FACIAL PARESIS AS A MANIFESTATION OF TUMOURS OF THE UPPER HALF OF THE CERVICAL SPINAL CORD.

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THE signs and symptoms of tumours of the upper four segments of the cervical portion of the spinal cord are both varied and numerous. This portion of the cord is not only the crossroad for pathways to and from the brain, but contains centres and tracts which have to do with the functions of certain cranial nerves, the sensory fifth, the vestibular and gustatory system. The diagnosis of tumours in this region is often very difficult. The clinical picture may be first manifested by such lower level symptoms as sciatic neuralgia. It may, on the other hand, present, among others, the clinical pictures of cerebral hæmorrhage, hereditary ataxia, combined sclerosis, Pott's disease, and pachymeningitis.

The variety of involvement of various cranial nerves described is striking. Choked disc, pupillary inequality, facial sensory disorders and absence of the corneal reflex, nystagmus and absence of the pharyngeal reflex have all been recorded. But the finding of facial paresis has been unusual and very little emphasized. We have found it referred to in Elsberg's book¹ in two cases only.

The material which we have analysed in order to ascertain whether this finding is usual consists of ten cases of cervical tumour, of which two were confirmed at autopsy (one extramedullary, one intramedullary). In seven of these cases, including the two autopsied cases, a lower facial paresis was noted. The facial nucleus and nerve were examined in these two cases, and found to be normal. There was no pathological evidence of tumour or of meningitis above the foramen magnum which could account for the paresis. The tumours in five of the seven cases were extramedullary, and in the other two, intramedullary.

These observations are not altogether easy to explain. The question of hereditary or acquired non-paralytic asymmetry can be ruled out. Previous disease which could have caused facial paralysis of the two recognised types, central and peripheral, was not present. Furthermore, the paralysis of the lower half of the face which was found precludes these types of paralysis,

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since it involves neither all of the facial muscles on one side as in the peripheral type, nor all of the infraocular muscles as in the central type, but is limited to the infrabuccal group. In none of the autopsied or non-autopsied cases was there any evidence of disease in the neighbourhood of the seventh nucleus or of its fibres and nerves.

We believe that some light may be thrown upon the matter from a consideration of the sensory supply to the face, the pathway of taste, and the comparative anatomy of the facial musculature.

The sensory supply to the face, head, and neck is derived not only from the trigeminal nerve, but also from the cervical plexus. The facial musculature is sometimes affected in lesions of the trigeminal nerve, reflexly. We have wondered whether the same may not occur when the cervical roots, and thus the sensory supply to the lower parts of the face, of the neck, and the back of the head are affected.

Another possible reflex pathway is through the gustatory system. This descends into the upper cervical region as the solitarius bundle. That impulses of taste produce movements of the facial musculature is well recognized. While in Utrecht visiting Prof. Cornelius Winkler's clinic, one of us (W.M.K.) saw a moving picture of an anencephalic infant in which quinine painted in the mouth produced an expression of displeasure, sugar an expression of pleasure. Moreover, we are well aware of similar pseudo-affective reflex movements, from the records of reaction of decerebrate animals (Bazett and Penfield²) and we know man responds in the same way, 'making a face' when he gets a badly-tasting mouthful. However, no evidence of taste disorder was recorded in our cases.

We mention all this without being in any way convinced that a reflex through the gustatory system is responsible for the paresis, but merely to state the anatomical pathways which we have considered in our attempt to explain our findings.

The comparative anatomy of the facial musculature throws some light upon the subject. The facial musculature is a derivative of the second hyoid arch and is part of the branchial muscular system. It has developed from the sphincter colli of fish and amphibia. It is also the only remaining part of the dermal musculature, a part of the body musculature which is skeletal in origin. In some animals this musculature envelopes the entire body, as in the hedgehog; in many, as the horse and cow, it is very extensive, and is the musculature which removes flies and other irritative things from the body by a twitch movement. In man, this musculature, the platysma, is limited to the head and neck and extends a variable distance downward on the front of the chest, as far as the nipple line in some. From the mouth downwards this muscle sheet is the most cervical portion of what remains of this dermal system of muscles, so much more extensive in many lower animals. Even in man it lies far below the anatomical limits of the head.

There is no evidence to show, from any viewpoint at present available, that the lower portion of this dermal musculature has a double nerve supply, i.e., spinal as well as by the seventh cranial nerve. No supply analogous to the double supply of the sternocleidomastoid has been shown.

We are left, therefore, with the idea that a reflex pathway within the nervous system from the cervical segments is the most probable cause of the facial paresis which we have found in tumours of the upper four cervical segments.

Further evidence, of a negative character, is that this paresis has not been observed in tumours below the fourth cervical segment, either in our series or the cases reported by Elsberg. Whatever centre or pathway is responsible for the sign is not present below C4.

That not all of the cases (25 per cent.) showed this sign indicates that some specific mechanism is affected.

SUMMARY.

1. Paresis of the infrabuccal portions of the facial musculature may occur in association with tumours of the upper four cervical segments, and, per contra, no such paresis appears in cases of tumours of the lower four cervical segments.

2. In two cases of a series of ten high cervical tumours which were autopsied, one intramedullary and one extramedullary, the seventh nucleus was normal, and there was no evidence of any abnormality in its region, or above the foramen magnum.

3. In seven cases, five of which were extramedullary, from the Montefiore series, and in two cases, both extramedullary, recorded by Elsberg, this facial paresis was present. In three cases, one intramedullary and two extramedullary, from the Montefiore series, the paresis was absent.

4. The phenomenon is probably due to disorder of a reflex pathway originating in this portion of the spinal cord and extending to the facial nucleus.

CASE REPORTS.

CASE I. I. B.—*Diagnosis*: Extramedullary neoplasm, upper level at C2.

Flattening of left side of face, nystagmus, weakness of left sternocleidomastoid, hyperæsthesia beginning at C1.

Autopsy: Osteofibroma of odontoid process. Facial nucleus normal. No pathological condition above foramen magnum.

History: Until the age of fourteen patient was a weakling. In June 1919 he began to lose control of his sphincters. This recurred at intervals of about every four weeks. Admitted to Mt. Sinai Hospital in June 1920, where attacks of dyspnoea and priapism were noted.

Neurological examination: Fundi hyperæmic. Disc margins blurred. Right pupil larger than left. Slight flattening of left side of face. Nystagmus in horizontal plane. Laboured rapid breathing. Left sternocleidomastoid and trapezius weaker than right. Tongue tremulous but not atrophied. General motor power fair. Gait spastic and ataxic. Dysdiadokokinesis on both sides. Deep reflexes exaggerated, more on left

than right. Bilateral Babinski sign. Area of hyperæsthesia beginning at C1 and extending to C4, below which there is hypæsthesia. Perianal anæsthesia.

Autopsy revealed osteofibroma of odontoid process, compressing cord posteriorly; below, descending pyramidal demyelination in both lateral columns and one anterior column. Above C1, degeneration of the columns of Goll and Burdach, and both dorsal spinocerebellar tracts. Sections at various levels through the seventh nucleus failed to reveal any abnormalities in cell structure.

CASE II. B. L.—*Diagnosis*: Intramedullary glioma, upper level at C3.

Drooping of left angle of mouth; hyperæsthesia beginning at C3.

Autopsy revealed intact seventh nerve nucleus. No pathological condition above foramen magnum.

History: Patient, age 61; present illness began in 1910 with severe cervical pain radiating to shoulders, followed by loss of sensation and by spastic paraplegia. Atrophy of muscles of left upper extremity. Hyperalgesia beginning at C3 and extending to C5. Hypalgesia and analgesia below.

Neurological examination: On admission, weakness of lower facial muscles on left with droop of angle of mouth. Marked spastic paralysis of both lower extremities. Upper extremities showed diffuse atrophy from shoulder girdle to hand. Bilateral Babinski sign; patellar and ankle clonus. Abdominals absent. Beginning at C3, there is hyperalgesia, with analgesia below C5 (temperature and touch).

Autopsy revealed an egg-shaped grayish tumour, covered anteriorly by a thin layer of cord tissue, occupying third, fourth and fifth cervical segments. No abnormalities of dura externally. Sections at levels through seventh nucleus revealed normal cellular structure. No other pathological changes above foramen magnum.

CASE III. M. N.—*Diagnosis*: Inoperable intramedullary glioma, upper level at C3.

Flattening of left face, nystagmus in right plane, slight ptosis of left eyelid, corneal hypæsthesia, pharyngeal and palatal hypæsthesia, hyperæsthesia beginning at C1.

History: Patient, age 16, female; present illness began 15 months before admission with weakness of left lower extremity and incontinence of urine; shooting pains at the back of the neck, weakness of left arm.

Laminectomy revealed intramedullary tumour.

Neurological examination: On admission, slightly irregular pupils, slight ptosis of eyelid, bilateral corneal hypæsthesia, left facial flattening, nystagmoid jerks in right lateral plane, slight pharyngeal hypæsthesia. Upper extremities, loss of voluntary power, bilateral wrist drop with atrophy of hand muscles. Level of hyperæsthesia and hyperalgesia, upper limit at C1, involving several segments. Sensation lost in lower extremities and greatly diminished in the upper. Bilateral ankle clonus and Babinski sign. Knee jerks exaggerated. Abdominals absent. Laboratory findings negative.

CASE IV. J. B.—*Diagnosis*: Endothelioma, upper level at C2.

Drooping of left angle of mouth, slight rotatory nystagmus, strabismus of right eye, right pupil larger than left, palatal reflex absent, hyperalgesia beginning at C3. Operation and removal.

History: Onset of illness eight months before admission, with sharp, lancinating pains along ulnar side of left hand soon followed by pain in left hip, travelling down leg. Difficulty in micturition and in walking.

Neurological examination: Right pupil larger than left, convergent strabismus of right eye, drooping of left angle of mouth, slight rotatory nystagmus and slight deviation of tongue to right, with fine tremor. Spastic gait; dragging of left foot. Marked wasting of musculature of upper extremities. Bilateral hyperreflexia. Abdominals absent. Babinski response on left. Hypalgesia below C3.

Operation : Endothelioma removed, upper pole extending to C2; attached by two posterior roots on left.

CASE V. A. P.—*Diagnosis* : Inoperable intramedullary glioma, upper level C3.

Paresis of lower right facial muscles, left corneal hypæsthesia, weakness of right sternocleidomastoid, hypalgesia below C3.

History : Patient developed stiffness in back of neck radiating to shoulders, which was soon followed by a subjective feeling of tightness in epigastrium and by tympanites. Legs and arms became weak; difficulty in micturition. Operated on at the Neurological Institute by Dr. Elsberg; cord found to be greatly enlarged.

Neurological examination : Right pupil larger than left. Left corneal hypæsthesia. Paresis of right lower facial muscles. Slight deviation of tongue to right. Upper extremities; atrophy of the pectoral girdles and weakness. Lower extremities; markedly diminished power, with hypotonia. Biceps and triceps reflexes greatly diminished. Tendon reflexes in lower extremities exaggerated, bilateral Babinski sign, ankle clonus, absent abdominals. Hypæsthesia below C3; hypalgesia and loss of vibratory sense below D4.

CASE VI. J. L.—*Diagnosis* : Extramedullary neoplasm, upper level at C4.

Immobility of left angle of mouth with slight flattening of left face, right pupil larger than left, hyperalgesia beginning at C4.

History : Patient, age 55; present illness began about ten years ago with pain in the lower cervical spine and progressive weakness of legs.

For three months before admission he was dyspnoic and had poor sphincteric control.

Neurological examination : Some immobility of the left angle of the mouth and slight flattening of left side of face; right pupil larger than left; tongue slightly tremulous. Lower extremities spastic; power greatly diminished. Upper extremities not affected grossly. Level of hyperalgesia at C4, C5 and C6, with hypalgesia below. Bilateral Babinski; ankle clonus. Biceps and triceps jerks exaggerated.

A diagnosis of extramedullary neoplasm was made. Patient refused operation and left the hospital unimproved.

CASE VII. A. K.—*Diagnosis* : Extramedullary neoplasm, C3-5.

Flattening of right cheek, divergent right strabismus, hypæsthesia below C3.

History : Patient, female, age 48; present illness began five years before admission with gradual development of pain in back of neck radiating up back of head and followed by stiffness of neck. Gradual weakness and stiffness of the right arm. Legs stiff and weak, more marked on the right side. During the last year, some difficulty in sphincteric control.

Neurological examination : Flattening of the right cheek. Inability to move head, especially to the right. Right arm held extended, with fibrillary twitchings and atrophy of shoulder girdle.

Lower extremities, flaccid paraplegia with double foot drop. Hypæsthesia below C3.

A diagnosis of extramedullary neoplasm from C3 to C5 made.

Patient died of uræmia; no autopsy.

CASE VIII. C. C.—*Diagnosis* : Intramedullary neoplasm, upper level at C2-4.

Slight drooping of left eyelid, slight deviation of tongue to left, hypæsthesia beginning at C3.

History : Patient, male, age 44; present illness began five years before admission with numbness in fingers of left hand, followed by numbness in right. One year later he developed stiffness and difficulty in walking, which soon became impossible. Upper extremities became very weak and stiff so that he was unable to feed himself. Four months later he began to lose bladder control.

Neurological examination : Left pupil larger than right, slight drooping of left lid, slight deviation of tongue to left. Marked weakness of both upper extremities, with fibrillary twitching and wasting. Lower extremities, bilateral spastic paraplegia. Deep reflexes of upper and lower extremities exaggerated. Abdominal reflexes absent. Bilateral Babinski sign and clonus. Hyperæsthesia at C3, with hypæsthesia below. Patient developed a temperature of 104°F., with dyspnœa, and death ensued. No autopsy.

X-ray revealed irregular bone destruction in transverse process of third cervical vertebra on the right, associated with bony arthritic changes.

CASE IX. S. B.—*Diagnosis* : Extramedullary neoplasm, upper level at C1.

Insufficiency of convergence; jaw deviation to right, bilateral corneal hypæsthesia, slight deviation of tongue to the left, hyperæsthesia beginning at C2.

History : Patient, female, age 39; onset of present illness about one year ago, with hoarseness and difficulty in articulation. The dysarthria was apparently due to dyspnœa rather than to definite involvement of the glottis. About three months ago occasional regurgitation of the food through the nose occurred. Weakness and stiffness of the right lower and upper extremities developed.

Neurological examination : Slight prominence of right eye, right pupil larger than left, some insufficiency of convergence; the jaw opened with some difficulty and deviated to the right. Bilateral corneal hypæsthesia. Air conduction greater than bone on the left. Tongue deviated slightly to the left, with a few fibrillary twitchings. Generalized hyperreflexia more marked on the right. Bilateral Babinski sign; ankle clonus on right and on left; area of hyperæsthesia in the distribution of C2.

Diagnosis of neoplasm (inoperable) of C1 and C2 made.

Patient discharged unimproved.

Laboratory findings were essentially negative.

CASE X. D. G.—*Diagnosis* : Extramedullary neoplasm, upper level C3-4 (clinical).

Nystagmus in right lateral plane, pharyngeal reflex diminished, tongue deviation slightly to right; hyperæsthesia beginning at C3.

History : Patient, male, age 45; present illness dates back nine months before admission, with pain in the left side of the neck which became so severe that it necessitated staying in bed. Soon after he noticed pain with a feeling of heaviness in the left arm, then weakness developed in left leg.

Neurological examination : Nystagmus in the right lateral plane, pharyngeal reflex diminished, slight protrusion of tongue to the right. Patient had a typical gait of a left hemiplegic, with limited movement of left upper extremity. Marked contracture at the elbow and flexion of fingers, atrophy of the left shoulder girdle. Movements of the neck not restricted. All deep reflexes exaggerated, with patellar and ankle clonus on the left but not well sustained on the right. Bilateral Babinski sign. Hyperalgesia on the left below C2, hypalgesia on the right below C2. Vibration sense impaired below clavicle on both sides. Diagnosis of extramedullary neoplasm at C3-4 was made. Operation was advised, which was refused by the patient, and he was therefore discharged unimproved.

CASE XI.*—*Diagnosis* : Extramedullary sarcoma, C1-4, presenting clinical picture of cervical spondylitis.

Slight weakness of right facial muscles, nystagmus to the right. Operation and recovery.

History : Patient, male, age 24; present illness began fourteen months ago with severe pains in the right shoulder. These gradually spread to the small of the back, and to the right hip and thigh. Right upper extremity became weak.

X-ray examination at that time revealed arthritic changes of the cervical vertebræ, and the case was treated as one of cervical Pott's disease.

Four months before operation the symptoms advanced rapidly, with loss of power of the right and then of the left lower extremity, and later of the left upper extremity. At first retention of urine, later incontinence. Bowels obstinately constipated.

Neurological examination: Distinct nystagmus to the right; slight weakness of the facial muscles. Breathing abnormal in character. Marked wasting of both shoulder girdles, more marked on the right, and atrophy of intrinsic muscles of hands. Motor power of all extremities diminished; right upper limb totally paralysed. Reflexes of right upper extremity not elicited, much diminished on the left. Abdominals and cremasterics absent. Reflexes of lower extremities increased, with bilateral Babinski sign and ankle clonus. A belt of hyperæsthesia round the entire neck and upper chest. Hypalgesia from C3 to L3, and analgesia below.

X-ray, arthritis of cervical vertebræ.

Operation: Laminectomy. Neoplasm exposed, extending from the fourth cervical segment to the foramen magnum.

Biopsy showed spindle-celled sarcoma.

CASE XII.*—*Diagnosis*: Extramedullary myxosarcoma, C4-7.

Pupils unequal, right larger than left, slight left facial weakness, left recurrent laryngeal paralysis, hypalgesia below D5.

History: Patient, male, age 59; symptoms began seven months before admission, with pain in the left shoulder extending down the arm. This was soon followed by weakness and atrophy of the limb. Four months later, girdle sensations round upper abdomen, followed by weakness of lower extremities, left greater than right. Increased frequency of urination.

Neurological examination: Pupils unequal, right larger than left; slight left facial weakness; left recurrent laryngeal paralysis; marked atrophy of left upper extremity and weakness. Biceps and radial reflexes not elicited. Triceps jerk very weak on left, but lively on right. Abdominals absent. Reflexes of lower extremities exaggerated with ankle clonus and bilateral Babinski sign. Some hyperæsthesia over upper chest. Pain and temperature senses diminished below level of fifth rib.

X-ray, spondylitis of cervical vertebræ.

Operation: Bluish tumour mass, 4 cm. long, extending from the fourth to the seventh cervical segment. Cord considerably flattened and pushed to the left.

Examination of tumour, myxosarcoma.

REFERENCES.

¹ ELSBERG, *Tumours of the Spinal Cord*, 1925.

² BAZETT and PENFOLD, "The decerebrate animal in the chronic and acute condition," *Brain*, 1922, xlv, 185.

* Taken from Elsberg's *Tumours of the Spinal Cord*.