# Intrascrotal Sarcoidosis in a Pediatric Patient

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## GOAL

To understand intrascrotal sarcoidosis to better manage patients with the condition

## LEARNING OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Differentiate the clinical manifestation of sarcoidosis in children and adults.
2. Identify histopathologic findings necessary for the diagnosis of sarcoidosis.
3. Evaluate the relationship between intrascrotal sarcoidosis and solid tissue tumors.

## INTENDED AUDIENCE

This CME activity is designed for dermatologists and generalists.

CME Test on page 130.

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*Sarcoidosis is a granulomatous disease with diverse skin manifestations and systemic involvement. We describe an adolescent boy who presented with unilateral scrotal swelling, constitutional symptoms, and a rash. Further workup demonstrated lymphadenopathy and pulmonary opacities. Granulomas caused by sarcoidosis were identified in biopsy specimens of the epididymis and skin lesions. No acid-fast organisms were found in biopsy specimens, and the patient had negative findings on placement of purified protein derivative (tuberculin). The concentration of angiotensin-converting enzyme was not elevated. We discuss pediatric sarcoidosis and, more specifically, intrascrotal sarcoidosis.*

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Sarcoidosis is a granulomatous multisystem disease of unknown pathogenesis characterized histologically by noncaseating granulomas. Most commonly diagnosed in patients aged 20 to 40 years, it has a prevalence in the United States of 5 per 100,000 in white individuals and 40 per 100,000 in black individuals.[1-3] It is considered rare in children; however, in mass screenings in countries such as Japan and Hungary, asymptomatic disease has been found with childhood prevalence similar to adults.[1] By 1990, 500 cases of pediatric sarcoidosis had been reported.[4] Most cases in the pediatric population occur in adolescents, with typical adult manifestations (eg, constitutional symptoms; lung, lymph node, ocular, or skin involvement).[1] There are 3 cases of pediatric sarcoidosis with scrotal enlargement.[4-6] To our knowledge, this is the fourth reported case of intrascrotal sarcoidosis in a pediatric patient.

## Case Report

A 14-year-old black adolescent boy who presented to his primary care physician with symptoms of dysuria was noted to have an enlarged right testicle. The patient complained of shortness of breath and a decreased appetite that had resulted in a 15-lb weight loss during the past 3 months. In the same period, he had a persistent asymptomatic skin rash on the abdomen, groin, and both legs. He had no history of fevers or night sweats, and he denied any joint pain or swelling. Prior to this incident, he had been healthy and had no pertinent medical history except an exposure to a family member with tuberculosis approximately 2 years earlier. He had no history of recent travel and was not sexually active. His immunizations were up-to-date.

On physical examination, the patient was found to have an enlarged right testicle with a palpable mass (Figure 1). He had no hepatosplenomegaly and no palpable peripheral lymphadenopathy. Breath sounds were normal. Examination of the skin showed multiple perifollicular hyperpigmented papules and plaques with mild scale scattered in an ichthyosiform pattern on the lower abdomen, lower legs, and dorsal areas of the feet (Figure 2). An annular plaque of hyperpigmented papules with peripheral scale was noted on the lower back. An ophthalmologic examination did not find any uveitis.

Laboratory studies were notable for a complete blood cell count within reference range with eosinophilia (18%), an elevated erythrocyte sedimentation rate (60 mm/h [reference range, 0–20 mm/h]), and concentrations of angiotensin-converting enzyme within reference range. Results of other laboratory studies included an elevated antinuclear antibody titer (1:40) with a speckled pattern, negative anti–double-stranded DNA, and C-reactive protein within reference range (2.1 mg/L [reference range, 0.08–3.1 mg/L]). Tests for antibodies to *Histoplasma capsulatum*, *Blastomyces dermatitidis*, and *Coccidioides immitis* were negative. Findings of serologic tests were nonreactive for syphilis and negative for human immunodeficiency virus and antineutrophil cytoplasmic autoantibodies. Two placements of purified protein derivative (tuberculin) both had negative results.

An ultrasound of the testicle showed epididymal enlargement. Chest radiographs showed possible thickening of the pleura of the left apex. Computed tomograms of the chest, abdomen, and pelvis showed lymphadenopathy of the neck, mediastinum, axilla,

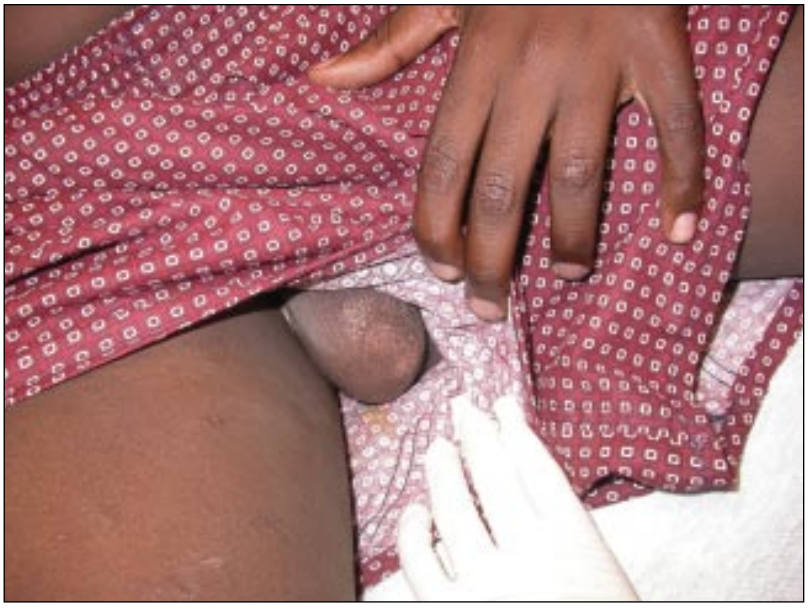

**Figure 1.** At initial presentation, the 14-year-old adolescent boy had a clinically enlarged right testicle.

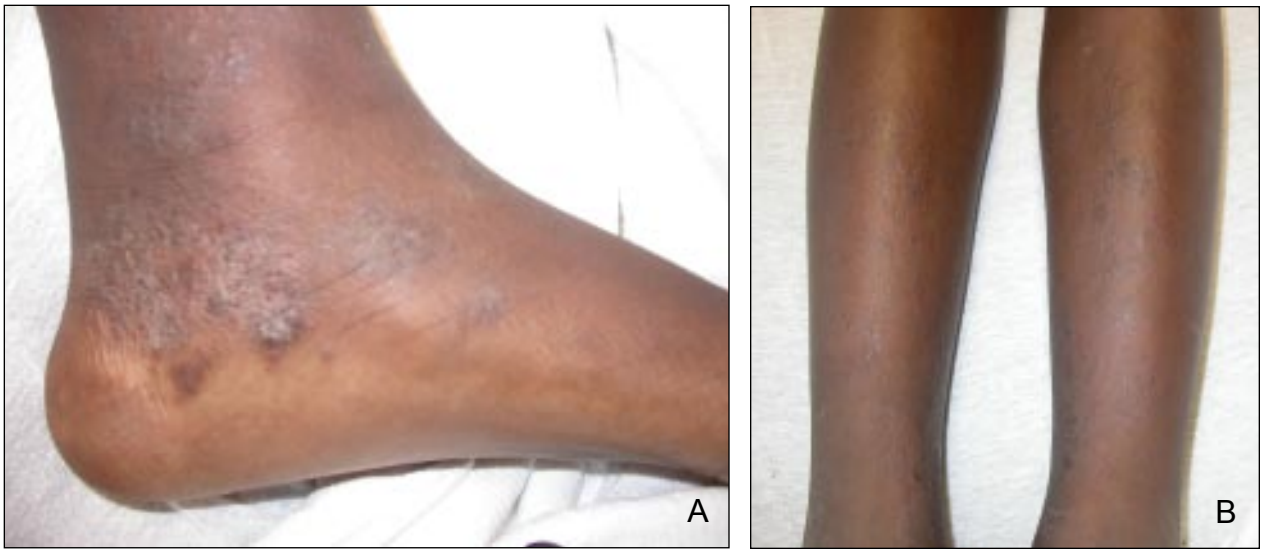


**Figure 2.** Multiple hyperpigmented papules and plaques with mild scale form an ichthyosiform pattern on the ankle (A) and on the bilateral anterior aspect of the lower legs (B).

mesentery, pelvis, and inguinal regions. Patchy bilateral pulmonary opacities and increased interstitial markings were found on computed tomograms of the chest. Pulmonary function tests demonstrated restrictive airway disease.

Histopathologic findings of an epididymal biopsy specimen showed noncaseating granulomas (Figure 3). Cultures and stains for acid-fast organisms were negative. Biopsy specimens of skin from the lower limbs also showed noncaseating naked tubercles consistent with sarcoidosis (Figures 4 and 5). Additional cultures and stains for acid-fast bacteria and fungi were again negative.

## Comment

Sarcoidosis is a granulomatous disease that can affect any organ system. It is most commonly identified in individuals aged 20 to 40 years. It has been reported in children, but the exact prevalence in childhood is unknown. In the United States, 70% of cases of pediatric sarcoidosis were reported in Virginia, North Carolina, South Carolina, or Arkansas.[3] Most childhood cases occur in preadolescent children or adolescents and less frequently in younger children.[1,3,7,8] In the pediatric population, manifestations occur in 2 different types of clinical presentations. Children younger than 5 years present with a triad of ocular, joint, and skin disease, but without the typical lung involvement, and their condition often is confused with juvenile idiopathic arthritis. Children aged 8 to 15 years have clinical manifestations similar to adults (eg, constitutional symptoms; lung, ocular, and skin involvement).[9-11]

Intrascrotal sarcoidosis was first described in 1937.[12] Reports of incidence have ranged from 0.2% to 5%.[13] In 2004, Kodama et al[14] conducted a comprehensive review of the medical literature and identified 60 reports of histologically proven sarcoidosis of the male reproductive tract. In these cases, the average age of patients was 33 years (age range, 2–67 years), 58% (35/60) of patients were black, and 78% (47/60) presented with either stage 1 or stage 2 disease. At initial presentation, 43% (26/60) of patients presented with intrascrotal abnormalities. The epididymis was the most commonly affected area (73%; 44/60), followed by the testes (47%; 28/60), spermatic cords (8%; 5/60), tunica albuginea (5%; 3/60), and prostate (3%; 2/60).[14]

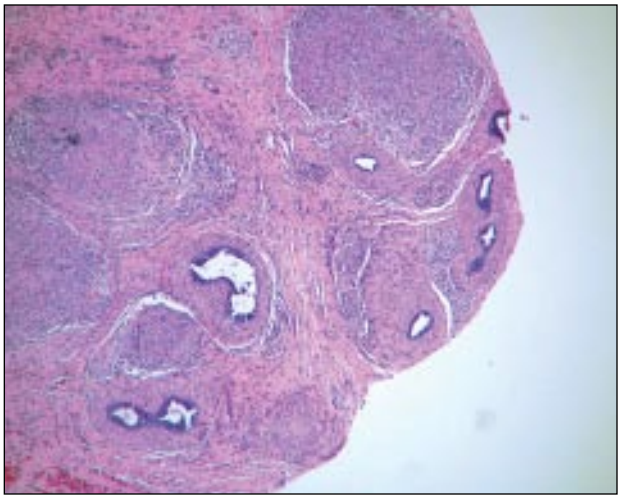

**Figure 3.** A biopsy specimen from the epididymis shows noncaseating granulomas (H&E, original magnification ×100).

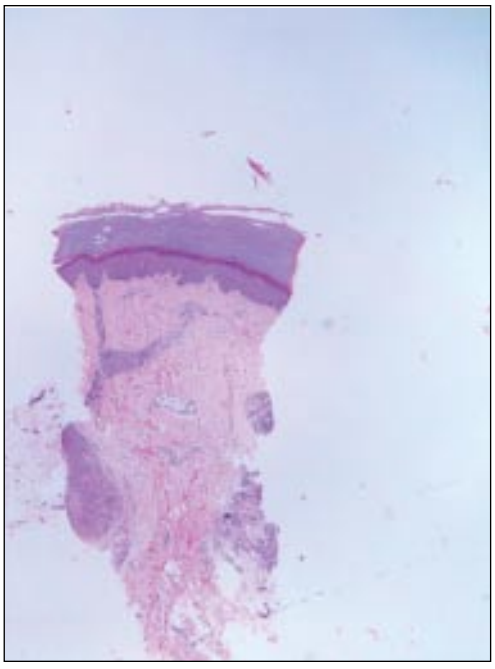

**Figure 4.** A cutaneous biopsy specimen shows noncaseating granulomas in the dermis (H&E, original magnification ×20).

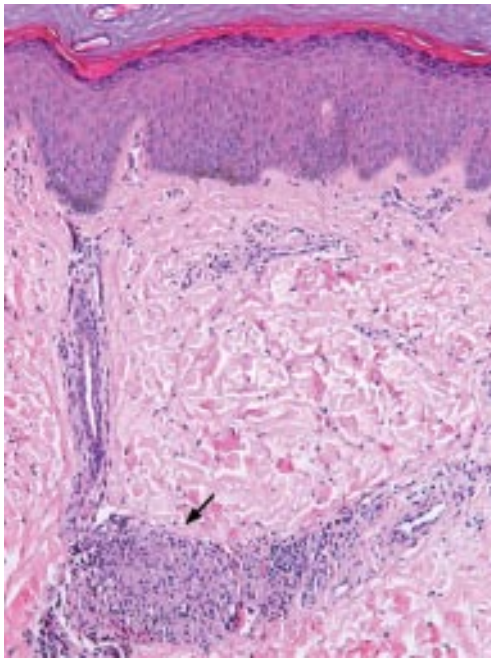

**Figure 5.** Dermal granuloma (arrow) (H&E, original magnification ×100).

Patients presenting with scrotal swelling or an intrascrotal mass should undergo appropriate imaging studies to determine the scrotal structure showing enlargement. One case report discussed a patient with sarcoidosis who presented with an enlarged scrotum that, after imaging, demonstrated involvement of only the scrotal skin.[15] Magnetic resonance imaging can help determine the scrotal structure involved; some features would suggest a nonmalignant process, such as sarcoidosis, but tissue pathology demonstrating noncaseating granulomas is still needed for accurate diagnosis.[14,16]

**Cases of Pediatric Intrascrotal Sarcoidosis**

| Reference | Organ Involved |
|---|---|
| Present report | Epididymis |
| Evans et al[4] | Epididymis, testicle |
| Gerstenhaber et al[5] | Epididymis |
| Weinberg and Ginsburg[6] | Epididymis |

In our recent review of the current English-language medical literature via a PubMed search for the terms *intrascrotal sarcoid*, *scrotal sarcoid*, *testicular sarcoid*, *epididymal sarcoid*, and *genital sarcoid*, we identified 3 pediatric cases of intrascrotal sarcoidosis. A review by Carmody and Sharma[17] cited a 1977 case report of a 17-year-old black adolescent boy with sarcoidosis of the epididymis.[5] As described in another report, a 5-year-old black boy presented with fever, arthritis, and a left facial droop that was followed by left and then right testicular swelling.[4] Small areas of hyperpigmentation were noted on the patient's abdomen. A biopsy specimen of the genitalia showed testicular and epididymal sarcoidosis.[4] In the third report, a 2-year-old boy presented with fever and bilateral scrotal swelling but no skin lesions. Biopsy specimens identified epididymal sarcoidosis.[6] Our case would be the fourth reported pediatric case of intrascrotal sarcoidosis (Table).

Some reports suggest a relationship between solid tissue tumors, particularly testicular carcinoma, in patients with sarcoidosis.[18] This association was reviewed in a retrospective study reported in 1998.[19] Compared with a general population of young white men, patients with sarcoidosis were found to have a 100-fold incidence of also having had a diagnosis of testicular carcinoma. The sarcoidosis did not involve the male reproductive tract in any of the patients. In the same report, 12 of 14 patients with testicular cancer had the tumor before diagnosis of sarcoidosis (mean interval between diagnoses,

59.5 months) and only 2 had the sarcoidosis before the testicular cancer. The authors commented that the intensive surveillance of patients after testicular cancer may be increasing the diagnosis of asymptomatic sarcoidosis and therefore causing a surveillance bias.[19] Other factors still need to be delineated by appropriate prospective studies.

## Conclusion

Our pediatric patient initially presented with unilateral intrascrotal swelling, constitutional symptoms, and a rash. Subsequent biopsies of the scrotal mass and skin lesions as well as a review of constitutional symptoms and characteristic lung findings led to a diagnosis of systemic sarcoidosis. Sarcoidosis presenting as an intrascrotal mass is rare, especially in pediatric patients. The epididymis is the most commonly affected intrascrotal structure. Patients of any age who present with intrascrotal or scrotal swelling should undergo imaging studies and biopsies to confirm the diagnosis.

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