BRAIN ABSCESS IN CYANOTIC CONGENITAL HEART DISEASE

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During the last century the so-called paradoxical brain abscess appearing as a complication of congenital heart disease has received special attention in the medical literature. As early as 1880 Ballet,1 taking into account the coincidence of these two conditions, stated: “Aussi bien pensons-nous qu’on ne saurait expliquer par une pure coincidence le fait de 6 cas de malformations du coeur s’accompagnant d’abcès du cerveau; que ce chiffre a son importance qui mérite de fixer l’attention, et qu’il est logique d’admettre une relation étiologique entre les lésions précédentes.”

Reviewing the frequency of brain abscesses in congenital heart disease by clinical and postmortem studies, it soon became clear that brain abscess as a complication of congenital heart disease is not altogether a rarity.6,8,12 The patients who underwent operation, however, are rare and even more so are those who were operated upon successfully.

Smolik et al.13 in 1946 reported the first case of successful surgical treatment. This was followed by the cases of Cohen et al.,5 Beller,2 Ingraham and Matson,9 Lafon et al.,10 Ley et al.,11 Weber,15 Campbell,4 and Gund.7

During the last 2 years we have observed 4 patients with congenital heart disease complicated by brain abscess, 3 of whom underwent successful surgery.

CASE REPORTS

Case 1. A 15-year-old girl was admitted to the neurosurgical ward with the chief complaints of fever, headaches, diplopia and visual disturbances. She was known to suffer from congenital heart disease, diagnosed as pulmonic stenosis with interauricular septal defect.

Two weeks prior to admission, she awoke from sleep with headache and diplopia. These manifestations appeared and ceased a number of times during the following days and she was admitted to another hospital for investigation. There was papillary hyperemia of the optic fundi. The blood contained 16,000 white blood cells/ c.mm. with a normal differential count; erythrocyte sedimentation rate was 37/80 mm. The cerebrospinal fluid contained 320/3 cells, mostly lymphocytes, and protein was 300 mg. per cent.

On the 5th day of hospitalization she complained of severe headache, blurred vision, nausea and vomiting, weakness in the right arm and leg, and she was seized with an attack of Jacksonian epilepsy involving the right hand and the right side of
the face. The cerebrospinal fluid then showed an increase to 500/3 cells, mostly lymphocytes, and protein was 750 mg. per cent. The fundi showed blurred papillary contour and venous engorgement. A tentative diagnosis of cerebral abscess was made and she was sent to us for further examination.

Examination. The patient was in poor condition, slightly cyanotic and somewhat lethargic. Positive neurological findings were slight stiffness of the neck, paresis of the right abducens nerve, hemiparesis of the right side, positive Babinski's sign, and bilateral papilledema (2 diopters) with right moderate homonymous hemianopsia. Electroencephalogram revealed gross disturbance of the left hemisphere, particularly in the mid and posterior zones.

Pneumoencephalography showed a space-occupying process in the left parieto-occipital region (Fig. 1).

![Fig. 1. Case 1. Pneumoencephalogram demonstrating dorsal part of left sella media and ventricular triangle pushed down.](image)

Operation. Under general anesthesia a left parieto-occipital osteoplastic flap was carried out. On exploring the occipital region with a ventricular needle, a cavity was encountered which yielded 50 cc. of yellowish-green pus. Penicillin was instilled into the operative cavity and the wound was closed without drainage and without fixation of the bone.

Bacteriological Report. The recovered exudate contained coliform bacilli especially sensitive to Sigmamycin.

Course. Treatment with Sigmamycin was continued and after 2 weeks her general condition improved considerably. The headaches and all other neurological signs disappeared; the electroencephalogram became normal. The patient's condition is satisfactory up to the present, 18 months after operation.

Case 2. A 16-year-old boy, known to suffer from tetralogy of Fallot proved by cardiac catheterization, was admitted to the medical ward because of violent headache. Shortly after admission he was seized with an attack of Jacksonian epilepsy involving the left half of the face. These attacks recurred four times the same day.

Besides the cardiological findings which were typical for tetralogy of Fallot, there were stiffness of the neck and slight left-sided hemiparesis. His temperature
was 38°C. Ocular fundi were normal; roentgenograms of the skull revealed no abnormalities; and electroencephalogram showed severe central and posterior disturbance on the right.

Cerebrospinal fluid contained 946/3 cells with 90 per cent polymorphonuclear cells and 10 per cent lymphocytes; Pandy was clearly positive; proteins were 100 mg. per cent; and culture yielded no growth. Blood studies revealed: erythrocyte sedimentation rate 20/25 mm.; count of white blood cells 25,300/c.mm., with differential count of 82 per cent polymorphonuclear cells, 4 per cent bandforms, 5 per cent monocytes, 9 per cent lymphocytes; hemoglobin 16.8 gm. per cent; and count of red blood cells 5,000,000/c.mm.

A tentative diagnosis of cerebral thrombosis or encephalitis was made in the medical ward and treatment with antibiotics (20,000,000 u. penicillin, 1 gr. streptomycin, 2 gm. i.v. Achromycin daily) was instituted. On the 6th day of hospitalization, visual disturbances in the form of blurring appeared, and examination of the fundi disclosed bilateral papilledema (3 diopters) and hemorrhages. The patient was transferred to our department.

**Examination.** The patient was cyanotic and markedly lethargic. Left hemiparesis, especially of the hand, together with signs of meningeal irritation were found. In view of his cardiac condition, the possibility of brain abscess was entertained.

Right-sided cerebral angiography demonstrated a space-occupying lesion in the left temporal region (Fig. 2).

**Operation.** Under general anesthesia a right temporoparietal osteoplastic flap was carried out, and exploration of the temporal region with a ventricular needle disclosed the presence of a nonencapsulated cavity from which 40 cc. of pus were removed. Penicillin was instilled and the wound was closed as in Case 1.

**Bacteriological Report.** Culture of the pus produced no growth.

**Course.** Recovery was uneventful and the patient regained consciousness, the neurological signs receded and the papilledema cleared up considerably. Electro-

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**Fig. 2. Case 2. Carotid arteriogram. (Left) Lateral position shows elevation of third part of sylvian group. (Right) Anteroposterior Towne position shows middle cerebral artery in normal position but there is an augmentation of blood vessels connecting the sylvian group with lateral parts of the cortex. Slight shifting of anterior cerebral artery to contralateral side.**
encephalography performed a month later showed improvement and much more so after 3 months.

For the following 11 months the patient's condition was satisfactory from the neurological point of view. He died during an operation for the repair of his cardiac defect.

Autopsy. The diagnosis of tetralogy of Fallot was confirmed. Multiple sections of the parietotemporal region revealed, at the site of the cerebral abscess discovered during operation, an indurated region, microscopic preparations of which showed a focus of scar tissue surrounded by a layer of reactive gliosis.

Case 3. An 11-year-old boy, known to suffer from Fallot's tetralogy, proved by cardiac catheterization, was admitted because of an attack of Jacksonian epilepsy.

![Fig. 3. Case 3. Carotid arteriogram. (Left) Lateral position shows the supraclinoid part of internal carotid artery depressed. Second and third parts of middle cerebral artery group are pushed down. Vessels of the candelabra are spread. (Right) Anteroposterior Towne position. The anterior cerebral artery is pushed towards the left side and the frontopolar sign is positive, indicating a right-sided frontal space-occupying process.]

Three months previously, he had lost consciousness for a few hours and was admitted to the medical ward of another hospital. Shortly afterwards he awoke but complained of a severe headache on the right side. During that day his temperature rose to 39.2°C. General and neurological findings were not significant, excepting those of congenital heart disease. The fundi were normal, and lumbar puncture yielded normal cerebrospinal fluid. The child was treated with antibiotics and after 1 week the fever and headache subsided. At the end of a fortnight the child was discharged.

A week prior to admission to our department the boy again complained of headache. On the day of admission he had an attack of Jacksonian epilepsy, which started in the left hand and spread to the left side of the face.

Examination. The patient was in good general condition but with a moderate degree of rigidity of the neck and left-sided spastic hemiparesis with extensor plantar reflexes.

The optic fundi were normal. Electroencephalography showed severe disturbance
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emanating from the right anterior quadrant. Blood studies revealed: hemoglobin 17 gm. per cent, red blood cells 6,156,000/c.mm., and white blood cells 12,300/c.mm. with a normal differential count. Cerebrospinal fluid contained 306/3 cells, mainly lymphocytes; protein was 148 mg. per cent. Right-sided arteriography showed a space-occupying lesion in the frontoparietal region (Fig. 3).

Operation. On the basis of these findings frontoparietal osteoplastic craniotomy was carried out. Upon exploration with a ventricular needle, hard tissue was encountered ~ cm. below the surface of the cortex. An incision was made in the frontal region and an encapsulated abscess, the size of a small tangerine, was removed. The wound was closed without drainage.

Bacteriologic Report. The pus was sterile.

Postoperative course was satisfactory; the hemiparesis cleared up and the patient remained in a satisfactory state of health.

Two months after operation the patient is still well but it is too early to state long-term prognosis.

Case 4. A girl aged 6 known to suffer from tetralogy of Fallot, proved by cardiac catheterization, was admitted to the medical ward because of an attack of severe headache and high fever (40°C.). She was treated with antibiotics. Three days prior to her transfer to the neurosurgical ward, motor aphasia developed and she had a right Jacksonian epileptic attack which resulted in a right-sided hemiplegia.

Examination. Her general condition was extremely poor. There were motor and sensory aphasia, hemiplegia, and pyrexia of 38–39°C. Hemoglobin was 18 gm. per cent, count of red blood cells was 9,000,000/c.mm., and of white blood cells 4,800/c.mm. with normal differential count. Electroencephalography revealed a lesion involving the entire left hemisphere. The patient’s condition continued to deteriorate and she died on the 2nd day after admission to our ward.

Necropsy. An encapsulated abscess was found, involving almost all of the left hemisphere (Fig. 4).

DISCUSSION

Brain abscess in cases of congenital heart disease occurs most frequently in patients suffering from the cyanotic variety of the latter condition. The two factors probably involved in the pathogenesis of brain abscess are cerebral damage and superimposed infection.

The polycythemia associated with the cyanotic type of congenital heart
disease may lead to thrombosis of the small vessels of the brain with resultant infarction and softening of the cerebral substance. In addition some degree of chronic anoxia of the brain may be operative in this type of congenital heart disease. Berthrong and Sabiston\(^3\) found 25 examples of cerebral softening among 135 necropsies of patients with cyanotic congenital heart disease. To this cerebral damage septic emboli are added, which, because of the cardiac lesion, enter the systemic circulation without passing through the lungs—thus brain abscesses are formed. Sometimes the source of the infection is obvious: infection of the upper respiratory tract, stomatitis, caries of the teeth, pharyngeal abscess, tonsillectomy, carbuncles—all may be the primary source of the infection. Quite often the source of the infection remains unknown. In these cases it is believed that organisms that normally inhabit the upper respiratory and gastrointestinal tracts find their way into the systemic circulation by way of the right-left shunt within the heart and thus reach the brain and, acting upon the pre-existing cerebral damage, readily proceed to formation of abscess.

It may be stated that, as a rule, the brain abscess in congenital heart disease is of an acute character and causes severe symptoms associated with increased intracranial pressure which endanger the patient's life. In these cases there is no formation of a capsule, and it is our policy to carry out an osteoplastic flap and aspirate the abscess and insufflate the cavity with penicillin, after which the flap is closed without drainage. This, while allowing decompression, admits the possibility of encapsulation which in case of recurrence can then be moved \textit{in toto} by the method of Vincent.\(^{14}\) In case of subacute or chronic abscess, there is enough time for the occurrence of encapsulation. In those cases in which the abscess is subcortical and not in the depth of the hemisphere, we remove it \textit{in toto}. This procedure is difficult when the abscess is large and deeply situated because of the risk of damage to the brain stem and basal ganglia. A safer treatment then would be tapping and antibiotics; if there is recurrence, the capsule has probably had time to thicken and the abscess has diminished in size. We think that then it may be justifiable to attempt complete removal.

In 2 of our cases the development was acute. At puncture we found pus without first encountering a capsule; the pus was aspirated. In Case 3 a capsule was encountered and the abscess was removed \textit{in toto}.

\textbf{SUMMARY}

Four cases of brain abscess complicating congenital heart disease are presented. Two of the patients recovered after aspiration and 1 after removal of the abscess \textit{in toto}. The fourth patient died before surgical intervention could be instituted. Pathogenesis and treatment of this condition are discussed.

\textbf{REFERENCES}