

*Diffused Cerebral Sclerosis.* By T. W. McDOWALL, M.D.,  
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In the course of my asylum experience, this is the best example of the disease I ever met with, and I am therefore tempted to place it on record.

Diffused cerebral sclerosis has not attracted much attention of late years, and is, as a rule, only referred to incidentally as one of the causes of idiocy. I know of no works where it is systematically described, except two, both American. The account given by Hammond is very full and interesting, that by Hamilton is more condensed. Hammond gives numerous references to the literature of the disease, mostly French and German.

At the time of making the post-mortem, I was not fully alive to the importance of a microscopic examination, not only of the cerebral convolutions, but also of the ganglia and spinal cord. Such an examination could not have failed to afford most valuable information as to the secondary affection of these parts. So, for some purposes, this record is admitted to be imperfect. Only one or two small portions of the frontal convolutions, where the disease appeared most advanced, were prepared for further study, and I must take this opportunity of thanking my friend Dr. Major for preparing the sections and giving me great assistance in arriving at a correct idea as to the nature of the morbid changes observed.

The following history is abridged from the Case Book :—

W. B., æt. 18, admitted 19th Sept., 1877. The medical certificate states that he is quite deaf and unable to answer questions; in fact, is quite idiotic. His mother says that he eats his shirts and clothes when he can get at them. *History* (from father).—He is the eldest of six children, and was a strong, healthy lad until about three years ago, when he sustained a severe injury of the head (left occipital region), by falling from the top of a lime kiln. He was able to walk home after the accident; but whilst telling his parents what had happened, he was seized by a succession of severe fits. For six months they recurred at irregular intervals, and during that time he was confined to bed, his mind gradually becoming weak. At last he began to improve, and became so well, both in body and mind, that he was almost fit for work again. Eighteen months ago he went to bed in his then usual condition; when he awoke he was paralysed on the left side. Since that time he has gradually become worse in every respect.

For the last six months he has shouted much at night. He is now, and has been for months, dirty in his habits. Since the paralysis

occurred, speech, with other manifestations of intelligence, has gradually left him. Except two vulgar words, he had said nothing for several months, when, last night, much to the surprise of his friends, he shouted "thimble."

*State on Admission.*—He is a typical case of organic dementia. Every trace of mind appears to be gone. He lies in bed all day, unable to move, a perfect skeleton. Whenever he can manage to get a piece of sheeting &c., near his mouth, he immediately begins chewing it. If care were not taken he might choke himself, for it is surprising to see the amount of cloth he can draw into his mouth in a few minutes. He frequently swears: this is his only manifestation of intelligence. All his limbs are contracted; his arms are strongly flexed across the chest and his knees almost touch his chin. The fingers are flexed, and press against the palm. When he wishes to get hold of a sheet he can stretch out his arm slightly, and catch it by bringing the thumb and side of index finger together. Grinding of the teeth is nearly constant. As he lies in his bed he rolls his head from side to side, and utters a sound something between moaning and humming. The attendant says that the song, "Not for Joe," can be occasionally recognised, but, as a rule, the humming is meaningless. He takes his food readily, but without evincing any pleasure; his jaws move freely, and he swallows mince, &c., with ease.

There is no squinting, but left pupil is slightly larger than right.

There is no external indications of any head injury, but there is a purulent discharge from the left ear.

5th Oct. Still confined to bed: wet and dirty. At night he is generally very noisy, shouting and screaming as if in pain.

2nd Nov. He has rapidly become more feeble; bedsores have appeared, and he is rapidly sinking. To-day he had two fits, but beyond the fact that they were severe, no description of them can be obtained from the attendant.

5th Nov. Died at noon.

*Post-mortem Examination* 22 hours after death. Weather cold and unsettled. Post-mortem rigidity well marked in the lower limbs. The body is much emaciated, and there is an extensive bed sore over the left trochanter, and there are smaller ones over the right and left iliac crests. There is no mark of injury. A considerable quantity of thick purulent discharge has escaped from the right ear.

On reflecting the scalp, a marked inequality on the surface of the left frontal eminence was observed. It has the appearance of an old punctured wound. Calvarium is of average thickness and density. The punctured depression observed on the outer table has not implicated the inner one.

During the removal of the brain, a large quantity of serum escaped. The arachnoid is slightly opaque in every direction, and is everywhere adherent to the convolutions. There is no trace of atheroma of the vessels at the base. The consistence of the brain is quite altered; this change is specially marked in the convolutions of the frontal and

parietal regions which feel quite hard. The whole of the cerebral hemispheres appear atrophied, even the occipital and temporo-sphenoidal lobes; but these are less affected than the superior and anterior. On section the grey matter is much atrophied, dark, and hard to the touch in most places. The white matter is also firmer than normal. In the middle of the right centrum ovale, on a level with the floor of the lateral ventricle, there are two small patches of softening. Both lateral ventricles are much distended by fluid, and their lining membrane roughened. In the floor of the right lateral ventricle, at the junction of the corpus striatum and optic thalamus, there is a patch of yellow softening. Superficially its area is very small, but it penetrates rather deeply, about  $\frac{1}{2}$  inch. The floor of the fourth ventricle is markedly granular.

*Heart.*—Very small, otherwise normal.

*Lungs.*—Somewhat œdematous and congested posteriorly.

*Liver.*—Gall-bladder distended by dark thick bile; normal but firm.

*Spleen.*—Normal but firm.

*Kidneys.*—Normal.

*Bladder.*—Considerably distended and coats thickened.

*Intestines* distended by flatus; numerous common thread-worms.

*Weights.*—Brain,  $38\frac{1}{2}$  oz.; heart,  $3\frac{1}{4}$  oz.; right lung, 10 oz.; left  $16\frac{1}{2}$  oz.; liver,  $18\frac{1}{2}$  oz.; spleen,  $1\frac{1}{2}$  oz.; kidneys, each,  $2\frac{1}{2}$  oz.

*Remarks.*—In his work on diseases of the nervous system, Dr. Hammond quotes at length several cases recorded by Pinel the younger, and others. It is worthy of special remark that they are all, with one exception, idiots in whom the disease began in early infancy. The exception is that of a gentleman, æt. 43, who had been ill six months, and in whom Dr. Hammond diagnosed diffused cerebral sclerosis; but, the patient being alive, no post-mortem notes are forthcoming to certify the correctness of his conclusion, and I must confess that the short history he gives does not conclusively set the question at rest.

Its predisposing causes are unknown, and the same may almost be said of its exciting causes; but “injuries of the skull from falls or blows and hæmorrhagic cysts appear to have some influence in originating the disease, but more generally it is developed, so far as we can perceive, spontaneously.” P. 278.

Relative to its morbid anatomy, it may be remarked in passing, that Pinel makes the curious statement that the diseased process does not invade the grey matter. This is not in accordance with my experience. In the present case the hardening and atrophy of the grey matter were very marked, and the same was true in some idiots’ brains I have

from time to time seen, but in these the lesion did not by any means involve such a wide area.

The microscopic characters of the lesion are well known, and to these I would not have referred, had it not been for the fact that the section prepared for me by Dr. Major was studded by those curious bodies first described by Dr. Batty Tuke as miliary sclerosis. It is now several years since I had the pleasure of examining Dr. Tuke's sections, and whilst I could not deny the accuracy with which he described the appearances, I always had a doubt as to the correctness of the name he had attached to them. During the eighteen months I worked as pathologist at the West Riding Asylum I never succeeded in getting a section containing the bodies described by him. During a late visit there, however, I saw several sections in which they existed in a variety of conditions, some of them still undescribed. Opinion differed widely as to their nature and conditions of development, and it was even suggested that they might be the products of putrefaction. It is, however, satisfactory to know that Dr. Bevan Lewis has taken the subject up; we may, therefore, look forward to some further additions to our knowledge.

As the section interested me, I submitted it to several microscopists. Dr. Batty Tuke agreed that it was an illustration of what he had described, miliary sclerosis in the first stage, a morbid condition he had often noticed. He considered that along with the history of the case the preparation was very interesting.

On consulting Dr. J. J. Brown, of the Fife and Kinross Asylum, he had the great kindness to prepare a complete report of the appearances. From his letter I make the following extracts:—

“The spots are, I think, unilocular miliary sclerosis (B. Tuke), and are quite distinctly seen to be such in the large spots; their margins are irregular, and the brain substance just external to the margins is slightly of a deeper hue, which is the case in miliary sclerosis. This, of course, is not well marked, as the disease is not far advanced; or perhaps I should rather say, the spots being small have not caused the same amount of compression of the surrounding brain-substance as is the case in larger spots, and so the brain-substance takes on a deeper stain. The spots themselves are unstained, which puts amyloid bodies out of the question, as they, as a rule, stain deeply; besides, their margins are different.

“These miliary spots do not take on the stain in your

slide, which is the case with this disease, but they are not of the transparent appearance which characterises colloid bodies. Colloid bodies have clearly defined, regular margins; they are as like boiled sago grains as they well can be. By varying the focus and shades of light the larger spots have a slightly yellow tinge, just resembling miliary spots when large or advanced in age.

"There are some smaller spots in this section which are more difficult to deal with, they being smaller, more circular, more transparent, and altogether resembling colloid bodies. But these, I think, are the miliary spots in the first stage of the disease. Curiously enough, the last two brains I have examined have been exactly similar to this slide of yours, and I was very much puzzled by these smaller, somewhat transparent spots, with almost circular margins; but from an examination of many sections and various stainings, I believe they are the early stage of miliary sclerosis, and exactly the same degeneration as exists in your case. At first I thought they might be albuminoid, but I have not been able to make this out."

The accompanying illustration is only intended to show the position and shape of some of the spots. They measure from .018 to .023 mm., and appear covered by a fine fibrillar network on shallow focusing. Had a complete drawing been made it would have shown, 1st, the fibrous transformation of the outer part of the first layer of the cortex; 2nd, large and numerous Deiter cells in the same region; 3rd, spots of miliary sclerosis scattered throughout the grey matter.

In concluding the record of this case, it may be well for me to point out the utter confusion which now exists in the terms in use in the literature of sclerosis. Microscopists attach to "unilocular" and "multilocular" certain meanings; systematic writers, as Charcot, attach others; and to make matters perfectly hopeless, in translating Charcot's work on Sclerosis, the writer confounds the terms. In the original there is "unilocular," in the translation it appears as "multilocular." Again, by some writers miliary sclerosis is confounded with grey degeneration or diffused sclerosis. (See Dr. Kesteven's paper, "Miliary Sclerosis," in the "Brit. and For. Med. Chir. Rev.," July, 1874).

#### DESCRIPTION OF PLATE.

Fig. I.—First (outermost) cortical layer, showing spots of miliary sclerosis. 450 diameters.

Fig. II.—Portion of the same section at the level of the third cortical layer. Patches of miliary sclerosis with crenated edges, are seen scattered among the nerve cells.