Corticosteroids in Sarcoidosis

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KEYWORDS

• Sarcoidosis • Corticosteroids • Treatment • Toxicity

KEY POINTS

- Corticosteroids are almost universally effective for the treatment of sarcoidosis. The treatment of sarcoidosis is indicated if the disease causes a dangerous health situation or significantly impairs the patient's quality of life.
- Treatment should not be based on biomarkers of active granulomatous inflammation.
- Pulmonary sarcoidosis can usually be adequately treated with modest doses of corticosteroids.
- It is unusual for patients to be refractory to corticosteroid therapy. Alternative medications are almost exclusively used because of the frequent development of corticosteroid toxicity.

INTRODUCTION

Corticosteroids are considered the drug of choice for the treatment of almost all forms of sarcoidosis. Nonetheless, there is considerable controversy and confusion concerning the use of corticosteroids in the management of sarcoidosis for several reasons. First, sarcoidosis may be a self-limiting disease that may spontaneously remit and/or never cause significant clinical problems. In such patients, toxicity from corticosteroid treatment may cause more harm than that caused by the natural course of the disease. Second, the optimal dose of corticosteroids has not been established for sarcoidosis. Third, the duration of therapy depends on the natural course of the disease, which is highly variable and often unpredictable. Fourth, the indications for adding a corticosteroid-sparing agent have not been standardized. This article discusses an approach to corticosteroid therapy in sarcoidosis based on the available clinical data plus our understanding of the disease.

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INDICATIONS TO TREAT SARCOIDOSIS

There are 2 indications for the treatment of sarcoidosis: (1) the development of a dangerous health situation; and (2) significant worsening of quality of life. **Table 1** lists several situations of danger resulting from sarcoidosis. Note that many of these relate to the development of fibrosis, which, unlike active granulomatous inflammation, is not corticosteroid responsive. Furthermore, most of the entities listed in **Table 1** are rare manifestations of sarcoidosis. Therefore, the overwhelmingly most common reason to treat sarcoidosis is for significant worsening of quality of life. **Fig. 1** shows the processes leading to significant quality-of-life impairment in sarcoidosis.[1] First, granulomatous inflammation occurs. This inflammation may not lead to any physiologic disturbance or the development of significant symptoms. For example, patients with pulmonary sarcoidosis who have bilateral hilar adenopathy and a normal lung parenchyma (stage I radiograph) often have no pulmonary symptoms, normal pulmonary function, and a benign clinical course.[2,3] Even if sarcoidosis causes a physiologic disturbance, it may have no significant clinical consequences and cause minimal to no symptoms.[4,5] Therefore, markers of active granulomatous inflammation, such as increased angiotensin-converting enzyme levels, pulmonary opacities on lung imaging, and positive PET scans, are not indications for treatment. Neither are the asymptomatic minor pulmonary function abnormalities that are very typically observed.[4,5] Note that the presence of quality-of-life impairment is a necessary but insufficient requirement for the treatment of sarcoidosis because (1) pulmonary symptoms may be caused by an alternative process, or (2) the quality-of-life impairment may be the result of previous granulomatous inflammation resulting in fibrosis that does not respond to antigranulomatous therapy.[6] Identifying biomarkers of granulomatous inflammation and physiologic abnormalities may provide useful evidence to support

Table 1
Dangerous health situations from sarcoidosis

Conditions	Estimated Frequency (%)	Corticosteroid Responsive
Sudden death, severe arrhythmia, severe left ventricular dysfunction from cardiac sarcoidosis	1	Yes
End-stage fibrocystic sarcoidosis (FVC<50% of predicted)	<5	No or minimally
Sarcoidosis-associated pulmonary hypertension	5	No
Optic neuritis	<5	Yes
Severe neurosarcoidosis	1	Yes
Hemoptysis from pulmonary mycetoma	<1	No
Vitamin D dysregulation causing renal failure, severe nephrolithiasis	<1	Yes
Upper airway obstruction	<1	Usually surgical resection is required if airway obstruction is critical
Endobronchial airway obstruction	<1	Usually not (significant fibrosis present)

Abbreviation: FVC, forced vital capacity.