

A Rare Case of Pulmonary Sarcoidosis With Extracranial Carotid System Involvement

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Introduction

Sarcoidosis is a disease of unknown etiology, whose pathogenesis appears to involve genetic, immunologic, infectious, and environmental factors. It was first described in 1889 by Ernest Besnier and is also called Besnier-Boeck-Schaumann disease.^{1,2}

In sarcoidosis, a noninfectious inflammatory process leads to the formation of granulomas, which predominantly affect the lungs and intrathoracic lymph nodes in approximately 90% to 95% of cases. Close to 30% of patients with sarcoidosis present with extrapulmonary involvement, which may be the primary manifestation of the disease or occur in isolation.³⁻⁵

Involvement of cervical vessels is an uncommon manifestation and seldom described in the literature. This is a case report and literature review aiming to understand and highlight the need to assess possible extrapulmonary sites using multimodal imaging, as well as the monitoring of these sites after initiation of drug treatment, with a focus on ultrasound findings.

Case report

A 61-year-old woman with complaints of headache and bilateral neck pain for approximately 6 months in association with weight loss and asthenia attended an appointment with a rheumatologist, who requested laboratory tests and an ultrasound of the carotid and vertebral arteries as initial diagnostic tests. Vascular ultrasound (VUS) of the carotid and vertebral arteries showed marked diffuse bilateral perivascular thickening along the common carotid artery, without involvement of the internal and external branches. The vascular lumen of the cervical vessels was also preserved (Figures 1 and 2).

Keywords

Endosonography; Carotid Arteries; Sarcoidosis

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The patient's past pathological history included previous surgery for meningioma 2 years ago. The histological report indicated the presence of noncaseating granulomatous inflammation, with sarcoidosis being the leading diagnostic hypothesis (Figure 3). The patient then underwent computed tomography of the chest, with findings consistent with pulmonary sarcoidosis.

Treatment with oral prednisone was initiated. There was improvement of systemic symptoms and regression of edema and cervical pain after 2 months of treatment. VUS of the carotid and vertebral arteries was performed 6 months after the start of drug treatment and demonstrated partial regression of perivascular edema bilaterally, and ultrasound follow-up was scheduled every 6 months (Figure 4).

Discussion

Sarcoidosis is a multisystem inflammatory disease with an heterogenous presentation and clinical course, which can affect any organ and progress from spontaneous remission to multiple organ dysfunction. This heterogeneity, associated with the overlap of symptoms with other more common diseases, often leads to diagnostic uncertainty and treatment delays. The diagnosis of sarcoidosis is based on three parameters: a) clinical and imaging presentations consistent with the disease; b) evidence of noncaseating granulomas; and c) exclusion of alternative diagnoses. However, diagnostic confirmation may be challenging in the absence of pronounced chest or skin manifestations.

In case of neck pain, other diseases must be discarded, such as carotid or vertebral dissection, Takayasu's arteritis, thyroiditis, or transient perivascular inflammation of the carotid artery (TIPIC syndrome).⁶

In this case report, B-mode ultrasound images in the transverse and longitudinal views showed extensive perivascular involvement, including the common carotid arteries but sparing the internal carotid arteries. Some features of sarcoidosis overlap with those of Takayasu's arteritis, such as long-term fatigue, weight loss, neck pain, and preservation of the carotid branches. However, in the case presented in this report, the lesions were extravascular, the intima-media complex was preserved, and the vessel lumen was not reduced, unlike echocardiographic findings of Takayasu's arteritis, in which there is a concentric thickening of the intima-media complex involving the posterior and anterior vessel walls, leading to a reduction in the vessel lumen.⁷

Case Report

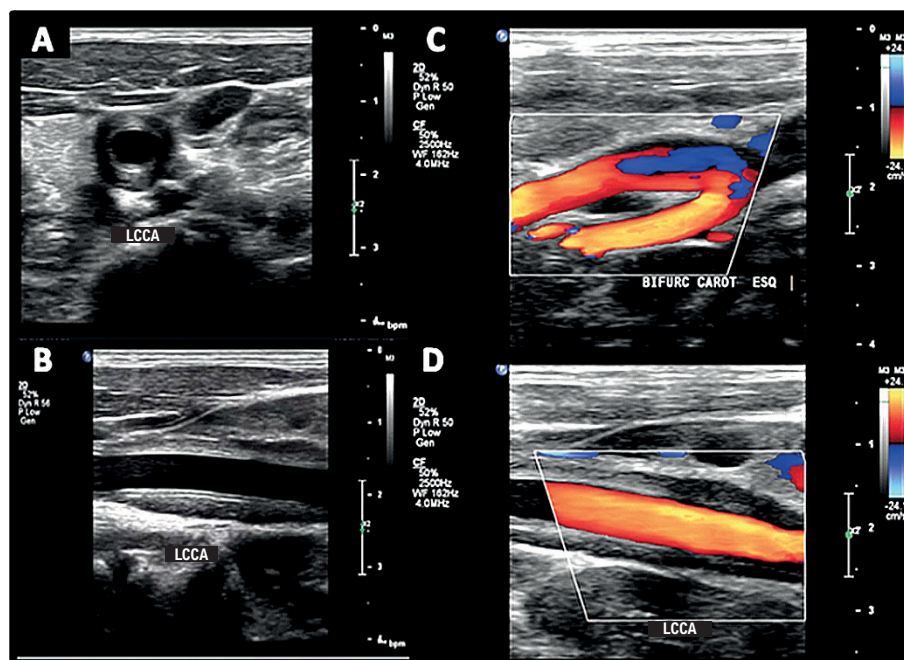


Figure 1 – B-mode ultrasound images of the left common carotid artery demonstrating the presence of significant circumferential perivascular thickening in the (A) transverse and (B) longitudinal views. Color flow mapping showing (C and D) preserved flow and perivascular thickening along the common carotid artery without involvement of the internal and external carotid arteries. LCCA: left common carotid artery.

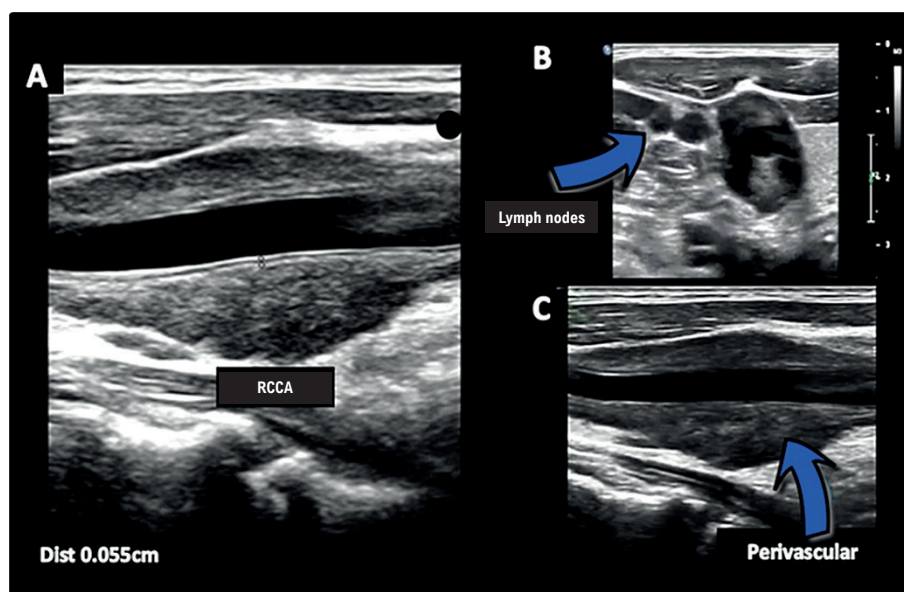


Figure 2 – Ultrasound of the right common carotid artery demonstrating A) preserved intima-media thickness in the longitudinal view, B) cervical lymphadenopathy in the transverse view, and C) perivascular thickening in the longitudinal view. RCCA: right common carotid artery.

Case Report

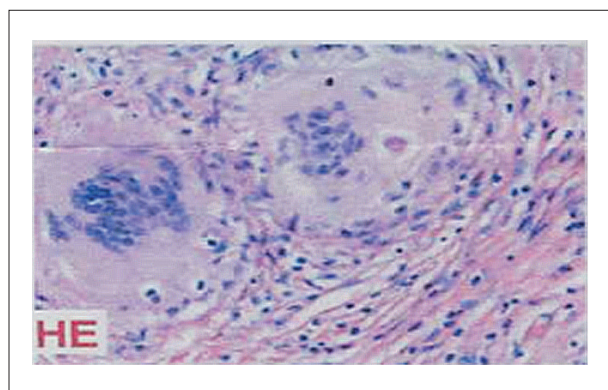


Figure 3 – Immunohistochemistry of the meninges identified areas of fibrosis, formation of noncaseating epithelioid granulomas, and structures suggestive of asteroid and Schumann bodies, findings consistent with a predominantly histiocytic granulomatous inflammatory process.

Perivascular inflammation of the carotid arteries is described in TIPIC syndrome, a rare condition characterized by neck pain and echocardiographic findings similar to those described in our patient. Its differentiation from sarcoidosis was made based on the location, the persistence of the inflammatory process, and the presence of cervical lymphadenopathy. The most frequent site of carotid artery involvement in patients with TIPIC is the carotid bifurcation, more precisely the bulbar region.⁸

Corticosteroid therapy remains the first-line treatment for sarcoidosis, except when contraindicated.⁹ If the disease is not controlled in the induction phase (generally within 3 to 6 months), the use of immunosuppressants as second-line treatment can be considered.¹⁰

Conclusion

Sarcoidosis is an inflammatory disease with a challenging diagnosis, in which vascular involvement is extremely rare. The VUS proved to be a reliable tool in the detection of perivascular inflammatory involvement of the extracranial carotid artery and cervical lymph nodes, which was crucial in the differential diagnosis between sarcoidosis and other nonatherosclerotic diseases involving the cervical vessels.

Author Contribution

Conception and design of the research and writing of the manuscript: Barros FS; acquisition of data and analysis and interpretation of the data: Barros FS, Silva HAGP, Barros DS; critical revision of the manuscript for intellectual content: Barros FS, Santos SN.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

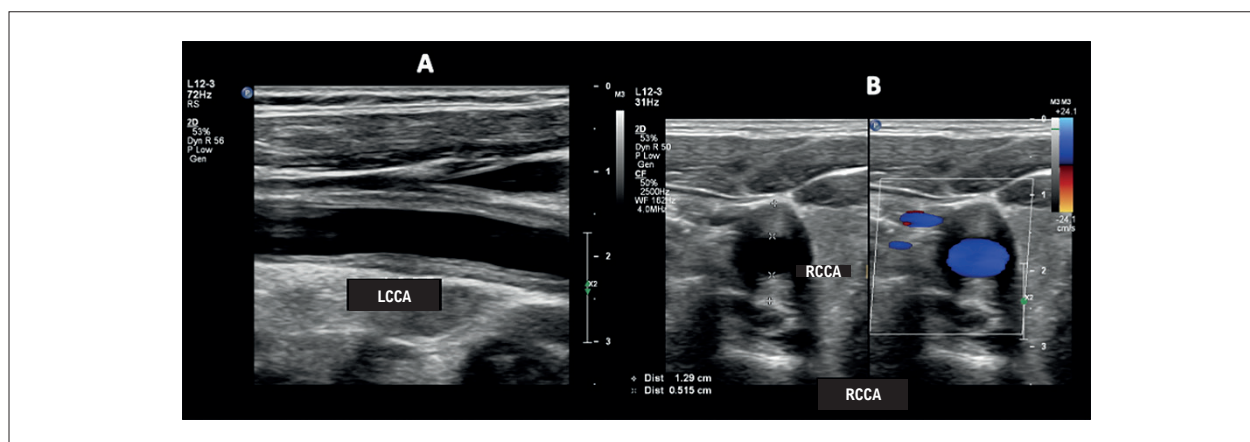


Figure 4 – Ultrasound of the common carotid arteries demonstrating a reduction in perivascular thickening bilaterally, after the start of clinical treatment.

References

1. Besnier EH. Lupus Pernio de La Face; Synovites Fongueuses (Scrofulo-Tuberculeuses) Symétriques Des Extrémités Supérieures. *Ann Derm Syph.* 1889;10:545-9.
2. Hem E. Boeck's sarcoidosis--a centennial. *Int J Dermatol.* 2000;39(7):545-9. doi: 10.1046/j.1365-4362.2000.00397.x.
3. Kouranos V, Wells AU. The Role of Primary Care in Sarcoidosis. *Curr Opin Pulm Med.* 2023;29(5):493-500. doi: 10.1097/MCP.0000000000000991.
4. Costabel U. Sarcoidosis: Clinical Update. *Eur Respir J Suppl.* 2001;32:56s-68s. doi: 10.1183/09031936.01.18s320056.
5. Crouser ED, Maier LA, Wilson KC, Bonham CA, Morgenthau AS, Patterson KC, et al. Diagnosis and Detection of Sarcoidosis. An Official American Thoracic Society Clinical Practice Guideline. *Am J Respir Crit Care Med.* 2020;201(8):e26-e51. doi: 10.1164/rccm.202002-0251ST.
6. Albricker ACL, Freire CMV, Santos SND, Alcantara ML, Cantisano AL, Porto CLL, et al. Recommendation Update for Vascular Ultrasound Evaluation of Carotid and Vertebral Artery Disease: DIC, CBR and SABCV - 2023. *Arq Bras Cardiol.* 2023;120(10):e20230695. doi: 10.36660/abc.20230695.
7. Brkic A, Terslev L, Møller Døhn U, Torp-Pedersen S, Schmidt WA, Diamantopoulos AP. Clinical Applicability of Ultrasound in Systemic Large Vessel Vasculitides. *Arthritis Rheumatol.* 2019;71(11):1780-7. doi: 10.1002/art.41039.
8. Barros FS, Santos SN, Barros DS, Cunha SS, Albricker AL, Petisco ACG. Transient Perivascular Inflammation of the Carotid Artery (TIPIC): Vascular Ultrasonography Role. *Arq Bras Cardiol: Imagem Cardiovasc.* 2023;36(2):e20230019. doi: 10.36660/abcimg.20230019i.
9. Baughman RP, Valeyre D, Korsten P, Mathioudakis AG, Wuyts WA, Wells A, et al. ERS Clinical Practice Guidelines on Treatment of Sarcoidosis. *Eur Respir J.* 2021;58(6):2004079. doi: 10.1183/13993003.04079-2020.
10. Hamzeh N, Voelker A, Forssén A, Gottschall EB, Rose C, Mroz P, et al. Efficacy of Mycophenolate Mofetil in Sarcoidosis. *Respir Med.* 2014;108(11):1663-9. doi: 10.1016/j.rmed.2014.09.013.



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