Cardiac panniculitis. A novel cause of reversible atrioventricular block

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Introduction
Panniculitis is a group of heterogeneous inflammatory diseases involving subcutaneous fat. The specific diagnosis requires histopathologic study because different etiologies usually show the same clinical appearance, presenting much like erythematous nodules generally located on the lower limbs. It can be caused by infectious, autoimmune, and oncological conditions and it is frequently considered idiopathic. Systemic panniculitis with cardiac involvement is a rare condition. We report a unique case presenting as complete AVB and evidence of reversibility with treatment.

Case report
A 76-year-old woman went to the hospital after syncope. She had been under study since 3 months prior for cutaneous nodules on her leg skin and receiving treatment with corticosteroids (25 mg prednisone daily). Skin biopsy revealed subcutaneous neutrophilic infiltrate with lobular distribution, minimal dermal involvement, leukocytoclastic nuclei, fat necrosis, and reactive stromal fibroblasts. She presented intermittent chest pain. She did not have any other cardiovascular history and did not take any drug that affected atrioventricular conduction. Her body mass index was 27 kg/m². She had a history of type 2 diabetes mellitus under oral antidiabetic treatment and hypertension.

Electrocardiography (Figure 1) showed complete AVB with escape rhythm at 40 beats per minute, occasionally alternating with occasionally conducted sinus beats. Blood electrolytes were normal and renal function mildly impaired. Troponin (13.65 μg/L, reference range <0.06) and N-terminal pro-B-type natriuretic peptide (7894 pg/mL, reference range 0–125) were increased.

Echocardiography revealed moderate left ventricular hypertrophy and pseudonormal filling with preserved systolic function without wall motion abnormalities, pericardial effusion, or valvulopathies. First, atropine was administered with successful response, so sinus rhythm was restored without alterations suggestive of ischemia and isoprenaline perfusion was initiated. Owing to insufficient response and initial suspicion of myocardial sarcoidosis, a bolus of corticosteroids (1 mg/kg) was administered, with subsequent recovery of atrioventricular conduction with narrow QRS. A definitive pacemaker was implanted.

Coronary angiography ruled out epicardial coronary lesions. In the same procedure, an endomyocardial biopsy (EMB) was taken from the left ventricle. EMB (Figure 2) showed inflammation of interstitial adipose tissue of the myocardium with mild lesion of muscular fibers. Although EMB avoided the conduction system, partial response to atropine suggests the inflammation was affecting the atrioventricular node. No vascular changes, granulomas, or microorganisms were identified. These findings were similar to those previously described in biopsies of the skin. All these findings together were pathognomonic of systemic nodular panniculitis.

KEY TEACHING POINTS
- Systemic panniculitis can affect the heart and cause atrioventricular block.
- Cardiac involvement can be demonstrated with endomyocardial biopsy.
- Treatment with corticosteroids and methotrexate improves atrioventricular block related to panniculitis with cardiac involvement.

KEYWORDS
Atrioventricular block; Case report; Endomyocardial biopsy; PET scan; Systemic panniculitis
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18F-FDG-PET/CT was performed to rule out an underlying disease (Figure 3). The association with myeloid disorders has been established in the setting of neutrophilic dermatoses and there are cases of neutrophilic panniculitis related to myeloid proliferations. Blood tests performed did not show any signs of myeloid disorder.

Because of minor uptake within the left breast and rectum, mammography and colonoscopy were performed, none of which showed pathologic findings.

Treatment with oral corticosteroids and methotrexate was started. Subsequently, inflammatory cutaneous lesions improved, lower uptake of FDG was reported (Figure 3), and pacemaker showed <1% of ventricular pacing. Although the patient was previously receiving corticosteroid treatment, the findings indicate a partial response to a more intense immunosuppressive treatment.

Discussion
Panniculitis with cardiac involvement is rare and its demonstration is difficult. Most previous cases demonstrate the involvement at autopsy, without prior cardiologic symptoms. Very variable pathologic findings have been described, such as isolated epicardial fat involvement, interstitial fibrosis without inflammatory activity, granulomas, vasculitis, and fibrinoid necrosis. In other cases, myocardial involvement has been assumed mainly owing to previous diagnosis of systemic panniculitis. This case represents important differences with previous cases, both clinically and pathologically. Few cases have previously shown perimyocardial inflammatory activity. None of them had been correlated with cardiology clinic or imaging findings; and although decrease of cutaneous lesions with corticosteroid treatment has been proved, the improvement of the AVB observed in our case constitutes a unique proof of visceral response to the treatment.

Specific characteristics of EMB gave the differential diagnosis of other panniculitis. Systemic lupus can rarely cause lobular panniculitis involving subcutaneous fat, but lupus cardiac involvement occurs with myocarditis without panniculitis. However, EMB did not show immune complexes, which are typical of lupus panniculitis.

A relevant limitation of the case is the lack of an EMB without signs of panniculitis after treatment, which makes it difficult to establish causality with panniculitis. However, the simultaneous improvement in auriculoventricular and skin lesions supports causality. Magnetic resonance imaging could not be performed because of recent pacemaker implantation, although it would have been useful to dismiss sarcoidosis.

Conclusion
This case is the first to represent systemic panniculitis with cardiac involvement demonstrated with EMB. In patients...
with suspected infiltrative etiology, an EMB could confirm the diagnosis of this potentially reversible cause of AVB.

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