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CLINICAL RESEARCH

Cardiac sarcoidosis: Diagnosis, therapeutic management and prognostic factors

Sarcoïdoses cardiaques : diagnostic, prise en charge thérapeutique et facteurs pronostiques

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KEYWORDS

Cardiac sarcoidosis;
Steroids;
Methotrexate;

Summary

Background. – Cardiac sarcoidosis (CS) is a severe localization, observed mostly in cardiology departments. Despite appropriate cardiological treatment, CS is a potentially life-threatening condition, and accounts for 13–85% of sarcoidosis-related deaths.

Abbreviations: CI, confidence interval; CS, cardiac sarcoidosis; ECG, electrocardiogram; IST, immunosuppressive therapy; MRI, magnetic resonance imaging; PET, positron emission tomography.

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Cyclophosphamide;
Relapse;
Survival rate

Aims. – This retrospective study aimed to give an overview of CS in a non-cardiac unit, to analyse the effect of first-line immunosuppressive treatment on outcome and survival and to evaluate factors associated with relapses.

Methods. – From 534 cases of sarcoidosis, we selected 59 patients with CS according to ‘‘2006 international criteria’’. We performed an in-depth analysis regarding symptoms, physical signs and cardiac investigation results. Patients were followed for a median period of 60 months.

Results. – The median age at cardiac signs was 42 years. Echocardiography abnormalities, isotopic defects and abnormal magnetic resonance imaging findings were observed in 81%, 84% and 92% of patients, respectively. First-line treatment included steroids alone in 24 patients and steroids plus immunosuppressive therapy in 35 patients. Forty-seven (80%) patients recovered; 12 stabilized or worsened. The recovery rate was 75% in the steroids alone group versus 83% in the steroids plus immunosuppressive therapy group. Five (9%) patients died during follow-up, with two deaths attributed to CS. The overall 1- and 5-year survival rates were 98% and 92%, respectively.

Conclusion. – Our series of patients with CS who received steroids alone or combined with immunosuppressive therapy had a good prognosis, with an overall 5-year survival rate of 92%. The recovery rate was 85%, with no significant difference between patients treated with steroids alone or plus immunosuppressive therapy.

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MOTS CLÉS

Sarcoïdose
cardiaque ;
Corticoïdes ;
Méthotrexate ;
Cyclophosphamide ;
Rechute taux de
guérison

Résumé

Contexte. – La sarcoïdose cardiaque est une localisation sévère, le plus souvent prise en charge en cardiologie. Malgré un traitement cardiologique approprié, le pronostic est sombre avec un taux de mortalité de 13 à 85 %.

Objectifs. – Étude rétrospective permettant de : (1) présenter les sarcoïdoses cardiaques en médecine interne ; (2) analyser l’impact des immunosuppresseurs sur le pronostic et la survie.

Méthodes. – Dans une cohorte de 534 sarcoïdoses, 59 patients répondaient aux critères internationaux de 2006. L’analyse a porté sur les signes cliniques et les investigations cardiologiques. Ils ont été suivis pendant une période médiane de 60 mois.

Résultats. – L’âge médian lors des signes cardiaques est de 42 ans. L’échocardiographie, les examens isotopiques, et l’IRM ont été anormaux respectivement dans 81 %, 84 % et 92 % des cas. De première intention, ils ont reçu soit de la prednisone dans 24 cas, soit de la prednisone associée à un immunosuppresseur dans 35 cas. Quatre-vingt pour cent ont évolué favorablement, 12 se sont stabilisés ou aggravés. Le taux de guérison a été de 75 % dans le groupe prednisone seule contre 83 % dans le groupe prednisone plus immunosuppresseur. Cinq patients sont décédés, deux sont en rapport avec la sarcoïdose cardiaque. L’espérance de vie à 1 et 5 ans est respectivement de 98 et 92 %.

Conclusion. – Dans cette série, les patients traités par prednisone seule ou associée à un autre immunosuppresseur ont un bon pronostic avec une espérance de vie à 5 ans de 92 %. Le taux de guérison ne diffère pas entre les deux groupes de traitement.

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Background

Evidence of non-caseating granulomas in affected organs characterizes sarcoidosis, which is described as a systemic chronic granulomatous disease of unknown aetiology. In patients with sarcoidosis, cardiac involvement has been reported in 3–40% of cases in clinical series according to diagnostic criteria and procedures [1–4], and in up to 30% of cases in pathology series [5]. Cardiac sarcoidosis (CS) is a potentially life-threatening condition, and accounts

for 13–25% of sarcoidosis-related deaths in the USA and 85% in Japan [6,7]. Most CS series are published by cardiology departments, with patients presenting predominant cardiac expression and receiving specific cardiac treatment combined, in most cases, with steroids [2,3,8,9]. However, CS is usually observed within the scope of severe multi-visceral sarcoidosis. The aim of the present study was to better characterize CS in patients seen in a non-cardiac unit and its related extracardiac sarcoidosis conditions. We also aimed to analyse the impact of immunosuppressive therapy

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