LYMPHOMA OF SPINAL NERVE ROOT

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(Received for publication August 6, 1958)

The central nervous system is not often involved by lymphomas. When this does occur, it is usually brought about by secondary invasion of the epidural space. According to Whisnant et al., such involvement takes place in 10 to 20 per cent of patients who have malignant lymphomas. Many cases have been reported since Murchison’s original article appeared in 1870. Intradural invasion, however, appears to be even rarer, and cases of primary lymphomas of the central nervous system are extremely unusual.

This report is based on a case which seems to be one of malignant lymphoma originating in the central nervous system and involving a spinal nerve root. Search of the pertinent medical literature has failed to uncover any similar instance.

CASE REPORT

#58-5162. M.S., a 75-year-old white woman, was admitted to the University of Kansas Medical Center in April 1958, complaining of numbness and weakness of the right leg.

In July, 1957 the patient had noted numbness in the right great toe upon awakening one morning. The condition became slowly progressive and eventually affected the entire right leg to the level of the knee. The sensory loss was accompanied by increasing muscular weakness of the extremity, most notably a foot drop. The patient was also bothered by a burning sensation over the anterior aspect of the leg.

The patient had been living in Indonesia for 2 years before her symptoms manifested themselves, and for some time thereafter, the patient had been living in Indonesia. She described the food as “poor” and lacking in vegetables. She had taken vitamin supplements regularly, however, and had felt well during this period.

Examination. The patient appeared well developed, well nourished, and not in severe distress. General physical findings were normal. There was some straightening of the normal lumbar curve associated with slight spasm of the paraspinal muscles on the right side. There was no vertebral tenderness. Range of motion of the spine was limited in all directions. She had considerable tenderness in the right gluteal area and over the course of the sciatic nerve. There was tenderness upon pressure over the right tibia. The skin of the right lower leg and ankle was smooth and glistening.

There was generalized weakness of the right lower extremity, especially of the dorsiflexors of the foot and toes. The patient’s muscle tone was greatly reduced and she had a right foot drop. She was barely able to walk with the aid of a cane. Right patellar and ankle jerks were absent. The other tendon reflexes were slightly hyperactive. There were no pathological reflexes. Lasègue’s sign was positive on the right side. Sensory abnormalities were also confined to the right leg: diminution of sensation to light touch and painful stimuli, decreased vibratory sense, and moderate hyperalgesia in the distributions of L5 and S1.
Superficial sensory losses were of the "stocking" type and extended from the knee downward. Position sense was intact. Skull was normal to auscultation and percussion. All the cranial nerves were intact. There were no cerebellar or meningeal signs. The patient's intellectual functions were not impaired.

Laboratory examinations, including hemogram, serology, fasting blood sugar, blood urea nitrogen, liver function tests, serum electrolytes, serum calcium, phosphorus and alkaline phosphatase, all gave findings well within normal limits. The erythrocyte sedimentation rate was 5 mm. in 1 hour (Wintrobe). Serum protein electrophoretic pattern was normal. Examination of urine failed to reveal the presence of Bence-Jones protein. The cerebrospinal fluid obtained at the time of myelography was found to contain 131 red blood cells and 209 lymphocytes per c. mm., 41 mg. of protein per 100 ml. and 25 mg. of sugar per 100 ml. Serology was negative and colloidal gold curve was 0011100000. The electrocardiogram was reported as normal. The clinical diagnosis was neuropathy of the sciatic nerve on the right, possibly from multiple myeloma because of the severe pain in the bones.

Roentgenograms of the cervical, thoracic and lumbosacral spines showed only generalized decalcification. Films of the right lower extremity and chest were reported as normal. On the 5th hospital day myelography was performed. The subarachnoid space was well visualized from the caudal limits of the sac to the level of T10. A bizarre filling defect was noted on the right side at the level of the 4th lumbar interspace (Fig. 1). There was no block to the flow of Pantopaque. Interpretation of the unusual myelographic defect ranged from enlargement of the nerve root caused by inflammatory or neoplastic disease to displacement and deformity secondary to extrinsic pressure by herniated nucleus pulposus, extradural neoplasm such as metastatic carcinoma, or intradural neurofibroma. The cerebrospinal fluid findings of elevated count of cells and low sugar were in favor of the first diagnostic possibility. Surgical exploration was felt to be mandatory in view of the patient's progressive disabling disease and of the diagnostic dilemma.

Fig. 1. Myelographic defect at level of L4–5 interspace on the right.
Operation. Laminectomy of L5 was performed on May 1, 1958. No pathology was noted in the extradural space. Intradural exploration showed that the myelographic defect was caused by two greatly enlarged nerve roots in close apposition to each other. These were interpreted by the surgeon as the sensory and motor roots of the 5th lumbar nerve. They were purplish brown in color and had a granular, turgid appearance. The pathologic change in the roots extended from their exit from the subarachnoid space rostrally for 3 cm. All the other roots appeared normal as did the pia arachnoid and the dura mater. The spinal fluid was clear and there were no adhesions between the roots. Electrical stimulation of the two diseased roots produced no response. The sensory root was excised and the motor root was left intact.

Pathological Reports. A frozen section was reported as showing "inflammatory infiltration of nerve root." Cultures obtained at the time of operation for pyogens, fungi, and mycobacterium were eventually reported as negative. A portion of the final pathological report follows (Dr. J. Boley): "Sections of the nerve reveal the entire trunk to be infiltrated with mononuclear basophilic cells, many of which are lymphocytes. Others have larger, more vesicular, oval or distorted nuclei and resemble lymphoblasts or reticulum cells; some of the latter contain nucleoli. Mitoses are noted and nuclear fragmentation is prominent. The endo-, epi- and perineurium are all involved, and the neural tissue appears compressed in some areas. Fibrosis is not evident. Diagnosis: lymphoma involving nerve root" (Fig. 2).

Postoperative Course. The patient made a satisfactory and uneventful recovery. She was re-examined extensively in an effort to discover neoplasia elsewhere, but none was found. She received a total of 3079 r (air dose) to the lower lumbar and sacral spines in seven divided doses. She was discharged on the 7th postoperative day, essentially free of pain. She was given a foot brace in an effort to improve her gait.

On June 16, 1958 the patient was re-examined. She reported that her original pain had gone although she still had mild pain in the hip on the right. She was able to walk well with the help of her brace and a cane. The foot drop persisted though she was now able to dorsiflex her right toes. The sensory deficit appeared to be slightly less extensive and the knee and ankle jerks were absent. The marked tenderness of the leg, which had been so striking preoperatively, was gone.

DISCUSSION

This case is interesting not only from the standpoint of the unusual pathology present, but also as a problem in differential diagnosis. The patient's history of progressive and ascending sensory loss and motor weakness in one lower extremity is highly suggestive of an intradural spinal cord tumor, probably of the cauda equina. The presence of burning pain is frequently associated with neuropathies caused by deficiency states or metabolic disease. Her 2-year residence in Indonesia, under poor dietary conditions, would lend strength to this supposition were it not for reliable evidence that she had taken supplementary vitamins. The exquisite pain in the bones and the tenderness that she exhibited are compatible with disease of the hematopoietic system, usually multiple myeloma, another known cause of bizarre neuropathies. Myelographic findings weighted in favor of a mechanical basis of the patient's symptoms.

The cerebrospinal fluid values in this case are of interest. The lymphocytic pleocytosis, low sugar, and normal protein are typical of the chronic (tuberculosis and Cryptococcus) and aseptic meningitides. Berg has recently reviewed the subject of decreased sugar in the spinal fluid (hypoglycorrhachia). He found that it was frequently associated with malignancies involving the central nervous system, particularly diffuse carcinomatosis or gliomatosis of the leptomeninges. He concluded that the presence of hypoglycorrhachia associated with signs of increased intracranial pressure, meningeal irritation, organic mental changes, or cranial-nerve or spinal-root signs may be taken as evidence of diffuse meningeal neoplasia when infection can be ruled out. Berg also felt that the development of meningeal signs and the finding of a
low spinal-fluid sugar in a patient known to have a lymphoma probably indicate a complicating Cryptococcus infection rather than meningeal spread of the neoplasm.

The pathological diagnosis of lymphoma made on the basis of the surgical biopsy raised the important question of the origin of such a neoplasm in the central nervous system. Almost all secondary lymphomas are confined to the spinal epidural space and do not involve the neural parenchyma or the nerve roots. It is interesting to note that in their review of lymphomatous involvement of the central nervous system, Whisnant et al.⁶ failed to mention nerve-root invasion except as part of a diffuse meningeal involvement.

Elsberg² mentioned having encountered 20 lymphosarcomas involving the spinal cord or roots, yet all but one were extradural. The one intradural lesion spread dif-
fusely through the leptomeninges. He also reported upon 3 reticulum-cell sarcomas, 2 of which were intradural and involved the cord and roots over many segments. In none of his cases was there actual invasion of the nerve root. Recently, Pool and Pava 5 found a metastatic lymphoma originating in the stomach which affected the intracranial portions of the 5th, 8th and 9th cranial nerves.

In our patient, no evidence of a neoplasm elsewhere in the body could be determined by clinical investigation. No extradural or other intradural pathology was seen. It is not unreasonable to postulate the existence of primary neural lymphatic tissue as the site of origin of this lesion. This possibility has been suggested previously by other writers. 8

SUMMARY

A 75-year-old patient had a foot drop and severe pain of the left hip and leg. The signs and symptoms pointed to neuropathy of the sciatic nerve. A myelogram showed an unusual defect and examination of the cerebrospinal fluid revealed a decreased sugar content. The diagnosis of a malignant lymphoma of the 5th lumbar nerve root was established at operation after examination of pathologic specimens. This case is believed to be a rare instance of lymphoma originating in the central nervous system.

ADDENDUM

The patient remained symptom free until July 25, 1958 when she was found to have signs of involvement of the left C8 nerve root. Relief was obtained with reontgen-ray therapy. Following this, she started a downhill course with signs of involvement of cervicothoracic nerve roots (C8-T5), return of severe pain and neurological deficit in the lumbosacral distribution and eventual right oculomotor and left facial palsy a few days before death, on Dec. 15, 1958.

Complete postmortem examination revealed the following: no evidence of lymphoma was found in any part of the body outside the nervous system. There was widespread, diffuse, perivascular infiltration by malignant lymphoma of the frontal white matter and the leptomeninges of brain and cerebellum. The subependymal regions of lateral, 3rd and 4th ventricles were densely infiltrated by lymphomatous tissue. Multiple levels of the spinal cord showed infiltration of the parenchyma by tumor cells. The right oculomotor nerve was hemorrhagic. No neoplastic cells were found in the left brachial plexus but there was widespread involvement of nearly all nerve roots and spinal ganglia at all levels. Metastatic lymphoma was also found in the pineal body.

The postmortem findings thus confirmed the fact that this tumor arose within the confines of the central nervous system. Final diagnosis was malignant lymphoma of the reticulum-cell sarcoma type.

REFERENCES