Xanthogranulomas of the choroid plexus in a neuro-epileptic child

Case report

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Bilateral xanthogranulomas arising from both choroid plexuses were found in a 6-year-old child with epileptic seizures and psychomotor retardation. Both masses were surgically removed without adding any neurological deficits. The xanthogranulomas were fairly large, and the posterior part of both lateral ventricles were slightly dilated, but clinical symptoms and the location of the xanthogranulomas could not be clearly correlated in this case.

KEY WORDS • xanthogranuloma • choroid plexus • lateral ventricle

The first case of xanthogranuloma of the choroid plexus in humans was recorded by Blumer in 1900, although he designated it as a cholesteatomatous endothelioma. Since then, there have been several case reports and reviews of this entity. Choroid plexus xanthogranulomas are frequently encountered at postmortem examination, but they are usually asymptomatic and very few cases have been reported with surgical intervention. We are reporting an additional clinical case, with discussion of possible preoperative diagnosis in order to avoid undertaking inappropriate surgery in this benign condition.

Case Report

This 6-year-old girl was brought to the emergency service of this hospital on August 18, 1976, in a state of unconsciousness due to continuous convulsions of 2 hours’ duration. Endotracheal intubation, intravenous injection of diazepam in a total dose of 30 mg, and intrarectal administration of 30 ml of 5% chloral hydrate, succeeded in stopping the seizures. Her history showed some degree of developmental mental and language retardation; “minimal cerebral dysfunction” had been suspected by a pediatrician when she was 3 years old. Two months before admission, she had had an initial attack of a prolonged generalized convulsion which continued for 1 hour.

Examination. After complete recovery from the postictal amnestic state, moderate retardation of language for her age, inconsistency of handedness and footedness, and latent strabismus were observed. Her IQ estimation by the Tanaka-Binet scale was 72. The remainder of the neurological and physical examination was unremarkable. There were no signs suggesting Hand-Schüller-Christian disease. Laboratory data...
FIG. 1. Tomographic air study showing bilateral intraventricular masses in the ventricular trigones. Upper Left: Coronal pneumotomogram of the ventricles. Upper Right: Lateral pneumotomogram of the right lateral ventricle. Lower Right: Lateral pneumotomogram of the left lateral ventricle.

were all within the normal range, total cholesterol in the blood was 117 mg/dl; cholesterol esters, 85 mg/dl; β-lipoproteins, 218 mg/dl; and triglycerides, 103 mg/dl. Lumbar puncture revealed an opening pressure of 150 mm H₂O, with clear cerebrospinal fluid (CSF). Protein was 30 mg/dl, glucose was 60 mg/dl, and there were three mononuclear cells per cu mm. Cytologic study of the CSF did not reveal any abnormal cells.

Electroencephalography showed 4 to 5 Hz slow wave activity, most prominent from the right parietooccipital area, without definite spikes. Skull films showed a spherical area of punctate calcification, measuring approximately 2 cm in diameter, in the area of the right ventricular trigone. Very faint calcification was also visible in the area of the left trigone. Pneumoencephalography revealed a mass arising from the floor of each ventricular trigone, which protruded into the ventricle. The left mass was larger and almost fully occupied the ventricular trigone, and the posterior parts of both lateral ventricles were slightly dilated, likely due to an intermittent or partial obstruction of flow of CSF. On the lateral and coronal pneumoencephalotomograms, the right mass measured $2.0 \times 2.2 \times 3.4$ cm, and the left mass measured $2.5 \times 3.0 \times 3.5$ cm (Fig. 1). Selective catheter angiographic study was carried out through the femoral route. A few feeder vessels arising from the anterior and posterior choroidal arteries were visualized, but the masses were not stained angiographically. Technetium pertechnetate gamma-encephalography demonstrated a weak accumulation in the right ventricular mass, but the left mass was not stained, which was thought to mitigate against the diagnosis of choroid plexus papilloma or ependymoma. A computerized tomographic (CT) scan confirmed nearly symmetrical masses with punctate calcification and central low density located in both ventricular trigones. Intravenous administration of 100 ml of 30% Conray showed a marginal enhancement in the right mass and less marked, irregular marginal enhancement in the left mass (Fig. 2). The absorption coef-
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icient in the central low-density zone was between 6 and 8, and suggested liquid content.

Operations. On September 21, a right temporal craniotomy was carried out. A corticotomy in the right middle temporal gyrus was placed and the lateral ventricle was opened. An ovoid, smooth-surfaced tumor was found in the trigone. The tumor was attached to the choroid glomus and small arteries and veins from the choroid plexus supplied the mass. There were no adhesions, nor was there invasion of the ventricular wall. The tumor was removed intact with a small part of the choroid plexus (Fig. 3). On October 5, a left temporal craniotomy was performed. Through a middle temporal corticotomy, a similar but somewhat larger tumor was found at the left ventricular trigone. The tumor was attached only to the choroid glomus, and it was removed intact without difficulty.

Postoperative Course. The clinical course following both operations was uneventful. No additional neurological deficit developed. Pneumoencephalography 1 month after the second operation did not reveal significant change in size of the ventricles.

Pathological Examination. Macroscopically, both tumors were similar in appearance. The right tumor measured 1.8 x 2.0 x 3.0 cm, and the left tumor 2.5 x 3.0 x 3.2 cm. Each had a smooth surface with moderate vascularity and was yellowish-red in color. Both tumors had a central cystic cavity which contained yellow, syrupy fluid. Cholesterol crystals were not visible in the fluid; chemical study was not done. Histologically, the tumor in each side was covered with choroid plexus epithelium. Aggregations of foamy (xanthoma) cells with small eccentric nuclei and amorphous or fine granular, eosinophilic materials were the main components of the tumor. Some foamy cells in the boundary with the amorphous substance showed degenerative changes, suggesting that the amorphous substance is the product of degeneration of foamy cells. Some foamy cells had intracellular calcified depositions, and a number of small spherical calcified bodies were found to be scattered in the amorphous substance (Fig. 4). Interlacing strands of loose fibrous tissue separated aggregations of foamy cells. Cholesterol crystals or clefts and foreign body giant cells were not found. Neither hemosiderin granules nor Gamma bodies were present. There was no leucocytic or lymphocytic infiltration. Histological diagnosis was xanthogranuloma (xanthoma) of the choroid plexus.

Discussion

It has long been recognized that xanthoma or cholesteatoma occurs commonly in the equine brain and sometimes produces a clinical syndrome in horses known as the staggers. However, the first human case was
not reported in the medical literature until 1900. During the early half of this century, this lesion in the choroid plexus was regarded as rare in the human. Several cases where xanthogranulomas of the choroid plexus were found at postmortem examination were reported."^^\textsuperscript{3,5,6,8,12} Most early authors thought these tumefactions in the choroid plexus were related to hypercholesteremia, atherosclerosis, and/or diabetes. Various designations, such as plexus cholesteatoma, cholesteatomatous endotheliomata, cholesterolinose, cholesteatoma vasculosa, xanthoma, cholesterol granuloma, and xanthogranuloma, were used by various authors, and confusion in terminology developed. Shuangshoti and Netsky,"^^\textsuperscript{10} for several reasons, recommended the use of the term “xanthogranuloma” for this tumefaction in the choroid plexus.

In 1950, Wolf, "^^\textit{et al.},"^^\textsuperscript{13} found 20 cases of choroid plexus xanthogranulomas in 1181 consecutive autopsies. They concluded that such lesions are not infrequent in humans and that the lesions are not related to hypercholesteremia, atherosclerosis, or diabetes. The paucity of clinical reports of choroid plexus xanthogranulomas is due to the fact that most of the tumors are too small to produce clinical symptoms. The average size of choroid plexus xanthogranulomas in Wolf’s series was 2 to 4 mm, and the average size of xanthomas and cholesterol granulomas in Ayres and Haymaker’s series"^^\textsuperscript{1} based on materials in the Armed Forces Institute of Pathology were 4 mm and 12 mm, respectively. In 1957, Rosner"^^\textsuperscript{a} reported the first case of xanthogranuloma that developed clinical manifestations of a tumor and was successfully removed by surgery. Morello, "^^\textit{et al.},"^^\textsuperscript{7} reported the surgical removal of fairly large xanthogranulomas from both choroid plexuses in an 8-year-old boy with Hand-Schüller-Christian disease. Jaer, "^^\textit{et al.},"^^\textsuperscript{4} described an 18-year-old girl in whom xanthogranuloma arising from the foramen of Monro produced ventricular obstruction and hydrocephalus. Shuangshoti, "^^\textit{et al.},"^^\textsuperscript{11} described a case of combined colloid cyst and xanthogranuloma in the third ventricle. Xanthogranulomas in these cases were also successfully removed by surgery.

Xanthogranuloma of the choroid plexus is a non-neoplastic lesion that has been an attractive target of pathological investigation, but its etiology and pathogenesis are still somewhat obscure and its natural history is unknown. It is an interesting fact that all of the reported cases with clinical symptoms and surgical intervention were in the pediatric and adolescent age group, whereas xanthogranulomas found incidentally at autopsy were all in an older age group. In our case, bilateral xanthogranulomas were perhaps incidentally found in clinical examination of an epileptic child. Both tumefactions were fairly large and the posterior parts of both lateral ventricles were slightly dilated, but it was impossible to correlate with certainty the clinical symptoms of prolonged convulsions and developmental retardation of mentation and language with the existence of xanthogranulomas in the choroid plexus.
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We could not make the diagnosis of xanthogranuloma before operation and histological examination in this case. However, with the use of modern techniques, especially the CT scan, preoperative diagnosis of xanthogranuloma may be possible even in cases without symptoms. The characteristic location in the lateral ventricles, frequent bilaterality of lesions, punctate calcification on x-ray films, poor accumulation of radioactivity on gamma-encephalogram, and little vascularity on angiogram are diagnostic clues that suggest xanthogranuloma.

In this case, surgery was undertaken because lateral ventricles were believed to be enlarged posteriorly, suggesting hydrocephalus. Whether this was the case is uncertain since postoperative pneumoencephalography did not show conclusively a change in ventricular size. In the case of silent xanthogranuloma, justification for surgical intervention is controversial. When a convincing diagnosis of xanthogranuloma is made, this benign lesion should be left intact unless it is behaving as a mass or interfering with the flow of CSF.

References

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