REPORT OF A CASE OF SPINA BIFIDA OCCULTA IN CERVICAL REGION

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Spina bifida occurs not infrequently as shown by the fact that in England during the year 1882, 647 deaths were reported as due to this condition. Presumably most of these deaths occurred shortly after birth and were due to the more marked instances of failure of the vertebral plates to close resulting in meningocele or possibly meningocele.

All varieties of spina bifida occur most frequently in the lumbosacral region, whereas, among 385 cases which had been operated upon, Moore¹ found only 9½ per cent. occurring in the cervical region. It has been estimated that of all varieties in all regions the occult comprises only 5 per cent.² Brickner,³ in 1909, collected altogether 85 cases of spina bifida occulta from the literature. He failed to speak of the proportion of cases in which the cervical region was involved. From these references it seems obvious that the type referred to in the title of this paper is uncommon and the brief presentation of a case may be justified.

CASE I.N., 13 years of age.

History.—A family history was obtained for two generations only. The father is 43 years old and is "tuberculous." The mother died at 38 years, 10 days following childbirth. According to the patient the mother was "paralyzed," the paralysis beginning one year before her death and gradually involving the upper and later the lower extremities. No further details concerning this disorder were learned. A sister of the mother had no knowledge of it. A brother 21 years of age is "healthy." A brother 10 years of age is "nervous." A brother died at 3 years, cause unknown, and a sister died in infancy. Of these the oldest brother is the only one born before the patient.

As far as known the birth of the patient was at full term, not difficult and followed an uneventful pregnancy. She cried a great deal as an infant and was constipated, but otherwise seemed well.

¹ Jas. E. Moore, *Journal of Surgery, Gynecology and Obstetrics*, August, 1905.

² Augustus Thorndike, *Reference Handbook of Medical Sciences*, 1917.

³ Walter M. Brickner, *Medical Record*, May, 1909.

She walked between one and two years of age and the aunt believed that talking and eruption of the teeth occurred at the "usual time."

She had diphtheria, measles, chickenpox and whooping cough at intervals after the age of 6, but none of these was followed by sequelæ.

CASE I. N., Dimple with tuft of hair between fourth and fifth cervical spines.

She attended school and learned rapidly; at 13 years was in the seventh grade. She made friends easily and was of a happy, cheerful disposition.

It was noted from birth that the patient had a small depression in the median line on back of neck out of which grew a small tuft of hair. Although the upper extremities seemed to develop normally the patient never was able to use the hands as well as the ordi-

nary child. She could sweep, carry or push objects of weight and play ordinary games in which finer movements of the hands were not involved. She was not clumsy except in buttoning the clothing, in tying laces, in doing fancy work, etc., but in these and other activities in which finer movements were necessary she was so slow as to preclude the doing of any but necessary tasks. She dressed and undressed herself, but did so slowly and was aided whenever speed was desired. Piano playing was attempted but quickly dropped as being too difficult.

Examination.—The girl was of moderate height for age and well nourished. She was slightly inclined to obesity. The tonsils were large and cryptic. The cardiac dulness extended about a

CASE I. N. Diagram sketched from X-ray plate showing cleft in third cervical vertebra.

centimeter to left of mid clavicular line and there was heard a soft blowing systolic murmur over apex; not transmitted. There were no other evidences suggestive of visceral disease.

On the posterior surface of the neck in median line between the fourth and fifth cervical spines was found a depression in skin about an eighth of an inch in depth, out of which a small tuft of hair half an inch in length was growing.

Skiagraph showed no defect in the fourth or fifth cervical vertebrae but a cleft in the arch of the third.

Both hands were sufficiently well developed that to casual inspection no abnormality was noticed. On examination, however, it was noted that the fingers were hyperextensible. This was more marked on left side; occasionally the fingers on this side were

slightly hyperextended even at rest. This was a little more marked in little and ring fingers than in the remaining two. Each finger was of ordinary contour and there was no evidence of trophic disturbance. Muscular power was deficient on both sides in little and ring fingers. This was noted in flexion and extension but more particularly in abduction and adduction. She could not quite approximate little finger to ring finger when latter was adducted on either side. Weakness was not demonstrable in simpler movements of index and middle fingers. The grip on right side registered 26 with dynamometer and 22 on the left. The defect was evident in all fingers of both hands in finer movements. She was very slow and clumsy in approximating in turn the fingers to thumb and in buttoning clothing. This difficulty became more marked when eyes were closed. In fact it seemed impossible for her to button a coat placed on her lap unless she was looking at it. In spite of this apparent defect of coördination the finger-finger and finger-nose tests were well done, except for error of three quarters of an inch on first two trials with left hand in latter tests. With eyes closed she accurately followed one hand moved passively about with the index finger of the other hand and correctly imitated with disengaged fingers the position in which the examiner had placed the fingers of other hand.

There were no subjective symptoms of disturbed sensibility and careful examination of parts involved failed to show defect in tactile, pain or thermal sensation either in acuity or in rapidity of conduction of stimuli.

The triceps, biceps and supinator longus reflexes were moderately brisk and were equal on the two sides. There was no flaccidity, spasticity or tremor demonstrable.

Except as mentioned the patient was quite well developed. Particular attention was given to the feet, but there was no suggestion of club-foot or other type of malformation.

The neurological examination, except for the evidences of defect of innervation of fingers, gave negative results. The special senses were normal. Although tactile, pain and thermal sensibility was carefully tested over entire surface of body no defect was found. Heel-to-knee test was well done, Rombergism was not present and sense of position was normal. There were no palsies or involuntary movements. The knee-jerks and ankle-jerks were moderately brisk and equal on the right and left sides. Ankle-clonus and the Babinski, Oppenheim, Gordon and Chaddock reflexes were not present.

Although no detailed discussion of the case will be attempted brief reference to some of the facts in the case may be made. Brickner⁴ states in regard to his cases that "heredity played no rôle and coincident congenital deformities were rarely found." No obvious congenital defects, other than those related to the spina bifida, were present in this case. In regard to heredity a tare is suggested in the facts that the father is tuberculous and that, of the two brothers

⁴ Ibid.

⁵ Gordon Holmes, *British Medical Journal*, 1915.

and the sister born after the patient, one is "nervous" and the others dead. The statement of the patient in regard to the illness which preceded the death of the mother may also be remembered in this connection although too indefinite to permit any deductions.

According to a number of writers there is, in some of the cases of spina bifida occulta, an increase in the peripheral disturbance, presumably due to adhesions involving the roots, as, with growth, there is proportionate shortening of the cord in the canal and consequent dragging upon the roots. In this case there is no history of such increase which may be taken as presumptive evidence that if adhesions exist they do not involve the roots.

In regard to site of the lesion there may be some difference of opinion. The peripheral disturbance was not only definitely motor in type but the inability to button coat with eyes closed although she could do so slowly, with eyes intent upon the task, suggests a disturbance in the deep sensibility even in the absence of other demonstrable evidences of involvement of afferent fibers. The absence of atrophy and of tactile, pain and temperature disturbance speaks for a cord rather than nerve lesion. The peripheral distribution of the motor disturbance is segmental in type corresponding approximately to the seventh and eighth cervical and the first dorsal roots which enter the cord a segment or two above the intervertebral foramina from which they take exit. The term "approximately" must be used, as it was impossible exactly to define the limits of the parts involved.

An attempt to correlate the peripheral disturbance with the defect in development of the third cervical vertebra is hardly possible, but as a matter of fact there need be no direct relationship between the vertebral cleft and the peripheral disturbance. It may be said rather that the cleft shows evidence of a general tendency to failure of development in this region. The fact that the dimple lies between the fourth and fifth cervical spines is evidence that the defect is not limited to the third cervical vertebra and parts immediately adjoining. It is possible that the actual lesion in the cord is caused by a defect in the region of the lower cervical vertebræ not visible in the X-ray. Norman Sharpe⁵ cites a case of spina bifida occulta in which although the cleft was present in all vertebræ from the fifth cervical to the sixth dorsal the peripheral disturbance was limited to tingling of the hands and fingers and areas of hypalgesia and hypesthesia on the outer side of left upper arm. Here too the disproportion between the vertebral defect with the clinical evidences of neural involvement is obvious.

⁵ Norman Sharpe, *Annals of Surgery*, Feb., 1916.