DESTRUCTION OF CERVICAL VERTEBRA BY SOLITARY NEUROFIBROMA
REPORT OF A CASE WITH QUADRIPLEGIA

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The neoplasms most frequently encountered in the spine, spinal cord or spinal nerve roots are those that grow from the spinal nerves. Various pathological terms have been given to these tumors, namely, fibromas, perineurial fibromas, neurofibromas, neurilemmomas and schwannomas. There have been differences of opinion as to their origin. Stout and others felt that their pathological characteristics indicate that these tumors are composed of, or are derived from, the schwannian cells of the nerve sheath. As to their incidence, the tumors fall into two large groups; (1) the neurofibromas of von Recklinghausen’s disease, which are part of a systemic disorder, and (2) the neurofibroma that occurs as a solitary neoplasm without other stigmata of neurofibromatosis. The pathology of the tumor in the two groups, however, is the same.

The involvement of bone by these tumors occurs in one of three different ways: (a) the bone is invaded and destroyed by the neighboring tumor; (b) the tumor begins as a primary growth within the bone; and (c) the bone is destroyed from pressure atrophy without invasion by the adjacent soft tumor.

Changes in bone associated with von Recklinghausen’s disease are a common occurrence. According to Hodges et al., they are found in 7 per cent of all cases of this disease. They were classified in 1924 by Brooks and Lehman who described lesions causing irregularities in the outline of the bone, tumors destroying the interior of the bone, called subperiosteal bone cysts, and pedunculated subperiosteal tumors. The fundamental cause of these changes in the bone, according to Brooks and Lehman, is the development of a neurofibroma in a nerve that supplies the periosteum. As the tumor enlarges, pressure is exerted on the cortex of the bone, causing erosion and formation of pits, which are filled with neoplastic tissue. Radiologically, they may take on the appearance of a cyst or a thin shell of cortex covering the fibrous tumor tissue. Diagnosis is made easily in the presence of other stigmata of the systemic disease, including the characteristic pigmented lesions of the skin, the hereditary factor and others.

Changes in the bone concomitant with solitary neurofibroma are similar to those described above. The changes encountered most frequently with neurofibromas of the spinal nerve roots are erosion and atrophy of bone caused by pressure. Atrophy, deformity or destruction of one or more vertebral pedicles, enlargement of an intervertebral foramen or deep indentation in the body of the vertebra are associated so commonly with this tumor as to be almost a part of its clinical syndrome. On the other hand, infiltration and destruction of bone by a solitary neurofibroma are extremely rare. When the so-called dumb-bell tumor invades and destroys the vertebra, the roentgen-ray appearance is that of a metastatic malignant growth. This diagnostic error is made most frequently in cases of tumors arising

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from the thoracic nerve roots. The extraspinal extension of the tumor into the thoracic cavity is misdiagnosed as cancer of the lung and the erosion of the bone is attributed to metastasis. Such a tumor was described by Poulis;[16] it took origin from the 7th cervical nerve root and extended into the thoracic cavity. The "malignant" lesion was treated by roentgen therapy, but a benign tumor was found at autopsy. Alexander and James[1] described a paraplegic patient with a large neurofibroma which had eroded the 3rd and 4th thoracic vertebrae extensively. Diagnosis of a malignant tumor was followed by roentgen therapy. The tumor was finally removed surgically with good recovery. Bucy[2] has had a similar case. The available literature includes only 2 cases reported as primary neurofibroma of the vertebra (Conley and Miller,[7] and DeSanto and Burgess[8]). In these cases there was roentgenographic evidence of extensive bone destruction having the appearance of a trabeculated cyst without evidence of a concomitant tumor of an adjacent nerve root. Each of these tumors was located in the 1st sacral vertebra and was removed surgically.

This paper describes a patient with a solitary dumb-bell neurofibroma of the left 3rd cervical nerve root which invaded and destroyed two vertebral bodies. A report of a similar case could not be found in the available medical literature. The serious consequences resulting from this lesion, the method employed in treatment and the end results obtained justify a report of the case.

CASE REPORT

Q.H. #143,941. S.N., a 33-year-old male banker from Bangkok, Thailand, was admitted to the Queen's Hospital on Feb. 3, 1958 by ambulance from the airport. He had flown from Bangkok to Honolulu.

The patient was referred by Dr. Udom Poshakrisha who gave the following history. "The patient first noticed a slow-growing mass on the left side of his neck in 1951 causing radiating pain in the left shoulder. In June 1952 the mass was removed under local anesthesia. It was the size of a pigeon's egg, inseparable from the 4th cervical nerve root and histologically was a neurilemmoma. Within 6 months after removal, the tumor had recurred to the size of the thumb. A second operation was advised but the patient refused." He was not seen again by Dr. Udom until October 1955 when he was admitted to Siriraj Hospital, Bangkok with signs of spinal-cord involvement. "He walked with a spastic gait; both upper extremities were involved, the left much more than the right. The left hemidiaphragm was paralyzed. The tumor on the left side of the neck was now the size of a hen's egg. The tendon reflexes were hyperactive with bilateral ankle clonus and Babinski sign. There was hypothermia of the entire body below the clavicle.

"Pantopaque myelography revealed a complete block of the spinal canal at the level of C5. A laminectomy from C2 to C5 inclusive was performed under endotracheal anesthesia on Oct. 7, 1955. The spinal cord at the C2-C4 level showed distinct fusiform swelling with obliteration of the normal superficial landmarks. There were no pulsations. The cord was reported to have a finely granular appearance with alteration of the normal vascular pattern at this level. There was no visible enlargement of the motor or sensory nerve groups to suggest an intradural invasion of the neurilemmoma. The operator, suspecting an intramedullary tumor, decided to carry the exploration no farther than the decompressive laminectomy.

"Following this operation the patient showed temporary improvement in the use of his upper extremities, but was unable to walk and remained confined to bed thereafter. He was given a 3-week course of roentgen-ray therapy to his cervical spine (amount not recorded). The tumor on the left side of his neck was not removed. For the next 27 months the patient was entirely bedridden. The function of all four extremities decreased slowly over the first 6 months until he was completely helpless and unable to care for himself. His bowel and bladder, however, remained functional until November 1957. Further surgery on his cervical