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Intradural, extramedullary spinal sarcoidosis: report of a rare case and review of the literature

Bernhard Schaller, MD[a,*], Thomas Kruschat, MA[a], Holger Schmidt, MD[b], Wolfgang Brück, MD[c], Michael Buchfelder, MD[a], Hans C. Ludwig, MD[a]

[a]*Department of Neurosurgery, University of Göttingen, Robert Koch Strasse 40, Göttingen 37075, Germany*
[b]*Department of Neurology, University of Göttingen, Robert Koch Strasse 40, Göttingen 37075, Germany*
[c]*Department of Neuropathology, University of Göttingen, Robert Koch Strasse 40, Göttingen 37075, Germany*

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Abstract

BACKGROUND: Spinal sarcoidosis represents a rare subgroup of neurosarcoidosis. Most spinal sarcoid lesions are intramedullary, and only eight cases of intradural, extramedullary sarcoid lesions have been reported hitherto. We describe the complete entity of intradural, extramedullary spinal sarcoidosis.

METHODS: A 39-year-old woman presented with a 2-year history of slowly progressive numbness and paresthesia of the right medial upper arm followed by brachialgia and cephalgia. Magnetic resonance imaging revealed an intradural extramedullary T1 isointense and T2 hyperintense mass lesion near the right C3 nerve root exit C3, extending into the right neuroforamen. The cervical cord was not severely compressed. The mass lesion showed a high T2 signal. After intravenous administration of gadolinium–diethylenetriaminepentaacetic acid, there was marked enhancement of the process.

RESULTS: The patient underwent a C2 to C3 laminoplasty and total resection of the intradural extramedullary tumor. The lesion encroached along the right neuroforamen, involving the C3 nerve root, and was grossly adherent to some of the rootlets. Histopathological examination of the specimen showed a noncaseating granulomatosis consistent with sarcoidosis. Postoperative testing did not reveal systemic involvement of sarcoidosis but bilateral hilar and mediastinal lymphadenopathy with normal lung parenchyma. The patient was therefore treated with corticosteroids. During the follow-up period of 16 months, the patient made a satisfactory recovery, though with persistent, distally pronounced sensory disturbance in the C3 or C4 root areas, and returned to work full-time.

CONCLUSION: An extramedullary sarcoid lesion is rare. Unlike intramedullary sarcoid lesions, it can be totally removed. Even if systemic sarcoidosis is present, the patient can have an excellent recovery. From a review of the literature, we can hypothesize that intradural extramedullary spinal sarcoidosis may represent a very early stage of spinal sarcoidosis progressing to intramedullary intradural spinal sarcoidosis. The intradural extramedullary spinal sarcoidosis can be subdivided into a peripheral and a central subtype. Positive histological examination is required to establish the diagnosis since other diagnostic tests are unspecific.

Keywords: Sarcoidosis; Extramedullary; Intradural; Spinal tumor

Introduction

Sarcoidosis represents a systemic disease of unknown origin characterized by the histopathological finding of noncaseating giant-cell granulomata disrupting the architecture and function of the surrounding tissue [1]. Several large case series have found nervous system involvement of sarcoidosis to be present in up to 5% of all cases [1]. Autopsies series suggest frequent subclinical involvement

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* Corresponding author. Department of Neurosurgery, University of Göttingen, Robert Koch Strasse 40, Göttingen 37075, Germany. Tel.: 0049-551-39-60-39.

E-mail address: neuroscience_imaging_journal@yahoo.de (B. Schaller)

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of sarcoidosis [2]. Spinal sarcoidosis was first described by Longcope in 1941 [3], as an autopsy finding in a patient with cervical cord involvement. Since then, spinal syndromes are reported at clinical presentation in 6% to 8% of patients with neurosarcoidosis [4]. However, intradural, extramedullary sarcoid masses represent a very rare picture of neurosarcoidosis [4–11], so that only 8 cases have been reported in the literature to date.

We present here the complete entity of an intradural, extramedullary spinal sarcoidosis with its common characteristics.

Case report

History

A 39-year-old woman presented with a 2-year history of slowly progressive numbness and paresthesia of the right, medial upper arm followed by brachialgia and cephalgia corresponding to the C3 and C5 root distributions dermatomes on the right side. Her general medical history was uneventful.

Examination

Neurological examination revealed a pronounced sensory disturbance of C3 to C5 on the right side. Her score for cervical myelopathy, according to the criteria proposed by the Japanese Orthopedic Association, was 15 of 17 points (motor: 8/8, sensory: 4/6, bladder and bowel dysfunction: 3/3). Laboratory investigations disclosed normal values. Subsequently, an autoantibody screen (including antinuclear antibodies and antineutrophil cytoplasmic antibodies) as well as syphilis and *Brucella* serology proved to be normal. The cerebrospinal fluid (CSF) analysis revealed an elevated protein level of 1,249 mg/L (normal 180–480 mg/L), lymphocytosis of 29×10^6/L ($<3\times10^6$/L), and normal serum angiotensin-converting enzyme (ACE). Early morning urine and CSF samples were negative for acid-fast bacilli, and a Mantoux test was also negative. Nerve conduction studies showed an involvement of the sensory nerves C3 to C5 at peripheral level. Cervical X-ray found no abnormalities. Magnetic resonance imaging (MRI) studies of the spine revealed an intradural extramedullary T1 isointense and T2 hyperintense mass near the right C3 nerve root exit C3, extending into the right neuroforamen (Fig. 1A–C). The cervical cord was not severely compressed and had a high T2 signal. After intravenous administration of gadolinium–diethylenetriaminepentaacetic acid there was marked gadolinium enhancement of the mass. Computed tomography of the chest disclosed bilateral hilar and mediastinal lymphadenopathy with normal lung parenchyma. Biopsy findings of a hilus lymph node were consistent with sarcoidosis. Dexamethasone 4 mg once a day was commenced, and consent for surgical therapy was obtained.

Operation

The patient underwent an open-door laminotomy C2 to C3 that was stemmed caudally. A firm, red-brown intradural but extramedullary lesion was found adherent to the dura from just below C3. The dentate ligament covered the tumor and was therefore severed. Beneath the dura, the mass was twined with the right C3 anterior and posterior nerve rootlets as well as the right C4 posterior nerve rootlets after the dura was opened radially (Fig. 2). Therefore the rootlets were cut at the origin of the cord and the mass was completely removed. The dura was closed and the laminotomy reinserted.

Postoperative course

The postoperative course was uneventful. During a follow-up period of 16 months, the patient returned to full-time work. Neurological examination showed distally pronounced sensory disturbance in the right C3 and C4 dermatomas. Repeated MRI studies of the cervical spine and brain disclosed complete resolution of the lesions (Fig. 1D), and cerebrospinal fluid analysis showed a normalization of cell content (3×10^6/L) and a decrease of the protein level to 372 mg/L. Nerve conduction studies showed no significant improvement compared with baseline investigations.

After excluding a tuberculosis infection, the patient was treated with oral prednisone 50 mg daily, leading to about 50% regression of the sensory symptoms within the first 3 months. As the patient planned to become pregnant, prednisone was gradually tapered off 12 months after the operation.

Histology

Histopathologic examination of the operative specimen revealed an irregular portion of rubbery, tan-colored tissue measuring $1.0\times0.5\times0.5$ cm. Granulomatous inflammation surrounded the nerve, with associated perineuronal fibrosis. The giant cells contained haphazard arrangement of multiple nuclei. Special stains for acid-fast bacilli were negative. The histologic appearance was regarded as entirely consistent with scattered noncaseating granulomatous inflammation mainly consisting of epitheloid histiocytes and lymphocytes (Fig. 3). Plasma cells as well as T cells were identified, and a predominance of CD3-positive T cells was demonstrated.

Literature review: methodology of literature search

For the purpose of this literature review, Medline and Embase databases were searched for trials, case series, or case reports published between 1966 and December 2004. The key-word terms used for search were: sarcoidosis, extramedullary, intramedullar, intradural, extradural, and spinal tumor. The study or report had to be published or