ISOTOPIC CISTERNOGRAPHY IN STURGE-WEBER SYNDROME

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Isotopic cisternography using radioiodinated serum albumin (IHSA) introduced into the cerebrospinal fluid (CSF) was first described by DiChiro (*1*) in 1964. It is recognized as a valuable diagnostic technique, particularly in normal pressure hydrocephalus (*2–4*), CSF leaks (*5*), subarachnoidal blocks (*6*) and subdural hematoma (*7*).

In other reports and texts the normal pattern of CSF circulation has been described. This communication briefly presents a patient with an incomplete form of Sturge-Weber syndrome and an abnormal pattern of CSF circulation. The possible mechanism and diagnostic applications are discussed.

CASE HISTORY

An 8-month-old white male infant (Case 800-00-4124; RB) was admitted to Harrisburg Hospital on July 8, 1969, for further evaluation of a seizure disorder. Past history revealed that the patient had his first seizure at approximately 6 months of age, associated with fever. A second seizure occurred shortly before admission. There was no family history of seizures.

Physical examination revealed a reddish rash similar to pigmented nevi involving the left side of the face and a portion of the right side of the head. This appeared to be a port wine stain (Fig. 1). No other unusual physical findings were noted.

Spinal tap was done and was reported as normal. Routine laboratory studies were all within normal limits. The skull x-ray was normal, and EEG revealed a moderate voltage tracing with variable slow background consisting of theta and delta activity. The background was considerably better defined over the right hemisphere than the left where the voltage was practically flat. The infant was treated with phenobarbital and had no seizures during his hospitalization.

The possibility of a subdural hematoma was of great clinical concern because of an abnormal brain scan done with 3 mCi ^{99m}Tc-pertechnetate (Fig. 2). Since cisternography might demonstrate the presence of a subdural hematoma versus abnormal vasculature, a study was performed.

The patient received 15 μCi ^{131}I-serum albumin containing 0.2 mg albumin by lumbar puncture. Six hours later anterior and left lateral scans of the head were performed on the rectilinear scanner (Fig. 3). At 24-hr anterior, posterior and left lateral projection scans were performed (Fig. 4).

DISCUSSION

In normal subjects, the pattern of IHSA flow in CSF circulation is assumed to be that of normal

Received Dec. 3, 1969; original accepted Feb. 5, 1970.

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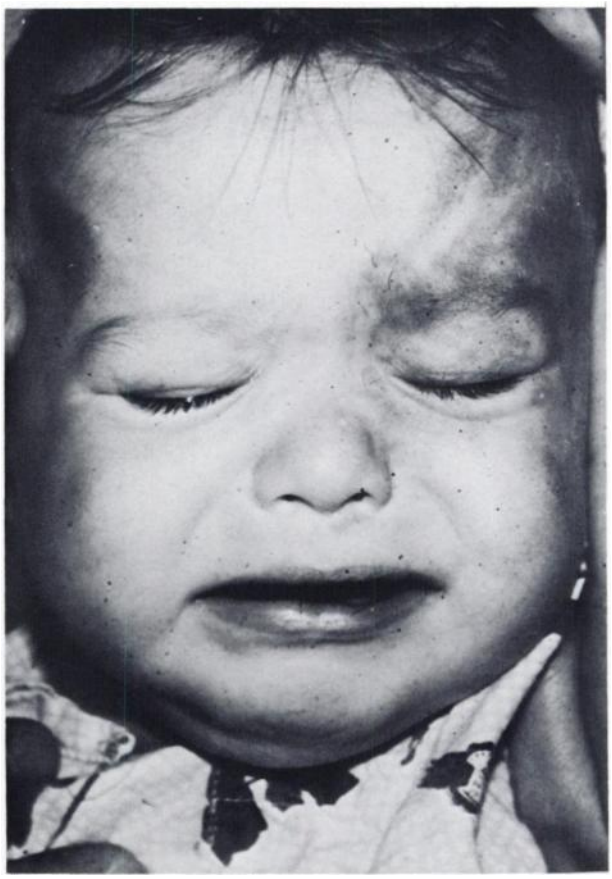

FIG. 1. Patient's face showing hemangiomas involving left forehead and upper eyelid.

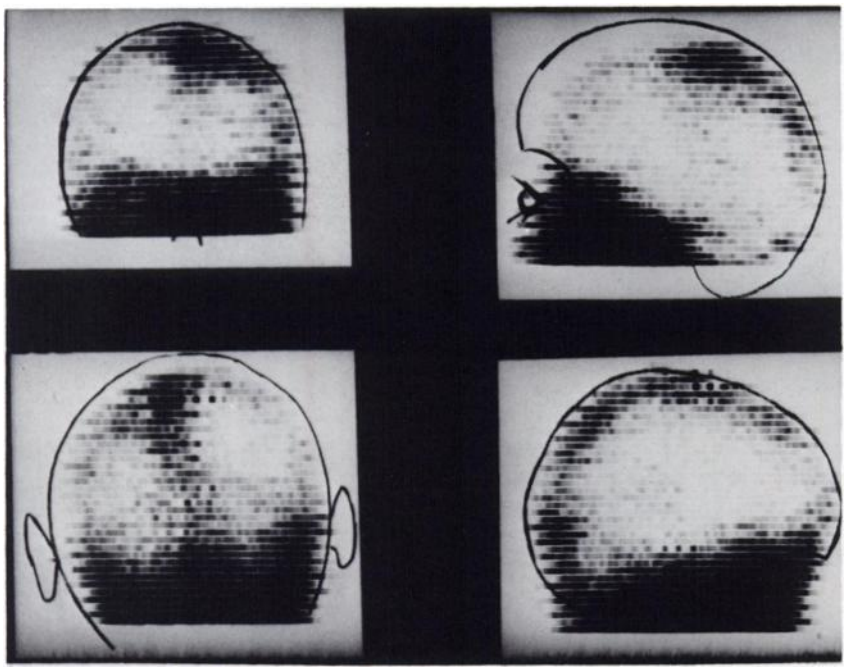

FIG. 2. Brain scan taken 1 hr after injection of 3 mCi ^{99m}Tc. Abnormality at left parietal cortex is demonstrated in anterior (upper left), left lateral (upper right) and posterior (lower left) views. Suggestive increase of radioactivity is seen in occipital lobe area in right lateral view (lower right).

CSF albumin. In our normal studies results similar to those of Tator *et al* (*2*) and others have been demonstrated. The IHSA appears in the subarachnoidal spaces of the head in approximately 1 hr. Symmetrical concentration of radioactivity in basal cisterns and the bilateral sylvian cisterns is seen at 3 hr. At the end of 24 hr most of the radioactivity is located in the parasagittal area over the hemispheres. Small concentrations remain in the basal cisterns and suprasella cistern.

In our patient a marked increase of concentration of radioactivity in the left sylvian cistern as well as over the left hemisphere and left parasagittal area at 6 hr (Fig. 3) was demonstrated. At the end of 24 hr an abnormally high concentration of radioactivity is distributed over the left hemisphere and the left parasagittal area (Fig. 4). These suggest an increased rate of CSF absorption and/or circulation in that region.

The exact mechanism of increased absorption and/or rapid circulation of CSF in this patient with Sturge-Weber syndrome is not known. However, this abnormal study suggests that the basic mechanism is related to the pathophysiology of the disease. It

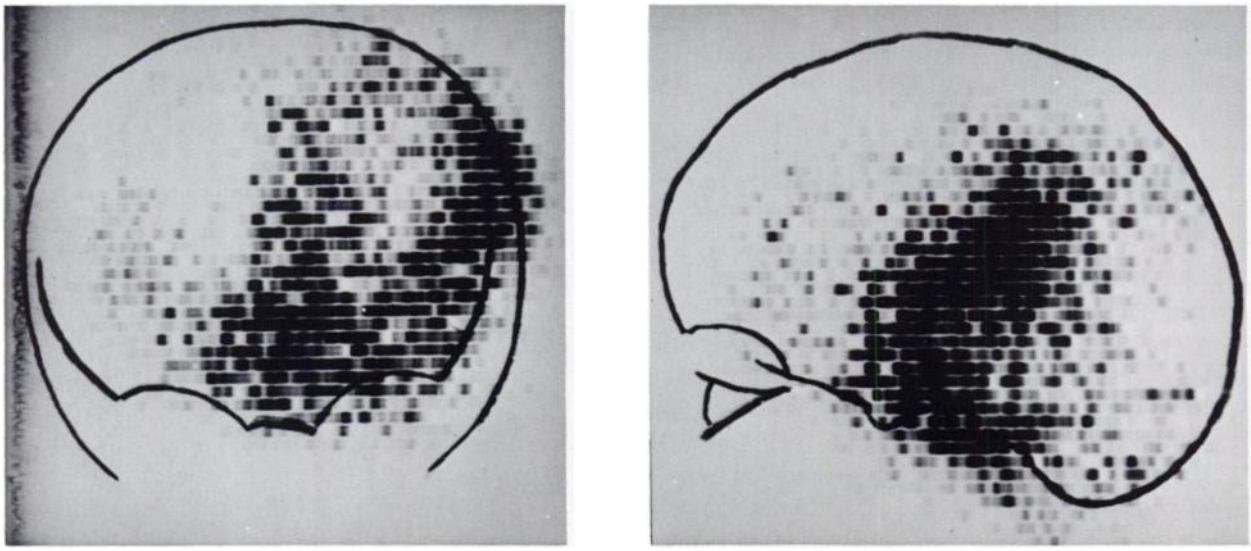

FIG. 3. Cisternography at 6 hr reveals asymmetric circulation with increase over left parietal cortex. Anterior view on left and left lateral view on right.

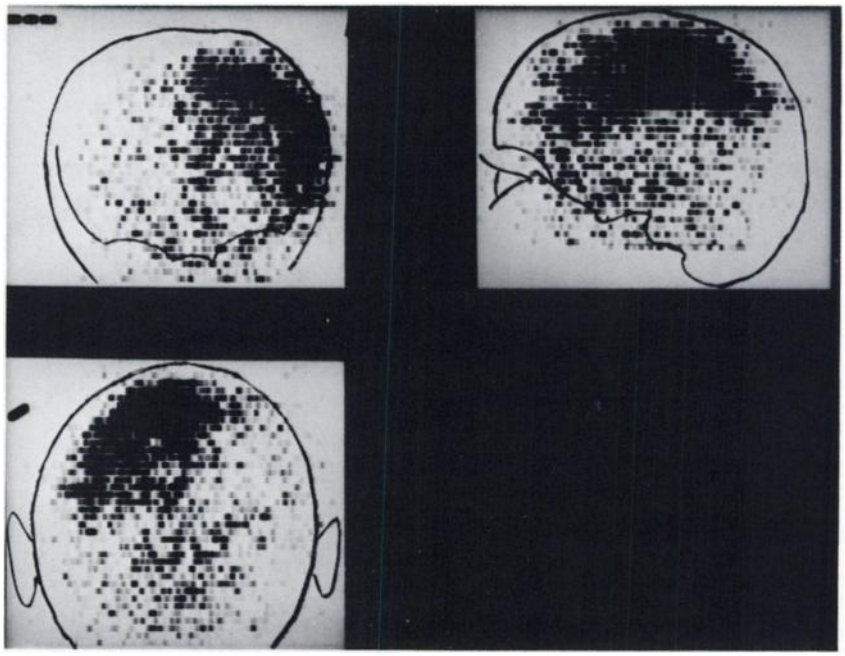

FIG. 4. Cisternography at 24 hr reveals clearing of basal cisterns and localization over left parietal cortex. In posterior view faint concentration to right of midline may correspond to area noted in brain scan.

is well known that the brain pathology of Sturge-Weber syndrome is a capillary venous hemangiomatosis involving the piarachnoid of the parieto-occipital region (*8*). It is suggested that the vascular anomalies produce changes in CSF circulation. It is quite possible that abnormal venous hemangiomatosis of the leptomeninges area increase the rate of CSF absorption, probably due to increased vascularity of absorption area. The brain in Sturge-Weber syndrome is also different from normal (*9*). The anoxic brain may clear CSF fluid at a slow rate.

Unlike other central nervous system disorders the diagnosis of Sturge-Weber syndrome is not difficult in the presence of classical symptoms and signs: (1) vascular nevi (port wine stain) on the face, (2) seizures and (3) typical "tram-line" intracranial calcification (*10*).

With atypical or incomplete forms, especially without typical intracranial calcification or neurologic manifestation, the diagnosis may be difficult. At times the clinician may be faced with the need to differentiate it from other intracranial diseases, such as subdural hematoma. As in this instance, isotopic cisternography may be helpful. The usual cisternographic abnormality reported with a subdural hematoma is an asymmetric decrease of radioactivity on the same side as the increased radioactivity on the brain scan. The cisternographic demonstration of an increase in radioactivity on the side of the lesion conclusively excludes the presence of subdural hematoma. No previous report of this observation in Sturge-Weber syndrome has been reported.

SUMMARY

This case report suggests that cisternography may be a useful adjunct in the diagnosis of atypical Sturge-Weber syndrome.

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