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LEFT LOWER LIMB PARESIS SECONDARY TO SYMPTOMATIC SPINAL CORD LESION AS THE INAUGURAL MANIFESTATION OF SARCOIDOSIS: CASE REPORT WITH REVIEW OF LITERATURE.

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ABSTRACT Sarcoidosis is a granulomatous disease that mainly affects the lungs. The specific histopathological findings in sarcoidosis are epithelioid and gigantocellular granulomas without caseous necrosis. The lung is the most commonly affected organ, with extrapulmonary involvement occurring in about half of sarcoidosis patients. Sarcoidosis can affect any organ, and when patients present with central or peripheral nervous system involvement, they are diagnosed with neurosarcoidosis. Sarcoidosis-associated myelopathy (SAM) has been described in patients with sarcoidosis, and the clinical and radiological manifestations are diverse and non-specific. The diagnosis of SAM can be very difficult due to multiple mimickers that can cause similarity in clinical and radiological features. Here we report the case of a 51-year-old woman with a history of diabetes who presented with slight and progressive left lower limb deficit with sensory complaints in four limbs secondary to an extensive and tumefactive lesion in the cervico-thoracic region of the spinal cord related to sarcoidosis.

KEYWORDS : sarcoidosis, myelopathy, granulomas, neurosarcoidosis

INTRODUCTION:

Neurosarcoidosis (NS) is a neurological entity related to central and/or peripheral nervous system involvement with sarcoidosis. Sarcoidosis can affect any organ, and is characterised histologically by epithelioid and gigantocellular granulomas without caseous necrosis. It mainly affects the lymphoreticular system and the lymphatic components of organs such as the liver, spleen and parotid glands [1]. NS lesions may appear as small or large nodular lesions of the central nervous system (CNS) parenchyma as well as meningeal involvement in the form of leptomeningitis or pachymeningitis. Neurological manifestations are variable and non-specific. Various regions of nervous system could be involved by sarcoidosis such as: cranial nerves, brain parenchyma, meninges, brainstem, cerebellum, spinal cord, nerve roots, and muscles [2].

The lungs are the most commonly affected organ, with extrapulmonary involvement seen in around half of sarcoidosis patients [2]. The CNS is reported to be affected in 5-15% of sarcoidosis patients [3]. Sarcoidosis-associated myelopathy (SAM) referred to sarcoidosis of the spinal cord is rarely reported in the literature and presents with variable clinical and radiological features [2,4]. The diagnosis is often challenging as it can mimic several diseases such as multiple sclerosis and neuromyelitis optica spectrum disease. Here we report a 51-year-old woman which complained from chronic slight paresis of left lower limb and sensitive symptoms of 4 limbs secondary to sarcoidosis of spinal cord.

Case report:

51-year-old female patient with type 2 diabetes, on oral antidiabetic medication for 25 years. For the past 2 years, she has had chronic and progressive sensory disturbances in all 4 limbs, but predominantly in the lower limbs, with paresthesia and sensation of walking on cotton and spikes. For the past 6 months, she has noticed muscle weakness in her left lower limb (LL), which has progressed slowly without affecting her walking. There was no evidence of sphincter dysfunction, no rachialgia, no evidence of visual disturbance and no change in general condition.

Clinical examination revealed a conscious, eupneic patient with normal blood pressure and heart rate. Neurological examination revealed a slight proximodistal paresis of the left lower limb with thermoalgic hypoesthesia and vibratory hypoesthesia of the LL. Tendon reflexes were presents and symmetrical in the upper limbs and absent in the LL. There was no dysmetria or cognitive impairment. Cranial nerve examination was unremarkable. The rest of the somatic examination was normal.

In view of this clinical presentation and in the context of a long history of diabetes mellitus, we opted for an EMG study, which showed features of a severe axonal length-dependent sensitivomotor neuropathy, probably due to diabetes. However, this neuropathy did not explain the paresis of the left LL or the complaints of walking on cottons and on spikes. Therefore, a spinal cord MRI was performed and was consistent with extensive longitudinally and transversely myelopathy, showing hyperintensity from C5 to T1 on T2 in sagittal sections and centrally distributed on axial sections, with postero-lateral contrast enhancement on the left (figure 1). There was a marked fusiform thickening of the spinal cord in the cervico-dorsal region. Brain MRI was also performed and was unremarkable. Brain MRI was also carried out and was unremarkable.

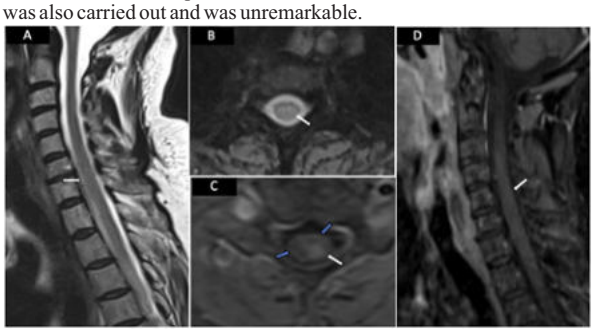

Figure 1: Cervico-thoracic spinal MRI in sagittal T2 (A), axial T2 (B), with post-contrast T1 FAT-SAT axial (C), and post-contrast T1 FAT-SAT sagittal (D); demonstrating tumefaction with extensive hyperintense signal (white arrows) longitudinally from C5 to T1 and centrally with enhancement that is posterior and lateralized to the left in the axial image (C), associated with subpial enhancement (blue arrows). Axial sequences (with and without contrast) were taken at the T1 level.

Biological investigations were consistent with hyperglycemia and an elevated level of enzyme-converting angiotensin (ECA) in serum, while a large number of paraclinical tests were normal (complete blood count, prothrombotic tests, liver and kidney tests, autoantibody tests, tumour markers, anti-AQP4, anti-MOG, syphilis serology, hepatitis and HIV tests). CSF analysis was normal. A CT scan of the chest was performed and showed small mediastinal adenopathies. Labial biopsy showed non-caseating granulomas (Figure 2).

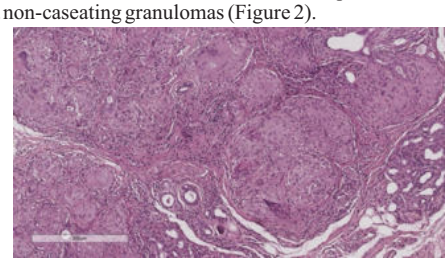

Figure 2: Anatomopathological study of a biopsy of the accessory salivary glands showing gigantocellular epithelioid granulomas without caseous necrosis consistent with sarcoidosis.

A diagnosis of chronic myelopathy in the setting of systemic sarcoidosis was made. The patient was started on pulse intravenous methylprednisolone (240 mg/day for 7 days) and subsequently discharged on prednisone 40 mg/day with azathioprine (150 mg/day). At 3 months, the course of the disease was characterised by a regression of sensory complaints and a recovery of the left LL deficit.

DISCUSSION:

Our case shows that sarcoidosis can be manifested by neurological damage in the form of chronic myelopathy. In a study conducted by Murphy et al., this finding was reported in 49 (79%) cases among the 62 patients included in his work on SAM [4]. The estimated incidence of myelopathy related to sarcoidosis is between 1.0 and 35.5 per 100,000 people around the world, with variations between population and ethnic groups[5]. In a previous meta-analysis of neurosarcoidosis studies, spinal cord involvement in sarcoidosis was reported in 18% of patients [6].

The pattern of onset of SAM is classified into 4 types: hyperacute: <6 hours; acute: 6-48 hours; subacute: 6-48 hours; subacute: 2-21 days; chronic >3 weeks [4]. Chronic presentations are reported in 81% of cases of SAM, with a predominance of sensory symptoms, as in our case. Criteria outlined by the Neurosarcoidosis Consortium Consensus Group in 2018, distinguish 2 subtypes of NS: defined, when pathological granulomatous disease in the nervous system is confirmed; and probable, when pathological systemic granulomatous disease is confirmed [4]. According to these criteria, our case corresponded to the probable subtype. Given the potential risk of neurological deterioration associated with spinal cord biopsy, it should be reserved only if other sites are not identified on 18F-FDG PET scan, if symptoms worsen despite empiric treatment, or if there is a high likelihood of malignancy [5]. We didn't perform the biopsy of the spinal cord lesion in our patient because there was a strong evidence for a sarcoidosis etiology.

There are several causes of chronic myelopathy that may mimic SAM and they are including etiologies like Sjögren's disease, multiple sclerosis, neuromyelitis optica, tuberculosis, Behçet's disease, metabolic disorders and malignancy [5]. These etiologies were unlikely in our case given the clinical presentation, biological findings and radiological pattern of lesions already reported to be associated with sarcoidosis.

Concerning pattern MRI lesion described in SAM are various: longitudinally extensive myelitis (>3 segments), Short tumefactive myelitis, Spinal meningitis/meningoradiculitis, Anterior myelitis with disc degeneration, and other aspects. The first radiologic pattern is in accordance with our finding, with predominantly dorsal subpial ± meningeal enhancement was the most reported in literature [4]. Experts suggest that dorsal-predominant enhancement or the trident sign in a tumefactive lesion should be considered clues to SAM. In our patient we didn't found this sign. The distribution of contrast enhancement on axial post contrast T1 FAT-SAT that was lateralized to left explained the paresis of left LL of our patient because the enhancement is located in the pathway of left pyramidal tract. Fusiform thickening of the spinal cord, as seen in our case, most commonly in the thoracic or cervical spine, involving three or more spinal segments, has been reported in the literature [7].

In Murphy's study, CSF testing revealed the presence of pleocytosis and hyperproteinorachia in some cases, and the presence of CSF-restricted oligoclonal bands (OCB) in approximately 20% of samples tested [4]. The latter finding of OCB in patients with SAM should alert the clinicians to interpret these results with caution when evaluating patients with inflammatory CNS involvement.

In general, there is no standardized protocol for the treatment of SAM. Experts agree that first-line treatment should be with glucocorticoids, which may be combined with second-line treatment with methotrexate or azathioprine. For refractory or very severe forms, third-line treatment with anti-tumour necrosis factor alpha (TNF-α) therapy such as infliximab may be indicated [5].

CONCLUSION:

Sarcoidosis-associated myelopathy is a disabling chronic disease that can be difficult to diagnose. The gold standard for diagnosis is histopathological evidence of CNS involvement. Clinical manifestations are variable and non-specific. Thus, the clinicians should adopt a correct clinical approach when evaluating patients with neurological manifestations. There are different patterns of radiologic findings in sarcoidosis-associated myelopathy, however some radiological lesions could suggest sarcoidosis disease. Our case highlights that when assessing diabetic patients with peripheral diabetic neuropathy, which doesn't explain the patients' symptoms, further investigations in these patients are needed.

REFERENCES:

1. Tetikkurt C. Neurosarcoidosis. *Journal of Neurology*. Published online 2018.
2. Bradshaw MJ, Pawate S, Koth LL, Cho TA, Gelfand JM. Neurosarcoidosis: Pathophysiology, Diagnosis, and Treatment. *Neurol Neuroimmunol Neuroinflamm*. 2021;8(6):e1084. doi:10.1212/NXI.0000000000001084
3. Cação G, Branco A, Meireles M, et al. Neurosarcoidosis according to Zajicek and Scolding criteria: 15 probable and definite cases, their treatment and outcomes. *Journal of the Neurological Sciences*. 2017;379:84-88. doi:10.1016/j.jns.2017.05.055
4. Murphy OC, Salazar-Camelo A, Jimenez JA, et al. Clinical and MRI phenotypes of sarcoidosis-associated myelopathy. *Neurol Neuroimmunol Neuroinflamm*. 2020;7(4):e722. doi:10.1212/NXI.0000000000000722
5. Nolte JYC, Ten Dam L, Van De Beek D, Brouwer MC. Clinical characteristics and outcome of neurosarcoidosis associated myelitis: A retrospective cohort study and review of the literature. *Euro J of Neurology*. 2022;29(6):1763-1770. doi:10.1111/ene.15295
6. Fritz D, Van De Beek D, Brouwer MC. Clinical features, treatment and outcome in neurosarcoidosis: systematic review and meta-analysis. *BMC Neurol*. 2016;16(1):220. doi:10.1186/s12883-016-0741-x
7. Hebel R, Dubaniewicz-Wybieralska M, Dubaniewicz A. Overview of neurosarcoidosis: recent advances. *J Neurol*. 2015;262(2):258-267. doi:10.1007/s00415-014-7482-9