

REVIEW **OPEN ACCESS**

Atrial Arrhythmias in Cardiac Sarcoidosis—Case Presentation and Systematic Review

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ABSTRACT

Sarcoidosis is a rare idiopathic, multiorgan disease with clinical cardiac involvement in approximately 5% of cases. Diagnosis and management of cardiac sarcoidosis (CS) have mainly focused on impaired LV function, ventricular arrhythmias, and atrio-ventricular conduction disease. Atrial arrhythmias (AA) may be an under-recognized manifestation of sarcoidosis, and the purpose of the study was to perform a systematic review of the current literature, with an emphasis on the prevalence and management of AA in CS. AA occur as a result of direct infiltration of granulomatous disease in the atria or secondary to atrial myopathic changes from pulmonary or ventricular disease. Positron emission tomography (PET) is the modality of choice for surveillance of disease activity in the atria. AA occur frequently, in up to 40% in patients with CS, and may be associated with frequent hospitalization and reduction in quality of life. Moreover, AA may be the predominant cardiac manifestation, and there should be some consideration to include AA as part of future iterations of the diagnostic criteria for CS. Contemporary treatment involves a combination of immunosuppression, anti-arrhythmic therapy, and catheter ablation, but recurrence occurs more frequently than in patients with non-CS AA. Further systematic studies, likely in the form of multi-center observational cohort studies, are required to inform guidelines on the specific management of AA in CS.

1 | Introduction

Sarcoidosis is an idiopathic, multiorgan granulomatous disease with an estimated Caucasian prevalence of 10–40 per 100,000 [1, 2]. The heart is clinically affected in approximately 5% of pulmonary/systemic sarcoidosis cases [3, 4], although post-mortem screening and imaging studies estimate a 20%–70% incidence of silent cardiac disease [3, 5–7].

The diagnosis of cardiac sarcoidosis (CS) is based on histological and clinical findings. The diagnostic guidelines endorsed by

the Japanese Circulation Society are widely used [8]. The Heart Rhythm Society (HRS) has also published a consensus statement on the diagnosis and management of arrhythmias associated with CS [9].

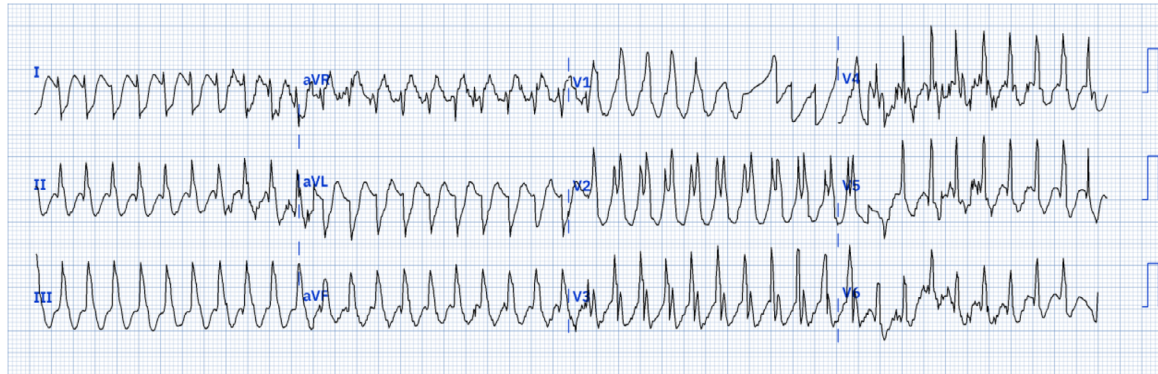
The most recognized cardiac manifestations of sarcoidosis are conduction system abnormalities, heart failure with reduced ejection fraction, ventricular arrhythmias, and sudden cardiac death [2, 10]. However, atrial arrhythmias (AA) may be an under-recognized manifestation of sarcoidosis and are not part of the diagnostic guidelines from the Japanese Circulation Society

Abbreviations: CS, cardiac sarcoidosis; AA, atrial arrhythmias; LGE, late gadolinium enhancement; PET, positron emission tomography.

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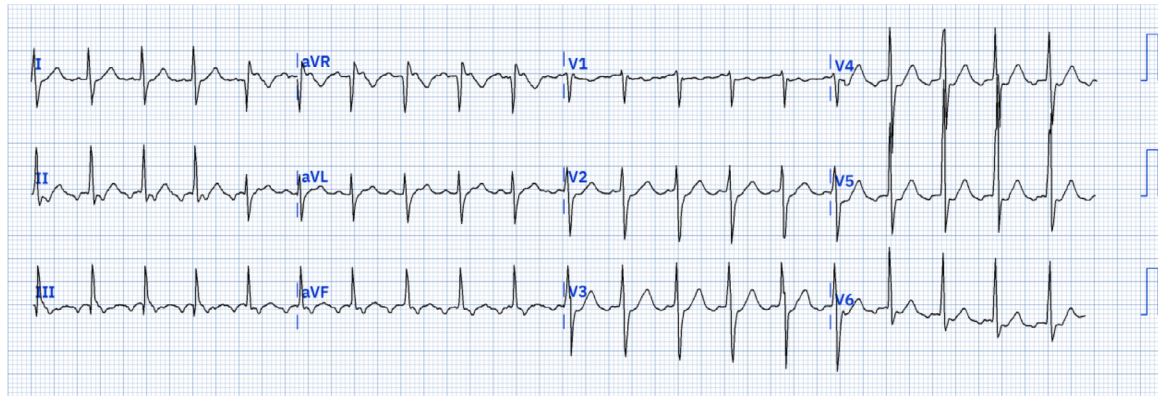


FIGURE 1 | ECGs. (Figure 1a) Wide-QRS complex tachycardia at a rate of 250 bpm. Right-inferior axis with right bundle branch block-type (RBBB) morphology. (Figure 1b) Subsequent ECG (Figure 1b) showing narrow complex tachycardia with an atrial rate of 250 bpm and 2:1 AV conduction. [Color figure can be viewed at [wileyonlinelibrary.com](https://onlinelibrary.wiley.com/doi/10.1111/pace.70062)]

or Heart Rhythm Society, although they represent a “possible” criterion in the World Association of Sarcoidosis and Other Granulomatous Diseases definition (WASOG) [11]. AA may contribute significantly to morbidity in patients with CS.

We present a case study illustrating the practical challenges of managing AA in patients with CS, followed by a systematic review of the current literature, focused on the prevalence and management of AA in CS.

2 | Case Presentation

A Caucasian male first presented at the age of 31 years old with palpitations and presyncope. He had been diagnosed with pulmonary sarcoidosis 2 years prior to the onset of cardiac symptoms, based on biopsy of a pulmonary nodule. He was taking Prednisone 25 mg OD. Cardiac MRI (CMR) at the time of diagnosis was normal.

Initial ECG showed a wide complex tachycardia (WCT) (Figure 1a,b) at a rate of 250 bpm, with typical right bundle branch block (RBBB) and right inferior axis. Subsequent ECG showed a narrow complex rhythm with 2:1 AV block. The atrial rate during the second ECG was also 250 bpm, suggesting that the original WCT was atrial flutter with aberrant conduction.

Repeat CMR (Figure 2) showed normal LV function and a small area of patch late gadolinium enhancement (LGE) in the basal posterior LV myocardium. There was mild biatrial dilation and a suggestion of possible LGE in the right atrium.

Cardiac positron emission tomography (PET) imaging suggested mildly increased uptake in the right atrium (Figure 2).

An electrophysiology study was performed. Anterograde AV conduction was normal with an HV interval of 40 ms. Ventricular tachycardia was not inducible; however, several different atrial flutters and atrial fibrillation were observed. Voltage mapping of the right atrium (RA) was markedly abnormal with extensive scarring, slow conduction, and fractionated electrograms (EGMs) in the lateral and superior RA (Figure 3a). Empirical cavo-tricuspid isthmus ablation was performed. The patient was discharged on Sotalol, Apixaban, and Prednisone.

Over the next 12 months, the patient remained highly symptomatic of palpitations requiring 10 admissions to the hospital for cardioversion of AA. Medical management was maximized with an increased dose of Sotalol and the addition of Methotrexate as a steroid-sparing agent.

A second EP study was then performed (Figure 3b). Wide antral circumferential radiofrequency (RF) ablation of the pulmonary veins was performed to treat paroxysmal AF. Left atrial voltage

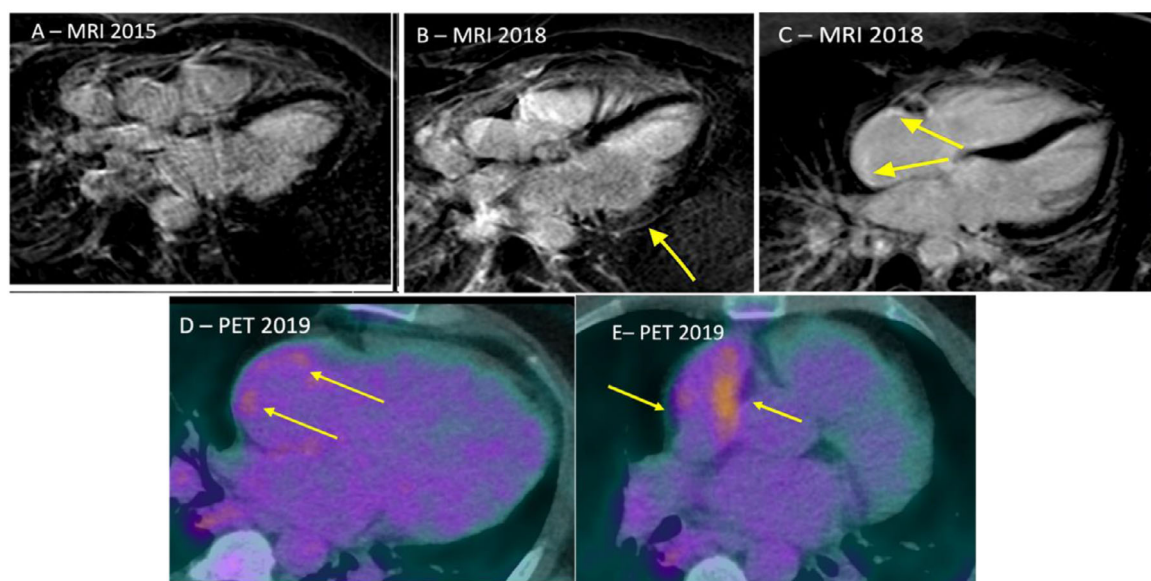


FIGURE 2 | Evolution of cardiac imaging—Cardiac MRI and PET. (a–c) Cardiac MRI (LVOT view). (a) Normal MRI in 2015. (b) Repeat MRI in 2018 showing patchy late gadolinium enhancement (LGE) of the basal lateral LV (arrow). (c) Possible LGE in the right atrium (arrows). (d, e) Axial fused PET-CT images in 2019. Uptake in the RA wall was mildly elevated (SUVmax ranging from 3.2 to 4.6) compared to the blood pool (SUVmax 3.0) as well as the LV myocardium where there was complete suppression of background FDG uptake. [Color figure can be viewed at wileyonlinelibrary.com]

was normal. Atrial flutter was easily inducible; pacing manoeuvres confirmed re-entry related to the scar in the lateral right atrium. Extensive ablation was performed in this region, and no further tachycardia could be induced at the end of the procedure.

Repeat PET scan (22 months after the first, performed on immunosuppression and in sinus rhythm) showed no active cardiac inflammation. Borderline uptake in the lateral right atrium was suggested but this was interpreted as either the effect of recent ablation or artefact related to suboptimal adherence to dietary protocol.

Three months later, the patient re-presented with rapidly conducted atrial flutter, and he had developed symptoms of depression related to his repeated admissions to the hospital. He was taken to the EP lab for a third time (Figure 3c). Atrial flutter at a rate of 160 bpm was induced. High-density activation mapping and entrainment mapping confirmed the recurrence of scar-related re-entrant arrhythmia. RF ablation was performed in residual areas of slow conduction within the scar and extended from the scar to the IVC and from the scar to the anterior tricuspid annulus (Figure 3d,e).

The patient was weaned off Sotalol and remains on immunosuppression. He has had no further atrial arrhythmia after 60 months of follow-up.

3 | Systematic Review

3.1 | Methods

This systematic review was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) principles and 2020 Statement [12].

A systematic search of articles using PubMed, EMBASE, Scopus Web of Science, and Cochrane Database of Systematic reviews and Cochrane Library was performed using the following search term “Cardiac Sarcoid* AND (arrhythmia OR tachycardia OR tachyarrhythmia OR atrial)”. The search was supplemented by manually reviewing bibliographies of published studies. We also performed an additional search using the search term “sarcoid myocarditis” in place of “Cardiac Sarcoid*” to ensure inclusion of CS cases that may have been coded as sarcoid myocarditis.

Articles not in the English language or where the full text was not available were excluded.

Each search result was individually reviewed by one of the authors for relevance to the topic of AA in CS.

3.2 | Results

Three hundred and fifty-three records were returned from the initial search criteria. Three hundred and thirty-seven records were screened for suitability, with 62 papers deemed relevant to the topic (Figure 4). Of the included manuscripts, there were 19 cohort studies, 20 case reports/case series, 19 reviews, two editorials, and two consensus guidelines. There were no systematic reviews on this subject in the literature. There were no clinical trials comparing different treatment options.

3.3 | Prevalence of Atrial Arrhythmias in CS

The prevalence of AA in sarcoidosis was based on two types of studies, those analyzing large administrative databases and smaller cohort studies with patient-level data (Table 1). Reports

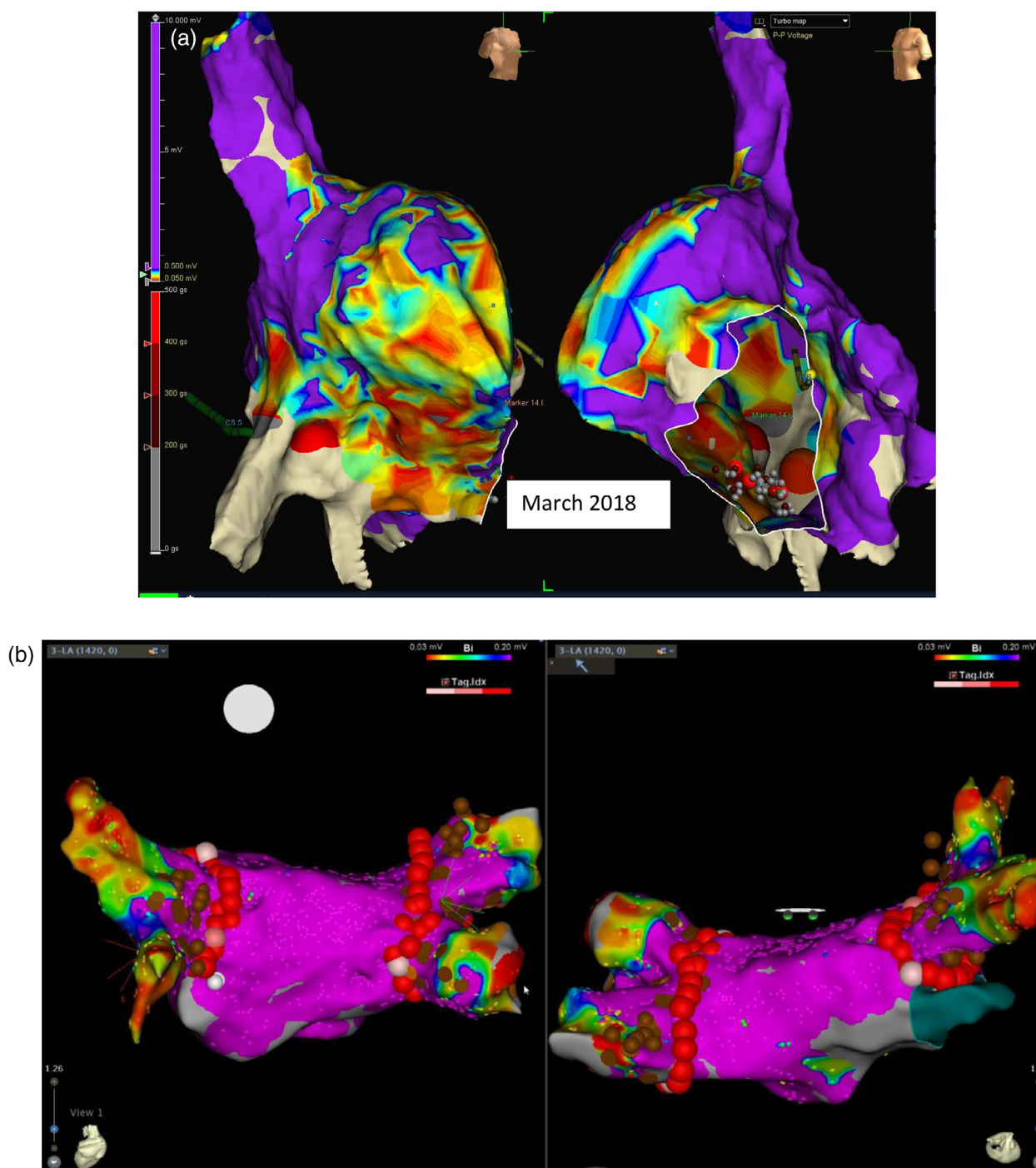


FIGURE 3 | Electrophysiology study and mapping. (Figure 3a) First EP study. Electro-anatomical map (Abbott Ensight NavX) of the right atrium showing diffuse low voltage in the lateral aspect of the right atrium. Right-anterior-oblique (left panel) and left-anterior-oblique projections (right panel). Purple areas indicate normal myocardial voltage (>0.5 mV) while red areas indicate very low voltage (<0.05 mV). Ablation was performed at the cavo-tricuspid isthmus as indicated by the grey and red balls. (Figure 3b) Second EP study with ablation. Electro-anatomical map (J&J CARTO) of left atrium. Posterior view (left panel) and superior view (right panel). The bipolar voltage of the left atrial was normal (>0.5 mV; purple). Circumferential pulmonary vein isolation was performed using radiofrequency ablation (lesions shown in red). (Figure 3c) Repeat voltage mapping of the right atrium (J&J CARTO) showed widespread patchy scar in the lateral right atrium. Right-anterior-oblique view. Please note, voltage scale in Figure 3a (Abbott Ensight NavX) and Figure 3c (J&J CARTO) are slightly different. (Figure 3d) Radiofrequency ablation (J&J CARTO) performed in areas with borderline voltage (0.03–0.2 mV) within lateral scar of right atrium and extended to inferior vena cava and lateral tricuspid annulus (red balls). Right-anterior-oblique (left panel) and inferior left-anterior-oblique projections (right panel). (Figure 3e) Third EP study for recurrent atrial flutter. High density mapping (Abbott Ensight NavX) of right atrium again showing reduced voltage in the right atrium (with unexcitable scar shown in grey; voltage <0.037 mV; unable to be captured with pacing). Further widespread ablation was performed from area of low voltage to the IVC and anteriorly to the tricuspid annulus. [Color figure can be viewed at [wileyonlinelibrary.com](https://onlinelibrary.wiley.com)]

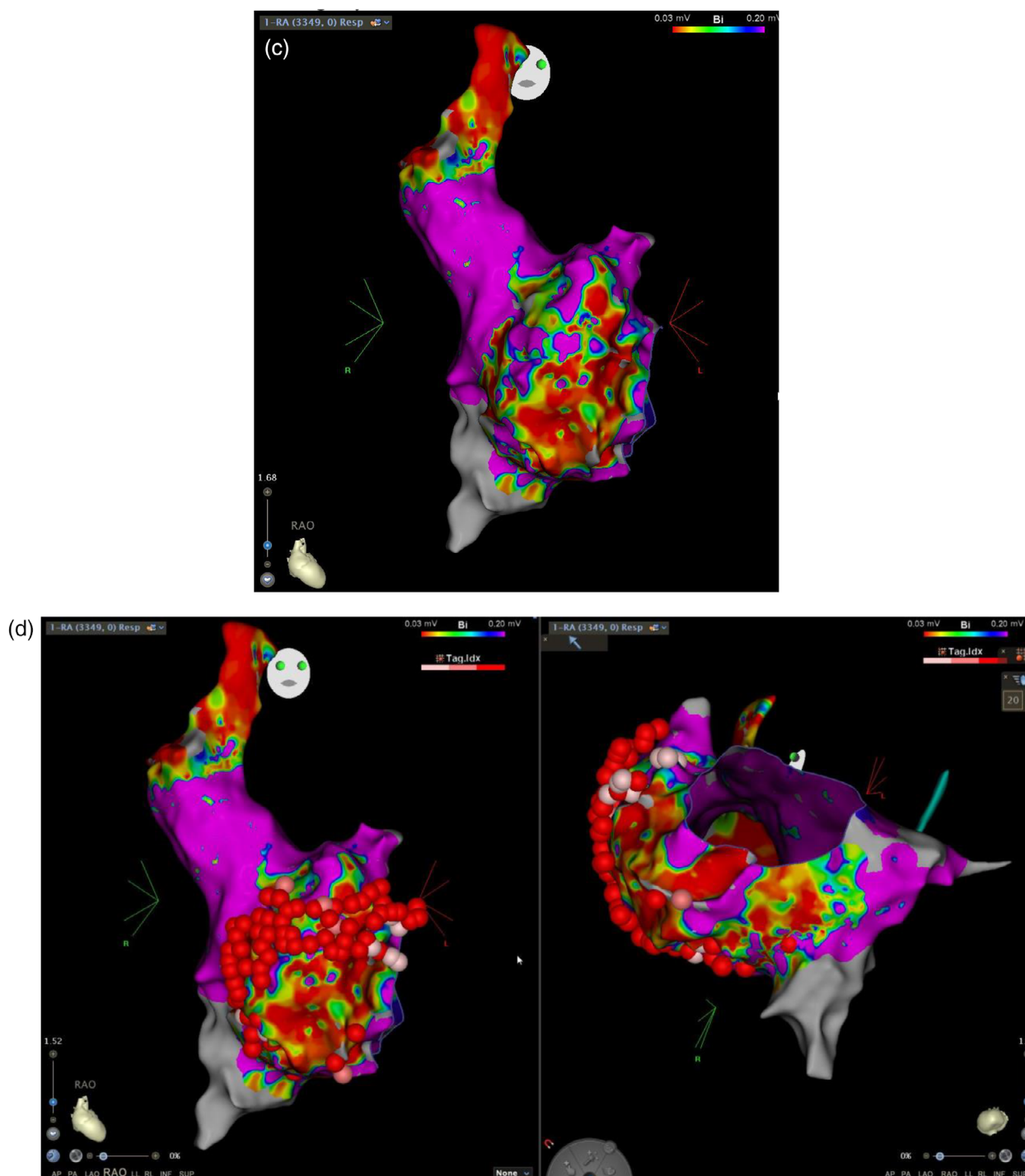


FIGURE 3 | (Continued)

were limited by variable definitions, and symptom status was often not reported.

Desai et al. interrogated the US National Inpatient database and reviewed almost 370,000 sarcoid-related admissions; 11% of the admissions were complicated by AF [13]. Gonuguntla et al. [14] also found an 18% rate of AF in over 9000 patients hospitalized for sarcoidosis over a 4-year period in the USA.

Several case series have examined the specific prevalence of AA in CS patients. Viles-Gonzalez et al. assessed 100 patients

with biopsy-proven CS for AA [15]. The prevalence of AA was 32%, with atrial fibrillation being the most common (18%). Of interest, all but four patients were asymptomatic. A sub-study of the CHASM-CS trial (cardiac sarcoidosis multicenter prospective cohort study) specifically reported on CS patients with implanted devices [16]. Device-detected AA (defined as atrial high-rate episodes >190 bpm) was assessed prospectively for an average of 4 years. A third of enrolled patients (11/33) developed AA over this period. Burden was generally low (as defined by duration and frequency of episodes), with only two patients having episodes >24 h duration. The mechanism of arrhythmias was not reported.

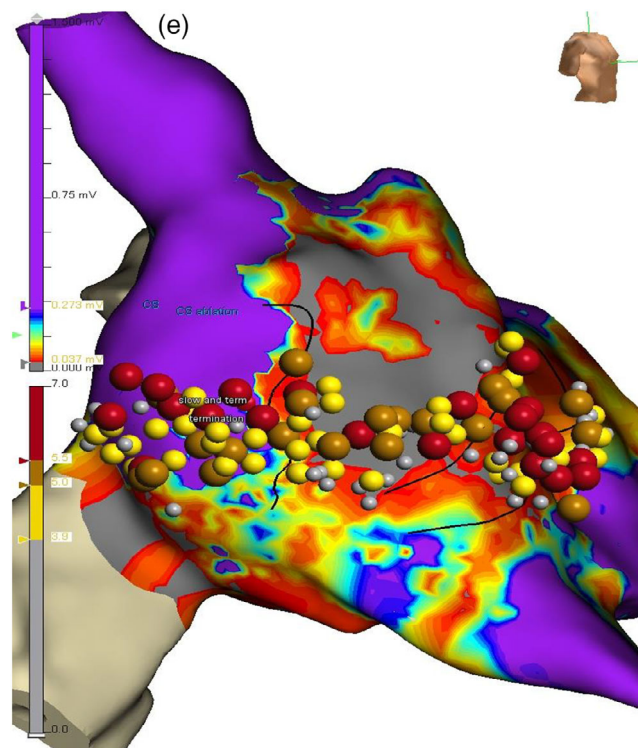


FIGURE 3 | (Continued)

TABLE 1 | Prevalence of atrial arrhythmias in cardiac sarcoid.

Study	Population studied	Key findings
Desai et al. [13]	US National Inpatient database 2010–2014	369,285 sarcoidosis-related admissions. AF complicated 11% of the admissions.
Gonuguntla et al. [14]	US Nationwide Inpatient from 2010 to 2014	9063 patients with CS (53.11 ± 11.28 years; men 51.8% and black 52.5%). 18% rate of AF in patients hospitalized for sarcoidosis over 4 years.
Viles-Gonzalez et al. [15]	100 patients with CS	100 patients with biopsy-proven cardiac sarcoidosis. Prevalence of atrial arrhythmias (AA) was 32%, with AF being the most common (18%). Majority of patients asymptomatic.
CHASM-CS trial [16]	33 CS patients with implanted devices	33% patients developed atrial arrhythmias. Only two patients had episodes >24 h duration. 100% reduction in burden after CS was successfully treated.
Cain et al. [17]	192 patients with extracardiac sarcoidosis	CS patients were evaluated by CMR imaging from January 2007 to July 2011. Atrial arrhythmias were documented in 16 patients (36%) with myocardial LGE and 11 patients (12%) without myocardial LGE ($p = 0.002$).
Yodogawa et al. [18]	62 CS patients who underwent PET/CT	25 patients (40.3%) had AA—4 patients had atrial tachycardia (AT), 23 patients had AF. Eighteen patients with AA had atrial FDG uptake on PET/CT, whereas 14 patients without AA had atrial FDG uptake (72.0% vs. 37.8%, $p = 0.017$).
Nordenswan et al. [19]	351 CS patients in Finland, compared with 28 patients with Giant Cell myocarditis.	1% CS patients had AA. 43% prevalence of AVB in CS patients.

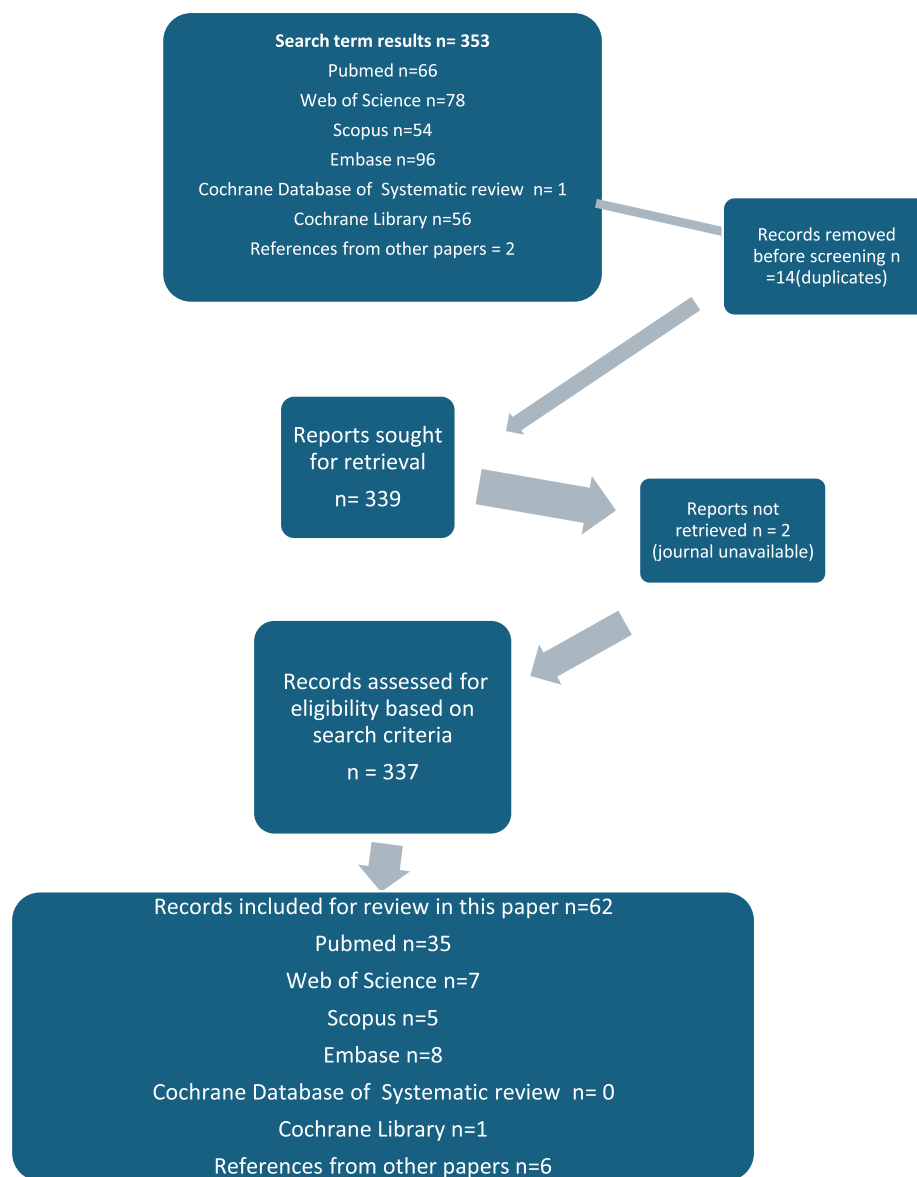


FIGURE 4 | PRISMA method search results. [Color figure can be viewed at wileyonlinelibrary.com]

Cain et al. used a combined cardiac MRI and electrophysiology approach to assess for atrial arrhythmia in 135 patients with known, biopsy-proven, extracardiac sarcoidosis [17]. In patients with LGE consistent with CS, 36% had AA (vs. 12% in patients without overt cardiac involvement/LGE). Interestingly, atrial arrhythmia was more frequently documented than ventricular arrhythmia in the CS group.

Yodogawa and colleagues [18] reported a 40% prevalence of AA in a cohort of 62 CS patients. FDG uptake on PET/CT was strongly associated with the presence of AA, with the majority of cases due to AF. Several small cohort studies (including use of Holter monitoring) found a prevalence of AA of 20%–25% [20–22].

In summary, there is a moderate burden of AA in patients with sarcoidosis (10%–20%), with higher incidence in patients with known cardiac involvement (30%–40%). However, AA are an unusual initial presentation of CS and thus do not currently

feature in any of the diagnostic guidelines [8, 9] or screening tools for CS [23].

3.4 | Factors Associated With AA in Cardiac Sarcoidosis

Studies have identified several factors that predispose to AA in the sarcoid-affected heart. In particular, abnormalities on imaging studies appeared to be strong predictors of AA.

Although the detection of atrial scar is technically challenging in CMR, Cain and colleagues [17] reported that the presence of ventricular myocardial LGE was independently associated with AA. Indeed, AAs were three times more prevalent in the sarcoidosis patients with cardiac involvement on MRI.

Atrial involvement can be inferred by volumetric analysis, and more specifically by atrial uptake of 18F-fluorodeoxyglucose

(FDG) on PET. Atrial enlargement has been reported by several authors as independently associated with atrial arrhythmia in CS [15, 24]. Atrial dilatation may be caused by direct granuloma deposition in the left atrium [7, 25, 26] or secondary to LV systolic or diastolic dysfunction and pulmonary hypertension [27]. These mechanisms are shared with several of the inflammatory cardiomyopathies [28].

Atrial uptake of FDG on PET scanning has been shown to be strongly associated with the presence of AA in CS. On multivariate analysis, Yodogawa et al. demonstrated an odds ratio of 7.23 (CI 1.01–1.31, p 0.04) for AA in the presence of atrial FDG uptake [18]. In a substudy of CHASM-CS [16], AAs were more common in the presence of atrial FDG—27% (3/11) versus 4.5% (1/22 patients). In the same paper, there was no association between ventricular FDG uptake and AAs. The 5-year incidence of atrial fibrillation was significantly higher in CS patients with atrial FDG-uptake than those without (55% vs. 18% p < 0.001) [29].

The specific interplay between AF, atrial remodeling, and inflammation on imaging was examined by Habibi et al. [30]. They conducted a small, retrospective study of 50 patients with CS who had undergone both PET and MRI scanning. CS-associated AF was correlated with larger LA volume, reduced LA ejection and strain on MRI, and a higher SUV on PET. All patients with AF had atrial FDG avidity. Chik et al. [31] also demonstrated the use of these complementary modalities in a case report of CS dominated by AA, notably sinus arrest, AF, and typical atrial flutter. Cardiac MRI demonstrated diffuse LGE in the right atrium and both ventricles, corresponding with biatrial hypermetabolism on PET-CT. High-density electro-anatomical mapping at the EP study demonstrated a low-voltage scar in the posterior right atrium.

Viles-Gonzalez et al. also identified systemic hypertension and diastolic dysfunction as additional clinical predictors [15]. The possible influence of racial background was highlighted in one cohort study reporting that the incidence of CS-related AF was lower in white Americans than African Americans [32].

3.5 | Types of AA

3.5.1 | Atrial Fibrillation

Sama and colleagues [33] performed a comprehensive meta-analysis in 2023 on the prevalence and incidence of AF in CS. They included eight studies (totaling 978 patients) directly reporting prevalence, and nine studies (545 patients) from which incidence could be inferred. Prevalence was estimated at 23% with over 80% of affected patients having paroxysmal AF. Overall, the pooled incidence was 10.6%. As would be expected, the study showed a higher incidence of AF with increasing length of follow-up. Other work has suggested a lower prevalence of around 18% [15].

Atrial fibrillation is probably the most common arrhythmia in CS patients. A large registry-based long-term outcomes study from Denmark showed patients with CS had a 10-year risk of AF of 3.4% and a hazard ratio of 3.25, when compared to matched controls [34]. In hospitalized patients with CS, rates of atrial fibrillation were higher in females [35], and mortality was higher

in those with AF than those without [13]. However, in a study of patients presenting for catheter ablation of AF, CS was rare—only two patients of 1574 undergoing ablation for AF were found to have CS (based on pre-ablation cardiac CT or MRI scans) [36]. In contrast, a cohort study by Kumar et al. [37] also examined patients presenting with AF at their center over 3 years. They carried out PET scans in 45 of 153 patients who had lone-AF under the age of 65 years. Of these 53% (24 patients) had atrial uptake consistent with atrial inflammation, and of these 15 patients (63%) had biopsy-proven atrial myocarditis. The final diagnosis was CS in 12 of these 15 patients. The authors conclude that atrial myocarditis is a prominent cause of early onset, lone and recurrent AA; the most common cause of which is CS, and it should be considered in these type of cases. It is likely that the discrepancy between studies is related to the higher sensitivity of the PET scan for identifying atrial inflammation.

The etiology of CS-associated AF differs from non-CS associated AF. In the sarcoid heart, AF is related to atrial granulomas with associated inflammation rather than scarring [24, 38, 39]. This process has been confirmed on post-mortem examination of the CS heart [40]. Secondary atrial dilatation and elevated atrial end-diastolic pressures from pulmonary involvement and LV involvement also play a role [24].

3.5.2 | Non-AF Atrial Arrhythmia

Electrophysiological assessment of non-AF AA in CS found a predominance of abnormal automaticity as the arrhythmia mechanism—47% automaticity, 42% re-entrant, and 11% trigger activity (15 patients studied) [41]. Patchy atrial scar, as demonstrated by low voltage areas on electroanatomical mapping (EAM) and FDG-quiet regions on PET scanning, can predispose to macro-re-entry circuits and atrial flutter [25, 42–44]. Both typical [31] and atypical flutters [43, 45] have been reported, including rare cases of biatrial flutter related to extensive scarring of the atrial septum [43].

Two cases of sinus arrest have been reported, presumably related to infiltration of the sinus node. In a report of a 42-year-old with sinus arrest and severely prolonged sinus node recovery time, there was evidence of scarring in the anatomical region of the sinoatrial nodal on EAM and cardiac MRI [31]. Kim and colleagues [46] also reported a case of atrial standstill as the first presentation of clinical cardiac sarcoid in a young patient. The right atrium was electrically silent and had severe fibrosis on biopsy. LGE was evident in both atria.

3.6 | Management of AA in CS

Existing literature and guidelines have largely focused on the management of conduction system disease and ventricular arrhythmias in sarcoidosis. Heart block may be responsive to immunosuppression (in ~50% cases), and it has a Class IIa recommendation in the HRS guidelines [2, 5, 9, 47, 48]. However, the natural history of CS is unpredictable with a variable relapse rate. Device therapy is recommended in cases of conduction system disease and ventricular arrhythmia (Class I-IIa). Imaging

may assist in selecting a location for pacing leads away from sites with known inflammation or scar.

3.6.1 | Immunosuppression

Immunosuppression may have a role in lowering the burden of AA [6, 22]. However, all of the evidence is based on case reports/series rather than randomized trials [2, 31, 45, 49–53, 54–57]. Hence, current guidelines do not formally recommend immunosuppression but state that a trial of immunosuppression in AF may be useful [9]. There is some emerging data that treatment with corticosteroids increases ventricular arrhythmia burden in some patients [58]. This is also plausible for AA, as steroids can increase blood pressure and cause fluid retention, but further studies would be required to evaluate the potential association between steroid therapy and AA.

3.6.2 | Anti-Arrhythmic Drugs (AAD)

Similarly, there are no randomized data or specific guidance recommendations on the anti-arrhythmic management for CS-related AA. Class 1 anti-arrhythmic drugs should generally be avoided in patients with CS-associated ventricular myocardial scarring and/or LV dysfunction [9]. Otherwise, the standard rate and rhythm control medications, including amiodarone, may be used.

3.6.3 | Anticoagulation

Risk stratification for thromboembolism/stroke in CS patients with atrial fibrillation should be completed using standard guidelines for non-valvular AF [9]. There is no evidence that patients with CS are at a higher risk compared to non-CS patients [9, 59].

3.6.4 | Ablation

The results of catheter ablation for AA in CS were reported by Willner et al. in 2014 [60] and Willy et al. in 2019 [61]. Willner et al. reported a cohort of 100 consecutive CS patients. Amongst 32 patients with symptomatic AAs, nine underwent EP study and ablation. Left AA were more common than right, and ablation was performed for atrial fibrillation, typical and atypical flutter. During the follow-up of 4 years, two patients experienced recurrence.

Willy et al. reported on the results of 16 catheter ablations in 37 patients with CS related AA [61]. The predominant arrhythmia was atrial flutter, and seven patients underwent RF ablation of cavo-tricuspid isthmus-dependent atrial flutter. Five patients underwent pulmonary vein isolations (PVI) for atrial fibrillation, all using cryoablation. During follow-up of 2.5 years, two patients underwent repeat ablation—one for flutter and one for AF. A hybrid surgical-percutaneous approach has also been described with surgical MAZE ablation for CS-related AF and percutaneous catheter ablation for concomitant refractory atypical atrial flutter [62].

At this stage, there is limited data on whether catheter ablation is as effective in CS-associated AF and whether the standard PVI strategy is the optimal strategy [24, 59]. Data from a Mount Sinai PVI cohort found that 80% of CS patients with AF had the usual pulmonary vein triggers and were AF free after 18 months; however, the cohort was small, comprising only five patients [60].

3.6.5 | Surveillance

In addition to standard clinical follow-up, FDG-PET has also been shown to be useful in disease monitoring in patients with arrhythmic CS [63]. Low voltage on EP mapping has been shown to correlate with areas of high avidity on PET. Kersey et al. [64] used the modality for tracking disease progression and response to immunosuppression in a quantitative manner using standard uptake values (SUV), a measure of glucose avidity and inflammation. Intuitively, FDG-PET may be useful for monitoring disease activity in the atria.

4 | Discussion

AA are an under-appreciated manifestation of sarcoidosis. Based on observational reports, the frequency of AA is actually higher than ventricular arrhythmias [17], estimated as 10%–20% in unselected patients with sarcoidosis and up to 30%–40% in selected patients with known cardiac involvement [15]. Although many patients may have asymptomatic AAs (often detected on device surveillance), some cases may be associated with significant morbidity related to frequent hospitalization and associated reduction in quality of life [65] and even depression, as demonstrated in the present case. AA are associated with increased 2-year mortality in CS patients with an ICD in situ.

CS-related AA may be related to active inflammation or established atrial scarring, resulting in a spectrum of AAs, including AF and both non-re-entrant and re-entrant AA. Either atria may be involved, and there may be additional contribution from LV dysfunction and pulmonary hypertension [9].

Atrial involvement is usually inferred by the presence of atrial dilatation on echocardiography [14, 24] and/or LGE in the ventricular myocardium on CMR [17]. Inflammation specifically affecting the atria may be detected by FDG-PET [22] and correlate with low voltage and abnormal electrophysiological substrate in the EP study [31].

However, the thin walls of the atrium make imaging assessment challenging for both LGE on CMRI and PET uptake. Moreover, it should be noted that AA in of itself can cause a mild increase in FDG-PET uptake [66–68]. Hence, it is important to look at the whole-body PET scan and to correlate atrial findings across imaging modalities. LGE on cardiac MRI is particularly useful in this regard [29]. Atrial imaging should also be carefully interpreted in order to differentiate true atrial wall enhancement from stagnant blood along the endocardium. For identification of abnormal atrial FDG uptake and atrial inflammation, a target-to-background ratio of ≥ 1.5 is recommended [29]. ECG gating of images can help in correctly localizing the uptake to the atria. Finally, caution is required in inferring the presence of tissue

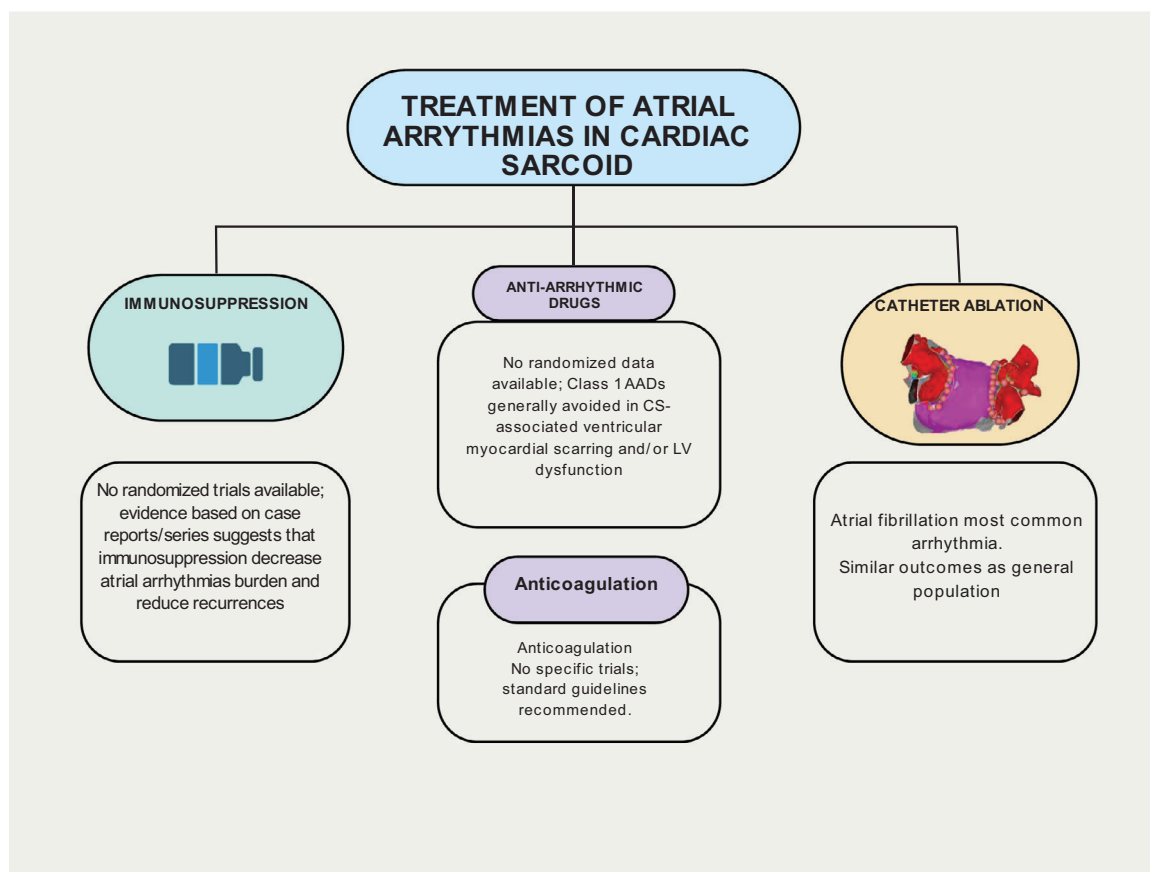


FIGURE 5 | Central illustration. Treatment of atrial arrhythmias in cardiac sarcoid. [Color figure can be viewed at [wileyonlinelibrary.com](https://onlinelibrary.wiley.com/doi/10.1111/pace.70062)]

pathology (i.e., granulomatous inflammation and/or scarring) based on abnormal imaging findings such as FDG uptake, acknowledging that atrial granulomatous disease has been rarely demonstrated in autopsy studies to date [69].

Data and guidelines informing the specific management of AA are limited in comparison with conduction system disease or ventricular arrhythmia in CS [9]. Management of AA in CS is multi-faceted, as summarized in Figure 5.

Assuming a common inflammatory etiology, the utility of immunosuppression for AA is largely extrapolated from reports of steroid-responsive conduction system disease [47] and/or ventricular arrhythmia. Immunosuppressive treatment may be empirical or based on the finding of atrial FDG-avidity on PET, with the latter allowing for quantitative surveillance during follow-up [63, 64]. Adjunctive measures include anti-arrhythmic therapy, device therapy (to prevent bradycardia), and catheter ablation [9]. However, data for these interventions are limited, and recurrence may occur, as demonstrated in the present case. Specifically, it is not known whether the recurrence of AA following catheter ablation in CS is due to a more resistant substrate or progression of disease during follow-up.

5 | Conclusions

AA occur frequently in patients with CS. They may be the predominant cardiac manifestation and be associated with sig-

nificant morbidity. As such, there should be some consideration to include AA as part of future iterations of diagnostic criteria for CS. Contemporary treatment involves a combination of immunosuppression, anti-arrhythmic therapy, and catheter ablation, but recurrence occurs more frequently than in patients with non-CS AA. Further systematic studies, likely in the form of multi-center observational cohort studies, are required to inform guidelines on the specific management of AA in CS.

Author Contributions

All authors were involved in the clinical management of the patient in the case study. **Emily C. Hodkinson**: literature search, drafting and re-drafting manuscript, coordination of paper, figure production, final approval of manuscript. **Luis Quininir**: literature search, reviewing manuscript, figure and table production, final approval of manuscript. **Edward Hsiao**: providing PET images, reviewing manuscript, final approval of manuscript. **Rajesh Puranik**: providing MRI images, reviewing manuscript, final approval of manuscript. **Raymond W. Sy**: Conception and design of paper, reviewing manuscript, final approval of manuscript.

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Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

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