



Neurosarcoidosis involving cervical nerve root with unusual MRI findings: a case report and systematic literature review

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Abstract

Background Neurosarcoidosis is rare, and among its manifestations, nerve root involvement has been reported in only a few cases. Therefore, magnetic resonance imaging (MRI) findings of neurosarcoidosis, particularly those involving nerve roots, are scarce in the literature.

Methods We presented the case of neurosarcoidosis involving cervical nerve roots and cranial nerves, alongside a systematic literature review.

Results A 28-year-old female suddenly developed right facial numbness as well as left upper extremity and left hand pain. Initial brain and spine MRI showed a bulging mass of T2 iso-to-high signal intensity in the left Meckel's cave/trigeminal nerve, as well as diffuse enlargement of the right C6 and C7 nerve roots. Follow-up MRI at 2 months revealed a reduction in the size of the initial lesion and the appearance of new similar lesions on the contralateral side (right Meckel's cave, left C3–C8 nerve roots). In particular, the lesions involving the nerve roots demonstrated central enlargement along the nerve roots, without involvement of the adjacent spinal cord. All these lesions exhibited enhancement, leading to the differentiation between sarcoidosis and lymphoma. Sarcoidosis was subsequently confirmed through biopsy of a hilar lymph node.

Conclusions This report presents a distinctive MRI feature of neurosarcoidosis involving spinal nerve roots, representing the first of its kind, and describes the evolution of MRI findings throughout the clinical course.

Keywords Sarcoidosis · Neurosarcoidosis · Spinal nerve root · Cranial nerve · MRI

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Introduction

Sarcoidosis is a rare chronic inflammatory disease that involves multiple organ systems, and approximately 5–15% of sarcoidosis patients show central nervous system (CNS) involvement [1–4]. In particular, spinal neurosarcoidosis accounts for less than 1% of all sarcoidosis, and within neurosarcoidosis, it is below 10% [1, 5], and only a few cases of spinal nerve root involvement have been reported [4–7]. Neurological symptoms manifest in approximately 50–70% of cases of neurosarcoidosis [8–10]. Patients with neurosarcoidosis involving the spinal nerve root show radiculopathy resulting from the direct compression of the nerve roots or ischemic changes due to granuloma and inflammation [11, 12].

Biopsies are often too invasive and difficult to perform for a confirmative diagnosis of sarcoidosis with CNS involvement [12]. Neurosarcoidosis is therefore usually diagnosed based on evidence of CNS inflammation on magnetic resonance imaging (MRI) or cerebrospinal fluid (CSF)

examination and histologic confirmation at other sites [10]. On MRI, common findings of neurosarcoidosis exhibit characteristic infiltrative linear and nodular enhancement along the meninges or leptomeninges [3, 8].

Here we present an exceptionally rare case of neurosarcoidosis involving the cervical spinal nerve roots with unusual MRI findings, and provide a systematic literature review.

Case presentation

A 28-year-old woman suddenly developed an olfactory disorder 3 months before presenting at our institution. The patient had undergone brain MRI (Fig. 1a) and cervical (C-) spine MRI at a different hospital (Fig. 2a, b). A mass of T2 iso-to-high signal intensity (SI) with a bulging appearance was observed in the left Meckel's cave, recognized by the obliteration of the CSF signal. Right C6 and C7 nerve roots revealed diffuse enlargement with T2 high SI compared to the normal left. The patient was prescribed corticosteroids, and her symptoms improved.

However, two months later, the patient suddenly developed numbness in the right face half and reported a tingling sensation and pain in the left upper extremity and the left hand, especially in the second and third fingers. She visited our hospital's neurology outpatient clinic, and physical examination revealed decreased sensation to light touch in the right forehead, cheek, and chin, without evidence of infection or neoplasm. On the same day, the patient underwent contrast-enhanced brain MRI. Compared with the initial brain MRI, we observed a decrease in the size of the soft tissue mass involving the left Meckel's cave, and newly appearing similar lesions with uniform enhancement in the right Meckel's cave. We also observed thickening and enhancement of the cistern segment of the right trigeminal nerve (CN V) (Fig. 1b, c). After 4 days, the patient underwent C-spine MRI, contrast-enhanced chest CT, and abdominopelvic CT. Compared with the initial spine MRI two months earlier, the hypertrophy involving the right C6 and C7 nerve roots had decreased, but other nerve roots were newly enlarged, at the left C3, C4, C6–8, and right T1 nerve (Fig. 2c–f). The lesions involving CNs and multiple cervical nerve roots show a diffuse central enlargement, with homogeneous enhancement. However, there was no involvement of the adjacent spinal cord by the lesions. Given the response to steroid therapy and recurrent nature, sarcoidosis was strongly suspected. However, based solely on the MRI findings, differentiation between sarcoidosis, lymphoma, or neurofibroma was challenging. Chest CT showed multiple enlarged and conglomerate lymphadenopathies in the right supraclavicular area, mediastinum, and both hilar and interlobar areas, which appeared to be sarcoidosis (Fig. 3a, b). Subsequent whole-body PET-CT and brain scans using

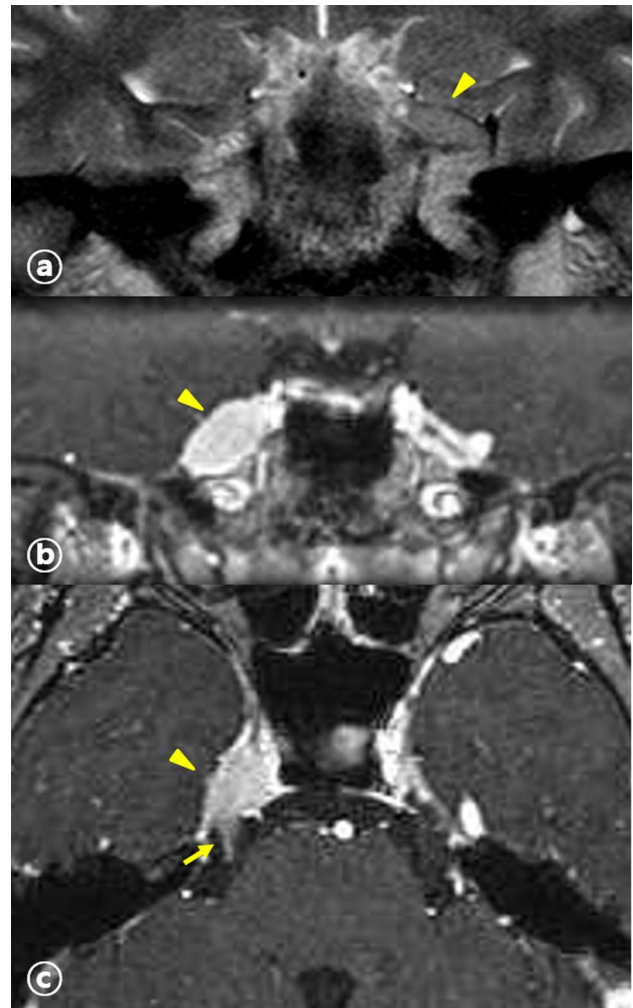


Fig. 1 Initial brain MRI with coronal T2-WI (a) and follow-up MRI with contrast-enhanced coronal (b) and axial (c) T1-WI. A T2 iso-to-high SI mass (*arrowhead*) with a bulging appearance can be seen in the left Meckel's cave, as indicated by the obliteration of the CSF signal (compare to the normal Meckel's cave findings on the right). Two months later, an interval decreases in the size of the soft tissue mass involving the left Meckel's cave was observed, as well as newly appeared lesions with enhancement in the right Meckel's cave (*arrowhead*) and thickening and enhancement along the cistern segment of the right trigeminal nerve (*arrow*). (WI, weighted image; SI, signal intensity; CSF, cerebrospinal fluid)

fludeoxyglucose injections revealed hypermetabolic lesions in the right Meckel's cave and neural foramen, suggesting cranial and peripheral nerve root involvement. Hypermetabolic lymphadenopathy was also observed (Fig. 3c–e). Considering the pulmonary involvement and neurological complications associated with CN involvement, we suspected sarcoidosis or lymphoma.

The patient was diagnosed with sarcoidosis based on thoracic lymph node biopsy, which revealed the formation of non-caseating small granulomas within lymph node structure (Fig. 4). She was prescribed corticosteroid, and

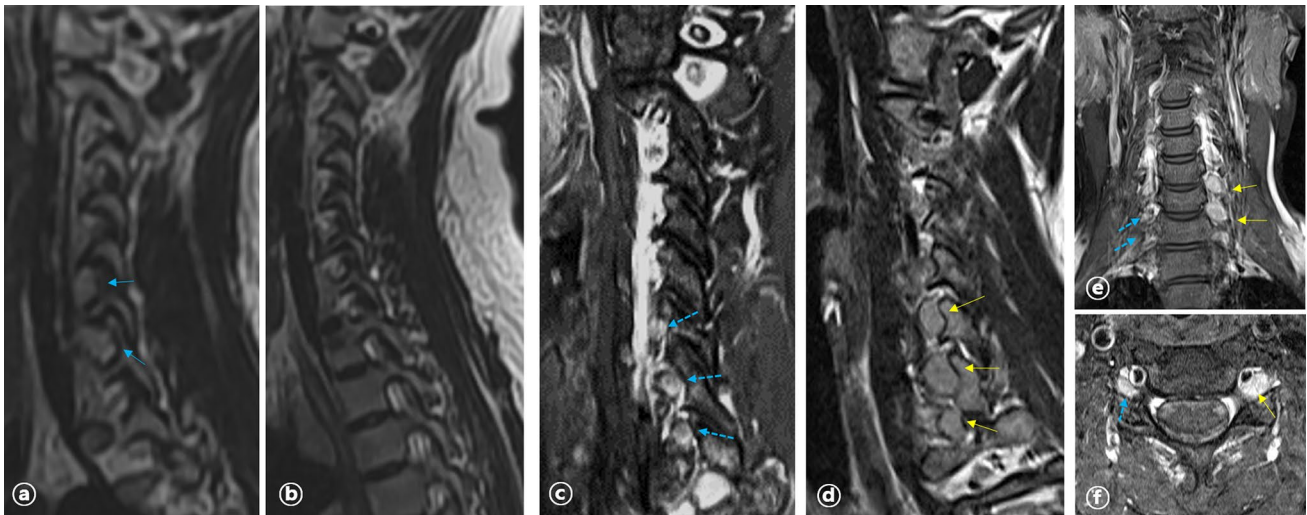


Fig. 2 Initial (a, b) and follow-up (c–f) cervical spine MRI. On sagittal T2-WI, the right C6 and C7 nerve roots exhibited diffuse enlargement in the neural foraminal area, as indicated by homogeneous T2 high SI (arrows) (a) compared to the left (b). Two months later, the nerve root enlargement had partially improved (dotted arrows) on

right sagittal T2-WI (c), left sagittal (d), coronal (e), and axial scans (f) of contrast-enhanced T1-WI. However, other nerve roots revealed new diffuse and central enlargement, involving the left C3, C4 (not shown), and C6–8 nerve roots (arrows). (WI, weighted image; SI, signal intensity)

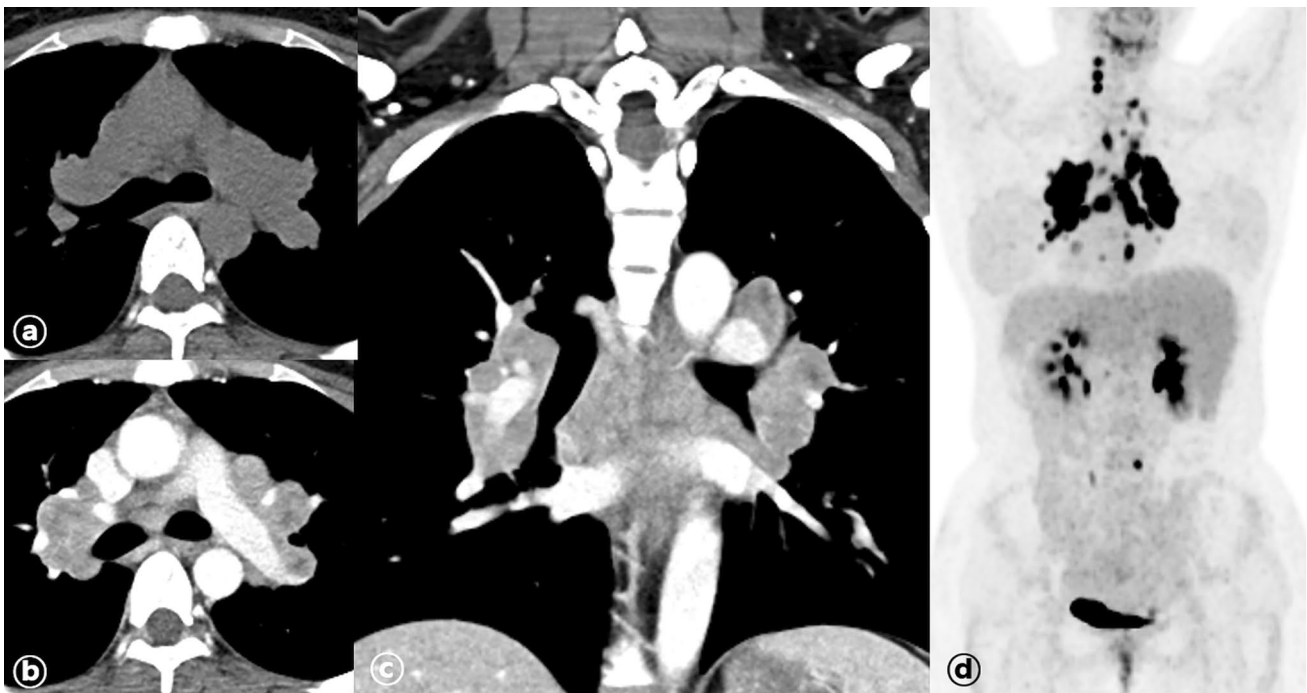


Fig. 3 Chest CT of the non-enhanced axial plane (a) and the contrast-enhanced axial (b) and coronal planes (c). Multiple enlarged and conglomerate lymphadenopathies were observed in the right supra-

clavicular, right paraesophageal, mediastinum, and both hilar and interlobar area and suspected to be sarcoidosis. Whole-body PET-CT (d) showed multifocal hypermetabolic lesions

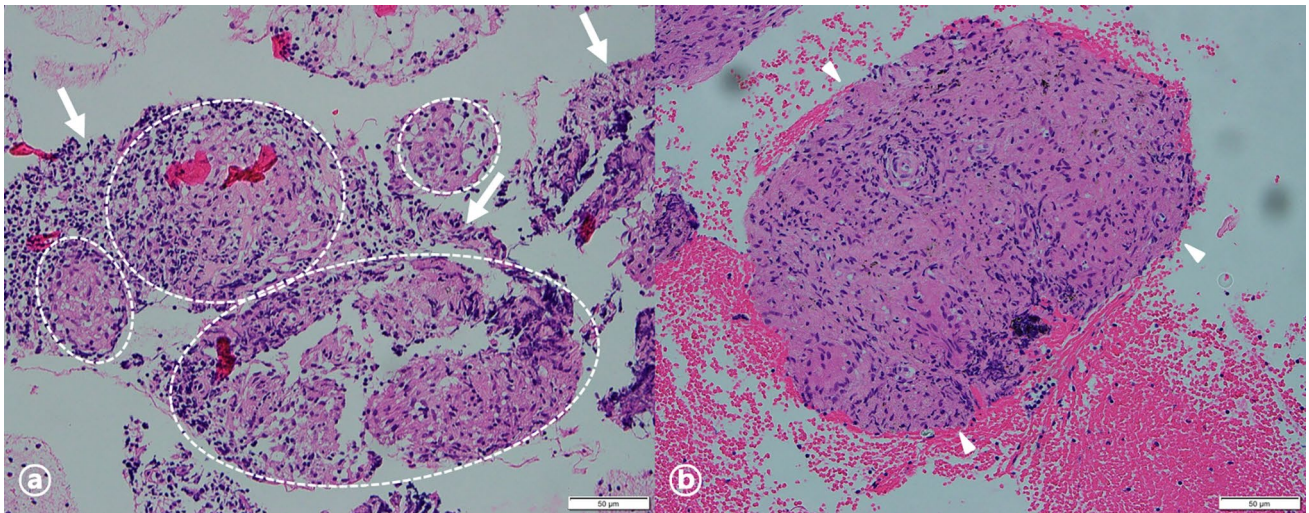


Fig. 4 Photomicrographs (H&E $\times 20$) obtained from the mediastinal and hilar lymph node biopsy depict **a** the formation of non-caseating small granulomas (*dashed circles*) within the lymph node structure (*arrows*), and **b** a clearly visible granuloma (*arrowheads*)

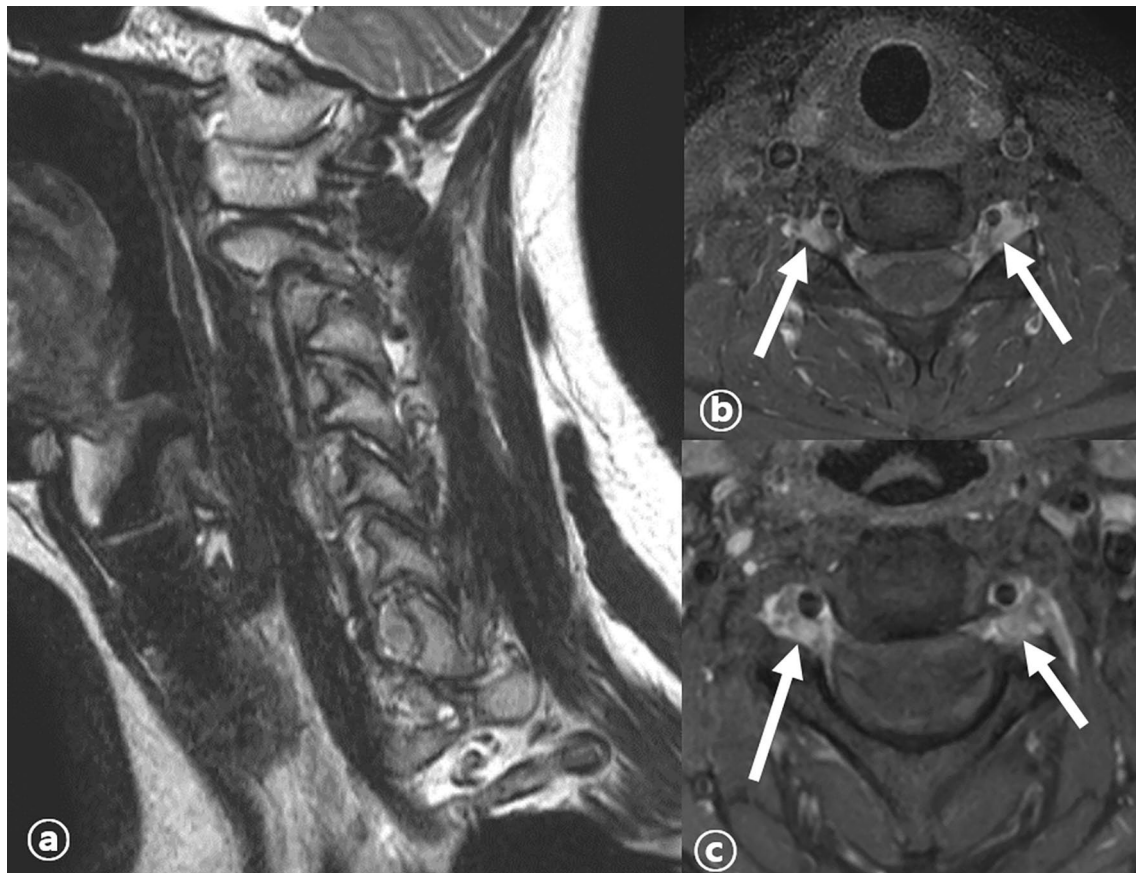


Fig. 5 Cervical spine MRI: left oblique sagittal T2-weighted image (**a**) and axial contrast-enhanced T1-weighted image (**b**, **c**). The previously enlarged soft tissue lesions involving mainly the left-side nerve roots were decreased in size (*arrows*)

follow-up spinal MRI showed that the previously enlarged soft tissue lesions involving mainly the left-side nerve roots had decreased in size (Fig. 5).

Systematic review

We utilized the PubMed search system following PRISMA guidelines to identify articles published in English with full text available after 1990. The inclusion criteria involved studies encompassing cases of neurosarcoidosis in adult patients that involved the infiltration of nerve roots and underwent spinal MRI. Exclusion criteria comprised studies that did not contain at least one case including demographic information, those with ambiguous diagnoses due to other concurrent medical conditions, those that mentioned sarcoidosis for differential diagnosis, and studies that did not conduct MRI.

The search terms used were 'neurosarcoidosis' or 'sarcoidosis' in the title or abstract, combined with 'nerve root' or 'spinal root.' Using the PubMed search engine with the query ("sarcoidosis"[Title/Abstract] OR "neurosarcoidosis"[Title/Abstract]) AND ("nerve root"[Title/Abstract] OR "spinal root"[Title/Abstract]), two radiologists independently conducted searches. The collected data from each researcher were then cross-verified for agreement, resulting in the identification of a total of 16 articles. Articles excluded per the criteria included one review article without demographic information, one study associated with lung cancer, three studies used for differential diagnosis, two studies without MRI, and additionally a case where sarcoidosis only involved the cauda equina, resulting in a final selection of 9 studies encompassing 18 cases (Table 1) [4–7, 9, 11, 13–15].

The mean age was approximately 41.78 years (range: 22 to 63 years, SD 13.0). There were 8 male patients (44.4%) and 10 female patients (55.6%). The distribution of invaded nerve root levels showed 12 cases at the cervical level (66.7%), 9 cases at the thoracic level (50.0%), and 11 cases at the lumbar level (61.1%), indicating a relatively balanced distribution with the cervical level being the most prevalent. In 13 cases (72.2%), spinal cord invasion was concurrent. Direct pathological confirmation through surgery or nerve root biopsy was conducted in 6 cases (33.3%). Out of the 18 cases, 9 did not show MRI findings or presented normal observations. Then, among the 9 cases with revealed MRI findings, the majority (7 cases, 77.8%) exhibited meningeal enhancement, peripheral enhancement around the cord, and concomitant nerve root enhancement. These enhancements appeared as multifocal linear or nodular enhancements along the meninges. All cases showing these features were concomitant with cord invasion (Chi-square analysis, $p = 0.047$). The remaining two cases (22.2%) depicted an enhancing mass.

Involvement in other neurological regions was observed in 10 cases (55.6%), with 16 cases (88.9%) showing improvement upon follow-up, while two cases (11.1%) showed no significant relapse or changes.

Discussion

We described a case of neurosarcoidosis that involved the extremely rare spinal nerve root and the relatively uncommon trigeminal nerve that responded well to treatment, as confirmed on serial follow-up C-spine and brain MRI. The most widely accepted diagnostic criteria for neurosarcoidosis are those proposed by Zajicek et al. [1, 8, 10]: Definite neurosarcoidosis is defined as the presence of direct neural tissue; probable neurosarcoidosis means that neurologic inflammation is present on MRI or CSF with evidence of systemic sarcoidosis; and possible neurosarcoidosis manifests as a typical clinical presentation; no other criteria were met definite diagnosis of neurosarcoidosis in the current case but other potential etiologies were excluded. As the probable diagnosis of neurosarcoidosis is based on imaging findings, an exact characterization of the MRI findings of neurosarcoidosis is needed.

This case represents neurosarcoidosis involving the cervical nerve roots, demonstrating MRI findings not previously reported. In most studies, MRI findings of neurosarcoidosis affecting nerve roots, the predominant observations were multiple linear or nodular enhancements along the meninges, and the dorsal root showed enhancement frequently. All these cases presented intramedullary or meningeal lesions in the adjacent spinal cord [3–7, 13, 14]. However, our case stands out as there was no adjacent spinal cord lesion. The nerve roots enlarge significantly as they pass through the neural foramen, displaying homogeneous contrast enhancement, which is an exceptionally distinctive finding. The pattern of the lesions seemed to be centralized within the nerve root, differing from the expected peripheral location along the meninges, a feature not previously reported in isolation. In contrast, the MRI findings associated with CN involvement have been reported relatively distinctive characteristics. This involvement was marked by diffuse nerve enlargement and enhancement [3, 16]. In our case, the CN involvement showed similar characteristics. The imaging pattern of nerve root involvement in our case seemed remarkably akin to the imaging observations associated with CN involvement. While most cases of neurosarcoidosis exhibit granulomatous inflammation pathologically infiltrating the leptomeninges, it can also infiltrate the parenchymal brain or the intramedullary portion of the spinal cord [8, 16]. The imaging findings in our case suggest that sarcoidosis infiltrated the substance of the nerve root, resulting in

Table 1 A systematic literature review of nerve root involvement on MRI

No	Authors	Year	Age	Sex	Nerve root level	Spinal cord lesion	Diagnostic confirmation	Nerve root MRI finding	Other neural lesions	Follow-up
1	Christoforidis [13]	1999	39	M	L	(-)	Biopsy (lymph node)	CE (not shown)	Optic nerve, brain	Improvement on nerve root
			34	M	L	(-)	Biopsy (not definite site)	CE (not shown)	Optic chiasm, brain	Resolution on nerve root
			23	M	L	(-)	Biopsy (not definite site)	T2 high SI; contrast NA (not shown)	Optic chiasm, brain	No change on nerve root
2	Koffman [14]	1999	30	M	T10-L2	C, T, L	Biopsy (L2 nerve root)	Dorsal nerve root CE, infiltrative CE	Brain	Improvement
3	Bode [6]	2001	49	F	Thoracolumbar, cauda equina	C, T, L	Biopsy (T12/L1 nerve root)	Meningeal CE, multifocal nodular CE, left T12/L1 root CE mass	Optic nerve, brain stem	Persistent visual loss, no visible new symptom
4	Moore [4]	2001	58	F	C, L	C, L	Biopsy (lumbar nerve root)	Meningeal CE, multifocal linear and nodular CE	Cranial nerves, basal meninges	Partial improving state
5	Bose [15]	2002	33	F	C5-6 (C6)	(-)	Excision	Extramedullary enhancing mass in neural foramen (like a Schwannoma)	(-)	No recurrence
6	Shono [11]	2004	60	F	T	(-)	Biopsy (lymph node)	Normal	(-)	No recurrence
7	Schaller [9]	2006	39	F	C3 (right)	C3	Excision	Intradural extramedullary enhancing mass	(-)	No recurrence
8	Kobayashi [7]	2013	22	M	C1-L4 (not definite)	C1-L4	Biopsy (skin)	CE (not shown)	Brain	CR on cord lesion
			25	M	C1-S2 (not definite)	C1-S2	Biopsy (lung)	CE (not shown)	(-)	CR on cord lesion
			35	M	C1-S2, cauda equina (not definite)	C1-S2	CSF study	Meningeal CE, multifocal linear and nodular CE	Uveitis, brain	PR on cord lesion
9	Deng [5]	2018	48	M	C2-6 and upper T, left L5, cauda equina	C2-6 and upper T, C2-T1	CSF study	Dorsal nerve root CE, meningeal CE, multifocal linear and nodular CE	(-)	6 months later, resolving inflammation of the cord
			54	F	C2-T1	C2-T1	Biopsy (cervical cord)	Dorsal nerve root CE, meningeal CE, multifocal linear and nodular CE	(-)	After treatment, clinically stable
			49	F	C1-T6 (not definite)	C1-T6	Biopsy (axilla)	CE (not shown)	(-)	PR on cord lesion
			54	F	C3-C7 (not definite)	C3-C7	CSF study	CE (not shown)	Uveitis	PR on cord lesion
			63	F	C5-6 (not definite)	C5-6	CSF study	CE (not shown)	Uveitis	wax and wane on cord lesion

Table 1 (continued)

No	Authors	Year	Age	Sex	Nerve root level	Spinal cord lesion	Diagnostic confirmation	Nerve root MRI finding	Other neural lesions	Follow-up
		37	F		C2-T11	C2-T11	Biopsy (mediastinal)	Dorsal nerve root CE, meningeal CE, multifocal linear and nodular CE	(-)	After treatment, clinically stable

†M male; F female; C cervical; T thoracic; L lumbar; CE contrast enhancement; SI signal intensity; NA not applicable; CSF cerebrospinal fluid; CR complete remission; PR partial remission

thickening and enlargement, distinct from the involvement of the spinal cord. This is considered an exceedingly rare imaging observation of nerve root involvement.

Neurosarcoidosis typically does not spontaneously remit, except in the case of facial nerve palsy. The goal of treatment is the relief of acute inflammation and restoration of neurological symptoms [8, 10]. However, since cervical nerve root involvement in neurosarcoidosis is very rare, few studies have reported changes in MRI findings according to the patient's clinical course [9]. In the case described here, initial lesions involving the CN and cervical nerve roots improved, while others newly appeared on the contralateral side; this was observed on three spinal MRI scans obtained over the course of two months, during which the patient received steroid treatment. Based on the observed changes in MRI findings and the clinical course of the patient, sarcoidosis was the reasonable diagnosis in this case.

Facial nerve and optic nerve are commonly involved in neurosarcoidosis, but trigeminal nerve involvement is relatively rare [16]. In our patient, a homogeneously enhancing soft tissue mass was observed in the right Meckel's cave, involving the trigeminal nerve through the dural lining of the Meckel's cave. These findings also correlated with the patient's facial numbness.

Conclusion

This case report presents a rare instance of neurosarcoidosis involving cervical spinal nerve roots with unusual MRI findings: diffuse enlargement with homogeneous contrast enhancement in nerve roots, particularly within the neural foramen. Additionally, we describe the evolution of the patient's follow-up MRI findings throughout the clinical course. These distinctive MRI findings of neurosarcoidosis involving nerve roots would assist clinicians in recognizing and diagnosing such lesions, as well as monitoring the patient's clinical progression and response to treatment.

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Authors' contributions Conceptualization: Eun Kyung Khil; methodology: Kyoung Yeon Lee, Eun Kyung Khil, Seun Ah Lee, Joon Woo Lee; formal analysis and investigation: Kyoung Yeon Lee, Eun Kyung Khil; resources: Seun Ah Lee, Joon Woo Lee, Eugene Lee; supervision: Eun Kyung Khil, Seun Ah Lee, Joon Woo Lee, Eugene Lee; validation: Seun Ah Lee, Eun Kyung Khil; visualization: Kyoung Yeon Lee, Eun Kyung Khil, Seun Ah Lee; writing—original draft: Kyoung Yeon Lee, Eun Kyung Khil; writing—review and editing: Kyoung Yeon Lee, Eun Kyung Khil.

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Declarations

Conflict of interest The authors have no competing interests to declare that are relevant to the content of this article.

Ethics approval Approval was obtained from the ethics committee of Hallym University Dontan Sacred Heart Hospital. The procedures used in this study adhere to the tenets of the Declaration of Helsinki.

Informed consent The participant has consented to the submission of the case report to the journal.

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