CASE REPORTS AND TECHNICAL NOTE

BILATERAL CERVICAL NEUROFIBROMATA PRESENTING
AS CERVICAL SPONDYLOSIS

TWO UNUSUAL CASES*

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Disease of the cervical spinal cord can have multiple etiologies, both extrinsic and intrinsic. When syndromes of the root occur in conjunction with the myelopathy, the gamut of diagnostic possibilities is narrowed. Extrinsic tumors, such as neurofibromata and meningioma acting as localized masses, can cause symptoms and signs of both root and long-tract involvement. Cervical spondylosis, a well recognized process so adequately described in recent years, also causes protein signs and symptoms involving spinal cord and roots. In the older age groups, especially when degenerative bone changes are present and no other systemic or radiologic changes are noted, spondylosis is the leading diagnostic possibility. Further diagnostic tests, such as myelography, most often will delineate the lesion and clarify the etiology.

However, the presence of spurs extending into the spinal canal may not be the cause of the signs involving root and the spinal cord even when they are at the correct anatomical level. This occurred recently in 2 patients having osteoarthritic spurs in whom the true and unusual diagnosis of bilateral small neurofibromata on anterior nerve roots at the same level was not recognized until they were exposed at operation.

CASE REPORTS

Case 1. M.K., a 73-year-old white male, 2 years before admission had first noticed numbness in the finger tips of the left hand and had begun to drop small objects. Several months later paresthesias and slight weakness of the left hand and arm developed and progressed very slowly. One year prior to admission he noticed mild "weakness in the knees" and fell once or twice. Three months after this, his gait became somewhat unsteady and paresthesias occurred in the right fingers and up to the elbow in the left arm. Six months before admission, he noted numbness in the big toes of both feet, and this spread gradually to all toes.

Past History. In 1940, he had resection of a benign papilloma of the bladder.

In 1946, mild diabetes mellitus was first noted. This was controlled adequately by diet until 1 year previously when small doses of insulin were required.

In 1960, he had segmental resection of papillary carcinoma of the bladder.

Examination. The gait was moderately spastic with mild unsteadiness. Generalized weakness of 10–30 per cent was present in all extremities. It was more marked proximally than distally, greater on the left side than on the right, and greater in the arms than in the legs. Winging of the scapulae also was present. The biceps reflexes were hypoactive on the left and hyperactive on the right. Knee jerks were hyperactive. Ankle jerks and hamstring reflexes were normal. There were no pathological toe signs.

Sense of position was decreased moderately in the right foot and less so in the left foot. It also was decreased in the hands, but only slightly, more on the right than the left. Perception of vibration was decreased markedly in both arms to the elbows and slightly decreased in the feet. Sensations of pain and touch were decreased moderately in both legs, more on the left than the right. There was more marked decrease in both hands and on the dorsum of the left arm to the elbow. Perception of temperature was decreased slightly on the left side of the body.

Laboratory findings were: Hemogram, normal; ESR, 4 mm./hr.; FBS, 96 mg. per cent; alkaline phosphatase, 11 King-Armstrong units (normal 5–15); acid phosphatase, 0.80 Bessey-Lowry units (normal 0.10–0.90); cerebrospinal-fluid protein, 160 mg. per cent with 1 white blood cell.

Roentgenograms of the cervical spine showed marked degenerative disease of the intervertebral discs from C3 to T1 (Fig. 1). Some bony ankylosis was present between C4 and C5. No large osteophytes or other abnormalities of the intervertebral foramina were seen.

Course. Myelography was performed (Figs. 2 and 3). There was a partial block between C4 and C5. From
C3 to C5, the opaque column showed discontinuity and lifting, and there were indentations of the column bilaterally at C6 and C7 and T1 and T2.

Operation. On Nov. 10, 1961, laminectomy from C3 to C6 was performed with the patient in the prone position. The dura mater filled the bony canal, but there was no evidence of any significant abnormality. When the dura mater was opened, the cord appeared grayish, somewhat flattened and slightly atrophic in the region of C3 and C4. The right edge of the cord then was carefully retracted medially preparatory to cutting the dentate ligaments. A small nodule, about 4 mm. in diameter (Fig. 4), was seen on the ventral C4 root making a small indentation in the ventrolateral aspect of the cord. The entire root was not involved, and it was possible to save about one quarter of the fibers after resection of the tumor. The dentate ligaments then were cut and the left side of the cord was inspected. Again there was no evidence of a tumor until the edge of the cord was retracted. Here the entire ventral C4 root was involved by a tumor, 5 mm. in diameter, again indenting the ventrolateral aspect of the cord. The tumor was resected and the dentate ligaments were cut. A dental tool was passed under the cord in the exposed area. Hard ridges were felt at the interspaces, but none of them was sufficiently large to compress the spinal cord or nerve roots.

Course. Postoperatively there was improvement in strength of the lower extremities with decrease in the spasticity and improvement in gait. There was marked weakness in the distribution of the C4 motor root, but function was improving well with physiotherapy. Paresthesias and numbness of the hands improved consider-
ably, and the right arm and hand were normal when the patient was discharged on Dec. 9, 1961. There still was slight decrease in perception of pain and touch in the left forearm and fourth and fifth fingers.

Pathological Diagnosis. Bilateral schwannomas.

Case 2. J.S., a 40-year-old white male, was first seen in August, 1959. Two years previously, when bowling, the right leg gave away and he fell. Upon getting up, both legs were slightly weak, the right more so than the left; and his arms also felt slightly weak. A limp developed on the right and he walked stairs with difficulty but unaided. Occasionally the right leg shook spontaneously at night. In addition to the motor dysfunction, numbness appeared in both legs to the mid-thighs and in all fingers, especially the first and second fingers on both sides. One year prior to admission, for a 2-month period, he had episodes of burning sensation in the lower region of the girdle. He never had any pain.

Past History. He had been treated for 1 month during World War II for “conversion hysteria” manifested by headaches, nausea and incoordination.

Examination. Gait was spastic, and tandem walking was poor. The right biceps and deltoid muscles were decreased 25 per cent in strength, and the left biceps muscle about 20 per cent. Grips of the hand were diminished slightly in power. There was 50 per cent loss of strength in the hamstrings, tibialis anticus and ilio- psoas and 25 per cent in the quadriceps femoris muscles. The biceps reflexes were hypoactive, and the triceps were normal. Deep tendon reflexes in the lower extremities were hyperactive, more so on the right than the left. There was a positive Babinski’s sign bilaterally.

There was decreased sense of position in the right foot. Very slight decrease in sensations of pain, touch and temperature was present in the first and second fingers and ulnar half of both hands.

Laboratory findings were: Hemogram and urinalysis, normal; cerebrospinal-fluid protein, 43 mg. per cent with 1 white blood cell.

Roentgenograms showed mild narrowing of the C5-C6 and C6-C7 interspaces with formation of osteophytes, both anteriorly and posteriorly. Some straightening of the normal cervical curvature was present (Fig. 5).

Course. Myelography demonstrated a large defect at C5 and C6 with indentation of the column of oil ventrally and displacement of the column dorsally (Figs. 6 and 7). There were smaller indentations ventrally at C4 and C5 and at C6 and C7. Posterior bony lipping rather than intervertebral discs appeared to be the cause of these defects.

Operation. Laminectomy was performed from C4 to C7. When the dura mater was opened, the cord did not appear remarkable other than a slight discoloration in the region of the posterior columns. When the edge of the cord then was retracted medially preparatory to cutting the dentate ligaments, a tumor, 8 mm. in size and involving about five fibers of the ventral root of C6, was found on the left side. A similar tumor, 1 cm. in diameter, was found on the ventral root of C6 on the right. The tumors were resected, and then definite bilateral indentation of the spinal cord could be seen. A dental tool was passed under the cord and a small ventral spur was felt.

Pathological Diagnosis. Schwannoma with proliferation of sheath cells in partially degenerated roots.

Course. Postoperatively the patient had an epidural hemorrhage which was evacuated, but it delayed his recovery. Two years after operation, he was able to work 60 hours a week, although there still was some impairment of strength of the right extremities.

DISCUSSION

Since the excellent descriptions of Brain et al. and Spillane and Lloyd,31 cervical spondylosis has been a well recognized clinical entity. Although it
most often is found in the older age groups, patients have been reported as young as 14 years of age. The symptoms may be present for only a few months or many years. The signs and symptoms are protean, involving cord, roots or both, and often may resemble extramedullary tumors of the cord, though motor symptoms more often predominate compared to sensory symptoms in spondylosis. The protein in the cerebrospinal fluid may be as high as 160 mg. per cent. The primary pathological defect is in the intervertebral disc which degenerates and promotes formation of osteophytes in the periphery of the vertebral body. Osteophytes may project posteriorly into the vertebral canal or intervertebral foramen causing pressure upon the spinal cord and nerve roots. These degenerative changes often are seen in roentgenograms of the older age groups, even when clinical symptoms are not present. The significance of these changes arises in patients with symptoms, and myelography frequently is necessary to determine whether or not the spinal cord or roots are being compressed by the spurs or whether another pathological process is present.

The symptoms, signs, plain roentgenograms and the myelographic findings at the proper level in our 2 cases were most compatible with the diagnosis of cervical spondylosis as outlined above.

Two principal forms of so-called neurofibromata must be distinguished. The first is the true neurofibroma which usually is a manifestation of von Recklinghausen’s disease, though there may be a central form with multiple lesions and no peripheral manifestations. There is diffuse proliferation of Schwann cells and connective-tissue fibers are arranged haphazardly with a loose texture.

The second type of tumor has been called a neurilemroma by Stout or a schwannoma by Russell and Rubinstein. The great majority are solitary and usually are unassociated with neurofibromatosis though multiple lesions can be considered an aspect of von Recklinghausen’s disease. These tumors are lesions of the posterior roots and only rarely is a motor root involved.

Location of the lesion in the cervical region does not appear to be of any help in the differential diagnosis. Elsberg found that all areas of the spinal canal had an equal predilection for neurofibromata though in one series the cervical region was the least common site. However, root pains, usually unilateral, rather than cord symptoms usually are one of the first indications of the presence of this tumor.

Duration of symptoms may be from several months to years with a neurofibroma as in cervical spondylosis though the content of protein in the cerebrospinal fluid tends to be higher in the former.

In 40 to 65 per cent of the cases, the neurofibroma cause changes in the plain roentgen-ray films. These consist of erosion of the pedicle,
lamina or the body of the vertebrae, widening of the interpediculate distance, or enlargement of the intervertebral foramina. The myelographic changes are characteristic: a rounded concave defect with displacement of the spinal cord, and there may be a total block.\textsuperscript{2,13}

Differentiation between cervical spondylosis and neurofibromata should not be difficult. However, the 2 cases presented here were very unusual in several aspects. First, the tumors were multiple without evidence of generalized neurofibromatosis. Second, they were bilateral on roots at the same level. Third, the roots involved were motor. Fourth, both cases presented as fairly typical examples of moderately severe cervical spondylosis. Roentgen-ray examination, including myelography, tended to confirm this diagnosis.

Disease of the cervical spinal cord and roots has protean manifestations and great care must be taken to avoid diagnostic pitfalls. At operation, the dura mater must be opened in all cases of spondylosis when the spurs do not appear adequate to produce the clinical findings.

**SUMMARY**

1. Two unusual cases of bilateral schwannomas on the same cervical motor root are presented. Neither patient had other indications of neurofibromatosis. Both patients initially were regarded as having cervical spondylosis, both from their clinical and radiological manifestations including myelography, and an accurate diagnosis was not made until the lesions were exposed at operation.

2. The problems of cervical spondylosis and extrinsic cervical spinal tumors are discussed and an attempt is made to compare and differentiate between these lesions.

3. The pathology of cervical-nerve tumors is presented briefly and the differences between the usually single schwannoma (or neurilemmoma) and multiple neurofibromas of von Recklinghausen's disease are highlighted.

**REFERENCES**


