Short Notes and Clinical Cases.

ERYTHROMELALGIA, CAUSALGIA, AND ALLIED CONDITIONS.

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IN considering this group of conditions the following case presents many features of interest.

A pensioner, A. T., age 41, came under my care on Jan. 17, 1921, complaining of continuous severe burning pain in both hands, and pain and swelling of the left leg and foot. The pain in the hands was so severe as to render them quite useless, being aggravated if he touched anything dry; and he constantly held wet rags in his hands. In the left leg and foot there was a continuous dull pain; and if in the dependent position, or on walking, there was considerable swelling.

On Examination.—The patient was a well-nourished man of excellent physique. The hands were blue, cold to the touch, and somewhat swelled, pitting on pressure. The discoloration was sharply demarcated at the wrists, the discoloured parts only being painful. The skin was glossy and the nails were thin, the blue colour showing through. The hands were tender, the patient resisting any attempt to touch them. The pain was not affected by movement or by the position of the limbs. On his being persuaded to undergo examination, sensation was found to be normal—heat, cold, pain, tactile, and two-point discrimination tests being used. All movements were normal.

The left leg and foot were oedematous below the knee, particularly round the ankle and in the plantar region at the roots of the toes. There was no discoloration, the skin and nails being normal in colour. On touching the foot there was some tenderness, which was severe on the sole, interfering very much with walking. The whole limb was slightly atrophied, the calf, thigh, and buttock being affected. The only limitation of movement was of dorsiflexion of the ankle. Sensation was normal.

The knee-jerks were brisk but equal. The other reflexes, super-

ficial and deep, were normal. The left pupil was slightly larger than the right, but there was no other abnormality. The pulse-rate was 140 per minute. The urine was normal. The electrical reactions of the muscles were normal. X-ray examination of the left leg and foot on March 30 showed "atrophic changes in the lower half of the tibia and in the bones of the foot". The mental condition was good. The patient was cheerful. There was no depression or 'nervousness'. Sleep was good.

History.—The patient is an old soldier who served in the Boer War, when he received a gunshot wound in the right calf which soon healed and has given no trouble since. While serving in India he had a slight attack of malaria, which also has never given rise to any trouble since. (Blood taken from one of the fingers of the left hand and examined microscopically showed no abnormality.) He was perfectly well till Aug. 26, 1914. On that date he received a superficial bullet wound over the external condyle of the left femur, two inches above the situation of the external popliteal nerve. The injury was merely a skin wound an inch long, and bled profusely. On receiving it he "felt a sharp pain shoot down from the knee to the heel and burst there all over the foot". He collapsed and was made a prisoner. After a few days there was no discomfort of the left leg.

While a prisoner in Germany, on three occasions within about four months he received the following atrocious punishment for petty offences. It consisted in being pegged down in the snow, flat on his back, with his hands above his head, for four hours daily for three consecutive days. He was secured to the ground by means of ropes attached round the wrists and ankles, and to pegs driven into the ground. The hands were entirely uncovered. He was able to move the wrists on first being pegged down, but after some time the ropes tightened and constricted the wrists. This was particularly so on one occasion, when "the hands ballooned up and felt they would burst". Following this treatment, the hands became blue, cold, and puffy, from a sharply demarcated line where the constriction had occurred. They constantly 'burned' all over, and he was unable to tolerate heat or dryness with them, or handle anything dry. He had constantly to keep wet rags in the palms, and periodically to put the hands into cold water. The left leg became painful, and swelled on walking or on hanging down. This state had persisted since the winter of 1914–15, but summer weather or heat had always aggravated the pain and swelling, while cold weather had slightly ameliorated the condition.

In the interval between 1914 and 1921 he had had treatment in Switzerland. This had included electrical treatment, thiosinamine injections, and various drugs, all of which had had no effect.

Treatment.—This apparently hopeless case has responded in a remarkable and gratifying manner to treatment. In January and February, 1921, he was given, on six occasions, suggestions under hypnosis that the pain would leave the hands and left leg, and that he would be able to use them normally. There was progressive improvement in the condition of the hands; and in fact, on this form of treatment being discontinued at the end of February (he has not been hypnotized since), he had full use of the hands. They were no longer tender, and the blueness had given place to a red discoloration which was not sharply demarcated at the wrist, being more obvious in the fingers and gradually shading off to the normal colour in the hand. During last summer—an exceptionally hot one—he had very little trouble on account of the hands, only exceptionally in hot weather having to use a wet rag because of some discomfort.

As to the left leg, the sole became much less tender, and, accordingly, the gait better; but the leg continued to swell to a tremendous amount on walking. In April, 1921, a course of calcium lactate was tried, but this had no effect. Following this an elastic stocking was supplied, and the patient was given extract. thyroid. siccum (gr. $1\frac{1}{2}$ a day). Since then there has been further progressive improvement. The pain in the foot, though persisting, is now very slight, and even on walking considerable distances without his elastic stocking there is only a little œdema. The pulse-rate also, on the administration of the thyroid, fell gradually to 96, at which it has remained. On Oct. 4, 1921, the left leg was again x-rayed, and the report was: "Slight though definite improvement in the condition of the tibia and tarsals".

The present condition is: The fingers are more red than normal, the discoloration shading off gradually in the hand. The nails still look thinner than normal. There is no tenderness or coldness to touch. The patient does not complain of any pain, and uses the hands normally. On warming them in front of a fire he complains of discomfort owing to 'tingling'. Sensation is normal. In the left leg there is still slight continuous deep-seated pain and a little œdema. No tenderness is complained of. All the movements are normal.

Remarks.—I have described this case in such detail, not only because of the extraordinary history and the unorthodox methods adopted in treatment, but also because of its bearing on the theories of the pathology of erythromelalgia and similar conditions.

The condition of the hands conformed in every particular to that described by Paget and Weir Mitchell, and named erythromelalgia by the latter in 1898; except that the symptoms were not made worse by the parts hanging down. Weir Mitchell suggested as a cause peripheral neuritis; others (e.g., Lewin and Benda) ascribe the

origin to the cerebrospinal axis, and Cassirer to the sympathetic system; while more recently it has been ascribed to changes in the walls of the smaller arteries. This last has been described in a number of cases (Barlow, Batty Shaw, and Parkes Weber). In the case described, the only condition that would give rise to the symptoms and their disappearance is a vasoconstriction of the arterioles of the hand. The radial arteries throughout presented no abnormality. This raises the question as to whether, in such cases as are described by Barlow and Batty Shaw, the initial change was a vasoconstriction, with organic changes supervening later. That we do get organic changes secondary to impaired function, whatever the origin of the impaired function, whether physiogenic or psychogenic, is, I think, generally admitted. The progress of the case described suggests that, in the cases which have been subjected to microscopic examination and in which considerable thickening of the arteries is described, we may have a secondary organic change supervening on a chronic vasoconstriction.

The similarity of the condition of the hands to that described by Head as 'glossy skin' is very noticeable. This symptom group, consisting of persistent hyperæsthesia and trophic disturbances of the skin, is ascribed to nerve irritation, following a partial nerve lesion. In this case there had been no such lesion, the condition following exposure to cold and constriction of the wrists.

The left leg presented a very different appearance from that of the hands. The condition was one of angioneurotic oedema without vasomotor phenomena, but with tenderness and bone changes. The origin is rather more complicated than in the case of the hands, because of the wound over the external condyle described above; the fact that the changes were also proximal to the ankle where the constriction had been applied; and because the exposure had not been so extreme or the constriction so severe as with the hands. The patient had woollen stockings on the feet when exposed, and had never felt the constriction so severely round the ankles as round the wrists. Again, only one leg was affected, and that the one which had not long before been slightly wounded.

The history of a pain shooting down a limb and then radiating out ('bursting') is often obtained in wounds of a limb in which there is injury to a nerve. In this case, the site of the wound precludes the possibility of any nerve lesion as a direct result of the wound. A frequent sequel in cases of nerve injury is causalgia (thermalgia of Stopford), i.e., a constant severe burning pain such as my patient had in both hands where he had not been wounded, but which was absent from the foot. Atrophy of the bones has been described also in cases of partial injury to peripheral nerves (e.g., Goldscheider).

This has been shown (by Raymond and Onanoff) to be reflex in nature, depending on irritation of afferent nerves, and not to occur in the rabbit if the limb is subjected to irritation after the division of the posterior roots. The correspondence between these traumatic cases and those produced by tabes and syringomyelia is commented on by Turney. But, again, in the case described there is no evidence of either nerve lesion or central trouble. Moreover, the course of the case rules out either possibility.

It is difficult to see how the wound could have played any part in causing the later symptoms, in spite of the fact that only the wounded leg was affected. The condition of the hands and leg, I think, was due to the constriction and exposure to cold. The constriction to the hands being more severe, the hands were rendered comparatively bloodless, and vasocontraction finally occurred; while in the ankle, it not being constricted to the same amount, the outflow of blood was more affected than the inflow, and stagnation, followed by increased permeability, supervened and led to the œdema.

As to the long duration, the fact that suggestion has practically cured the condition of the hands and ameliorated that of the foot, points to psychogenic causes having maintained the condition.

In spite, then, of the observations on a number of cases of erythromelalgia and causalgia in which organic changes have been demonstrated, it seems wrong to assume such changes in every case. Where organic changes do occur, they may at times be secondary to disturbances of function, and not causes of the disturbances of function. Once established, a vicious circle will be formed; but in their establishment it seems likely that psychogenic factors play an important part.

Cassirer has collected 130 cases of erythromelalgia, some of which were associated with diverse organic conditions—tabes, tumours, multiple sclerosis, arteriosclerosis, etc.—and which were relieved by the treatment of the underlying conditions (e.g., syphilis). In many of his cases the symptoms persisted where there was no evidence of any organic factors. In such cases psychotherapy should be considered. That the sympathetic nervous system was involved is obvious if only from the marked improvement that has followed the administration of thyroid. This does not detract from the importance of psychogenic factors. "The emotions find their expression through the sympathetic nervous system" (Langdon Brown).

Finally, it is interesting to note that, in certain cases of causalgia, the Committee upon Injuries of the Nervous System, in their report to the Medical Research Council, recommend psychotherapeutic treatment. That recommendation is fully supported by the observations in this paper.

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