasia (4/6, 66%) while systemic disorders (8/30, 26%) or unknown causes (8/30, 23%) are responsible for most of the more diffuse atrophies involving both the cerebrum and cerebellum. In this situation, CT is a useful method in detecting the injury. The cerebellum, as the cerebrum, appears to be prone to injury resulting from a heterogenous group of disorders. These disorders frequently give no signs indicating cerebellar involvement. In addition, CT is particularly indicated in children showing one or more cerebellar signs or symptoms since it usually reveals important diagnostic information of these patients.

Summary

Of 1105 childhood cases who were evaluated by computerized tomography (CT) in a two-year interval, 93 who had posterior fossa abnormalities are reviewed. The cerebellar atrophies, either alone or accompanied by cerebral atrophy, were the most common morphological diagnoses. The clinical picture, etiology, and developmental state of the cases are discussed in relation to the CT findings.

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