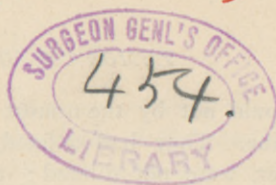


MILLS (C.K.)



TUMOR OF THE PONS VAROLII, WITH CONJUGATE DEVIATION OF THE EYES AND ROTATION OF THE HEAD.*

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R. C., æt. 32, single, groom, had for several years been intemperate, and had a history of syphilis. About five years before coming under observation he had twice been thrown from a horse and kicked on the head. After the occurrence of these accidents he began frequently to suffer from severe headache which always came on at night. He also had at times spells of dizziness. Four weeks before coming for treatment he fell on the ice, striking his head. At the time he noticed no ill effects from the fall, but a week later, while grooming a horse, he became dizzy and fell to the ground, but did not lose consciousness. A few days later his eyes began to trouble him, and he also noticed a slight loss of power in his right arm and leg. Such was the history obtained from the patient, whose memory was defective, but I think it probable that his ocular and paretic symptoms were of longer standing than a few weeks.

On admission to the Philadelphia Hospital he was able to walk about the wards and even go out of doors, but he was weak, anæmic, and apathetic. The right side of the forehead wrinkled more promptly than the left. The lower part of the right side of the face, and the right arm and leg were paretic, but decided paralysis and contractures were not present. Sensation was diminished in the left side of the face and in the right limbs, but owing to the patient's mental condition, his answers with reference to sensation were somewhat confusing and conflicting. Hearing, smell, and taste were preserved.

Both eyes were kept constantly directed to the right. The pa-

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tient could not by the utmost effort bring them even to the median line. They had a fixed, staring expression. The pupils, however, were not dilated; they were at this time equal and about normal in size. Dr. E. O. Shakespeare, ophthalmologist to the Philadelphia Hospital, examined the eyes for me, and the following notes were made by him: "The corneæ and other media were transparent. There was a conjugate deviation of the optic axes to the right. The power of accommodation was not greatly impaired, and in the act of accommodation there was an associated convergence of the optic axes and the usual contraction of the pupils. In attempted movements of the eyes to the left, the right eye turned slightly, the left eye scarcely at all. The right lid showed a slight tendency to ptosis. This was most noticeable in attempts to raise the eyes above the horizontal meridian. Ophthalmoscopic examination of the left eye gave the following results: Fundus seen with a $+\frac{1}{24}$ glass. It was of a pale reddish-yellow color. The outline of the disc was distinct, but not as marked, or as regular, as normal. It was more or less opaque and slightly hyperæmic. The arteries were scarcely distinguishable from the veins by their color. The former were a little contracted, but were regular in their course. The view of the whole fundus was, however, slightly veiled. In consequence of the extreme deviation of the eyes to the right, the right eye could not be satisfactorily examined by the ophthalmoscope."

On cutting the patient's hair close to the head, a scar about one inch and a half in length was found in the scalp of the left side of the head. Its direction was from behind forward, and from above downward, at a slight angle, its posterior end being three and a quarter inches in almost a direct line above the external auditory meatus. It corresponded to the middle region of the squamous portion of the temporal bone. The bone beneath the scar appeared to have in it a cleft. Two slight scars were found in the scalp of the parietal region of the right side.

The patient was placed upon potassium iodide, tonics, and nourishment. He got weaker from day to day, however, his ocular and parietic symptoms remaining about the same. He was compelled because of weakness and dizziness to stay in bed. His nose began to bleed, the blood sometimes escaping from one nostril and sometimes from the other. In spite of local and internal remedies, such as ice, alum, iron, ergot, erigeron, gallic acid, etc., the epistaxis persisted until the death of the patient, the bleeding sometimes stopping for an hour or two, apparently without refer-

ence to treatment. He became extremely anæmic; and died of general exhaustion. A few notes were made on his condition the day before his death. The limbs of both sides appeared to be about equally helpless. The mouth was now drawn very slightly to the right. Little could be made out certainly with reference to sensation. He still appeared, however, to be less sensitive to impressions on the left side of the face and in the limbs of the right side. Both pupils were small, the left a little smaller than the right. The eyes still looked to the right; the deviation, however, was not quite as great as when he was first admitted. The conjunctiva of the right eye, from the cornea to the internal canthus, was much injected.

Autopsy.—The scalp was found adherent to the skull in the line of the scar in the left squamoso-temporal region. A narrow fissure was present in the skull beneath the scar. The internal table of the skull was fissured for the distance of half an inch, the fracture corresponding to a portion of the external cleft. The fracture was a simple crack or break, no bone being depressed or displaced. The dura mater was slightly adherent along the internal fissure, and exactly beneath the point of adhesion, on the inner surface of the dura mater, was a hard, yellowish tumor, no larger than a pea. It was attached below to the pia mater, and caused a slight depression in the first temporal convolution, about the junction of its middle and posterior thirds, and half way between the parallel fissure and the horizontal branch of the Sylvian fissure. No other lesion of the surface of the brain, or of the ganglia, centrum ovale, or cranial nerves, was discovered. The pia mater of the middle region of the base was hyperæmic and not quite transparent. On exposing the floor of the fourth ventricle a distinct bulging of its left upper portion was observed. This proved to be due to a tumor about half an inch in diameter. It was situated in the body of the pons, both the anterior and posterior surfaces of the latter retaining their integrity. It was distinctly limited to the left upper

quarter of the pons, coming close to, but not crossing, the median line. It was found on section to be of firm consistence and of a greenish-gray color.

Tubercular deposits were found at the apex of the left lung. The heart walls were a little softened. The liver was intensely cirrhotic, and a small whitish tumor was embedded in the upper surface of its left lobe. The spleen was soft and about twice the normal size. Both kidneys were fatty.

The tumor of the pons was examined microscopically by Drs. J. H. C. Simes and H. Formad, who concluded that it was a gumma.

Both eyeballs and the optic nerves were carefully removed and placed in the hands of Dr. E. O. Shakespeare, who furnished me with the following report upon the microscopical examination of the optic nerves:

“After proper hardening, thin sections of the anterior third of the optic nerve, including its entrance into the eye, were made so that the sections were longitudinal to the course of the nerve. In one of the eyes the optic disc was slightly more prominent than normal. The walls, both of the arteries and veins, of the optic papilla were somewhat sclerosed. Their lining endothelium was slightly irritated. The connective tissue between the nerve bundles of the papilla was in a state of considerable cellular hyperplasia. Their corresponding capillary blood-vessels were apparently more numerous than usual, while their walls were surrounded by numerous leucocytes. As the position of the lamina cribrosa was approached, the cellular hyperplasia was found to increase, and large numbers of nuclei were present upon the fibres of the lamina cribrosa itself. This cellular multiplication extended far back of the nerve entrance into the eyeball. The subvagal and subdural spaces of the sheath of the optic nerve were considerably enlarged, and

in the anterior portion, adjacent to the eyeball, the walls and the enclosed fibrous trabeculæ were in a state of inflammatory irritation. The nerve from the other eye was in practically the same condition. From the examination, it would appear that there was present a descending neuritis of subacute character."

Remarks.—The peculiar ocular symptoms present in this case were doubtless due to the tumor of the pons Varolii. Conjugate deviation of the eyes, with rotation of the head, is a condition often present in the early stages of apoplectic attacks. The patient is found with both eyes turned to one side and slightly upward, as if looking over one or the other shoulder, the head and neck being usually rotated in the same direction. Sometimes the deviation is slight, sometimes it is marked. Frequently the muscles of the neck on one side are rigid. The eyes are commonly motionless, but occasionally exhibit oscillations. This sign, well known to neurologists, usually disappears in a few hours or days, although it occasionally persists for a long time.

Vulpian was probably the first to study thoroughly conjugate deviation. The sign, when associated with disease of the pons, was supposed by him and by others to be connected in some way with the rotatory manifestations exhibited by animals after certain injuries to the pons. Transverse section across the longitudinal fibres of the anterior portions of the pons produces, according to Schiff, deviation of the anterior limbs (as in section of a cerebral peduncle), with extreme flexion of the body in a horizontal plane toward the opposite side, and very imperfect movements of the posterior limbs on the other side. Rotation in a very small circle develops in consequence of this paralysis (Rosenthal's "Diseases of the Nervous System," vol. i, p. 125). The movements of partial rotation are caused, according to Schiff, by a partial lesion of the most

posterior of the transverse fibres of the pons, which is followed in animals by rotation of the cervical vertebræ (with the lateral part of the head directed downward, the snout directed obliquely upward and to the side).

This lateral deviation, both of head and eyes, occurs, however, not only from lesions of the pons and cerebellar peduncles, but also from disease or injury of various parts of the cerebrum—of the cortex, centrum ovale, ganglia, capsules, and cerebral peduncles. It is always a matter of interest, and sometimes of importance, with reference especially to prognosis, to determine what is the probable seat of lesion as indicated by the deviation and rotation.

Lockhart Clarke, Prevost, Brown-Séquard, and Bastian, among others, have devoted considerable attention to this subject. To Prevost we owe an interesting memoir. Bastian, in his work on "Paralysis from Brain Disease," summarizes the subject up to the date of publication (1875). Ferrier, Priestly Smith, and Hughlings Jackson have investigated the relations which cortical lesions bear to the deviation of the eyes and head.

It has been pointed out by several of the observers alluded to that when the lesion is of the cerebrum the deviation is usually toward the side of the brain affected, and therefore away from the side of the body which is paralyzed. In a case of ordinary left hemiplegia, it is toward the right; in one of right hemiplegia, toward the left. In several cases of limited disease of the pons, however, it has been observed that the deviation has been away from the side of the lesion. In the case here recorded, the conjugate deviation was to the right, while the tumor was entirely to the left of the median line, thus carrying out what appears to be the usual rule with reference to lesions of the pons.

During the life of the patient, it was a question whether the case was not one of oculomotor monoplegia or mono-

spasm from lesion of cortical centres. I believe, with Hughlings Jackson, that ocular, and, indeed, all other movements, are in some way represented in the cerebral convolutions. In the *British Medical Journal* for June 2, 1877, Jackson discusses the subject of disorders of ocular movements from disease of nerve centres. The right corpus striatum is damaged, left hemiplegia results, and the eyes and head often turn to the right for some hours or days. The healthy nervous arrangement for this lateral movement has been likened by Foville to the arrangement of reins for driving two horses. What occurs in lateral deviation is analogous to dropping one rein; the other pulls the heads of both horses to one side. The lateral deviation shows, according to Jackson, that after the nerve fibres of the ocular nerve-trunks have entered the central nervous system, they are probably redistributed into several centres. The nerve fibres of the ocular muscles are rearranged in each cerebral hemisphere in complete ways for particular movements of both eyeballs. There is no such thing as paralysis of the muscles supplied by the third nerve or sixth nerve from disease above the crus cerebri, but the movement for turning the two eyes is represented still higher than the corpus striatum.

Ferrier found that irritation of a certain limited area of the surface of the brain of the monkey, corresponding to a region in the brain of man at the base of the first frontal, and extending partly into the second frontal, convolution, caused elevation of the eyelids, dilatation of the pupils, conjugate deviation of the eyes, and turning of the head to the opposite side.

Priestly Smith (*Ophthalmological Hospital Reports*, vol. ix, p. 428) concludes that the chief coördinations in the brain of ocular movements are of four kinds: 1. Movements of both eyes to the right. 2. Movements of both

eyes to the left. 3. Movements of both eyes downward and inward, narrowing of the pupils, and contraction of the ciliary muscles, producing increased convergence and accommodation. 4. Movements of both eyes upward and outward, producing diminished convergence, and accompanied by, though not actively producing, widening of the pupils and relaxation of accommodation. These several forms of compound movements are produced by the action of distinct brain centres, and disease may destroy or irritate one or other of the four, and leave the others intact.

A few cases are on record in which conjugate deviation of the eyes alone has occurred, constituting, according to Ferrier, what may be regarded either as unilateral oculomotor monoplegia or monospasm. Five such cases, or, rather, supposed cases, for an autopsy was held in only one instance, have been collected by Ferrier ("The Localization of Cerebral Disease," New York, G. P. Putnam's Sons, 1879). Three of these were reported by Priestly Smith, whom I have just quoted. In the first case, after an attack of pain in the head, giddiness, and vomiting, the eyes became persistently turned to the right, with complete inability to turn them to the left. The right side of the forehead was marked with wrinkles; the left eyelids were more open than the right; there was frequent winking of the eyelids, and synchronous but imperfect action of the left. Gradually the right eye recovered its mobility to the left, while the outward motion of the left eye still continued very imperfect, and caused double vision. Three months after the first appearance of the symptoms the patient became affected with left hemiplegia. A fortnight later the right side became paretic. In a second case, the symptoms noticeable were deviation of the eyes to the right, facial paralysis on the left, and some paralysis in the left limbs. In a third case, severe pain in the right side of the head

and face had been followed by "squinting of both eyes to the right." When first seen, twelve months after the attack, the left external rectus appeared to be paralyzed. It is suggested, however, that as both eyes had at first been turned to the right, the eye symptoms are explicable, as in the two preceding cases, by the recovery of the right eye, while the outward movement of the left remains paralyzed.

Dr. Carroll, of Staten Island, furnished Dr. Ferrier with the particulars of another case. A child, aged five months, fell six feet, and was stunned for a few minutes. No paralysis occurred, but conjugate deviation of the eyes and rotation of the head to the right, with, at first, dilatation of the pupils, were noticed. A linear fracture was detected in the right parietal bone, about midway between the squamous and sagittal sutures, and intersecting a vertical line drawn upward from the auditory meatus. Pressure at the seat of injury caused a distinct increase of the deviation. Ferrier supposed the symptoms to be accounted for by unantagonized action of the left centre, from hemorrhagic lesion of the right.

Chouppe, quoted by Landouzy, relates the case of a lad, aged 19, who showed symptoms of tubercular meningitis, in which, in addition to pain, vomiting, etc., the most remarkable symptom was a rotation of the head and eyes to the right. After death a patch of disease, free from granulation, and quite superficial, of the size of a franc piece, was found in the "superior part of the middle frontal convolution" of the left hemisphere. Ferrier thinks that the seat of the lesion probably corresponded with the oculomotor centre in the brain of the monkey.

I have taken the liberty to quote a condensed account of these cases in order to fully bring forward the subject, the literature of which is as yet scanty. A similarity will be

observed between the symptoms presented by my case and those exhibited by some of the cases collected by Ferrier. In the first case, reported by Priestly Smith, the symptoms are strikingly similar to those shown by my patient—conjugate deviation of the eyes to the right, with complete inability to turn them to the left; more marked wrinkling of the right side of the forehead than of the left; hemiplegic or hemiparetic symptoms first of one side and then of the other. In the absence of an autopsy on the case of Priestly Smith, and in the light of the *post-mortem* examination here reported, it may, indeed, be considered doubtful whether his patient suffered from a cortical lesion.

It does not seem probable that the fissured skull, and the small meningeal tumor in connection with it, had any thing to do with the production of the ocular symptoms. The lesion was comparatively remote from the oculomotor centres of Ferrier, at the bases of the first and second frontal convolutions. It is true that efforts have been made to localize a centre for the levator palpebræ superioris muscle in the angular gyrus, and if such could be made out to exist in this region, it is probable that centres for other ocular movements would be in proximity. The weight of evidence, both physiological and pathological, is, however, against this localization. The meningeal tumor was, in addition, very small, and was a little too far forward for the angular gyrus proper. Both the ocular and other phenomena of the case are, I think, well accounted for by the pontine lesion.

Cases like that reported in the present paper are far from discouraging with reference to the local diagnosis of brain lesions. They serve simply to give additional zest to close investigation. I conclude, from a study of this case, that tumors limited to one-half of the upper portion of the body of the pons will cause conjugate deviation of the eyes and

rotation of the head away from the side of the lesion. It is quite likely that if the lesion is sufficiently limited the ocular deviation may stand alone. In the vast majority of cases, however, owing to the narrow limits of the pons and its position with reference to connecting tracts both from the cerebrum and cerebellum, other symptoms will be present. In this last fact we have the clue to the differential diagnosis of the pontine lesions from disease of the oculomotor centres of the convolutions.

The paralysis or spasm of face or limbs, that may be associated with the ocular symptoms, is more likely in cases of cortical disease to be unilateral than in lesions of the pons. The oculomotor centres of the cortex are near to the crural, brachial, and facial centres, and these may be involved in the same lesion, or may become involved by extension, and thus arise paralytic or spasmodic symptoms in face or limbs, or both, on the opposite side of the body. Both sides of the body would present symptoms only after extension of the lesion to both hemispheres, which is not likely to occur. Even when a lesion is strictly limited to one-half of the pons, the nuclei and tracts for both sides are so close together that in the case of tumors and hemorrhages the uninjured side will be more or less involved by pressure. In the patient whose history I have just given, the paresis was first noticed upon the right side, but both sides showed signs of paralysis before his death. Disturbances of sensation are more likely to be present in pontine lesions than in cases of cortical oculomotor disease. Such sensory disorders, according to Ladame, are to be found in about one-third of the cases of tumor of the pons. According to Rosenthal, with whom I entirely agree, careful examination will show them to be even more frequent. Supposing the bases of the first and second frontal convolutions to cover the true oculomotor centres, these are compara-

tively remote from the sensory zone, which is in the parieto-temporal and occipital regions. Some changes of sensibility were present in the case here recorded. Contraction of the pupils, varying in degree for the two eyes, pointed also to disease of the pons. Depressed farado-contractility and peculiarities of temperature would have helped to confirm the diagnosis of tumors of the pons, but these, by an unfortunate omission, in the present instance were not studied.