Cystic intracranial teratoma in an infant
Case report

RALPH A. W. LEHMAN, M.D. AND EMILIO TORRES-REYES, M.D.
Division of Neurological Surgery, and Mallinckrodt Institute of Radiology, Washington University School of Medicine, St. Louis, Missouri

Patients with intracranial tumors symptomatric within the first 2 months of life are seldom seen and rarely diagnosed before death.1,5 In only one case of such a tumor was a localizing contrast study performed.5 Two other cases were recognized by the presence of calcification on roentgenograms.2,5 All three of these patients were operated on. Only one survived beyond the first week following surgery. In that case, the operative procedure was limited to the evacuation of multiloculated cysts and biopsy of the cyst wall of a presumed teratoma.5

This report describes a hydrocephalic infant with a large calcified intracranial teratoma overlying the planum sphenoidale. Ventriculography, angiography, and surgical removal were performed. It is believed this is the first case in which a patient less than 3 months of age has been reported to survive the resection of the bulk of a verified intracranial tumor.

Case Report

An infant girl was born on December 4, 1968, at estimated full term. Prior to conception and during the first trimester of pregnancy, the mother had been taking birth control pills. In mid-April the mother was treated with sulfisoxazole. Because she developed abdominal pain and vomiting, gall bladder and upper gastrointestinal series were performed on June 18 and 19. Pregnancy was diagnosed in mid-July, at about 4 months of gestation, and oral contraceptives were discontinued.

At birth the patient weighed 6 lbs 3 oz. Head circumference was 33 cm. At 9 weeks of age, the child became irritable, vomited, no longer held her head up well, and stopped following objects with her eyes. Head circumference was 40 cm. A week later she became lethargic with irregular respirations and medial deviation of the right eye. The head circumference was now 45.5 cm.

Both parents and six brothers were in good health.

Examination. Upon admission on February 17, 1969, at age 10½ weeks, the patient's head circumference was 46.3 cm. There was slight prominence of the right frontal region. Sutures and fontanels were spread and tense. Transillumination revealed increased lucency of the left midparietal, right parietooccipital, and especially the right frontal regions. The child was lethargic and had poor head control. Pupils were equal and poorly reactive. The discs were atrophic. The eyes tended to deviate medially and would not focus. There was marked limitation of lateral gaze of the right eye and an occasional rapid jerk of the eye inferomedially. The right corneal reflex was diminished.

Hospital Course. A right subdural tap at
Cystic intracranial teratoma in an infant

the anterior rim of the fontanel released clear, colorless fluid with a protein content of 71 mg%. No fluid was obtained on the left side. Skull films revealed widened sutures, craniofacial disproportion, and a densely calcified mass chiefly on the right above the planum sphenoidale (Fig. 1 left). A liberal amount of radiolucent tissue outlined the mass, which measured $4.5 \times 4.5 \times 4.5$ cm (Fig. 1 right). There were four other separate areas of calcification. One was located 3 cm to the left of the main mass, two others were on the right at the vertex, and another was suprasellar. The tuberculum sellae was flattened. A left ventricular tap returned crystal clear, colorless fluid with a protein of 4 mg%. Air injected into the left ventricle showed poor communication with the right ventricle. The right ventricle was elevated by a suprasellar tumor of considerably larger size than the calcification. This indented the right side and the floor of the third ventricle. Air entered the spinal canal but not the intracranial cisterns. Air introduced into the right subdural space revealed a cyst. A right ventriculoperitoneal shunt was performed.

On February 25, a right carotid angiogram demonstrated that the right middle cerebral artery was displaced posteriorly and superiorly by the mass. The horizontal portion of the right anterior cerebral artery did not fill, and the pericallosal artery filled in retrograde fashion from the middle cerebral group. There was a diffuse homogeneous vascular blush in the mass of the tumor behind the calcification evident during the arterial and early venous phases (Fig. 2).

Operation. On the following day a right frontotemporal craniotomy was performed. The right frontal lobe extended only a few centimeters in front of the coronal suture. The remainder of the right anterior fossa was occupied by cysts containing clear colorless fluids of differing protein concentrations. Anteriorly in the middle fossa was a cyst containing viscous yellow fluid. Beneath the temporal lobe a separate cyst extended over the petrous ridge and behind the clivus. This contained extremely thick, mucoid, colorless fluid. On the floor of the anterior fossa lay a hard pale-white mass, firmly fixed to the planum sphenoidale, and embraced by very thin olfactory nerves. Attached to it were two small cysts of yellow fluid containing hair. Soft tumor extended posteriorly from this into the medial inferior aspect of the right frontal lobe. A plane of dissection was

![Fig. 1. Left: Anteroposterior skull film. Note punctate calcification to left of main mass as well as above it. Right: Lateral skull film. The radiolucent region surrounding the calcification is most evident in this view.](image-url)
created about the tumor up to the point where injury to the hypothalamus was feared and the mass amputated. Beneath was a large cyst of colorless, thick, mucoid fluid which was removed from the suprasellar area and from under the left frontal lobe. The right optic nerve was sacrificed during the dissection. After the mass had been removed, the left optic nerve, the optic chiasm, the third, fourth, fifth and sixth nerves bilaterally, as well as the pituitary stalk, diaphragma sella, and the preptineal cistern were visible (Fig. 3).

Pathology. Biopsies from four of the cysts revealed arachnoid-like tissue. The solid tumor weighed 40 gm. It was a lobulated mass with a few hard excrescences and contained numerous cysts filled with mucoid material and black hair. Microscopically, the tumor was composed of a large number of distinctly different tissues, including well-formed salivary gland, skin with sebaceous glands and hair, muscle, fat, bone, respiratory mucosa, peripheral nerve, and brain (both cerebral cortex and primitive cerebellum). Individual tissues appeared well differentiated and benign (Fig. 4).

Postoperative Course. By the third week postoperatively, eye movements were complete and conjugate, and pupillary reaction had been regained in the left eye. Air injection revealed communication between large subdural spaces on the right and left. The fluid protein count from this region was 572 mg%. Six weeks postoperatively the ventriculoperitoneal shunt was revised, a Rickham reservoir and an anterior limb being added to drain the right subdural collection. Ten weeks following craniotomy the child was discharged on diphenylhydantoin.

After discharge the patient gained weight and began to respond to the parents. She was readmitted 1 month later because of an increase in head size from 46 to 48 cm over a 2-week period. The child's behavior had not deteriorated.

Second Examination. On June 7, the head circumference was 48.5 cm and the fontanel was full but not tense. The head transilluminated as before. Eye findings were unchanged, and the right corneal reflex was absent. A tap through the Rickham reservoir recovered clear colorless fluid with a protein of 472 mg%. Air injected through the anterior catheter into the right prefrontal space entered freely. Air injected into the ventricular catheter demonstrated free communication of the right ventricle with the prefrontal region. The valve pumped well. Enlargement of the head was controlled by pumping the valve more frequently.

June 20 marked the onset of seizures in-
Cystic intracranial teratoma in an infant

volving both sides of the body. Blood sugar was 7 mg%. Sugar supplement, increased doses of anticonvulsants, and cortisone acetate were given. At this time the child was found to have an enlarged liver but no palpable spleen. At discharge, there were still occasional short jerking spells of the extremities; head circumference was 48 cm, and the child remained lethargic with poor head control. The patient died after a period of pneumonia and respiratory difficulty on April 26, 1970.

Postmortem Examination. Autopsy was limited to the head. Sections of the brain revealed cysts along the inferior medial aspect of the left frontal lobe as well as the rostral thalamus. Microscopically these were associated with mild gliosis. There was extensive loss of cortical neurons throughout the cortex. Encircling the base of the cerebellum and medulla was a thick cellular meningeal reaction without obvious inflammatory response. No residual tumor was identified.

Discussion

Tumors presenting in the neonatal period probably arise prenatally. This is almost certainly true of slow-growing tumors such as the teratoma described here. The history of maternal irradiation in the first trimester of pregnancy probably has no bearing on the development of the teratoma in this case. Despite the susceptibility of the early fetus to irradiation, the sensitivity of the fetal nervous system to this agent, and the possibility that fluoroscopy entails sufficient radiation to produce changes in the embryo,7 detailed reviews by Russell7 and Rugh8 mention neither teratoma nor intracranial tumor resulting from experimental or clinical irradiation of the fetus. Furthermore, review of the original reports of cases of teratomas in the newborn, collected from the literature by Greenhouse and Neuberger,4 failed to disclose any case in which the mother had been irradiated during the pregnancy. The role of oral contraceptives taken during the first trimester of pregnancy in the induction of tumor is uncertain but seems improbable. The clinical picture of intracranial teratomas of infancy has been well described in the excellent review by Greenhouse and Neuberger.

Much of the interest of this case derives from the roentgenological findings. The pres-
ence of a large intracranial calcification with areas of lucency both within and surrounding it led us to consider the diagnoses of dermoid and teratoma preoperatively. Calcification at sites distant from the main mass of calcified tumor and the striking transillumination of the head were clues favoring the diagnosis of teratoma.

At craniotomy, the solid portion of the tumor was amputated where it extended into the region of the lamina terminalis and hypothalamus. Residual tumor is therefore present at this point. Roentgenographically visible calcification and transillumination of the left side of the head indicated tumor in the left half of the cranium as well. It is not clear, however, that residual tumor means a bad prognosis in the case of cystic teratomas.

Ingraham and Bailey reported a case of presumed teratoma in an 8-week-old child who survived at least 18 months after an operation limited solely to the evacuation of cysts and biopsy of the cyst wall. No other reported case of intracranial teratoma in this age group has been operated. These authors mention two cases of intraspinal cystic teratomas with evacuation of cysts and incomplete removals. Both of these cases had good neurological recovery postoperatively and maintained this over follow-up periods of 4 and 10 years. It therefore seems likely that partial removal of intracranial teratoma with large cysts may also result in long-term survivals.

Summary

An infant girl with hydrocephalus due to a large cystic intracranial teratoma has been discussed. Unique features included marked asymmetrical transillumination of the head and roentgenograms revealing a large densely calcified mass surrounded by areas of radiolucency. Ventriculography, angiography and operative removal were carried out.

References


Received for publication November 20, 1969.
This work was supported in part by USPHS Grant NB 05580, and the Allen P. and Josephine B. Green Foundation. Dr. Torres-Reyes holds a Special Fellowship (NSRA 1-F11-NB-1824) as Instructor of Radiology from NINDS.

Address requests for reprints to: Ralph A. W. Lehman, M.D., Division of Neurological Surgery, Washington University School of Medicine, Barnes Hospital Plaza, St. Louis, Missouri 63110.