

## A CASE EXHIBITING SYMPTOMS OF FACIAL HEMIATROPHY AND JACKSONIAN SENSORY EPILEPSY.\*

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FOR the opportunity to study this case and present the patient here to-night I am indebted to my friend, Dr. R. W. Stewart.

For want of a better name, I have called the paretic phenomena he exhibits Jacksonian sensory epilepsy.

M.C., aged 29, moulder; married; has two children; denies syphilis. Five years ago he began to drink heavily, and continued the habit until two years ago, since which time he has been temperate. His family history is negative.

Ten years ago he noticed that the right anterior part of the scalp was dry, and that he perspired less in this than in other regions of the head. This condition continued unchanged up to the time of injury to be described now.

Four years ago he was kicked by a mule on the right forehead, just over the region which is now most atrophic. He was unconscious for about one minute. The skin was not broken, but the eye and forehead became blue and very swollen, and he was compelled to discontinue work for about a week, when he resumed his usual occupation.

Six months after this injury he had his hair cut very close, and, for the first time, he noticed a bare streak on the scalp, parallel with, and just to the right of the median line. It extended to the line of the hairy scalp on the forehead, and its area could just about be covered by the first two phalanges of the little finger; almost as

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\* This paper was read and the patient presented to the Philadelphia Neurological Society at the meeting held March 25, 1895.

large as you now see it. Over this area, and over a strip of skin, representing a continuation of this region, extending down to the right side of the nose, he noticed that he perspired much less than he did in other parts of the head and face.

Two and a half years ago, about the time of his first epileptoid attack, the patient states that the skin over the anterior two-thirds of the right half of the scalp began to waste, and has continued to do so up to the present time. About the same time he received a blow from an iron rod (not very severe) over the lower part of the right calf, just above the tendo Achillis. A few minutes later his first "spell" occurred in the following manner: He experienced a feeling of numbness and tingling in the left toes which he likens unto a feeling of "pins and needles." This paresthesia slowly ascended the leg, thigh and trunk, went down the left arm to the fingers, then up the left side of the neck and face, involved the left side of the tongue. The attack terminated in pain on the right side of the head, its intensity being greatest in the atrophic regions. When the paresthesia had reached the middle of the trunk in its upward march, it began to fade from the toes, and precisely as it had come it passed away. An absolute loss of muscular power passed over and faded from the left side, just as the paresthesia had, save that, in time, it was a little later. The attack proper lasted from twenty to thirty minutes, but the right-sided head pains continued for an hour or two. He was entirely conscious throughout the attack.

Subsequent attacks have been similar in character except that nearly half of them began in the left fingers. Those which began in this way marched down the left leg to the toes, and then up the neck and face, terminating in head pains, as in the first attack. In a few instances the paresthesia marched down the arm after having ascended it. Two attacks began in the left lip, marched down the arm and leg and returned up the leg and then pursued the usual course. Sometimes the head pains, in which attacks always terminate, last nearly a whole day. This pain is always strictly confined to the right side and is chiefly located in the atrophic regions. The pain varies greatly and is always proportionate to the severity of his attacks. There is considerable difference in the severity of his attacks, and they last from twenty to forty-five minutes. In every attack, no matter

how it begins, the entire left side of the body has been involved. Since the first attack he has averaged about one a month, although they come at very irregular intervals, *e. g.*, six or seven months intervened between the first and second attacks, and two attacks have occurred within three days.

One year ago he was seized with a general motor convulsion, lasting twenty minutes, during which blood tinged foam was noticed about his mouth, and after which he slept. He was, however, conscious during the onset of this attack, and states that it began with the ordinary sensory manifestations in the leg, which were more severe than usual. The last thing he noticed before unconsciousness ensued was that his leg was trembling violently (convulsive movements). (?)

Until the occurrence of this attack, he felt well in the interval between his paresthetic seizures. Since then he has been subject to attacks of dizziness and various ill-defined subjective sensations in the head, and also to certain vague fears. During the past year he has not felt able to work on this account. Since this general convulsion, his sexual appetite has declined until now it is almost absent.

Examination March 3, 1895.—The skin of the right forehead and anterior third of the right scalp is decidedly wasted. There is also slight wasting of the skin over the right side of the nose. This atrophy extends to the median line, but is strictly confined to the right side. The atrophy of the scalp is not even. There are spots where wasting is much more marked than at others. Besides the bald spot already described, there is another one which could be covered with a silver quarter dollar, situated a little back and external to this one. The margin of hairy scalp is a little further back and external to this one. The margin of hairy scalp is a little further back on the right than on the left side. The hair all over the atrophic region of the scalp is less abundant and somewhat coarser than on the other side. Sensation is present over this atrophic region. The orbicularis palpebrarum and the occipito-frontalis muscles are active—apparently as strong as on the affected side.

With the dynamometer he registers sixty-five with the right and sixty-two with the left hand. No disorder of sensation of any sort is present on any part of the body; no atrophy; gait natural; knee-jerks sluggish.



Dr. Frank Edsall, who has examined his eyes, reports as follows: Vision O. D. 20/xl with + 0.50 D. Sph. 0.50 Dcyl. ax 180° = 20/xxx O. S. 20/xxv; with + 0.25 Dcyl. ax 90° v = 20/xx. No mydriasis; no muscular abnormality detectable; no limitation of visual field for white in either eye; field for color not tested; small conus of pigment atrophy at the lower temporal quadrant of the right disc; entire disc slightly hazy and indistinct; vessels tortuous, temporal half of the disc a little paler than normal. In the left eye the disc is also a little paler in the temporal half, slight cupping of disc small spot of pigment atrophy at the temporal side of the disc. Disc slightly oval in vertical axis.

#### REMARKS.

The conditions present in the tissues of the right side

of the head leads me to diagnose the case as one of facial hemiatrophy. Apparently the mule kick he received four years ago hastened, if it did not start, the atrophic process. It must be borne in mind that the patient states that ten years ago he noticed the right side of his hairy scalp was crisp and dry; so for six years before the injury some malnutrition existed. When we "remember the fact that the nutrition of all parts except the muscles seems to depend on the posterior root fibres, to which the fifth nerve chiefly belongs, and that the influence of chronic lesions is to cause a slow wasting, distinct from the acute disturbance produced by irritation," (Gowers, *Disease Nervous System*, Vol. II., p. 870), we might suppose that some chronic affection of the fifth nerve or its nucleus, has been present for ten years in this man, and that it was made more active by the mule kick he received four years ago. In the case of facial hemiatrophy, now classic, in which Mendel performed an autopsy (*Deutscher Med. Ztschr.*, 1888, p. 407), degeneration of the descending root of the fifth nerve and of the cells of the locus ceruleus was found. Assuming that a similar condition is present in this man's fifth nerve and its nucleus, might we not suppose that the degenerated nucleus, acting reflexly as a source of irritation, has provoked these periodical sensory discharges? I must confess that I have been unable to demonstrate to my own satisfaction any exact mechanism which would on theoretical grounds definitely account for the attacks. The suggestions I have thrown out, even if adopted, would not explain very far and would leave very much unexplained. If we attempt to explain the attacks on the theory of an irritated sensory cortex, the fact that they begin in different parts of the body, arises at once before us as a difficulty in localization. Besides cortical trouble would not explain the atrophic condition present in the right face. (Bremer and Eskridge have recently reported cases showing cortical trophic influence).

These attacks are themselves, it seems to me, analogous to certain types of Jacksonian epilepsy, except that they are sensory instead of motor, and that there is no constant or signal symptom. The essential feature of epilepsy is the more or less sudden and vicarious discharge of morbidly unstable nerve cells. We are too much accustomed to think this discharge must necessarily be motor. Every now and then we see cases like

this, which seem to be clearly chiefly or wholly sensory. I am disposed to think the motor paralysis he describes as following closely after the paresthesia is more apparent than real. He tells me that on one or two occasions he walked some distance after the parasthesia had involved his leg. Because the leg feels "heavy" and very "large," he thinks he cannot use it. Indeed, it may be fairly doubted whether there is any real paralysis.

The morbid process, wherever or whatever it is, is likely extending. The general convulsions from which he suffered one year ago, the vertigo, the restlessness and uneasiness, the loss of sexual appetite—indicate this. I have as yet instituted no treatment, but have been holding under advisement the propriety of having excised the various branches of the fifth nerve, as recommended by one of the members of this society (Dr. Dercum) in a paper read before the society a few years ago, or of having the Gasserian ganglion itself excised, which, in operation as shown by Stewart, Keen, Rose, Hartly and others, can be done without any very great risk. Dr. Dercum's recommendation (*JOURNAL OF NERVOUS AND MENTAL DISEASE*, Volume XVII. p. 108) is based on the theory that facial hemiatrophy depends, not so much on loss as on perversion of the trophic influence of the fifth nerve. The question, too, as to whether trephining would be advisable also presents itself. Although this man does not, it seems to me, present symptoms which would justify a diagnosis of brain tumor, nor which would localize a focus of irritation, it is possible that trephining undertaken in an empirical way might result in some benefit. Yet, notwithstanding the absence of clear indications, it is just possible that a brain tumor may be present, for we know that in some instances of intracranial growths many, or nearly all the classical symptoms are absent.