

CASE REPORT

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## Sarcoid myositis with muscle weakness as a presenting symptom

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**Abstract** A 54-year-old woman complaining of muscle weakness and weight loss was admitted to our hospital with suspected polymyositis. Muscle biopsy revealed Langhans-type giant cells and noncaseating granulomas. Therefore, sarcoid myositis was diagnosed. The patient was treated with prednisolone, and the symptoms improved gradually. Generally, sarcoidosis is identified clinically in patients with foggy vision or mediastinal lymphadenopathy, but muscular weakness may be an infrequently observed initial symptom. Sarcoidosis should be considered in the differential diagnosis of polymyositis.

**Key words** Acute sarcoid myositis · Polymyositis · Prednisolone · Sarcoidosis

### Introduction

Sarcoidosis is a disease of unknown origin, characterized by granulomas in multiple organ systems. Generally, it affects the hilar lymph nodes bilaterally, eyes, and heart. Muscular symptoms do occur, but they appear occasionally as complications and only rarely as main symptoms. We report a case of sarcoidosis in which polymyositis was first suspected because of acute muscle weakness, a high creatine kinase (CK) level, and no hilar lymph node enlargement on chest X-ray examination. Acute sarcoid myositis was diagnosed on the basis of muscle biopsy.

### Case report

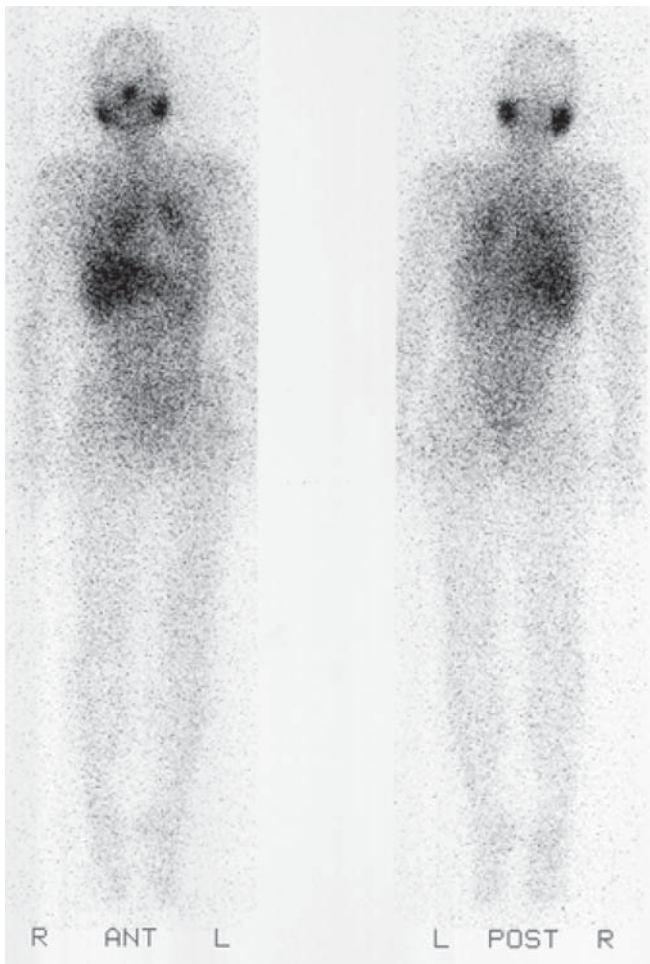
A 54-year-old woman, born and living in Ibaraki Prefecture, Japan, began to complain of impaired speech, general fatigue, and weight loss in mid-February 2005. In the beginning of March, she could not stand from a squatting position. She visited a clinic, and a high CK level was noted. She reported foggy vision beginning in May. Because the CK elevation did not improve, she consulted a health center on May 2. Chest computed tomography (CT) showed multiple, small nodular shadows. The patient was admitted to our hospital on May 9. Skin rash, lymphadenopathy, dry eye, and dry mouth were not found. Mild dysphasia was found, along with symmetrical proximal muscle weakness in her upper and lower extremities; the manual muscle testing score was 3/5 for the proximal muscles and 4/5 for the distal muscles. Aspirate aminotransferase, alanine aminotransferase, lactate dehydrogenase, CK, aldorase, and myoglobin levels were elevated at 168 IU/l (normal 8–40 IU/l), 247 IU/l (normal 4–40 IU/l), 1073 IU/l (normal 124–232 IU/l), 2241 U/l (normal 40–180 IU/l) (BB 0%, ALB 1%, MB 14%, MM 85%), 69.6 U/l (normal 2.7–7.5 U/l), and 2382.0 ng/ml (normal 0–60 ng/ml), respectively. Serum total protein, albumin, calcium, free T3, free T4, and thyroid stimulating hormone (TSH) levels were 7.2 g/dl (normal 6.5–8.2 g/dl), 3.4 g/dl (normal 4.2–5.2 g/dl), 10.6 mg/dl (normal 8.7–11.0 mg/dl), 6.2 pg/ml (normal 2.1–3.8 pg/ml), 1.39 ng/dl (normal 0.82–1.63 ng/dl), and 3.41  $\mu$ U/ml (normal 0.38–4.31  $\mu$ U/ml), respectively. Other laboratory values were within normal limits. Serum immunoglobulin G was slightly increased at 1764 mg/dl. The results of antinuclear antibody, anti-Jo-1 antibody, anti-SS-A antibody, and anti-SS-B antibody tests were negative, and the result of anticytoplasmic antibody test was positive. Arterial blood gas values were normal (pH 7.421, PaO<sub>2</sub> 78.7 torr, PaCO<sub>2</sub> 42.6 torr). Electrocardiographic examination showed a pulse rate of 75 beats per minute and regular sinus rhythm. Chest X-ray did not reveal an abnormal shadow. Chest CT revealed the enlargement of the mediastinal lymph nodes and hilar lymph nodes to 10 mm and multiple small nodular shadows.

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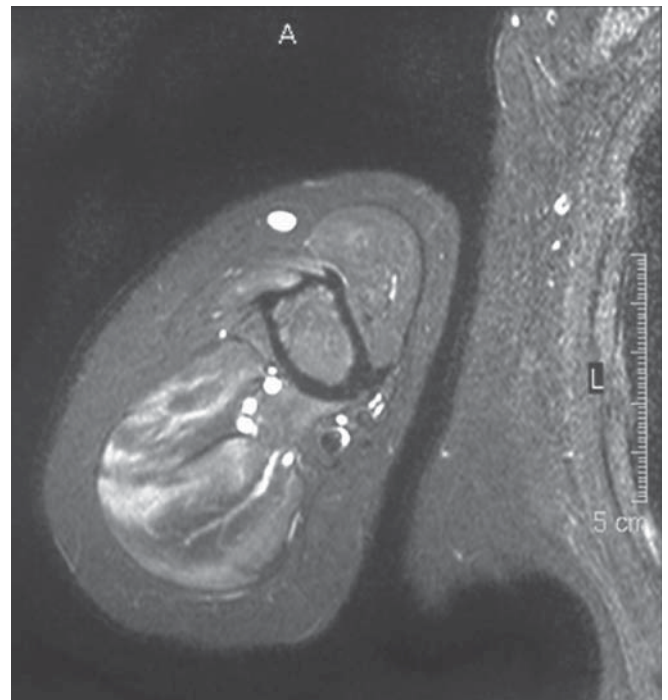
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**Fig. 1.** Ga-scintigraphy revealed strong accumulation in the hilar region and both parotid glands and no accumulation in muscles

Ga-scintigraphy revealed strong accumulation in the hilar region and bilateral parotid glands, and no accumulation in muscles (Fig. 1). Magnetic resonance imaging of the right arm revealed geographic regions of high signal intensity at the triceps muscle on short T1 inversion recovery images (Fig. 2). Therefore, sarcoidosis was strongly suspected. The angiotensin-converting enzyme (ACE) level was 25.0 U/l (normal 7.0–25.0 U/l), and the lysozyme level was 19.1 µg/ml (normal 5.0–10.2 µg/ml). Moreover, the tuberculin reaction was negative (1 mm × 2 mm). We consulted an ophthalmologist regarding the foggy vision. It was diagnosed as uveitis with mutton-fat keratic precipitates. Electromyography showed short, small, polyphasic motor unit potentials. A muscle biopsy specimen from the biceps muscle of the left arm contained Langhans-type giant cells in noncaseating granulomas (Fig. 3a). In addition, a muscle biopsy specimen showed the infiltration of lymphocytes at endomysial sites, necrosis of muscle fibers, and variations in fiber size (Fig. 3b). Therefore, acute sarcoid myositis was diagnosed from the muscular lesions and pathologic features according to the diagnostic criteria proposed by the Ministry of Health and Welfare of Japan (1989). Prednisolone was administered at 60 mg/day, beginning on May 26. The symptoms of



**Fig. 2.** Magnetic resonance imaging of the right arm revealed the geographic regions of high signal intensity at the triceps muscle on short T1 inversion recovery images

the patient improved within 1 week. The prednisolone was tapered gradually to 35 mg/day, and rehabilitation efforts improved the performance of the daily activities of the patient. The uveitis and eyesight also improved. On August 1, when the patient was transferred to a local hospital for further rehabilitation, she had some muscle weakness in the lower extremities and her CK level was 200 IU/ml (Fig. 4).

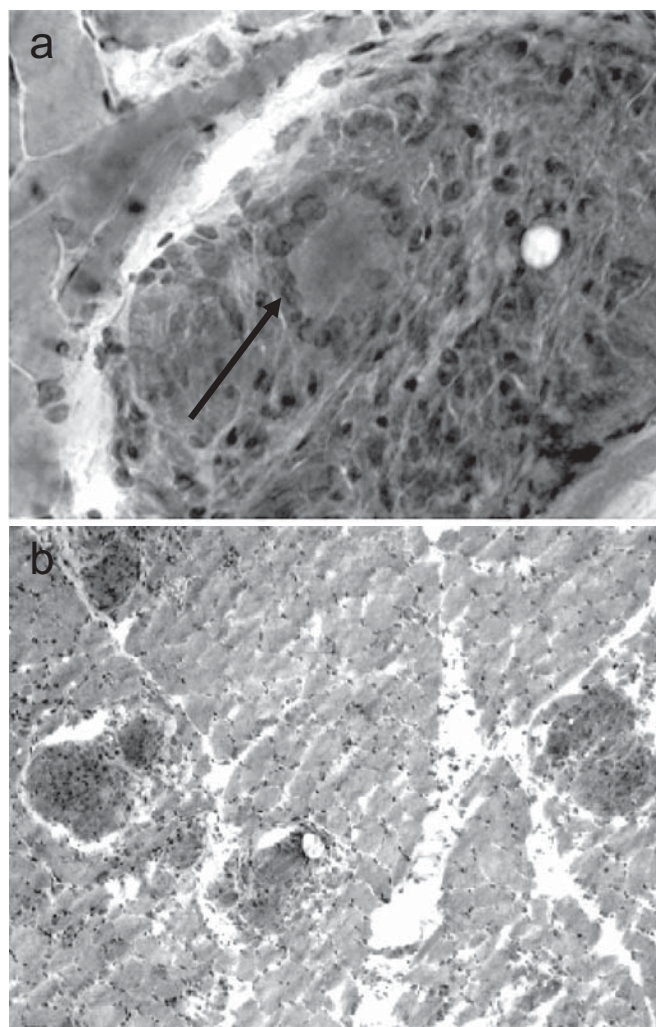
## Discussion

Sarcoid myositis is classified as symptomatic or asymptomatic.<sup>1–3</sup> Asymptomatic sarcoid myositis is relatively common, accounting for 50%–80% of cases of sarcoidosis.<sup>1,2</sup> Symptomatic sarcoid myositis is rare, accounting for less than 0.5% of cases of sarcoidosis.<sup>2,4,5</sup> Symptomatic sarcoid myositis is generally classified as one of three types: palpable nodular type, chronic type, or acute type.<sup>2,4</sup> The sarcoid myositis in our case was considered to be the acute type because of the acute course and marked CK elevation. In the English literature, we found only 14 cases of acute sarcoid myositis in which muscle biopsy specimens contained noncaseating granulomas (Table 1).<sup>2,4,6–17</sup> Twelve of the 14 patients were given steroids, and 10 of these 12 showed improvement. Our patient also had an excellent response to prednisolone. For patients with CK elevation and muscle weakness, sarcoid myositis as well as polymyositis should be considered. Such patients should be evaluated by means of chest X-ray, chest CT, ACE and lysozyme determinations, consultation with an ophthalmologist, electromyography, and muscle biopsy.

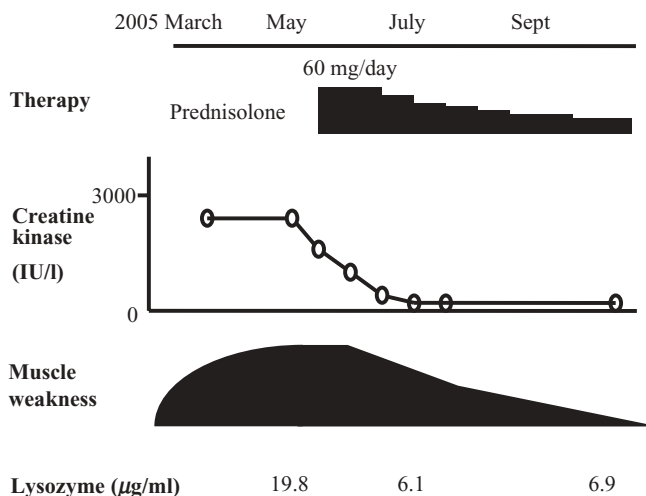
**Table 1.** Reported cases of acute sarcoid myositi

Authors	Year	Patient sex/age (years)	CK (IU/l)	Treatment	Outcome
McConkey <sup>6</sup>	1958	W/39	–	Cortisone	Relapsed
Harvey <sup>7</sup>	1959	M/27	–	None	Recovered
Silverstein and Siltzbach <sup>2</sup>	1969	W/28	–	Cortisone, 25 mg	Recovered
Douglas et al. <sup>4</sup>	1973	W/59	–	Prednisolone, 20 mg	Relapsed
Hewlett and Brownell <sup>8</sup>	1975	M/21	1 103	Prednisone, 60 mg	Recovered
Alpert et al. <sup>9</sup>	1979	W/15	1 650	Prednisone, 60 mg	Recovered
Ando et al. <sup>10</sup>	1985	M/27	3 040	Prednisone, 60 mg	Recovered
Jamal et al. <sup>11</sup>	1988	M/26	335	Prednisone, 30 mg	Recovered
Nidiry et al. <sup>12</sup>	1991	W/44	836	Prednisone, 30 mg	Recovered
Fonseca et al. <sup>13</sup>	1993	W/47	9 600	Prednisone, 60 mg	Recovered
Ost et al. <sup>14</sup>	1995	W/61	271	Prednisone, 60 mg	Recovered
Takanashi et al. <sup>15</sup>	1997	M/50	347	Steroid therapy <sup>a</sup>	Recovered
Pringle et al. <sup>16</sup>	1997	M/35	15 249	Unknown	Unknown
Takuma et al. <sup>17</sup>	2000	W/46	12 565	Prednisolone, 60 mg	Recovered
Our group	2006	W/54	2 241	Prednisolone, 60 mg	Recovered

CK, creatine kinase; W, Woman; M man

<sup>a</sup>Regimen not specified

**Fig. 3.** **a** Muscle biopsy specimen showing Langhans-type giant cells in noncaseating granulomas (arrow); (H&E, 400). **b** Muscle biopsy specimen showing infiltration of lymphocytes at endomysial sites, necrosis of muscle fibers, and variations in fiber size (H&E, ×100)

**Fig. 4.** Clinical course

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