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CASE REPORT

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A case report on the implantation of a leadless pacemaker in a patient with eosinophilic fasciitis and third-degree atrioventricular block

Li-Juan Huang[1], Jian-Shu Chen[2], Qi Zou[1], Yu-Zhe Song[1], Yi Yan[1], Zhao-Fen Wang[1], Hai-Juan Wang[1] and Peng Chang[2*]

Abstract

Eosinophilic fasciitis (EF) is a rare connective tissue disorder characterized by the involvement of the dermis, subcutaneous tissue, and fascia. The treatment for EF usually involves long-term use of glucocorticoids and immunosuppressants. Patients with EF are at risk of developing third-degree atrioventricular (AV) block during the course of the disease. The distinctive features of EF, the side effects of its treatment, and the inherent limitations of transvenous pacemakers (TVPs) present significant challenges in the management of patients with EF who also have third-degree AV block. We present the case of a 64-year-old Chinese male diagnosed with EF and concomitant third-degree AV block. Given the patient's skin tissue characteristics, the increased risk of infection associated with long-term immunosuppressive therapy, and the potential complications related to TVPs we chose to implant a leadless pacemaker(LP) in the apical region of the right ventricle. This case report underscores the importance of identifying potential cardiovascular complications in EF patients treated with corticosteroids and immunosuppressants. It also highlights the clinical benefit of LP implantation in managing patients with EF and third-degree AV block, especially in terms of minimizing device-related complications and infection risks. This study offers a fresh perspective on the treatment of EF patients who have third-degree AV block and advocates for the use of LPs as a preferred option for cardiac pacing in this patient group. Further research is warranted to evaluate the indications and potential benefits of LPs in a wider range of patients.

Clinical trial number

Not applicable.

Keywords Leadless pacemaker, Micra, Third-degree atrioventricular block, Eosinophilic fasciitis, Case report

*Correspondence:
Peng Chang
pchang@lzu.edu.cn
[1]Lanzhou University, Lanzhou 730030, China
[2]Department of Cardiology, Lanzhou University Second Hospital, Lanzhou 730030, China

Introduction

Eosinophilic fasciitis (EF) is a rare inflammatory condition characterized by skin hardening, tightness, and the distinctive 'peau d'orange' appearance due to subcutaneous fibrosis [1]. The progressive hardening of the skin often results in contraction and a reduction in wrinkles, impacting both the appearance and function of the affected areas. The cause of EF is not entirely clear, but it is widely accepted that autoimmune mechanisms play a central role in its development [2]. Immunosuppressive therapy and glucocorticoid treatment are the standard approaches for managing EF, intended to mitigate the inflammatory process and prevent disease progression [3]. Nevertheless, these treatment strategies increase the risk of infectious complications, which in turn present significant challenges in the clinical management of patients with EF who also experience third-degree atrioventricular (AV) block.

Medtronic's Micra leadless pacemaker (LP) gained FDA approval in 2016. This technology provides an innovative solution for patients in need of pacing therapy who are at high risk for complications associated with transvenous pacemakers (TVP). Compared to TVP, LP significantly lower the risk of infection and complications related to leads and pockets [4]. By reducing the risks of pocket infections, lead dislodgement, and mechanical complications, this type of LP is especially advantageous for patients with compromised immune systems or those with connective tissue diseases.

This case illustrates a unique clinical scenario involving a patient with EF who also has third-degree AV block, a severe arrhythmia that requires pacemaker implantation for treatment. The most commonly used type of pacemaker is the transvenous pacemaker. Considering the difficulties in pocket formation, the risk of device exposure, and the increased risk of infection, a LP was successfully implanted in this patient. This approach not only provides effective pacing support but also reduces the risks associated with the device. Compared to TVP, LPs have a shorter time on the market, are more expensive, and due to the rarity of cases involving EF with third-degree AV block, their application in similar cases has been insufficient. This case report emphasizes the potential benefits of LPs in managing cases of EF with third-degree AV block and advocates for the consideration of LPs in similar patient populations. It also demonstrates the advantages and clinical value of LPs, which could facilitate their broader application.

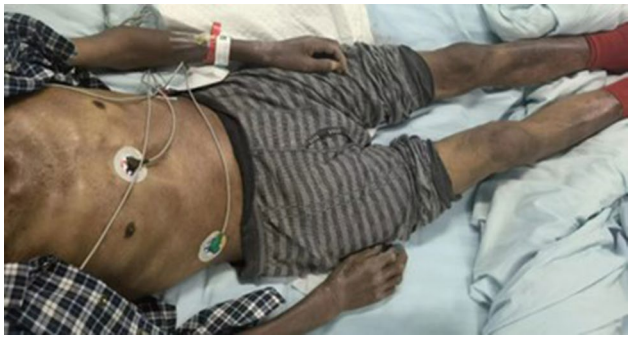


Fig. 1 Cutaneous hyperpigmentation was observed on the patient's thoracic wall and extremities

Case presentation

Seven months prior to current admission, the patient was hospitalized for swelling in the limbs, accompanied by chest tightness and shortness of breath. There were no recurrent oral ulcers, no Raynaud's phenomenon, and no joint or muscle discomfort. On physical examination, severe hyperpigmentation was noted across the body, with the skin appearing black and hardened to a texture resembling wood (Fig. 1). Relevant tests were performed to further evaluate the condition. The results indicated an elevated eosinophil count (2.15 × 10^9/L), positive antinuclear antibodies (ANA), and positivity for myositis-specific antibodies RO52 + + and SSA++. MRI of the proximal lower limbs demonstrated extensive effusions within the muscle interstices of both thighs, along with corresponding posterolateral subcutaneous effusions (Fig. 2). Electromyography (EMG) revealed myogenic changes and

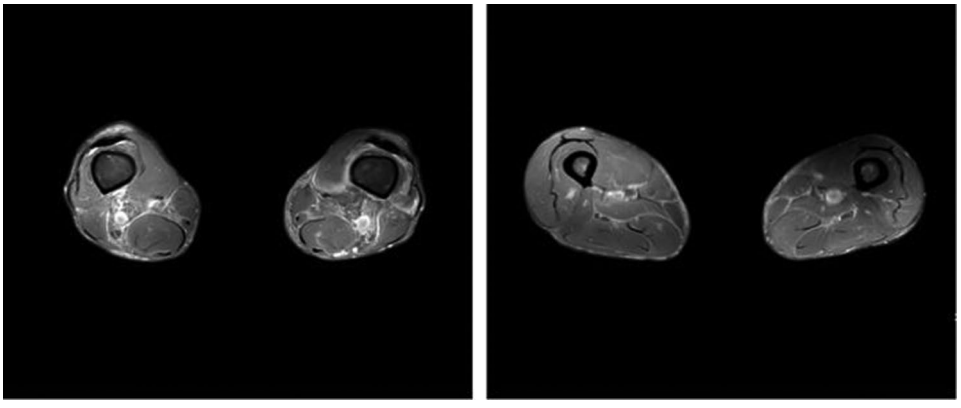


Fig. 2 MRI of the bilateral thigh muscles in the patient revealed inhomogeneous signal within the posterior muscle groups of both thighs, along with extensive intermuscular effusion, particularly at the distal levels of the thighs, and corresponding posterior and lateral subcutaneous effusion

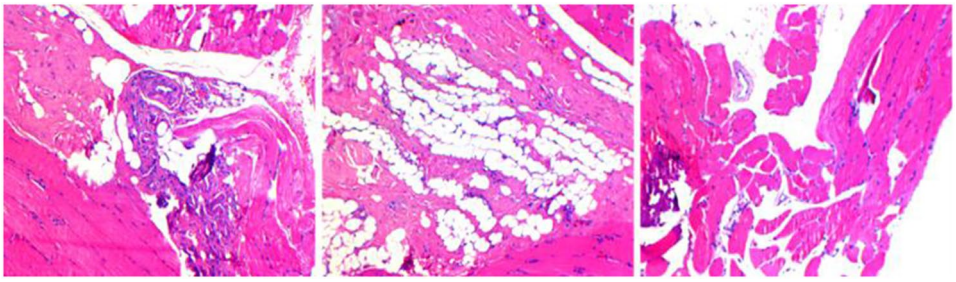
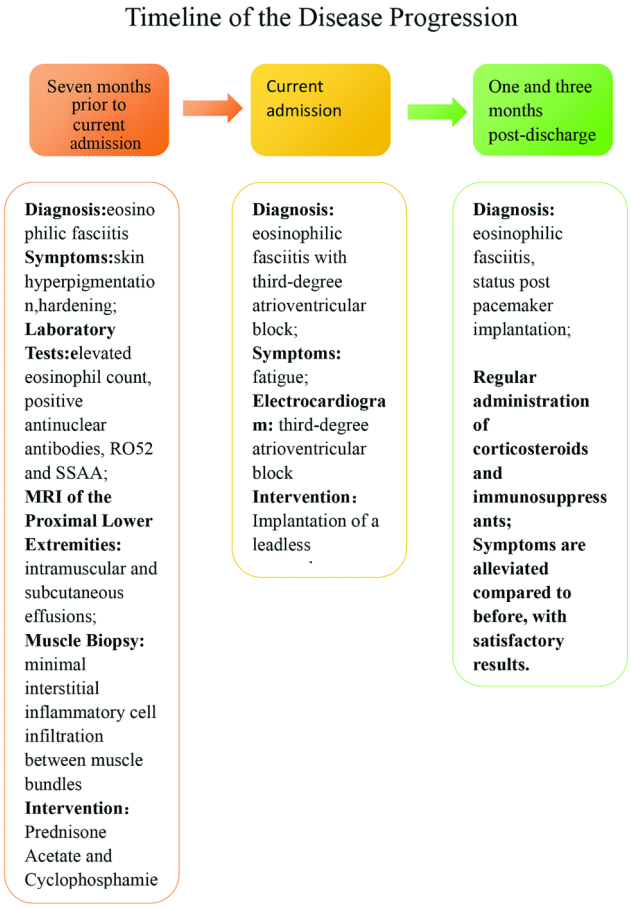


Fig. 3 It is the histopathological findings from the patient's fascial and muscle biopsy, with Hematoxylin and Eosin (H&E) staining. There is minimal inflammatory cell infiltration between the muscle fascicles (original magnification 200×)

peripheral nerve damage. A muscle biopsy from the left upper limb showed minimal interstitial inflammatory cell infiltration (Fig. 3). The patient was diagnosed with "EF" and was prescribed prednisone acetate and cyclophosphamide, which were taken regularly as an outpatient. Current admission (seven months later), the patient was admitted to the hospital due to unexplained fatigue and limitations in daily activities that had persisted for a week. An electrocardiogram revealed third-degree AV block (Fig. 4a). Combining this finding with the patient's medical history, a definitive diagnosis of "EF with third-degree AV block" was established. Considering the patient's medical history, a definitive diagnosis of "EF with third-degree AV block" was confirmed. In accordance with the 2021 ESC Guidelines for cardiac pacing and cardiac resynchronization therapy, this patient is eligible for pacemaker implantation. Given the patient's history of EF and long-term use of corticosteroids and immunosuppressants, the chronic inflammatory state may impact the body's immune response. The inflammation of the skin and fascia caused by EF results in hardening, which complicates tissue dissection during surgery and increases the difficulty of creating a pacemaker pocket. This hardening enhances the friction between the pacemaker pocket and surrounding tissues, making the skin over the pocket more prone to compression and injury, which can lead to pressure necrosis and a heightened risk of device exposure. As the disease progresses, patients may develop cardiac fibrosis, which can make it challenging for a conventional pacemaker to remain fixed, thereby increasing the risk of dislodgement. Following a thorough assessment, the decision was made to implant a LP. After obtaining consent from the patient and their family, a LP (Micra, Medtronic, Minneapolis, Minnesota, USA) was successfully implanted in the apical region of the right ventricle with the following parameters: Test parameters: pacing threshold 0.38 V, R wave 4.5mV, and impedance 580Ω. (Fig. 4b). Postoperative electrocardiogram (Fig. 4c). During the follow-up visits at 1 month and 3 months post-surgery, the patient reported no fatigue or other discomfort symptoms, the wound healed well without signs of infection, and the quality of life and exercise tolerance were both maintained at good levels.



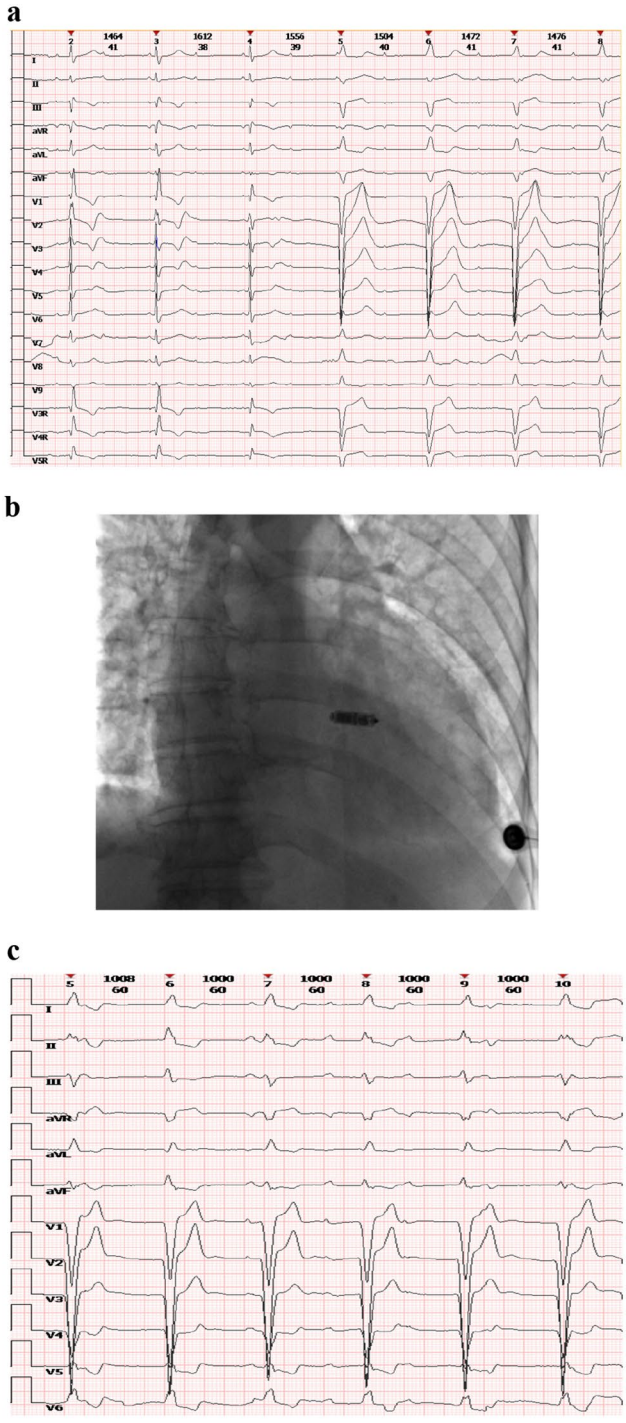


Fig. 4 **a** Third-degree atrioventricular block. **b** Successful Implantation of a Micra Leadless Pacemaker. **c** Post-implantation pacemaker electrocardiogram

Discussion

The signature biomarkers of EF include CCL4, CCL18, and CXCL9. These chemokines are upregulated in EF, potentially promoting the recruitment of inflammatory cells and increasing the risk of cardiovascular diseases [5]. To date, there are no clear research findings regarding whether EF can cause third-degree AV block.

The incidence of complications from TVP is high. Research by Clémenty et al. [6] found that the rate of complications with traditional cardiac pacemakers was 5.3%, with 89% of these complications being related to leads and pockets. The main issues included pocket bleeding, mechanical complications associated with leads or the generator, and pneumothorax. In a canine study involving transvenous pacemaker implantation, echocardiographic examinations revealed thrombus formation related to cardiac pacemaker leads in 10.4% of cases [7]. The rate of infection within the first year after implantation of cardiac implantable electronic devices is about 0.9%. For patients who develop an infection, the all-cause mortality rate within the subsequent 12 months is between 15% and 30% [8]. Furthermore, patients with transvenous pacemaker implants may also experience complications such as cardiac perforation and pacing-induced cardiomyopathy [9, 10].

In pacemaker-dependent patients, LPs provide a safer alternative, particularly in those with EF and third-degree AV block. Compared to TVP, LPs have a lower complication rate. Recent research indicates that the implantation of LPs reduces the incidence of cardiomyopathy, device-related complications, and the need for revisions, with a significantly lower risk of long-term reinterventions [4, 9, 11–14]. A 5-year long-term follow-up study of LPs revealed a very low incidence of major complications and system revisions, confirming their safety and reliability in clinical practice. Notably, no cases required LP removal due to infection during the study period, underscoring the advantage of LPs in reducing the risk of device-related infections [15]. An increasing body of research evidence suggests that LPs, as an alternative to traditional TVP, are not only viable but may also be safer in reducing the risk of certain complications. The aforementioned research suggests that complications such as cardiac injury and perforation may arise from the implantation of leadless pacemakers. Patients at risk of bleeding, those who cannot tolerate surgery, and those with abnormalities in the inferior vena cava pathway are contraindicated for leadless pacemaker implantation.

In this case, the patient is receiving long-term treatment with corticosteroids and immunosuppressants, which elevates the risk of opportunistic infections and complicates the therapeutic approach. Moreover, the chronic inflammatory state may impact the body's immune response. EF leads to inflammation of the skin and fascia, subsequently causing hardening that complicates tissue dissection during surgery and increases the difficulty of creating a pacemaker pocket. This hardening enhances the friction between the pacemaker pocket and surrounding tissues, making the skin over the pocket more prone to compression and injury, potentially leading to pressure necrosis

and the risk of device exposure. As the disease advances, patients may develop cardiac fibrosis, which can make it challenging for conventional pacemakers to remain fixed, thus increasing the risk of dislodgement. Given these considerations, we selected a LP for this patient with compromised immune function who might face tissue and structural challenges due to EF. This decision aims to minimize risks while ensuring the patient receives a stable and reliable pacing therapy solution.

Conclusion

This case highlights the complexity of managing EF with third-degree atrioventricular block, especially in patients undergoing long-term corticosteroid and immunosuppressive therapy. The use of a LP in this patient provided an effective therapeutic option, with potential advantages in reducing the risk of infection and device-related complications. However, given the limitations of this case report, including the short follow-up duration and the individualized nature of the patient's condition, we emphasize the need for further research to evaluate the broader application and long-term outcomes of LPs in similar high-risk populations. Our findings suggest that LPs may be a viable alternative for patients with compromised immune function and unique tissue challenges, but individualized assessment remains crucial.

Abbreviations

EF	Eosinophilic fasciitis.
AV	Atrioventricular.
IOM	Institute of Medicine
TVP	Transvenous pacemakers.
LP	Leadless pacemaker.

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Author contributions

H. contributed to patient follow-up, data collection, and manuscript draft. C.is in charge of securing funding and managing the project.C. and Z. were responsible for the development of the methodology and the peer review of the manuscript.S., Y., W. and W. are tasked with data management.All authors have reviewed the manuscript.

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Data availability

All the data regarding the findings are available within the manuscript.

Declarations

Ethics approval and consent to participate

This case report was approved by the Ethics Committee of Lanzhou University Second Hospital. This clinical case report was published with the informed consent of the patient.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests

The authors declare no competing interests.

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