AJNR

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AJNR Am J Neuroradiol 1987, 8 (4) 653-656
http://www.ajnr.org/content/8/4/653

Difficulties in Diagnosing Congenital Posterior Fossa Fluid Collections After Shunting Procedures

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The differential diagnosis of retrocerebellar fluid collections in infants is important because of the prognostic implications. Usually the diagnoses are easy; however, shunting of the lateral ventricles or of the fluid collections may alter the appearance of the lesions, precluding accurate diagnosis. Under such circumstances a careful study of all the sequential radiologic studies is necessary.

The differential diagnosis of posterior fossa fluid collections such as retrocerebellar cysts, Dandy-Walker syndrome, and mega cisterna magna is important because of the different prognostic implications of each. Usually the radiologic diagnoses are easy. Recently, however, we encountered two patients in whom the radiologic appearance of the fluid collections changed after cyst shunting in the first patient and after lateral ventricular shunting in the second patient. In these two patients the diagnoses would have been difficult if the sequential CT and MR studies had not been available. These cases provide the basis for this paper.

Case Reports

Case 1

A 2-year-old girl was admitted for enlargement of her head. A CT scan showed hydrocephalus and retro- and supracerebellar fluid collections in open communication with an enlarged fourth ventricle (Figs. 1A and 1B). Since no cerebellar vermis was defined, the diagnosis of Dandy-Walker syndrome was made, although the significance of a normally positioned torcula was not appreciated at the time. Metrizamide ventriculography showed free communication between the ventricles, the enlarged fourth ventricle, and the fluid collection. A cystoperitoneal shunt was inserted. Over the ensuing months, the fluid collection and the hydrocephalus decreased. A follow-up CT scan at the age of 3 years 9 months (Fig. 1C) showed marked decrease of the fluid collection with decompression of the fourth ventricle. An MR scan obtained at age 6 (Fig. 1D) demonstrated a normal cerebellum, a collapsed fourth ventricle, and a normally positioned torcula. The diagnosis was changed from Dandy-Walker syndrome to primary atresia of the outlet foramina of the fourth ventricle. The child did well on subsequent follow-up.

Case 2

A patient was originally admitted in 1978 at the age of 6 months for increasing head size. The diagnosis of a retrocerebellar cyst and hydrocephalus was made by CT. The lateral ventricles were effectively shunted and a repeat CT scan (Figs. 2A and 2B) confirmed the diagnosis of a retrocerebellar cyst. On an air ventriculogram performed 1 month later, there was no communication between the lateral ventricles and the third ventricle, presumably due to a ventriculitis after shunt insertion. A repeat CT scan obtained 26 months after the first scan showed marked dilatation of the fourth ventricle and the appearance now resembled Dandy-Walker syndrome (Fig. 2C). Subsequently, the posterior fossa fluid collection was shunted with a good result (Fig. 2D). However, over the next few years, many revisions of

Received September 22, 1986; accepted after revision January 20, 1987.

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AJNR 8:653–656, July/August 1987
0195–6108/87/0804–0653

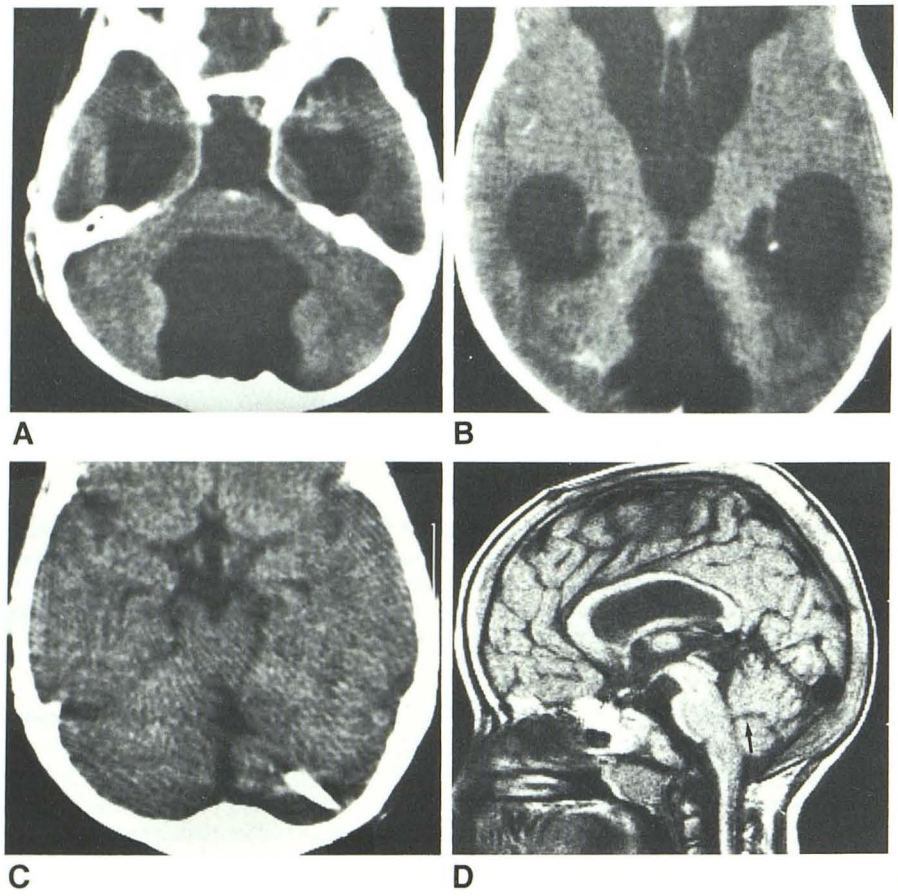

Fig. 1.—*A* and *B*, Axial CT scans show retrocerebellar fluid collection freely communicating with enlarged fourth ventricle. Note absence of a large part of vermis.

***C*, 20 months later, after effective shunting of "cyst," cerebellar hemispheres have markedly expanded.**

***D*, Sagittal MR scan (TR = 1 sec, TE = 30 msec) shows supratonsillar fissure (*arrow*) and collapsed fourth ventricle.**

both shunts were necessary. On the MR scan obtained at age 7 (Fig. 2E), the cerebellar vermis inferiorly was deformed but the fourth ventricle was normal, and a moderate-sized retrocerebellar fluid collection was seen. The torcula was elevated. The final diagnosis was that of a retrocerebellar cyst, possibly associated with some degree of the Dandy-Walker syndrome because of the deformity of the inferior vermis. The child did well on subsequent follow-up.

Discussion

The prognosis in patients with retrocerebellar cyst is usually better than that in patients with Dandy-Walker syndrome [1, 2]. This is probably because of the associated cerebral anomalies such as agenesis of the corpus callosum found with Dandy-Walker syndrome. Patients with cerebellar foramina atresia have been described as having posterior fossa dysfunction, which can remain even after surgical treatment [3, 4]. However, the second patient in our series performed well and had no neurologic sequelae after effective shunting of the fluid collection. Patients with the mega cisterna magna usually have no adverse clinical sequelae and no specific treatment or surgical intervention is necessary [5]. Although both our patients are doing well, accurate diagnoses are necessary because of the usually different prognostic implications.

A retrocerebellar cyst is usually easily differentiated from Dandy-Walker syndrome by CT or MR, where the position, size, and shape of the fourth ventricle and the integrity of the cerebellar vermis are key differential indicators [1, 6]. Atresia of the fourth ventricle foramina is diagnosed if the fourth ventricle is expanded in all directions, the vermis is stretched but has no defects, and the tentorium is not elevated [7]. Distinguishing between the different conditions may be difficult when alterations in fluid dynamics occur, due to either effective or ineffective CSF shunting procedures, and a careful analysis of all previous studies may be necessary to reach a final diagnosis.

Case 1 is considered to be a primary cerebellar foramen atresia in which it appears that cerebellar "growth" occurred after cyst shunting. This situation is akin to that after shunting of patients with lateral ventricular hydrocephalus, where the cerebral hemispheres can increase remarkably in size through an increase in the size of the periventricular white matter and a decrease in the amount of periventricular edema [8]. Probably the "growth" is also age related and is expected to be observed at an early age when both the cerebrum and cerebellum are still growing with an increase in the amount of myelin, neurons, and glial cells [9]. In this case, the initial CT scans suggested the diagnosis of Dandy-Walker syndrome, although in retrospect the absence of torcula elevation is

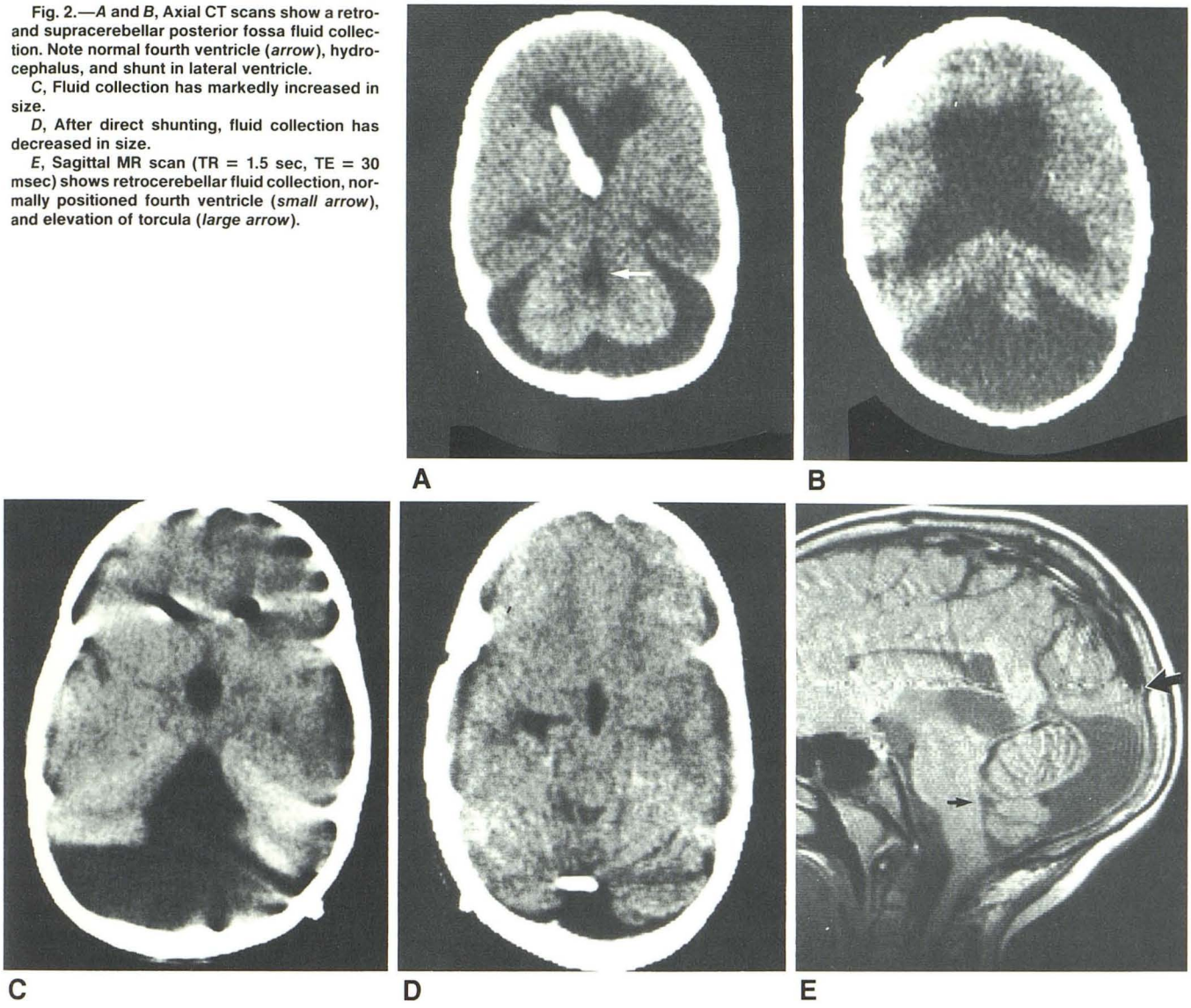

Fig. 2.—*A* and *B*, Axial CT scans show a retro- and supracerebellar posterior fossa fluid collection. Note normal fourth ventricle (*arrow*), hydrocephalus, and shunt in lateral ventricle.

***C*, Fluid collection has markedly increased in size.**

***D*, After direct shunting, fluid collection has decreased in size.**

***E*, Sagittal MR scan (TR = 1.5 sec, TE = 30 msec) shows retrocerebellar fluid collection, normally positioned fourth ventricle (*small arrow*), and elevation of torcula (*large arrow*).**

against this diagnosis. Had follow-up scans not been obtained, the diagnosis of atresia of the fourth ventricle foramina would not have been made.

In case 2, had only the scans obtained 26 months after the first scan been available, Dandy-Walker syndrome would have been diagnosed. In this case progressive expansion of the retrocerebellar cyst led to marked separation of the cerebellar tonsils. Effective therapy reduced the size of the cyst. The subsequent deformity of the inferior vermis is probably due to either pressure atrophy from the expanded fluid mass or partial agenesis of the inferior vermis as would be seen when a retrocerebellar cyst is associated with a minor degree of Dandy-Walker syndrome. This association has been discussed by Shaw and Alvord [10], who described a spectrum of changes in 15 autopsy cases of posterior fossa fluid collections ranging from typical retrocerebellar cyst to typical Dandy-Walker syndrome.

In conclusion, ventricular or cyst shunting of patients with retrocerebellar fluid collections can lead to confusing appearances on CT and MR scans. Only with the benefit of prior studies can the correct diagnosis be made.

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