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Subacute Myelopathy as the Presenting Manifestation of Sarcoidosis

By

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Summary

We studied a patient with progressive myelopathy, in whom systemic sarcoidosis was discovered. Involvement of other organs was asymptomatic. The myelopathy considerably improved after corticosteroid therapy. Spinal cord involvement in sarcoidosis is very uncommon, and its occurrence as a presenting manifestation is even rarer. Prompt recognition is emphasized, because it is a treatable condition.

The neurological manifestations of sarcoidosis have been estimated from 3.5 to 17% [8]. Among them spinal cord involvement is one of the rarest, and it is usually a late manifestation of the disease [8, 9, 17]. We studied a case in which spinal cord involvement was the presenting symptom, with a favourable evolution after corticosteroid therapy. The rarity of this presenting manifestation and the diagnostic difficulties incited us to report this case.

Case Report

A 31-year-old white man was in good health until he developed progressive dorsal pain 4 months before admission. By that time bilateral radicular T_8–T_9 pain, numbness of the lower limbs and increased urinary frequency and constipation had appeared. Neurological examination showed a decreased sensation to pinprick, light touch, temperature, position and vibration up to the level of T_7–T_8 bilaterally. The lower limbs were paretic with hyperactive tendon reflexes and bilateral Babinski signs. Superficial abdominal reflexes were absent. Walking was not possible. The disturbances predominated the right side. No evidence of polyneuropathy was present, and EMG studies were normal. The first lumbar puncture (LP) showed a clear CSF with an opening pressure of 32 cm H_2O and a normal Queckenstedt, 17 lympho-monocytes/mm^3 and a protein content of 620 mg/l. After 4 days a second LP showed 48.6 lympho-monocytes/

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mm^3 and a protein content of 335 mg/l. Glucose was not measured. Standard vertebral X-rays, a vertebral CT scan (T_4–T_9), 2 myelographies (metrizamide, Duroliopaque), spinal angiography (catheterization of intercostal arteries between T_4–T_9), and a brain CT scan were all normal. Because of the progressive disability without a clear diagnosis, a laminectomy at the level T_5–T_6 was performed (6 months after the initial symptoms) and showed an enlargement of the spinal cord without an extraspinal spinal tumour or arachnoid proliferation. An intramedullary glioma was suspected but no biopsy of the spinal cord was made. Further investigations were then performed: a liver needle biopsy showed sarcoid granulomas, as did biopsies of pulmonary hilar ganglia. The intradermal Kveim test was positive. Cultures for tuberculosis were negative. The ESR was 55 mm and blood anti-Kveim antibodies were positive with 1/5,120 dilution. Other standard examinations were normal. The diagnosis of sarcoidosis with spinal cord involvement was made and the patient was started on oral prednisone 100 mg/day. After 3 months walking was possible, with disappearance of sphincter disturbances and considerable improvement of pain. Neurological examination still showed decreased superficial and deep sensation below the level of T_7–T_8 and paresis of the lower limbs with hyperactive tendon reflexes, mainly on the right side. 13 months later (prednisone 40 mg/day, baclofen 30 mg/day) the improvement in gait and pain was still better, but without further changes in the neurological findings.

Discussion

Our patient showed a progressive dorsal myelopathy as the initial and isolated symptom of sarcoidosis. Involvement of other organs was present but remained asymptomatic. No biopsy of the spinal cord was made but progressive and persisting improvement after corticosteroid therapy is suggestive of sarcoidosis [5, 7, 8, 12].

Spinal cord sarcoidosis is one of the rarest neurological manifestations of the disease. We evaluated its clinical frequency around 0.43% in systemic sarcoidosis, according to the detailed published reviews (17/3,967 cases: review of the literature and personal series of Silverstein *et al.* [22], and studies of Wiederholt and Siekert [25], Schubert *et al.* [21], and Douglas and Maloney [10]). In a necropsy series the frequency may be higher, up to 20% [18]. As in our case the dorsal segments of the spinal cord are the most commonly involved [8]. The presenting symptoms are local pain and/or paraparesis, followed by incontinence, sensation disturbances, with progression towards irreversible paraplegia and frequently early death [9]. Generally spinal cord involvement appears during the course of known sarcoidosis [8, 9, 12, 25], but it may rarely occur as its presenting manifestation. We found only 10 such cases in the literature (Table 1). In some other cases spinal cord involvement was an early but not presenting manifestation of the disease [7, 13, 15, 16, 19, 26].

It appears that two main types of spinal cord sarcoidosis may be distinguishable. Firstly, a subacute myelopathy mimicking a glioma,

Table 1. *Sarcoidosis of the Spinal Cord as the Presenting Manifestation of the Disease*

	Age	Sex	Race	Clinical manifestation	Myelography	Level	Evolution
Azkanazy 1951	52	F	?	pm	—	T_1	death after 6 years
Gallenkamp and Suchenwirth 1971	29	M	W	pm	—	T_8	improvement after cst (FU = 6 months)
Banerjee and Hunt 1972	36	F	B	pm	block	C_4–C_7	improvement after cst (FU = 9 months)
Goebel 1975	22	F	W	tm	—	T_9	improvement after cst (FU = 2 years)
Buge *et al.* 1975	45	M	W	pm	normal	diffuse	improvement after cst (death after 3 years)
Nathan *et al* 1976 (case 1)	43	F	B	pm/tm	enlargement of spinal cord	lower C	death after a few days
Snyder *et al.* 1976	43	F	?	pm	—	C_4–C_5	progressive worsening (no cst) death after 6 years
Day and Sypert 1977 (case 2)	43	M	B	pm	partial block	C_5–C_6	improvement after cst, new worsening after 7 years
Bernstein and Rival 1978	31	M	B	pm	filling defect indicating intramedullary space-occupying lesion	T_1–T_8	?
Waxman and Sher 1979 (case 1)	26	M	B	pm	block	T_9–T_{12}	improvement after resection of spinal cord granulomas
Our case—1982	31	M	W	pm	normal	T_7–T_8	persisting improvement after cst (FU = 18 months)

cst = corticosteroid therapy; FU = follow-up; M = male; F = female; B = black; W = white; pm = progressive myelopathy; tm = transverse myelitis.

with progressive paresis, sensation and sphincter impairment, and pain, in which myelography may be normal (as in our case) or abnormal. Histological examinations have shown local or diffuse spinal infiltration by sarcoid granulomas [1, 2, 8]. Secondly, a more acute form, which is in fact a transverse myelitis, with sudden paraplegia and various sensation and sphincter disturbances.

Tetra/paraparesis may also be the presenting symptom without involvement of the spinal cord itself, but with spinal cord or cauda equina compression by arachnoid or intra-/extradural granulomas, or by vertebral collapse due to granulomas [3, 6, 11, 17, 25].

The CSF may show various modifications. Lympho-monocytic pleiocytosis is usual, up to 200–300 cells/mm^3 [7, 8], with moderately increased proteins and slightly decreased or normal glucose [7, 8].

As in spinal cord involvement, in known sarcoidosis the evolution of the presenting subacute myelopathy usually improves after corticosteroid therapy (see Table 1). The disease usually stops with partial recovery, but permanent sequelae almost always remain. In exceptional cases surgical repair may be worthwhile [24].

Our case shows that progressive myelopathy in otherwise healthy people may be due to sarcoidosis. In patients with unclear subacute myelopathy and an inflammatory CSF one should look for granulomas (liver, hilar ganglia) and perform an intradermal Kveim test. Detection of anti-Kveim antibodies in the blood and CSF may also be an important clue. Apart from spinal cord tumours, diagnosis should be made between other granulomatosis, parasitic cysts, leukemic infiltration, angioma and multiple sclerosis.

Sarcoidosis of the spinal cord as the presenting manifestation of the disease is a rare condition, and only 10 cases have been reported in the literature. However, an early diagnosis is very important, because a significant improvement may be achieved with adequate therapy.

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