Spinal cord sarcoidosis

Case report

TIMIR BANERJEE, M.D., AND WILLIAM E. HUNT, M.D.
Division of Neurological Surgery, The Ohio State University Hospitals, Columbus, Ohio

A case of spinal cord sarcoidosis simulating an intramedullary tumor is presented. Surgical intervention increased the neurological deficit which was subsequently decreased by steroid therapy. The common complications are discussed and the need for extensive physical therapy emphasized.

KEY WORDS spinal cord tumor · intramedullary tumor · sarcoid

SARCOIDOSIS, also known as Besnier-Boeck-Schaumann disease, is a non-caseating, chronic granulomatous disease of unknown etiology affecting multiple organs. This disease involves the nervous system in from 1% to 5% of the cases described in various series. In a series of 422 patients, James, et al., reported a 7% incidence of neurosarcoidosis; four of these cases had papilledema. In 1909 Heerfordt described cranial nerve palsies and diabetes insipidus in association with Uveo-parotid fever. It was soon recognized that 50% of the cases of Heerfordt's disease were associated with one or more of the following: cranial nerve palsies, peripheral neuritis, diabetes insipidus, hypersomnia, spinal cord involvement, and cerebellar deficits. Sarcoidosis may occur at any site within the brain or spinal cord. However, the meninges and the region of the floor of the third ventricle including the optic nerves and pituitary gland are most commonly affected. The spinal cord is rarely involved.

Case Report

A 36-year-old Negro woman had had pain and numbness in both legs for 4 months. She also had noticed paresthesia in the third, fourth and fifth fingers of the left hand, the fourth and fifth fingers of the right hand, and the ulnar aspect of the right forearm. Two weeks before admission she had an episode of incontinence of urine.

Examination. General examination was normal with the exception of bilaterally enlarged lacrimal glands. The patient was unable to walk. The left leg was weaker than the right. There were bilateral ankle clonus and extensor plantar responses. Knee jerks and bicep jerks were present but diminished. There was atrophy of both the thenar and hypothenar eminences of the left hand. The left hand grip was weaker than that of the right. There was bilateral hypesthesia and hypalgesia of the C-7, C-8, and T-1 segments. Radiographs of the cervical spine indicated mild degenerative disease at C5–6 and possibly at C6–7. Chest films showed bilateral hilar adenopathy; the right medial base had a positive silhouette sign and obliteration of the right cardiac border. There was electromyographic evidence of lower motor neuron disease in the C8–T1 distribution on the left side.

The purified protein derivative (PPD)
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Fig. 1. Myelogram showing a block to contrast medium and enlargement of the spinal cord.

and Histoskin tests were normal. The cerebrospinal fluid (CSF) was clear and colorless with 58 red blood cells and 8 white blood cells; the opening pressure was 150 mm of CSF, and Queckenstedt's test was normal. A myelogram showed evidence of narrowing of the spinal canal at the C6-7 level but the cord was widened at the C5-6 level (Fig. 1).

Operation. A preoperative diagnosis of intramedullary tumor was made, and laminectomy at C4-C7 was performed. A bluish discoloration of the dura was noted. Upon opening the dura a gelatinous tumor was seen within the spinal cord. The tentative diagnosis of malignant ependymoma was made. As much of the tumor as possible was removed by sharp dissection under the microscope.

Histological Examination. The specimen was a granulomatous lesion with central areas of necrosis surrounded by round cell infiltration, epithelioid cells, and giant cells (Fig. 2).

Postoperative Course. Examination 12 hours postoperatively revealed anesthesia to pinprick below C-4. Deep tendon reflexes were increased in both arms and legs with a sustained clonus and extensor plantar response in both legs. On the fourth postoperative day there was diminished sensation to touch and pinprick below C-6 and total paralyses of both legs. The patient was started on extensive physical therapy, and when the final pathological report was sarcoidosis, she was treated with Prednisone.

At 9 months postoperative the patient was able to pull herself to a sitting position in bed and could propel a wheel chair equipped with a plastic rim. She stated that she had slight pain perception in her toes and was maintaining her weight well. She had an indwelling catheter.

Discussion

There seems to be a sharp distinction between the diagnostic characteristics of the remittent and progressive forms of sarcoidosis. James in 1968 suggested that the chronic progressive form usually shows a negative Kveim reaction (sarcoid antigen skin test). In the remittent form there is involvement usually of multiple systems, and the associated neurological signs may be a part of the syndrome. In the chronic progressive form, if it selectively involves the nervous system, biopsy almost always is necessary for diagnosis of the disease. The Kveim test was not performed in our case.

In 1905, Winkler first reported the manifestation of this disease in the nervous system. Among 20 cases of sarcoidosis at autopsy, Askanazy found three with central nervous system lesions. The clues to sarcoid-
Fro. 2. Left: Low-power photomicrograph of the biopsied tissue showing granulomatous lesions. H & E, ×100. Right: High-power photomicrograph of a granulomatous lesion showing giant cells, round cell infiltration, and a central area of necrosis. H & E, × 600.

osis usually were obtained from examination or biopsy of other systems. One of his cases had a laminectomy of the cervical and thoracic regions; in contradistinction to our case the spinal cord was found to be small and had diffuse degeneration. At autopsy the spinal cord in the upper one-half of the thoracic region was grayish, small, and soft, while the atrophied portion was diffusely infiltrated by granulomatous nodules.

The case reported by MoldoveP had widespread sarcoid involvement with signs suggestive of possible invasion of the spinal cord or meninges. No myelogram was performed. No biopsy was taken. The patient improved on steroid therapy. The symptoms, signs, and operative findings in this patient resembled the case reported by Jefferson.

Wiederholt and Siekert, in 1965, reported 907 patients with sarcoid involvement, 3% to 5% of whom had neurological findings. One patient had paraparesis from cord compression by sarcoidosis of the arachnoid membrane. Walker presented seven cases with central nervous system involvement but none of them had an intramedullary mass. Moreover, each of his patients had x-ray evidence of a large mediastinal mass and changes in the hands.

This disease may run a benign self-limiting course, have remissions and exacerbations, remain stationary for years, run a progressively declining course, or cause sudden death. Three major complications are: pulmonary fibrosis, fibrotic uveitis, and nephrocalcinosis.

The CSF usually shows moderate pleocytosis when the central nervous system is involved. It is not unusual to have a high CSF protein content of 200 to 500 mg%; the CSF sugar is usually normal. In our patient the CSF protein was 76 mg% and the sugar 58 mg%. Transient meningeal reactions manifested by CSF changes without neurological symptoms and signs may occur much more frequently when part of the gen-
eralized systemic reaction. James, et al., suggested that the Kveim test, right scalene node biopsy, and aspiration liver biopsy are helpful in diagnosis when there is a multisystem involvement with neurological deficits.

Indications for steroids have been discussed by James and others and include uveitis, disfiguring skin lesions, worsening of chest x-ray, disabling breathlessness, neurological involvement, hypercalcemia, and glandular involvement (parotid, lacrimal, spleen).

Our patient did not have hypercalcemia and there was no evidence of uveitis or renal disease.

Nelson stated that 75% of his patients with active sarcoid had a positive Kveim test, and that the test was negative in all nine cases in which sarcoid was clinically not active.

Summary

A case of intramedullary tumor caused by sarcoidosis has been discussed. This is the second case reported in which definite ante-mortem tissue diagnosis of intramedullary sarcoid of the spinal cord was made. Resorting to tests other than biopsy of central nervous system tissue probably would confirm the diagnosis in cases where there is a high suspicion on clinical grounds. Surgical intervention even with the aid of a microscope increases the morbidity. The incidence of intramedullary tumors in association with sarcoidosis is not definitely known.

References

16. Winkler M: Quoted by Colover, reference 2

Address reprint requests to: Timir Banerjee, M.D., Division of Neurological Surgery, The Ohio State University Hospitals, 410 West 10th Avenue, Columbus, Ohio 43210.