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On a case of acute optic neuritis associated with acute myelitis.

By SEYMOUR J. SHARKEY, M.B., and J. B. LAWFORD.

G. T—, æt. 17, kitchen-maid, was admitted into the Royal London Ophthalmic Hospital, under the care of Mr. Streatfeild, on November 22nd, 1883.

There was nothing noteworthy in the patient's family history, except that she was one of sixteen children, seven of whom died as infants. Eight were alive and well, four of whom were younger than the patient. There were no grounds for suspecting syphilis. She had always been delicate, but never seriously ill. Menstruation was regular but profuse.

Vision began to fail on the 9th of November, about a fortnight before she came under observation, and by the 13th she was quite blind. She had had neither headache, sickness, paralysis, nor fits, nor could she suggest any cause which might have given rise to her illness.

On admission she was found to be a well-nourished girl, looking tolerably healthy, but anæmic. She had no complaint to make except of her blindness. All the functions seemed to be well performed. The thyroid gland was the seat of a simple hypertrophy, and the patient said it had been large as long as she could recollect.

On examination of the eyes the pupils were found to be unduly dilated and motionless to light, but there was no paralysis of ocular muscles. Well-marked optic neuritis was seen in both eyes, accompanied by much swelling of the discs and of the retina immediately surrounding them.

The veins were very tortuous, but there were no hæmorrhages and no choroidal changes. She had no perception of light. She was given a mixture containing ten grains of iodide of potassium, and in addition two grains of grey powder, and three of Dover's powder, three times a day.

On December 6th the pupils were found to act well to accommodation, but not to light.

On the 10th it was noted that no marked salivation had been produced. Both optic discs were decidedly swollen and their outlines lost, but the veins were not so tortuous as on admission, nor did the inflammatory swelling extend so far. There were no hæmorrhages. Five grains of blue pill and a quarter of a grain of the extract of opium were now given twice a day.

On the 12th, *i.e.* thirty-three days after her vision began to fail, she complained of weakness of the left leg, though she said she had noticed it coming on four days previously. On examination she was found to be unable to walk, although she could do so the day before. The left leg was almost powerless and slightly rigid, and sensation in it was impaired. The knee-jerk was much more marked than in the right leg. She had no pain in the head or elsewhere, no paralysis except in the left leg, and no abnormal mental symptoms.

13th.—Sensation almost absent in left leg and much impaired in right. Complete paralysis of left leg, slight loss of power in right. Plantar reflex absent on left side, fairly marked on right. Knee-jerk very marked on both sides. Slight œdema of both legs, more marked in left. No paralysis of arms or face; no headache, pain in back, or incontinence of urine.

14th.—Anæsthesia extending up the left side of the trunk, reaching as high as the nipple in front and to a finger's breadth below the spine of the scapula behind. Left leg powerless and quite anæsthetic; right partially anæsthetic and more or less paralysed. Urine passed in bed for the first time. No paralysis or anæsthesia of arms, no headache, delirium, or fits, and no paralysis of

cranial nerves. In the morning, shortly after the foregoing note was made, the patient became very excited, tossing her arms and right leg about, while the left leg remained motionless. When questioned she said that she had no pain, but that she could not speak. She appeared in great mental distress, but answered questions rationally and used the proper words. Soon she had a fit, but only the right arm and leg were convulsed, and she was not unconscious. The fit lasted about ten minutes.

In the afternoon of the same day she was much quieter, but still tossed her arms and right leg about; the latter was quite anæsthetic, while the left had not only lost sensation, but was likewise completely paralysed. There was marked flushing of the face during the period of excitement; the pupils were equal and dilated. The temperature at 5.30 p.m. was 100.2° F., the pulse 96 and regular, the bowels constipated.

15th.—Quiet all night; paralytic condition the same as yesterday. Emotional and excited at times. Evacuations passed involuntarily. No sickness, no headache; tongue coated. Pulse 106, regular; temperature 100° F. Lungs and heart normal. The patient was now admitted into St. Thomas's Hospital under the care of Dr. Bristowe.

19th.—Seventy-six ounces of urine were drawn off, and it was found to be clear and acid, but offensive and containing a trace of albumen.

22nd.—Examination showed that there was complete paralysis of both legs and loss of control over the evacuations, and, in addition, evident weakness of the left hand. Loss of sensation was found not only in the legs and over the greater part of the trunk, but likewise in the left forearm and hand on the ulnar side. There was no ankle-clonus; the knee-jerk was excessive on the left side, about natural on the right. No tendon reflexes obtained in the arms.

23rd.—Sensation impaired in right forearm and hand. Complaints made of pain in the lower part of the abdomen.

26th.—Urine muddy, offensive, alkaline, and containing triple phosphates, mucus, red blood-corpuscles, and leucocytes.

January 4th.—The condition of legs remained the same. Sensation was impaired in the forearms. The urine was offensive and contained pus. Dr. Kilner examined the muscles and nerves electrically and reported that they presented the "reaction of degeneration." The temperature, which had been only moderately raised up to December 29th, after that became very high and of an intermittent type, ranging from about 101° F. to 104° F. or 105° F. The patient also complained of pain in the epigastrium and right side.

After this but little alteration occurred except that the presence of peritonitis became evident, and slight double external strabismus was noticed. The patient died on January 10th, sixty-two days from the time when her vision first failed and twenty-nine days after the first appearance of symptoms of paralysis.

Autopsy (twenty-nine hours after death).—Body well nourished; subcutaneous fat abundant. On opening the abdominal cavity, acute peritonitis was found, though the inflammation was most intense in the pelvis. When the abdominal wall above the pubic symphysis was cut through the subperitoneal connective tissue in that region was seen to be infiltrated with pus; and this inflammation, though external to the bladder, appeared to have been caused by the acute cystitis which was present, the inflammatory process having spread through the wall of the viscus and attacked the connective tissue around. The peritonitis too seemed to have had a similar origin.

The ovaries were swollen and hyperæmic and the mucous membrane of the uterus was intensely congested.

In addition to very acute cystitis there was inflammation of the ureters and of the pelvis of both kidneys. The latter were much enlarged, hyperæmic, soft, and juicy. Their capsules were thin and peeled off easily, and groups of suppurating points were seen in the cortex beneath them.

These were the terminations of inflammatory streaks and lines of suppuration which radiated from the pelvis.

The left ventricle of the heart was slightly enlarged, but with this exception the intra-thoracic viscera were healthy.

The thyroid gland was considerably hypertrophied, but contained no cysts.

The liver was pale and soft, but otherwise normal.

The brain was in every respect normal except that the soft commissure was absent. No inflammation could be detected anywhere. The pituitary body was large, pale, and mottled.

The cerebral sinuses were natural and no abnormality of any of the cranial nerves was observed.

The spinal cord presented no abnormal appearances except over a space of two or three inches in length in the lower cervical and upper dorsal regions. Here it was intensely congested and much softened, but not diffuent. The section-surface was bright pink, and blood oozed from a great number of distended vessels. The cervical region above, and the dorsal and lumbar regions below, the softened part presented no evidences of disease. The membranes of the cord seemed healthy.

Microscopical examination of the spinal cord.—Sections of the spinal cord were cut, stained, and mounted in the usual way. Four regions were selected for this purpose:

1. The lower cervical region, where naked-eye changes due to disease were observed at the autopsy.

2. The upper cervical region, above the area of evident disease.

3. The upper lumbar region; and

4. The lumbar enlargement.

1. *Lower cervical region.*—On holding up a prepared section of this part of the cord towards the light it was seen to be almost homogeneous in appearance and to stain imperfectly. One could scarcely distinguish even the grey matter from the white. On further examination with the microscope the whole area of the section was

seen to be crowded with small cells, apparently leucocytes, which stained deeply with logwood. The vessels were greatly dilated, their sheaths were occupied by leucocytes, and many of the smaller ones were filled with these cells. The white substance of the nerves themselves was granular, and the large stellate and other cells were seen with great difficulty. Those which could be made out presented no processes, and their protoplasm in many instances had lost its natural granular appearance and was transformed into a homogeneous hyaline substance. All the nerve-cells and their nuclei took the staining either imperfectly or not at all. The pia mater had shared but slightly in the general inflammation. In short, that part of the cord which was soft and hyperæmic to the naked eye was the seat of an intense inflammatory process which had not, however, gone so far as to produce gross disorganisation.

2. *Cervical region above the seat of disease.*—Although at the post-mortem examination this part of the spinal cord presented no abnormal appearances, it did so after it had been hardened, and still more clearly after it had been stained and mounted. The columns of Goll, and these columns alone, were the seat of an acute inflammatory process, which was in an earlier stage than the myelitis below. Its distribution was just that of ascending degeneration.

3. *Upper lumbar region.*—Neither with the naked eye nor by the aid of the microscope could any disease be detected in this part of the cord, either in the lateral regions or elsewhere.

4. *The lumbar enlargement* presented morbid appearances to the naked eye (but only after the sections had been stained and mounted) in the columns of Goll. They appeared to be more transparent than the rest of the posterior columns, and resembled the central grey matter. The microscope showed that this was due to complete granular degeneration of the nerve-fibres; some of the vessels in this region were crowded with leucocytes, but otherwise there was but slight evidence of inflammation.

In the anterior cornua some of the vessels contained an abnormal number of leucocytes, and some of the ganglion cells were more homogeneous than they are in health. The membranes were everywhere healthy except in the lower cervical region, where the pia mater was seen to share in the general inflammation of the spinal cord.

Microscopical examination of the under surface of the frontal lobes of the brain, of the meninges about the chiasma and optic tracts, of the chiasma and tracts, and of the right optic nerve, disc, and retina.

Under surface of frontal lobes of brain.—Slight signs of inflammation were evident in the pia mater and in the small vessels entering the cortex, but the morbid changes could not be traced deeper than the two superficial layers of the grey matter. In the first and second layers, especially in the former, there seemed to be a larger number of connective-tissue corpuscles than usual.

Meninges about chiasma and optic tracts (teased) presented evident though slight signs of inflammation.

Optic tracts (transverse section).—Changes of equal degree were seen in both tracts. There was increase in the number of staining nuclei throughout them; near their periphery these nuclei formed a border two or three deep, which encircled the tracts. The small vessels showed distinct inflammatory changes. The larger nerve-cells in the brain substance on which the tracts rest appeared to be normal.

Chiasma (transverse section).—Marked signs of inflammation present. There was a great increase in the number of connective-tissue corpuscles throughout, and surrounding the small vessels were large accumulations of these cells. The coats of the small vessels were thickened, and studded with stained nuclei. Around most of the vessels were clear spaces, œdema spaces (?).

Right optic nerve to the naked eye appeared normal. In a transverse section, on a level with the optic

foramen thickening of the sheaths, especially of the inner, was observed. In the sheath-space there was a considerable amount of recent inflammatory material, as there was also in the small vessels. The trabeculæ in general were thickened, though they appeared more so in some places than in others. They enclosed many stained nuclei. The latter were also present in largely increased numbers throughout the bundles of nerve-fibres. Alongside the trabeculæ, between them and the nerve-fibres, were spaces containing a very faintly granular material which did not stain. These, which were probably lymph-spaces, were of greater width than is usually observed.

In transverse sections close to the globe were changes similar to those just described. The increase of stained nuclei appeared to be greater in the peripheral than in the central parts of the nerve.

Right disc and retina.—The sheath-space close to the disc was very slightly distended. Swelling, considerable in degree, involved the disc and the nerve-fibre layer for some distance towards the periphery. The stained nuclei were very numerous in the disc and in the two inner layers of the retina. Inflammatory changes were present in the vessels, and a large hæmorrhage was seen in the retina close to the disc, which involved all its structures except the nerve-fibre layer.

In short, microscopical examination proved that there was intense inflammation of the optic discs, nerves, and chiasma, and that it involved, though less severely, the optic tracts. The meninges about the chiasma and on the adjacent under surface of the frontal lobes presented slight evidences of inflammation.

The interest of the case just related lies in the association of an acute optic neuritis with acute inflammation of the spinal cord. Gowers in his work on 'Medical Ophthalmoscopy' refers to five cases recorded by Clifford Allbutt, Seguin, Noyes, Steffen, and Erb, in which spinal symptoms, apparently due to myelitis, were

present in connection with changes in the optic discs. Some of these, however, appear to have been uncertain in their pathology. It has long been known that affections of sight, which are generally due to slight changes in the discs and retina, occasionally occur in cases of spinal injury, and especially in injuries of the higher parts of the spinal column. But it is not so well known that optic neuritis is sometimes associated with spinal myelitis where no injury has been sustained.

In the 'Archives of Ophthalmology,' for 1882, No. II, edited by Knapp and Schweigger, a very interesting case is recorded by Julian J. Chisholm, M.D., which appears to correspond with ours except that it was much more rapidly fatal. Unfortunately an autopsy was not obtained. The patient was a man, *æt.* 28, healthy and robust. Without evident cause he suddenly felt pain on movement of the eyeballs, and his vision became slightly cloudy. By the third day he was completely blind. Then loss of power and sensation in the lower extremities supervened, and paralysis advancing upwards, similar to that in our patient, proved fatal on the twelfth day from the first sign of illness. His brain remained clear until a few hours before his death.

Dr. Dreschfeld published two cases, with accounts of the post-mortem examinations, in the 'Lancet' for 1882, and these are quoted by Dr. Gowers.

Case 1 was that of a married woman, *æt.* 38, in whom numbness and weakness occurred in the legs after exposure to cold three weeks before she came under observation. On admission into the hospital the lower extremities were found to be completely paralysed and the upper partially so, and she had retention of the evacuations. There was marked double optic neuritis, but vision was good.

Post-mortem examination revealed congestion of the cerebral membranes and excess of fluid in the ventricles. One and a half inches of the spinal cord at the cervical enlargement were exceedingly soft and of a yellowish colour.

Case 2 was that of a man, æt. 41, who had been intemperate and had had syphilis. One month before admission his sight failed and his legs began to get weak. On October 5th (apparently about a fortnight after he was taken ill) he was examined at the Manchester Eye Hospital and found to have optic neuritis, and at that time he could walk with the aid of a stick. On 13th he was quite blind, but could still walk with support. On 22nd he was admitted into the general hospital. He was quite blind, and the optic nerves were atrophied. He had complete paraplegia with analgesia of the legs, and with anæsthesia and analgesia of the trunk as high as the fourth rib. There was no affection of the arms but he had retention of urine and involuntary action of bowels. On November 2nd the patient died in a comatose condition, the legs having become anæsthetic, the patella reflexes having disappeared, and the intercostal muscles having ceased to act.

At the autopsy the membranes of the brain were normal and the brain itself quite healthy. The membranes of the cord were also intact. There was extensive central myelitis in the middle of the dorsal and in the upper lumbar regions, extending over about one and a half inches in each situation. The cord between these spots was slightly softened. The microscope showed that the brain, the chiasma, and the proximal part of the optic nerves were healthy; the peripheral portion of the latter contained an excess of fibrous tissue. Besides the pathological changes just mentioned there were patches of acute myelitis in the right side of the lower cervical and upper dorsal regions; a second patch just below this, and a third in the lower lumbar region affecting the posterior columns.

The conclusion which both Gowers and Dreschfeld draw with regard to these cases is that the optic neuritis and the myelitis are associated phenomena due to a common cause, but that neither depends directly on the other. Our case seems to point very strongly in this direction.

For the acute optic neuritis was present certainly one month before any symptoms of disease of the spinal cord appeared. Moreover, post-mortem examination showed that the spinal cord between the lower cervical region and the lumbar region was healthy, so that these centres of disease must have originated independently one of the other. The same may probably be asserted with regard to the optic nerves and the seat of disease in the cervical region, as there was no evidence of continuity between the inflammatory processes going on in these situations. Unfortunately, absolute proof of this is not at hand as the medulla oblongata was not kept for microscopical examination. Still the case may with great probability be said to be one of acute optic neuritis associated with acute disseminated myelitis. The cause which gave rise to these morbid changes, however, remains quite obscure.

Clinically such cases as these are of considerable importance. Before attention had been called to them the association of acute optic neuritis with paralytic phenomena would have justified the diagnosis of cerebral disease. And in the present instance, although the symptoms were not very intelligible from that point of view, the case was looked upon as probably one of brain disease. Acute optic neuritis will therefore have to lose some of its significance as a sign of intracranial affection.

With reference to prognosis our data are insufficient to allow any definite conclusions to be drawn. But as far as observations at present go, it seems that where acute disease of the optic nerves is associated with acute disease of the spinal cord the chances of a favourable termination are not great.

(June 5th, 1884.)

