CASE REPORTS AND TECHNICAL NOTE

CHOROID CYST OF BRAIN WITH LEUKEMIC CONTENTS
IN A PATIENT HAVING LEUKEMIA

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The lesion to be reported is unique so far as we are aware. In his series of 2,000 intracranial tumors Cushing did not mention such a condition, and during the past 35 years one of us (G.H.) has not encountered a similar case. In our registry of brain tumors we have called it a “leukemic” cyst, but this designation is doubtless incorrect, since the cyst was of choroidal origin, but its content was entirely leukemic material with the general appearance of ordinary milk.

Leukemic processes in the central nervous system are well known. Whether they occur as primary lesions, favoring the multicentric theory of leukemia, or as secondary, invasive lesions, we cannot say. In the literature most cases described are part of the terminal picture of leukemia. Diamond1 studied the brains of 14 patients who died of this malady. In 7 histories he found no clinical evidence of neurological disorder, yet all had histologic changes in the nervous system characteristic of leukemia, namely, infiltration of myelocytes (1) around and within blood vessels, (2) in subarachnoid spaces, (3) within brain parenchyma both circumscribed and diffuse lesions, (4) rarefaction and vacuolation of neurons and axons, and (5) proliferation of oligodendroglia. Such pathological changes produce a variety of symptoms and signs, usually suggesting diffuse instead of focal disease. Focal fits, however, were reported by Hellich2 in a patient 3 to 4 months after a diagnosis of leukemia was made. The brain in this case showed parenchymatous infiltration of leukemic cells in the cortex and thalamus.

The case here reported deserves attention because the clinical course was that of a brain tumor in the pararolandic area. However, the high leukocytosis with myelocytes on routine blood count was diagnostic of myelogenous leukemia. The extracortical cyst in the rolandic area found at craniotomy is apparently most unusual for leukemia. Histologically, the wall of the cyst was lined by tall, ciliated columnar cells instead of the expected glial tissue. This pointed to a papillary adenocystoma of choroidal origin. It is not easy to explain this finding as most choroidal cysts originate in or near the ventricular system. Since leukocytes and myelocytes crammed the cyst cavity, it is possible that leukemic cells infiltrated a congenital choroidal “cell rest,” causing its growth until signs of a space-consuming lesion developed.

REPORT OF CASE

F.B., a 29-year-old white male, was admitted on Oct. 4, 1934, complaining of spells of unconsciousness of 9 months’ duration. He had been relatively well until January 1934.

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when his first seizures began. There was an aura of tremor with twitchings of the left hand. This was followed by clonic spasms of the left arm and leg, proceeding to generalized convulsions with loss of consciousness. After the seizure the patient's left hand felt "queer" and weak. He had had a total of nine seizures in the previous 9 months. Two months before admission he became excessively irritable and somnolent. He took prolonged afternoon naps, although he slept as much as 10 hours during the night. About 1 month prior to admission his left hand and leg became weak.

Neurological Examination. Bilateral papilledema of 2 D. was present with normal fields on confrontation and perimetry. There was moderate loss of strength in the left hand grip. Reflexes were more active in the left arm and leg; there were no abnormal reflexes. A roentgenogram of the skull revealed an area of decreased radiodensity in the right frontal and central zones. The sella turcica was intact. In the course of routine blood studies a white blood cell count of 53,000 with 18 myelocytes was noted. The medical consultant found an enlarged spleen on physical examination, and he made a diagnosis of chronic myelogenous leukemia.

1st Operation. Despite the presence of leukemia there remained the possibility of a non-associated tumor. On Oct. 8, 1934, a pneumoventriculogram was made and showed downward displacement of the central portion of the right lateral ventricle with a shift from right to left.

Under local anesthesia a right frontoparietal bone flap was reflected, exposing a large cystic mass near the midline of the rolandic area. To quote from the surgeon's notes: "A rather cystic area could be felt through the dura mater near the midline and in the center of the exposed region. The dura was incised with the pedicle at the midline and upon reflecting it a large cystic lesion was seen involving the cortex near the midline immediately in the rolandic area. The cyst wall was rather thin and translucent, filled with an opaque grayish white fluid (Fig. 1). The lesion was different from those seen by any of the operative team. . . The cyst was then evacuated by suction through a needle attached to a syringe, approximately 60 cc. in all being obtained. A smear disclosed pus cells and myelocytes. A cell count of the fluid showed 1,918 white blood cells with 13 myelocytes per c. mm. The question, of course, arose whether this was due to a leukemic condition or an abscess. The entire cyst wall was carefully removed and stripped from the cortex fairly readily" (Fig. 2).

Pathological Report. The diagnosis of the cyst wall was adenocystoma, papillary type, of choroidal origin. The pathologist, Dr. Shields Warren, reported that the fluid removed from
the cyst was compatible with leukemia. A culture taken from the fluid at operation showed no growth after 3 days.

A further study of this cyst wall was made by Dr. S. J. Hicks. He reported as follows: "There are atypical mononuclear cells in the blood vessels of the cyst, and there are similar cells incorporated in some clotted material adherent to the lining of the cyst. These may certainly be interpreted as leukemic cells and represent some remains of the cyst content."

Course. The postoperative course was somewhat stormy. Although the patient aroused and was able to move the left side, the area over the bone flap swelled and was palpably riding.

2nd Operation. On Oct. 11, 1934, because of slowing pulse and increased drowsiness, the bone flap was elevated and a large extradural hematoma was removed.

Course. The patient rallied and, except for three persistent draining sinuses in the skin flap, his recovery was satisfactory. The leukemia caused these complications, predisposing to faulty blood clotting and lessened resistance to infection. After the 2nd operation the white blood cell count rose to 85,000. Fowler's solution was given, and by Nov. 7, 1934 the blood count dropped to 15,000. The draining sinuses granulated and at discharge (Nov. 9, 1934) the patient was ambulatory but the left side was weak. Seizures were controlled fairly well with sodium phenobarbital.

2nd Admission, June 27, 1935. Since his discharge the patient had had intermittent left-sided focal seizures followed by generalized convulsions. The wound was draining and he was febrile. Roentgenograms of the skull showed a moth-eaten appearance of the bone flap, indicating osteomyelitis.

3rd and 4th Operations. On June 29, 1935, the bone flap was completely removed and the area was left adequately decompressed. The skin flap bulged increasingly; so on July 3, 1935 it was reflected and a huge extradural clot was evacuated. After this the skin flap was partially resutured and a pressure dressing was applied.

Fig. 3. Photomicrograph of cyst lining to show tall ciliated columnar epithelium \((\times 500)\).
Course. The wound finally healed. The white blood cell count at this time was 120,000 with a great number of myelocytes. The spleen was palpable several fingerbreadths below the costal margin. Roentgen therapy was given to the long bones, and at the time of discharge on July 21, 1935 the white blood cell count had dropped to 7,750.

The course of the patient was followed at home by Dr. Malott of Scottsbluff, Nebraska, who reported that the patient had seizures, although less severe.

3rd Admission, April 1, 1936. Because of seizures and paresis of the left arm and leg, progressive debility, and a white blood cell count of 163,000, the patient was readmitted.

Neurological examination disclosed moderate weakness of the left arm and leg with hyperactive reflexes; the optic disks were not elevated. The leukemia was manifestly advanced.

After roentgen therapy the white blood cell count dropped to 7,000. The patient regained his strength and left the hospital on April 6, 1936.

Course. From April to July, 1936 the patient had three seizures. He was back at work. When seen at the Lahey Clinic in July 1936 the findings were unchanged from the previous examination. The white blood cell count was 14,850. Dr. Malott, on Oct. 29, 1936, reported that the patient had died. The course of events leading to his death was typical of leukemia. Until his death he had had an occasional left-sided seizure.

Autopsy Findings. The autopsy was done by Dr. Malott who removed the organs and sent them to the Pathological Laboratory of the New England Deaconess Hospital. On removing the brain Dr. Malott noted that the posterior portion of the resection was filled with approximately 2 ounces of grayish-green pus. The organs were examined by Dr. Shields Warren who reported changes characteristic of chronic myelogenous leukemia in the spleen, lymph nodes and bone marrow. The report on the brain was as follows: "Convolutions of the right hemisphere normal except in the right frontoparietal region where there is a large cystic space 4 by 3 by 3 cm. which appears to be lined by choroid membrane. No other abnormalities are noted. The histological section of this cortical cyst showed it to be lined by a single layer of tall, ciliated, columnar and flattened fusiform epithelium supported on a delicate basement membrane (Fig. 3). Small blood vessels scattered through the brain are partially filled with leukemoid cells." Dr. Warren's diagnosis was cortical cyst or papillary adenocystoma of choroid origin.

SUMMARY

Circumscribed infiltration of the brain with leukemic cells is rare compared with diffuse infiltration of the subarachnoid spaces and the epidural spinal spaces. In his series of 2000 intracranial tumors Cushing reported no such case. The lesion here reported is that of a choroid cyst containing leukemic material in a patient having chronic myelogenous leukemia, whose illness was ushered in with left jacksonian seizures. After craniotomy and removal of a cyst in the right rolandic area, followed by roentgen therapy, the patient survived 2 years.

We are greatly indebted to Dr. Shields Warren and to Dr. Samuel J. Hicks for their painstaking study of the pathological material from this patient.

REFERENCES
