PARADOXIC BRAIN ABSCESS IN CONGENITAL HEART DISEASE

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Paradigmatic brain abscess is the name given to a certain form of metastatic abscess occurring in patients without pulmonary suppuration, but with congenital cardiac defects permitting venous blood to circulate in the arterial system without passing through the lungs.

This type of brain abscess is rare, presumably because of the combination of factors that must exist before such lesions can develop. Compared with metastatic abscesses arising from chronic suppuration in the lungs, Ruhberg found that a paradoxical abscess occurred once for every 13 cases of metastasis from lung disease. The rarity of the condition is further emphasized by Hanna, who recorded 6 cases of paradoxical abscess in 160 fatal cases of congenital cardiac disease. Robbins found almost the same proportion (3 in 53), but Abbott’s percentage was much lower (7 in 1000).

Only 39 cases of paradoxical brain abscess could be found reported in the literature. Largely because of failure to recognize this condition and the extremely rapid course in some of these patients, they fared almost uniformly badly. Up to the present time, only 1 patient has been cured. This report deals with a patient who is apparently well 1 year after treatment.

CASE REPORT

The patient was a college student, aged 22 years. She first came to the Mount Sinai Hospital in 1946, with signs of a generalized infection. A complete investigation at that time established the diagnosis of subacute bacterial endocarditis (endarteritis). The basic cardiac lesion was tetralogy of Fallot with a patent ductus arteriosus, confirmed by angiocardiography. The infection responded well to penicillin treatment, and she was discharged, apparently cured. She had no further complaints until November, 1948, when a routine dental prophylaxis was carried out. Four days later she complained of malaise, nausea and chills; her temperature was 101°. For the next week she continued to run a low-grade fever and complained of headache. The day prior to admission she had what was apparently a generalized convulsion. No antibiotics were given before or after the dental work. A blood culture taken by her physician was negative.

Examination. Temperature was 100.4°, pulse rate 100, and B.P. 110/74. There was pronounced cyanosis. A continuous, to-and-fro, machinery-like murmur was heard at the pulmonic area; there was a thrill at the pulmonic area and at Erb’s point. A severe kyphoscoliosis was noted. The tip of the spleen was just palpable. She showed some drowsiness and loss of recent memory. There was repeated vomiting, and she complained of severe headache, worse on the right side. There was a Babinski sign bilaterally, with transient bilateral ankle clonus. On double simultaneous stimulation a left hemihypesthesia, without astereognosis was noted. There was early papilledema bilaterally, worse on the right, with marked venous engorgement. A complete left homonymous hemianopsia was found. There was bilateral periorbital edema and tenderness in the right frontotemporal region.

Hb. was 13.9 gm. WBC was 19,400, with a shift to the left. Sedimentation rate was normal on two occasions. Repeated blood cultures were sterile. X-ray of the skull showed no abnormality. EEG showed a large amount of very slow (1-2/sec) activity in the right posterior temporal region. A lumbar puncture yielded clear, colorless fluid, under a pressure of 220 mm.; it was otherwise normal. The blood and CSF Wassermann reaction was negative. X-ray of the
chest showed the typical boot-shaped heart of tetralogy of Fallot and marked kyphoscoliosis (Fig. 1).

A diagnosis of brain abscess was made and she was transferred to the neurosurgical service.

Treatment. Penicillin (aqueous procaine penicillin, 600,000 units a day) was started at the time of admission and given continuously thereafter. For a while it appeared that penicillin alone was going to control the infection. About 2 weeks later, however, the papilledema increased, the EEG abnormality became more pronounced, and the neurological signs advanced.

1st Operation. On Jan. 4, 1949 the senior author made a burr hole in the right temporal bone. With the exploring needle, an abscess cavity was encountered in the right temporal lobe, and 5 cc. of thick yellowish-white, odorless pus were aspirated. The abscess was not encapsulated and at one time the needle, of its own weight, fell into the temporal horn of the lateral ventricle. Penicillin solution was instilled into the cavity and the wound closed without drainage. A culture of the pus yielded no growth, and no organisms were seen on direct smear.

Course. For the next few days, the papilledema appeared to be receding and the patient felt well, but she soon complained of severe headache and began to vomit. The abscess was again aspirated and 5 cc. of thick reddish pus were obtained. Into the abscess cavity penicillin solution and 1 cc. of pantopaque (R) were injected.

X-rays of the skull taken after this procedure showed a spherical mass of pantopaque (R), about 1.5 cm. in diameter, deep in the right temporal lobe; droplets of pantopaque could be seen scattered throughout the ventricular system (Fig. 2).

Fig. 1. X-ray of the chest, showing typical heart shadow.

Fig. 2. Showing pantopaque in the right temporal lobe and scattered droplets in the ventricles.
The next day the patient felt worse; she continued to vomit and the papilledema increased.

2nd Operation. The following day a right temporal craniotomy was performed by Dr. Cohen and the abscess was entered through a transcortical incision in the inferior portion of the temporal lobe. The brain was very tense, and there was no capsule. There seemed to be several pockets within the abscess, each containing pus of a slightly different color and consistency. The entire abscess and the softened necrotic brain tissue surrounding it were removed by suction. A drain was placed in the cavity and brought out through a separate posterior stab wound.

Postoperative Course. This was discouraging at first, and there was little hope of her survival. The CSF was purulent and under great pressure for a long time after operation, and she required daily lumbar punctures to reduce the tense, bulging decompression. The EEG showed what appeared to be a larger focus of abnormality, and, with the other signs, led us to suspect multiple cerebral abscesses. For this reason, a pneumoencephalogram was performed.

The films (Fig. 3) showed only the distortion corresponding to the area of the original abscess. Antibiotic systemic therapy was continued. Eventually she felt better and the signs receded.

When penicillin was discontinued and there was no recurrence, she was discharged, apparently well, 4 months after admission. Her vision was moderately reduced and the fundi presented the picture of post-papilledema optic atrophy. There was a left homonymous hemianopsia, with some sparing of the upper left temporal field. A mild hemisensory loss was present, but there was no weakness.

When examined last, 1 year after discharge, she had essentially the same findings. The fundi had reverted almost to normal. She complained only of difficulty in walking out-of-doors, owing to the visual field defect. There was no evidence of infection. She was attending regular classes at a university and doing well in her studies.

DISCUSSION

A review of the literature indicates that paradoxic brain abscess has been found in 40 cases, including this one. We have arbitrarily eliminated 1 case so diagnosed, because generalized sepsis was present. Of this number, only 8 were recognized before death, 7 of them receiving suitable treatment.

Fig. 4 shows that in a good proportion of cases there was ample reason to suspect a localized process in the brain. Convulsions, generalized or focal, occurred in 11 cases, hemiparesis or its equivalents in 23, signs of increased intracranial pressure in 20, and meningeal signs in 17. It is sig-
significant that well over half the patients were stuporous at the time of admission; this may account for the fact that although there were 11 abscesses in the occipital lobes and 10 in the temporal, hemianopsia was reported only twice in the entire group. Aphasia, also, was mentioned in only 2 cases, although there was a total of 10 abscesses involving the dominant temporal or parietal lobes. Sensory changes were recorded in only 8 cases.

The laboratory findings are summarized in Fig. 5. The white blood count was elevated in 11 cases, and reported as normal in only 3. Blood cultures were negative in every case where this was done. The exact location of the abscess given by electroencephalography in our case made it unnecessary to do air studies. Most of the cases were reported before electroencephalography had reached its present refinement, and we can find a report of its having been done in only 1 case; there was diffuse slow activity, without any localizing characteristics. The cerebrospinal fluid was abnormal at some stage of the disease in almost every case in which such an examination was reported.

It is evident from the clinical observations in this series of cases that in every patient with congenital heart disease in which there is a right-left shunt (manifested by transient or permanent cyanosis), the development of any signs of central nervous system disease should raise the suspicion of brain abscess and immediate steps should be taken to establish the diagnosis and institute treatment.

Fig. 6 shows some of the characteristics of these abscesses as encountered at operation or postmortem examination. Paradoxic abscesses are almost always single, and in only 3 of the entire series was there more than one in the brain. Like other types of metastatic abscess, paradoxic lesions are rarely encapsulated; in 4 of the 40 cases there was a definite capsule, although the lack of it was actually mentioned in only 4. This absence of encapsulation undoubtedly accounts for the tendency of these lesions to rupture into the ventricles and the subarachnoid space; evidence of intraventricular rupture was found in 12 cases, although in 24 others it had definitely not occurred. Organisms could be recovered from most of these lesions. In our case, failure to culture bacteria from the abscess was attributed to penicillin given prior to the operation.
No preference for either side or any particular part of the brain was found in any of the reported cases, as shown in Fig. 7.

The treatment of paradoxic brain abscess is essentially a surgical problem, and the principles that apply to intracranial suppuration in general will hold in these cases. The liberal use of penicillin and, when indicated, other antibiotics, locally and systemically, is obvious.

Two cases previously reported from this hospital by Wechsler and Kaplan were correctly diagnosed, and, although adequate surgical treatment was undertaken, the patients died. Had penicillin and the newer antibiotics been available, they might have survived.

A patient of Grant’s reported by Hand was operated upon and apparently cured, but she died 6 weeks later of an abscess in another part of the brain.

The patient of Smolik, Blattner and Heys was cured by surgical drainage and sulfonamides, and represents the sole survival up to this time.

The best treatment, in view of all these observations, would seem to be prophylactic. Fig. 8 demonstrates that some antecedent illness was recorded in half of the cases in the literature in which the history was sufficiently detailed. The same precautions should be taken as for the prevention of bacterial endocarditis in rheumatic or congenital cardiac patients when a procedure such as tonsillectomy or tooth extraction is undertaken. It is conceivable that one injection of a long-acting penicillin preparation would have prevented this complication in our patient.

Most of the patients died soon after the onset of cerebral symptoms, half of them within 2 weeks.

A great deal has been written about paradoxic embolism in congenital heart disease. Unfortunately, its occurrence has been overemphasized in contrast to abscess, which, unlike embolism, is potentially curable. Hanna, for example, in 160 fatal cases of congenital cardiac disease, found only 1 case of paradoxic embolism, whereas there were 6 of brain abscess. Abbott found 13 instances of embolism and 7 of abscess in her 1000 cases.
The pathogenesis of paradoxical cerebral abscess has occupied considerable space in the literature. Various theories, all without adequate proof, have been advanced. Transient bacteremia must occur, and organisms must pass through with the venous blood, from the right to the left side of the heart, thus escaping the normal filtering or phagocytic action of the lung capillaries, and reaching the brain. It is significant, however, that in the recorded cases there were rarely abscesses in other organs, despite large right-left shunts.

There is some evidence to show that these patients have some form of pre-existing cerebral disease, whatever the cause. In Abbott's\textsuperscript{4} cases of tetralogy of Fallot, for example, cerebral disease was considered the primary cause of death in 15 per cent of the patients, and this figure was only slightly exceeded by cardiac insufficiency. Episodes of unconsciousness and convulsions are not unusual in patients with congenital heart disease, and in 2 of the reported cases of paradoxical abscess there was a history of such attacks for a much longer time than an abscess could have existed.

As far as actual knowledge is concerned, however, we know very little more than we did in 1850, when Ballet\textsuperscript{6} presented the first collected series of cases of this type. We should, therefore, like to borrow his conclusions. "As to the mechanism by which an abnormal communication between the chambers of the heart favors the development of cerebral suppuration, this is for the time being a matter of pure speculation, and any number of guesses might be made ... These theories are seductive because in the present state of affairs there is no proof that they are right or wrong. We should do better, for the time being, to stick to the clinical facts."

\textbf{ADDENDUM}

Since this paper was submitted for publication, two additional series of cases have appeared in the literature. The first\textsuperscript{8} records 2 cases, only 1 of which was verified. In this instance a right temporal lobe abscess due to hemolytic staphylococcus albus was responsible for the death of a patient with an interventricular septal defect and advanced pulmonary tuberculosis. Death occurred rapidly and there was no opportunity for adequate diagnostic study or treatment. Their other patient died, but permission for postmortem examination was not obtained. There is a comprehensive review of the literature.

The second report\textsuperscript{9} deals with 9 new cases from routine autopsy material. The cardiac and cerebral lesions were similar to those previously described in the literature.

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INTRANASAL ENCEPHALOCELE

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Encephalocele, or congenital protrusion of the brain and meninges through the skull, is not uncommon. The most frequent site of such a lesion is occipital. Ingram and Matson reported a series of 84 encephaloceles, of which 63 were occipital, 9 parietal, 6 frontal, 5 nasal, and only 1 presenting through a defect in the cribiform plate into the nasopharynx.

Fenger stated that of the anterior herniations, the so-called sincipital type is the more frequent, in which an external mass presents itself at the nasion in the midline, or laterally at the junction of cartilaginous and bony portions of the nose, or at the