Thoracic Intramedullary Sarcoidosis Mimicking an Intramedullary Tumor

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ABSTRACT

Sarcoidosis is a chronic, systemic granulomatous reticulosis of unknown origin, characterized by formation of hard tubercles and noncaseating granulomas1. Since other infectious diseases such as berylliosis, mycobacterium and fungal infections may present with a noncaseating granulomas, histological diagnosis of sarcoidosis is made using the elimination method. Central nervous system manifestations of sarcoidosis may be present in 5–10% of the cases2–5 involving cranial nerves, leptomeninges and third ventricle respectively. Any part of the central nervous system can be affected. Involvement of spinal cord in sarcoidosis is extremely rare and presents with only 0.3–0.4% in patients with systemic sarcoidosis2. Intramedullary sarcoidosis is a rare first manifestation of the disease and it can mimic an intramedullary tumor, which is often manifested with symptoms that initiate from spinal cord compression, resulting in paraparesis, sensory disorders and sphincter dysfunction6–11. We present a case of intramedullary sarcoidosis that mimics a tumor of the thoracic spinal cord. Clinical features, neuroradiological, pathohistological findings, laboratory analysis and surgical treatment of such a rare entity are being discussed.

Key words: intramedullary tumor, sarcoidosis, thoracic spine

Introduction

Sarcoidosis is a systemic disease of unknown origin characterized by formation of noncaseating granulomas1. Intramedullary sarcoidosis is a very rare entity that presents with the symptomatology similar to the intramedullary spinal tumors10. Clinical presentation of medulla spinalis compression by a granulomatous lesion is seldom the first manifestation of systemic sarcoidosis which is shown to be true in our patient. True diagnosis in these cases is therefore extremely difficult and it is not unusual to mistake it for tuberculosis, other granulomatous infectious diseases, intramedullary tumors or paraneoplastic myelopathy5,12. In this particular case it also took us almost a year before we postulated the right diagnosis and only after a thorough physical, diagnostic and laboratory assessment. MR imaging can help in establishing a diagnosis, but in our case it did not indicate the presence of an infective process.

Case Report

A 40-year-old male patient was referred, for the first time, to our Department of Neurosurgery with the progressive muscle weakness in lower limbs bilaterally accompanied by parestesias in umbilical region and loss of sensation of temperature and touch from Th10 downward. The symptoms started 8 months earlier. The patient was unable to walk without support and he developed bladder incontinence when he was admitted to our Department. Magnetic resonance imaging of the thoracic spine using a 1.5 Tesla superconductive imager revealed thickened portion of the thoracic spinal cord from Th8 to Th12 level with inhomogenously increased signal and dense inhomogenous enhancement after Gadolinium application both on T1 and T2-weighted images (Figure 1). Brain and cervical spine MRI were normal. Reviewing the medical documentation we have found that the patient referred to the general practitioner with enlarged
submandibular lymph nodes on the right side and the presentation of viral infection resembling the flu 4 years ago. Since there was no regression in the lymph nodes size for more than five months, the lymphatic nodule was surgically extirpated and pathohistological findings did not find any pathological changes. The lumbar puncture performed earlier by the neurologist and the routine analysis of the cerebrospinal fluid revealed no pathological findings.

Taken the clinical presentation and the MR findings in the consideration we have decided to treat the patient operatively. After the preoperative preparation the laminectomy of Th9–Th12 was done with the partial laminectomy of Th8 vertebrae. After opening the dura we have microneurosurgically performed myelotomy through the posterior medullar sulcus using CO2 laser. Infiltrative, softly dark grayish tumor-like tissue was revealed. The sample of this tissue was intraoperatively sent to pathohistologic analysis which revealed a few small granulomas, devoid of central necrosis consisting of centrally placed epitheloid cells, scattered Langhans giant cells and rim of lymphocytes (Figure 2).

No further reduction of this tumor-like tissue was done and the procedure was finished by the dural plasty using the Liodura in order to allow decompression. The postoperative recovery was uneventful and the patient was able to walk on his own again. In a three months period the patient developed enlarged lymph nodes on the neck which were percutaneously punctuated. All of them were positive on granulomatous infection.

Due to all said above and the development of nonspecific symptoms such as fatigue and the weight loss a patient was referred to and treated by a specialists for internal and infectious diseases.

The following assessment was completed:

- Mantoux test was negative. LE cells, AST, ASTA, latex, anti DNA, antinuclear factor (ANF), serology for fungal infections, Toxoplasmosis, Leishmaniosis, adenovirus, Epstein-Barr virus (EBV), Cytomegalovirus (CMV), Coxiella burnetti, Chlamydia pneumoniae, Mycoplasma pneumoniae, Parvovirus were all negative. C reactive protein (CRP) was 21, creatine phosphokinase (CPK) and lactate dehydrogenase (LDH) also were in the normal range. Immunohistochemical analysis were performed on CD 34, CD 20, CD3 which revealed granulomatous angiitis with the predominance of B cell population of lymphocytes, while T lymphocytes were present in lesser degree in the inflammatory infiltrate. Hypergammaglobulinemia was evident with IgG predomination both in

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**Fig 1.** Magnetic resonance (MR) imaging of a 40-year old man with thoracic spinal cord sarcoidosis, eight months after first onset of neurological symptoms. A) T1-weighted image in the sagittal plane shows thickened portion of the thoracic spinal cord (white arrows). B) T2-weighted image in the sagittal plane reveals inhomogenously increased signal in the thickened portion of the cord (white arrow), along with cord edema extending proximally (white arrowhead). C) T1-weighted image in the sagittal plane after application of gadolinium demonstrates dense inhomogenous enhancement, corresponding to the areas of increased signal intensity on T2-weighted images (white arrows). D) Gradient-echo T2-weighted image in the axial plane demonstrates thickened spinal cord, with abnormal high signal intensity affecting most of the cord.

**Fig 2.** Noncaseating granulomatous lesion (H&E, x100).

**Fig 3.** MR imaging of the same patient nine months later, after a multi-level laminectomy (black arrows) and spinal cord surgery. A) T2-weighted image in the sagittal plane shows reduced area of abnormal signal (white arrow) with less prominent spinal cord thickening in comparison with Fig. 1B. Spinal cord edema is no longer present (white arrowhead). B) T1-weighted image in the sagittal plane after application of gadolinium also shows reduction in size of the abnormality, with a number of smaller enhancing lesions. C) Fast spin-echoT2-weighted image in the axial plane at a similar level as in Fig. 1D shows definite regression, with minimal residual increased signal within the cord, and almost complete resolution of cord thickening.
the blood and in the CSF. CD34 reveals regular endothelium. All of the above is supporting the findings for non-infectious granulomatous.

The cerebrospinal fluid analysis revealed moderate lymphocytic pleocytosis (60 cells per mm$^3$: 84% mononuclear), slightly elevated protein, normal glucose ratio (CSF:plasma), normal levels of chlorides and hypercalcemia. Both serum and CSF levels of angitensin-converting enzyme (ACE) were significantly elevated. The CSF T4:T8 ratio, measured by flow cytometry, was elevated which also showed to be true in the work of Hawley and al.$^{13}$ and therefore supported the diagnosis of neurosarcoidosis.

No organisms including Koch’s Bacillus in cerebrospinal fluid were recovered on culture or gram stains.

Afterwards the gallium scintigraphy was done which showed gallium accumulation in lymph nodes in the neck, mediastinum and in the region of L1 and L2 vertebral. The trans-bronchial lung biopsy was performed together with submandibular lymph node biopsy and all the findings indicated a granulomatous inflammation.

The medical consilium including infectologist, pulmonologist, neurologist, neurosurgeon and the pathologist, using the elimination method, concluded that this medical condition could only be sarcoidosis so the patient was treated with postoperative immunosuppressive corticosteroid therapy (Medrol) for 10 weeks. A rapid clinical improvement of the patient together with the regression of enlarged lymph nodes, verified by MR scans was observed.

During the future treatment we have also observed the development of skin efflorescence but due to the previous contrast application required for MR scanning it was not possible to truly distinguish if it was allergic reaction or one more bizarre representation of systemic sarcoidosis.

**Conclusion**

Intraspinal sarcoidosis may mimic other neurologic medical entities, primarily intramedullary tumors$^{6-11}$. When suspecting on a granulomatous inflammatory process, radical surgery should be avoided since the tendency of spontaneous remission rate, after surgical reduction and prolonged use of corticosteroids in majority of patients lead to disease regression and even to full patient recovery.

**REFERENCES**


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**TORAKALNA INTRAMEDULARNA SARKOIDOZA KOJA IMITIRA INTRAMEDULARNI TUMOR**

**SAŽETAK**

Sarkoidoza je kronična, sistemska granulomatozna bolestitina, koja može se prezentirati s nekrozom i nekakve infekcije. Histolоška dijagnoza sarkoidoze postavlja se koja se radi jezičnog sustava izazivajuće u 5–10% slučajeva. Sarkoza je prezentirani simptomi srednjeg živčanog sustava u 5–10% slučajeva. Klinička simptoma u sarkozi je izrazito rijetko, i javlja se u samo 0.3–0.4% slučajeva kod bolesnika sa sistemskom sarokoidozom. Intramedularna sarkoidoza je rijetko prva manifestacija sistemskih bolesti, a tada kliničkom slikom može sličiti na intramedularni tumor koji se manifestira simptomima kompresije kralježnike moždine na sarkoidozi. U radu smo iznijeli slučaj intramedularne sarkoidoze koja je imitirala tumoralne kralježnike moždine. Prikazani su klinički simptomi, neuroradiološka dijagnostika, laboratorijski nalazi, patohistološki nalazi te kirurška procedura kod ove rješetke bolesti.