A Case of Endarteritis with Insanity and Aphasia. By Richard B. Mitchell, M.B., C.M., Assistant Medical Officer, Fife and Kinross District Lunatic Asylum.

(With Illustrations.)

(Read at a Quarterly Meeting of the Medico-Psychological Association held at Glasgow on March 8th, 1882.)

Mrs. N., æt. 40, married, teacher. Admitted to the Fife and Kinross District Asylum on June 9th, 1880.

History.—Her mother was said to be "weak-minded," but apart from this there was no hereditary predisposition to insanity, and the family history was good.

Her abilities had been fair, and she had received an average education. This was her first attack of insanity, and no cause for it was known. She had been married for 23 years, but had never had a child. She had been three years in India, but stood the climate very well. For the last two years previous to admission she had suffered from menorrhagia every fortnight. There was no history of venereal disease. Habits had always been very steady; she had always been a total abstainer, and "very religious." In January, 1879, she suddenly dropped her cup and saucer one morning, and she "seemed to have lost the power of feeling things," as very soon after that day it was noticed that her hands, but especially the right, trembled a good deal when she was fastening buttons or tying knots, and that she had difficulty in the performing of these acts. Now and then she complained of very severe pains in the calves of the legs, and she got very soon fatigued if she went for a walk outside. The motor symptoms went on gradually increasing for about six months, and then one day, on coming in from a walk, she was noticed to be confused mentally, and resembled in her general appearance "a person who had been drinking." This state lasted only a few seconds, and she then went about her work as usual.

In October new symptoms became developed. It was noticed that she spoke "as if her tongue were too big for her mouth," or "like a person who speaks thick from the effects of liquor," and she often made mistakes in words. Thus she would say, "What's the matter?" when she really wished to say, "What o'clock is it?" She also began to be childish in her manners. About this time twitchings of the lips, the tongue, and the muscles of the legs were often observed in addition to the trembling of the hands, and the pupils were noticed to be contracted and unequal. Her control over the movements of the lower limbs became diminished, so much so at times that she could scarcely walk.

During the next six months the gravity of the mental and motor symptoms continued, on the whole, to increase, but there were pretty frequent remissions. She often now had difficulty in expressing her-

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self in articulate speech, as she often said "yes" when she meant "no," and *vice versâ*. At the same time she often had less difficulty in spelling and pronouncing long words than short ones. The nervous and mental disturbance was more marked before each attack of menorrhagia. During March, 1880, she was treated in an infirmary. She could generally get up and go about the ward, and could generally feed herself, but was sometimes unable to do so from her inability to control her movements. She was subject to fits of screaming, and was sometimes very childish. It was observed that her speech had a *staccato* character. The case was at that time diagnosed as general paralysis of the insane.

Her state on admission to the Fife and Kinross Asylum, in June, 1880, was as follows:—

Thoracic and abdominal viscera healthy. Common sensibility and sensibility to pain pretty acute all over the body; plantar skin-reflex much exaggerated, and patellar tendon-reflex also, especially on the right side. Right pupil contracted, left normal size; both sluggish to action of light. Tip of tongue could not be protruded beyond the teeth. Sides of face equal as regards the lines of expression and motility, and patient looked fairly intelligent. There were slight fibrillary tremors in all the facial muscles. The chin was often jerked spasmodically up and down, or backwards and forwards. When patient was taken by surprise she often made a snorting sound in her throat, as if there were some paralysis of the soft palate. She snored heavily while asleep. Mastication and deglutition were performed clumsily, but without marked difficulty. Phonation normal. The fingers were semi-flexed, and resisted extension. The arms, legs, hands, and feet were almost constantly shaken with tremors, which were most marked on the right side. Voluntary movement was accompanied by extreme and uncontrollable spasmodic action in the limbs. Delicacy and precision of prehensile movements, and, in fact, of all manual movements, were much impaired. When asked to take hold of a light object, such as a pencil, she obeyed in a clutching, fumbling, uncertain way, and she could not hold it properly. There was a considerable degree of paresis in the upper and lower extremities, besides the more marked ataxy and tremors. She could not walk or stand without support; otherwise she could move her legs pretty freely, but in a most disorderly, ataxic style.

There was considerable emotionalism and mental enfeeblement, but she understood what was said to her if it was expressed distinctly. She took considerable interest in her surroundings, and seemed, on the whole, to be pretty well satisfied with them, but she was subject to periods of screaming for which there was no apparent cause.

As regards language, she had almost lost the power to express her thoughts in articulate speech. When questioned she tried hard to reply, but as a rule she only succeeded in uttering a string of short interjections, such as "Oh—oh—oh," or "Aye—aye—aye." She had

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scarcely any propositions besides "yes" and "no," but she had not always the propositional use even of these words; they were often as articulated by her mere "utterances." Thus, after saying "yes" or "no" *propositionally*, she often kept on uttering "yes" or "no" in reply to everything said to her, all the time apparently quite unconscious of the unsuitability of her replies in many of the cases. Often she uttered "no" when she meant "yes," and *vice versâ*. Many of her utterances occurred unexpectedly, and as a rule they were articulated in a slow, subdued manner, but they were sometimes suddenly "rapped out." As a rule, the more one tried to make her speak the more inarticulate she became. Sometimes she repeated a word uttered in her hearing, *but she could rarely repeat a word when asked to do so.* She would, on request, read aloud a few lines from a printed book with distinctly less difficulty than she showed in original propositionizing, but at the same time she resembled one who was only learning to read, and to whom the letters and words were still unfamiliar.

Even allowing for the ataxia, her power of pantomime was distinctly impaired. She aided her defective articulation by signs, but she nevertheless often shook her head for assent and nodded when she meant "No."

During the first ten days after admission the nervous and mental phenomena varied in degree from time to time, but motility, on the whole, improved so far that she could walk a few steps by herself and hold a spoon properly. Articulation also improved, and at three different times she said intelligently, "Good-night," "Good-morning," and "Some better." On one occasion, when asked her name, she said, "I won't tell you," and immediately afterwards, when asked to write, she said, "I couldn't." The average morning temperature was 98·2° F., evening 99·3°, and the maximum evening temperature 100·6°.

By the 29th June, three weeks after admission, the motor powers had improved considerably, and the intelligence was clearer; the tremors were much less marked; she could walk better, though very unsteadily, and she fed herself at all her meals. The tip of the tongue could be protruded beyond the teeth, and the articulation was much improved, although markedly halting and laborious, and at times "explosive" in character. She tried to write to her husband at this time after much coaxing. The result of the attempt occupied about five lines altogether, consisting chiefly of irregular scratches and obscure characters. She began, "*Dear*," "*Dear*" (very indistinctly written); then came a number of irregularly formed letters and scratches, which might stand for "*I am disappointed;*" then more illegible scratches, and then "*You come to see me*" (pretty distinctly written). "*Never*" was the last legible word, and between it and the signature (which was repeated and wrongly spelt both times) came a number of runic-looking marks.

After the above date a relapse occurred. The tremors again became

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very marked, as well as the general ataxy and paresis, and the articulation was much worse. She became more enfeebled mentally, and then became completely demented. Yet about the middle of September there were three occasions, but only three, on which she had utterances of high speech-value. She was asked how she had liked India, and she at once replied, "Oh—I—liked—it—first—r-rate," immediately afterwards adding, in the same drawling, syllabic way, "I—don't—know—how—ma-ny—I—h-had," when asked how large her class was. On the other two occasions about this time she said intelligently "Good-night" and "Good-morning." After this her utterances were limited to "Yes" and "No," and for the last six weeks of her life she was entirely mute. She recognised her husband shortly before her death, and showed her pleasure at seeing him by giving forth cries and inarticulate sounds.

Bed-sores formed over both trochanters 70 days before death. On December 25th she had an "epileptiform seizure." Three days afterwards rapid and violent spasmodic twitches of the muscles set in all over the body, and remained almost constantly for a week. There was now rigid flexed contracture of the limbs at all the joints. Patient became reduced to the last stage of weakness, and died on February 4th, 1881, after an illness lasting in all 25 months.

Autopsy 40 hours after death.—Body much emaciated. Bed-sore over each trochanter, one over right ilium, and a small one over right acromion process. Rigid flexed contracture of all the joints of the limbs, most marked in the lower limbs.

Heart healthy. Aorta very much thickened, and its inner coats thrown longitudinally into numerous folds.

Lungs.—Right slightly adherent at the base; the lower lobe contained a cavity the size of a hazel-nut, and in the apex was another cavity the size of a walnut, both containing a dark, inodorous fluid. Left healthy.

Liver cut more firmly than is usual, but appeared to be healthy. No evidence of waxy disease.

Spleen normal.

Kidneys.—Both cut very firmly. Pelvis of right was dilated and filled with urine, as was also the ureter leading from it.

An ovarian tumour, about the size of a fœtal head at the full time, was found in connection with the right ovary, filling up this side of the pelvis and compressing the ureter and other organs.

Spinal Canal.—Vertebral laminæ appeared somewhat harder than normal. Subarachnoid fluid increased in amount. Membranes of cord healthy.

Spinal Cord.—The cervical and upper two-thirds of the dorsal region seemed of normal consistence, but the lower third of the dorsal and the lumbar were firm and resistent, and could be, as a whole, lifted up by the middle almost without bending. Section of cervical and upper two-thirds of dorsal was pale, and showed the grey matter

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indistinctly defined from the white. Lower third of dorsal and lumbar cut crisply; periphery of section was grey, and was firm to the touch, this being most marked in the posterior and lateral columns of the lumbar region.

Cranium.—Skull-cap symmetrical; diploe diminished. Some roughness of inner surface of frontal bone on both sides at vertex. Dura mater somewhat thickened and anæmic. Pia mater much thickened and fibrous, opaque, milky, and anæmic; it stripped freely from the subjacent grey matter. Gyri were much atrophied generally; sulci wide, and filled by clear serous fluid. On right hemisphere, where second frontal gyrus joined the ascending frontal, there was a distinct localised atrophy the size of a filbert, and a similar atrophy the size of a hazel-nut occupied an exactly corresponding spot on the left hemisphere. The third left frontal gyrus was especially atrophied, and the gyri in its immediate neighbourhood were more atrophied than those of the corresponding region of the right hemisphere. The basilar artery and the vessels of the *circle of Willis* and its branches were studded with a number of oval, thickened, whitish spots varying in size from a pin's head to a split pea. The membranes at the base of the brain and the *lamina cinerea* were tough. Fourth and lateral ventricles had their ependyma thickened and covered with abundant granulations. Cerebrum on section cut crisply; white matter was pale, grey matter diminished in depth, the outer layer of it having a pale gelatinous, semi-transparent appearance, and felt firm and resistent to the touch; inner layer was congested. Cerebellum, pons, and medulla fairly normal on section.

Encephalon weighed 46 ounces.

Microscopical Examination.

Cerebrum.—The pia mater was everywhere much thickened, and in some places markedly so, as over Broca's convolution and those surrounding it where there were numerous fibrous layers, and abundance of nuclei between the layers. The vessels were thickened, and were dilated with blood. Some small arteries were distended with a yellow granular-looking material, and all those above $\frac{1}{400}$ of an inch in diameter presented on transverse section a wavy and crenated appearance, due to the inner coats being wrinkled and thrown longitudinally into folds. There were no miliary aneurisms. The neuroglia was everywhere much increased as regards its fibrillar element, but there was only a slight degree of proliferation of its nuclei. Patches of "miliary sclerosis" in the first stage were abundant throughout the convolutions, chiefly in the white matter. They varied in size from $\frac{1}{100}$ of an inch downwards, and the majority of them had a greenish yellow tinge, probably due to the chromic acid in which the preparations were hardened. In more than one case a patch was seen embraced by the two branches of a vessel at its bifurcation. In two cases a vessel could be dimly seen through a

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patch, and apparently bent down into a deeper plane by it. In many instances patches were seen in groups of three or four, fusing together by their borders.

A. *Frontal Lobes.*—There were numerous empty vacuoles in the more superficial layers of the grey matter. The nerve-cells were, as a rule, diminished in number and in size; they were generally surrounded by vacuoles, and they stained dimly with carmine. Some were rounded, some shrivelled, and most showed a greater or less degree of yellow granular degeneration, although in no case observed had this process gone on to cell-disintegration. In most cases the processes were very indistinct, and the nucleus and nucleolus invisible.

As a rule the perivascular canals were widely dilated, and proliferation of the nuclei of the perivascular sheath was well seen. The vessels themselves were, in many cases, highly tortuous; they were dilated in an irregular manner, and, in many places, were distended with blood corpuscles; their walls—between and outside of which lay numerous haimatoidin granules—were distinctly thickened, especially the muscular coat and the tunica adventitia.

Many of the smaller vessels were occluded, apparently by slow inflammatory changes in their inner walls, and these, along with the capillaries, exhibited well-marked proliferation of their nuclei.

In the deepest layers of the grey matter, and in the immediately subjacent white matter, a large number of round bodies were seen (see Fig. iii.) varying in size from $\frac{1}{200}$ of an inch downwards. They were deeply stained with carmine. The largest of them presented a rough, mulberry-like appearance, and were evidently composed of a number of bodies identical with, if not actually, amyloid corpuscles. In every case these aggregations were in the neighbourhood of a vessel.

All the above appearances were most marked in the third left frontal gyrus in its posterior portion.

In the grey matter of the right ascending frontal gyrus were a few unevenly oval empty spaces surrounded by a deeply stained area of brain-substance. There was hæmatine close to them, and the broken end of a vessel terminated in one of the spaces. A number of small, rounded, apparently non-nucleated bodies with short curved processes were seen in most sections; they were more frequent in the grey substance, and especially so in the posterior tip of the third left frontal gyrus.

B. *Parietal Lobes.*—The convolutions on the right side presented the same morbid appearances as those seen in the frontal lobes, only in a less marked degree. On the left side the cells stained much better, and were more healthy. In the fourth layer of the grey matter were one or two very large bloated pyramidal cells with the nucleus and nucleolus very sharply defined, but the contour of these cells was dim, and they were being extensively invaded by yellow granular degeneration.

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C. *Occipital Lobes.*—These presented a more normal appearance than the frontals and parietals. The cells stained deeply, and had sharply defined borders. The vessels, however, had the same appearances as are noted above, although in a less degree.

D. *Corpus striatum* showed great richness of the neurogliar nuclei, and a few of the bodies represented in Fig. iii. were present.

Cerebellum presented nothing abnormal beyond a few very small patches of miliary sclerosis.

Right Crus Cerebri.—Many of the multipolar cells of the *locus niger* were pigmented to an extreme degree, and some were represented merely by dark brown granular masses without poles.

Pons Varolii.—Many of the cells showed nucleus and nucleolus very distinctly, but contour and processes were not as a rule well defined. There were numerous patches of miliary sclerosis, and in one case a patch was seen pushing a vessel aside, and compressing it so as to entirely occlude its lumen. (Fig. i.)

Medulla Oblongata.—The olivary cells were, nearly all, affected by yellow granular degeneration: they did not stain at all well, and their processes were very indistinct. Some of the cells in the floor of the fourth ventricle were highly fuscous, several being represented merely by dark brown granular balls. Periphery of the grey tubercles of Rolando fairly healthy. The vessels near the nuclei of the hypoglossal nerves were in most cases dilated and engorged with blood. A few scattered *corpora amylacea* lay close to the floor of the fourth ventricle.

Spinal Cord.—Pia mater was not much thickened, but its vessels presented the same appearance on transverse section as those in the cerebral pia mater. The fibrous septa of the cord were increased in number and in size, and there was a marked general hyperplasia of the connective tissue elements at the expense of the nerve-tubes, this being most marked in the posterior columns, and in Goll's columns. This sclerosed condition was most evident in the lower third of the dorsal and in the lumbar portion of the cord. The cells of the "cornua" presented a fairly normal appearance, being only in a slight degree affected with yellow degeneration. Some of the cells contained a number of minute black granules. *Corpora amylacea* were pretty numerous, and were best seen over the origins of the nerve roots; they were most abundant in the lumbar portion of the cord. No part of the cord presented miliary sclerosis except the dorsal portion, but in it the patches were extremely numerous.

Middle Cervical Sympathetic Ganglion.—The nucleated fibrous capsule around each cell was very clearly seen, and was sharply defined. Many of the cells contained yellow granular material; some of them were filled with it, and in these cases the nucleus was invisible, and the processes and contour of cell were indistinct. The connective-tissue framework between the cell-capsules contained an abundance of fusiform and other nuclei.

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I.—As regards the patient's practically speechless condition, which is the most interesting feature of the symptomatology of this case. Failure of ideas and of the understanding had apparently not much to do with the speech affection. *She understood what was said to her.* Thus, for example, she attempted to read aloud when she was asked to do so, and she tried hard, and with an intelligent expression, to reply to questions. In short, she obeyed orders to the best of her ability, except when she was more than usually obstinate. It is therefore indicated that her "nervous arrangements for words used in understanding speech" were not so very gravely disturbed. But both an ataxic and an amnesic element seem to be present in the case.

That ataxy and paresis of the articulatory muscles accounted in part for the speech-phenomena observed seems certain.

For, 1st, the gravity of the speech-affection varied as a rule in the same ratio with the degree of ataxia displayed in the tongue, as well as with the degree of ataxia generally.

2nd. The speech was slow, laboured, and often "explosive" in character.

3rd. Patient became more inarticulate the harder she tried to speak.

4th. She was rarely able to repeat a word *when asked to do so.*

In harmony with these facts is the unhealthy state of the cells in the nuclei of orgin of the ninth nerves, and of those in the *corpus dentatum* of the medulla.

But it seems impossible that ataxy of the muscles of articulation alone can account for such manifestations as (*a*) her often saying "Yes" when she meant "No," and *vice versâ ;* (*b*) her often saying "Yes" or "No" indiscriminately in reply to all questions ; (*c*) her often nodding the head for dissent, and again shaking it for affirmation. Another element seems to come in here, and a further defect to be indicated, a defect apparently of an *amnesic* nature, viz., a form of "inco-ordinate defect of memory," in which "the person cannot recollect the words in which he wishes to express himself . . . . , and is moreover unconscious that he makes use of wrong words."* This would correspond

* Bastain on "Loss of Speech in Cerebral Disease."—"Med. Chir. Review," January, 1869, p. 212.

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FIG. 1.

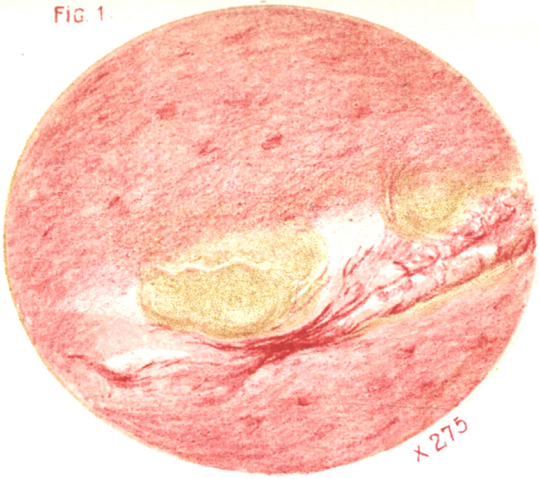


FIG. 2.

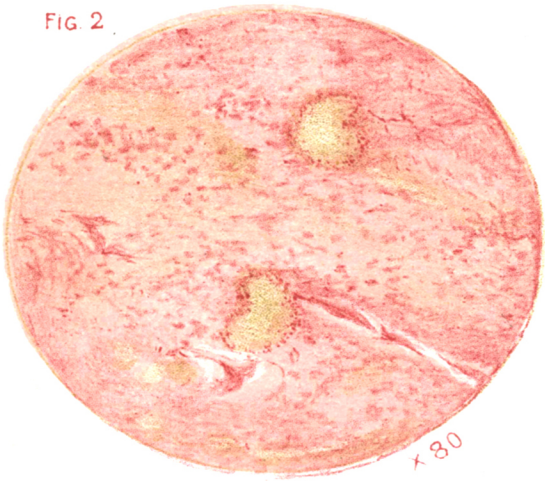
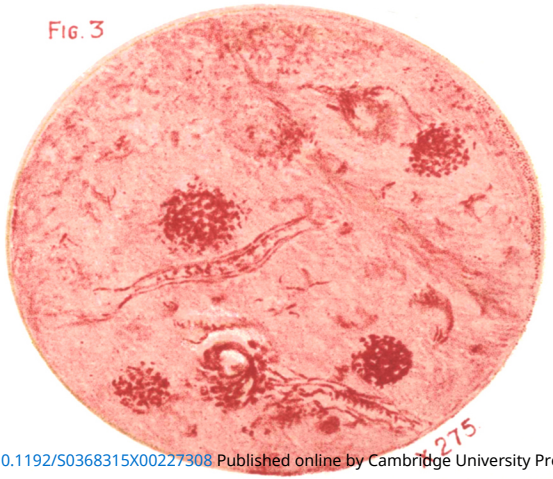


FIG. 3.



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To Illustrate Dr Mitchell's Case

with the lesions in the frontal gyri, among which Broca's convolution was distinctly the most affected. The "defect of speech" which the patient showed in October, 1879, several weeks before the establishment of mental symptoms at all, is evidently of the same nature (*i.e.*, amnesic), and was produced probably by disturbances in the intra-cranial blood-supply caused by the inflammatory changes in the minute blood vessels—changes that led ultimately to the occlusion of many of them.

II.—As regards the presence of miliary sclerosis in the cerebro-spinal system, it is worthy of note that

1st. The patches were numerous and distinct in sections that had been exposed to the action of alcohol for a period of not more than thirty hours altogether.

2nd. No trace of miliary sclerosis was detected in the corpus striatum, the medulla oblongata, and the cervical region of the cord, although these were exposed to the action of alcohol for exactly the same length of time and under precisely the same conditions as the right ascending frontal gyrus, left vertex, and other portions of the brain, and spinal cord, which were more or less thickly studded with well-marked examples of the lesion.

3rd. The patches were generally as distinct in those sections that had been exposed a *short* time to the action of the chromic acid used for hardening the nerve-tissues, as those exposed for a longer period.

III.—So far as I have been able to ascertain, the bodies represented in Fig. iii. have not been described previously. I have submitted several of the sections containing them to Dr. Batty Tuke, of Edinburgh, and he assures me that he does not remember having ever seen them before. It is difficult to suggest a theory of their origin. In some cases one was inclined to think that they represented the position of a vessel which had become obliterated, and that its space was now represented by a mass of proliferated nuclei.

EXPLANATION OF PLATE.

Fig. I. shows vessel pushed aside, and compressed by a mass of miliary sclerosis. × 275.

Fig. II. shows diffused masses of miliary sclerosis with induration of surrounding tissue. × 80.

Fig. III. shows aggregations of amyloid bodies or nuclei. × 275.

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