Sarcoidosis of the spinal cord

Case report

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A case of intramedullary sarcoidosis simulating a tumor of the cervical spinal cord is presented. Autopsy showed that the disease was limited to the cervical cord and hilar lymph nodes. The literature is reviewed and six cases of histologically documented spinal cord sarcoidosis are discussed.

KEY WORDS □ spinal cord sarcoidosis □ spinal cord mass

SARCOIDOSIS, a fairly common systemic granulomatous disease, is found most frequently in lymph nodes, lungs, liver, spleen, skin, eyes, phalangeal bones, and parotid glands, and rarely involves the nervous system. Siltzbach1 reported 311 cases of sarcoidosis of which 13 (4%) involved the nervous system. Although the leptomeninges and floor of the third ventricle are the most common sites when central nervous system involvement does occur,2 no part of the brain or spinal cord is inviolate. Relatively few cases of spinal cord involvement have been reported in the literature,1-4,6-8,10-12 and most of these patients had disseminated disease. We found only six cases with histological documentation,1,2,6-8,12 and only two with symptoms solely in the spinal cord.1,2

We are reporting another case of sarcoidosis which presented with spinal cord symptoms only. Autopsy showed the disease to be limited to the spinal cord and hilar lymph nodes.

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This 43-year-old woman was well until 1967 when she developed paresthesias of both legs and weakness in all extremities. She was subsequently hospitalized and found to have an intramedullary mass at the C4-5 level, two-thirds of which was surgically removed. Histological examination of the lesion showed a noncaseating, granulomatous inflammation of the spinal cord parenchyma (Fig. 1); special stains for acid-fast organisms, fungi, and bacteria were negative. The patient was treated with isoniazid, prednisone, and pyridoxine, with some initial improvement. However, over the next 2 years she developed flaccid weakness and wasting of the forearms, spasticity of the lower extremities with hyperactive deep tendon reflexes, and sensory disturbances such as varying degrees of paresthesia, decreased pain and touch sensation below the level of C-5, and loss of position sense in all extremities. Over
the next 4 years her weakness progressed slowly but bowel and bladder function remained intact.

**Course.** In April, 1973, she was admitted to our hospital for treatment of a large pustular lesion in the right groin and thigh. Her neurological status at that time was stable. Her sensorium and cranial nerves were intact. Motor examination revealed markedly decreased strength in the proximal arm muscles, absence of movement of the lower extremities, hyperactive knee reflexes, and bilateral Babinski reflexes. Sensation was absent below the T-12 level. The pustular lesion was diagnosed as necrotizing fasciitis and failed to respond to debridement and intensive antibiotic therapy. The patient's condition became septic; osteomyelitis developed in the right femoral head and septic arthritis in the right hip. The femoral head and proximal two-thirds of the femur were both removed. The patient remained febrile postoperatively, became progressively lethargic, hypotensive, and died from sepsis in August, 1973.

**Postmortem Examination.** The main pathological findings at autopsy were limited to the spinal cord and the hilar lymph nodes. The hilar lymph nodes were enlarged, firm, and gray-black. Histologically, the nodes were replaced throughout by a diffuse nodular fibrosis. Around the periphery, however, were granulomas containing giant cells and epithelioid histiocytes. Special stains for acid-fast organisms, fungi, and bacteria were negative. The leptomeninges in the lower cervical and upper thoracic segments, however, were fibrotic and adherent to the dura. In addition, the spinal cord showed enlargement in this area with a cross-sectional diameter of 2 cm (Fig. 2). On cut
section, the entire parenchyma appeared to be replaced by a homogeneous, firm, gray, gelatinous substance which extended from the lower segments of the cervical cord to the level of T-2. Grossly, the lesion resembled a glioma. Histological examination showed an area of inactive, fibrotic granulomas replacing the cord parenchyma (Fig. 3). The overlying leptomeninges and dura showed scattered mononuclear cell infiltrates. Sections from the thoracic, lumbar, and sacral area of the spinal cord and cerebellum, medulla, hypothalamus, and frontal lobe failed to show any evidence of active or healed granulomas in the subarachnoid space or parenchyma. Wallerian degeneration of the pyramidal tracts, however, was seen in sections from the lower thoracic and lumbar regions.

Discussion

Only rarely has involvement of the spinal cord by sarcoidosis been reported in the literature. Colover found some cases of sarcoidosis that suggested spinal cord lesions clinically. Critchley and Phillips observed a patient with sarcoid disease who developed signs of a low cervical cord lesion. Moldover reported a case of a 24-year-old man with widespread sarcoidosis and clinical evidence of spinal cord involvement. This patient was relieved of his symptoms by steroid therapy. None of these reports, however, included histological confirmation. Cases of spinal cord sarcoidosis verified by biopsy or autopsy are very rare. Longcope included in a table of distribution of lesions one case of cervical cord sarcoidosis found at autopsy, but no further description was given. Erickson, et al., reported a patient with multisystem disease whose cord was grossly normal but microscopically was found to contain parenchymal infiltration of giant cells and epithelioid histiocytes throughout the cervical, thoracic, and lumbar regions. Of the five cases reported by Askhanazy, three were limited to the central nervous system. One of these patients, a 52-year-old woman, presented with signs of a cord lesion at the level of T-1. At laminectomy the cord appeared small and a diagnosis of diffuse degeneration was made. She remained bedridden and died 6 years later. Autopsy showed an atrophic cord diffusely infiltrated by noncasing, acid-fast, negative granulomatous nodules. The leptomeninges were also involved but this was thought to be secondary to the cord lesion. There was no evidence of sarcoidosis in any other organ system. Jefferson mentioned a case in which myelography demonstrated a mid-cervical cord lesion. At surgery the lesion appeared to be a glioma, but a biopsy proved it to be a granulomatous lesion. The patient died 1 week later from respiratory arrest following a seizure. Autopsy showed a normal brain, but typical sarcoid lesions were found in the cervical cord, hilar lymph nodes, lungs, and liver. An interesting case of sarcoid arachnoiditis causing compression of the lower spinal cord with subsequent development of a cauda equina syndrome was reported by Wood and Bream. Lysis of the adhesions found at surgery and cortisone therapy led to recovery. Due to the normal appearance of the cord, it was not biopsied. However, examination of the spinal arachnoid showed chronic granulomatous inflammation. In 1972, Banerjee and Hunt reported a case which was initially diagnosed as an intramedullary malignant ependymoma until histology proved the lesion to be granulomatous inflammation.

Because of the similarities of the clinical course, radiographic evidence, and even gross
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appearance between sarcoidosis and tumors of the spinal cord, the distinction usually is not possible except by diagnostic biopsy. Since neural sarcoidosis, at least in a few reported cases, has shown a very good response to steroid treatment,\textsuperscript{6,9,10,12} it is important to establish the diagnosis and hence the treatment early in the course of the disease.

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References


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