GANGLIONEUROMATA OF THE SPINE ASSOCIATED WITH VON RECKLINGHAUSEN'S DISEASE

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Within the neurodermatoses is included von Recklinghausen's neurofibromatosis, a heredofamilial disease consisting of hyperplasia and neoplasia of neuroectodermal tissue. Essentially, neurofibromatosis consists of cutaneous pigmentation associated with tumors of either the peripheral or central nervous system, or both.

Neurofibromata containing ganglion cells occasionally have been reported in association with von Recklinghausen's disease. Cases of multiple dumb-bell ganglioneuromata of the cervical spine with spinal-cord compression, associated with typical findings of von Recklinghausen's disease, have rarely been reported previously.

CASE REPORT

On July 29, 1958, a 44-year-old white woman was admitted to the Medical School Hospital with the complaint of difficulty with movement of the right extremities.

She had been well until 4 years previously, when there was gradual onset of clumsiness, weakness, numbness, tingling, dull aching and sharp shooting pains in the right lower extremity. Gradually the right upper extremity became involved. Two years before admission she noticed slight similar symptoms on the left. Six months before admission, and especially the previous 6 weeks, the right-sided symptoms became severe. There were no other complaints relating to the central nervous system.

There was no pertinent past history. Her father and two of her three sons have brown macules of the skin; otherwise they are well.

EXAMINATION. The patient was well nourished, alert, and in moderate distress, complaining of pain in the right lower extremity, which was in spastic flexion. She was unable to walk, and her right upper extremity revealed marked motor loss, muscle atrophy and spasticity. There were café-au-lait macules, most numerous on the extremities, varying in size from 1 mm. to 3-4 cm. in diameter (Fig. 1). Several pedunculated cutaneous tumors were present. The blood pressure was 150/105, and general physical findings otherwise were noncontributory. Neurologic examination revealed positive findings only caudal to the high cervical area: hyperreflexia of all tendon reflexes, greater on the right; bilateral extensor plantar response; clonus of the right lower extremity, severe flexor spasm of the right lower extremity, severe weakness of both right extremities, especially of the right hand, and generalized moderate atrophy of the muscles on the right, most marked of the right hand; a diffuse loss of perception of sharp pain and touch was present, the level being at C3–C4.

Plain roentgenograms of the spine demonstrated widening of the intervertebral foramens (cervical and lumbar). Pantopaque myelography revealed a cobblestone pattern of the cervical area, indicating the presence of extramedullary, intradural masses, the area of most intense involvement extending from C5 at least as far cephalad as C2 (Figs. 2 and 3). (Similar radioluencies were found at the lumbar level, but were less marked.) The only other pertinent laboratory finding was urinary catecholamines, within normal limits (9 micrograms per 100 ml.).

Clinical diagnosis: von Recklinghausen's disease, with multiple neurofibroma compressing the cervical spinal cord.

Operation. On Aug. 5, 1958 cervical laminectomy (from C1 to C5 inclusive) and modified suboccipital craniectomy were done. After removing the arch of the atlas, the cord began to pulsate. Just below the medulla the cord was about 1 1/2 cm. in diameter. Beneath the arch of the atlas there was an extradural tumor, 1 to 1 1/2 cm. in diameter, to the right of the spinal cord.
Figs. 2 and 3. Roentgenograms of cervical spine during Pantopaque myelography. (Left) Left oblique view, showing intradural, extramedullary filling defects. (Right) Posteroanterior view, showing filling defects, and almost total block to flow at lower cervical level.

(Fig. 4). It was dissected free. After removal of the posterior rim of the foramen magnum, this tumor was seen extending through the dura mater and upward along the medulla, and five additional tumors were removed, these extending between C2 and C5 (Fig. 5). The tumors had been attached to nerve roots, mainly the dorsal roots, which appeared to run directly into the sheath of the tumor.

Postoperative Course. There was severe pain in the right lower extremity. The preoperative spasm persisted. On Sept. 3, 1958 the patient had a left spinothalamic tractotomy at T3-T4. Upon recovery, the pain gradually abated. As the flexor spasm decreased, she gradually became ambulatory. A pedunculated, soft tumor was removed from the ventral surface of the left foot.

Pathological Report. The tumors removed from the spine were ganglioneuromata. Grossly, these tumors were well encapsulated (Fig. 5). Microscopically, well-differentiated ganglion cells were seen (Fig. 6). Silver impregnated sections disclosed a meshwork of nerve fibers. The tumor of the left foot was a dermatofibroma, silver stains revealing no nerve cells or fibers.

Course. The patient was discharged on Sept. 13, 1958, ambulatory. There was partial loss of position and vibratory sense on the right, especially of the hand. The tendon reflexes of the extremities were equal and normally active, except for hyperactivity of reflexes of the right lower extremity. The plantar response was extensor on the right. The spasm of the right lower extremity had decreased markedly. There was a dermatomal distribution of sensory loss on the right of C2 and (subtotal) C3, and on the left of C3.

COMMENT

Synonyms for ganglioneuroma are gangliocytoma, ganglioglioma, true neuroma, glioneuroblastoma, neuroastrocytoma, ganglioma, Purkinjeoma, etc. By anatomical location three classes are distinguished: (1) of the cerebral hemispheres, brain stem and spinal cord; (2) of the cerebellum; and (3) of the sympathetic trunk.

The age of greatest incidence is in the first three decades. Occurrence is more frequent in females. The incidence is lower, e.g., in Cushing’s series it was 0.2 per cent; in Zülch’s series (4,000 cases) it was 0.4 per cent, which was 0.7 per cent of the neuroectodermal tumors. The site in order of frequency in Zülch’s series was (1) cerebral hemispheres, (2) tuber cinereum and third ventricle, (3)