

9. Saigal S, Zunyk O, Larke B, Chernesky M. The outcome of children with congenital cytomegalovirus infection—A long term follow up study. *Am J Dis Child* 1982,136: 896-901.
10. Deka RC. Management of hearing im-
paired children. *Indian Pediatr* 1993, 30: 977-979.
11. Brady MT, Milian JD, Anderson DC, *et al.* Use of deglycerolized RBC to prevent post-transfusion infection with cytomegalovirus in neonates. *J Inf Dis* 1984,150: 334-339.

Sarcoidosis

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Sarcoidosis is a chronic multisystem disease of unknown etiology, usually affecting adults. Only a few reports describing the clinical features and course of sarcoidosis in children have been published. Only one case in a child has previously been reported from this country(1). The rarity of the condition prompts us to report the clinical features in two such patients.

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Case Reports

Case I: Two and a half year old girl presented with fever, weight loss and cough of 5 months duration. The weight and height were below the fifth percentile for the age. Examination showed marked pallor. The liver and spleen were palpable 5 cm and 3 cm, respectively below the costal margin. Rest of the systemic examination was normal. Investigations showed a hemoglobin level of 7 g/dl, total leucocyte count of 7800/cu mm with 60% polymorphonuclear leucocytes, 30% lymphocytes and 10% eosinophils. The blood levels of transaminases, alkaline phosphatase, serum proteins, creatinine, calcium and phosphate were normal. The liver biopsy showed non-caseating granulomatous lesions. An X-ray film of the chest showed bilateral enlarged hilar lymph nodes. The Mantoux test (using 1 TU injected intradermally) and VDRL test were negative.

A diagnosis of disseminated tuberculosis was made and the patient treated with isoniazid (5 mg/kg), rifampicin (10 mg/kg) and pyrazinamide (30 mg/kg) daily for 2 months. Subsequently pyrazinamide was stopped and therapy continued with rifampicin and isoniazid.

Three months after starting therapy the child still had remittent fever, worsening of cough and showed respiratory distress. There was swelling, pain and restriction of movement in both knee joints. She also showed a nonpruritic erythematous maculopapular rash over the trunk and the extremities. Slit lamp examination of the eyes showed posterior synechiae in the right iris. The liver and spleen had enlarged to 8 cm and 10 cm, respectively. Auscultation of the chest showed bilateral fine crepitations.

Investigations showed a hemoglobin level of 7.6 g/dl, total count of 8400/cu mm with 70% polymorphonuclear cells and 28% lymphocytes. Urine examination showed 8-10 red cells per high power field and the 24-hour urinary calcium excretion was 6 mg/kg. The blood level of urea was 16 mg/dl, creatinine 0.4 mg/dl, calcium 11.6 mg/dl (normal 9-11 mg/dl) and phosphate 4.5 mg/dl.¹ A repeat chest radiograph showed bilateral enlarged hilar lymphnodes and streaky infiltrates in the pulmonary parenchyma. Gastric aspirate for acid fast bacilli was negative on three occasions. Biopsy of the skin rash showed non-caseating granulomatous lesions. The liver and skin biopsies did not show any fungal hyphae within the granulomas. Cultures of the urine and blood for bacteria and fungi were sterile. Ultrasonography of abdomen showed hepatosplenomegaly and a small calculus in right renal pelvis.

In presence of non-caseating granulomas in the liver and skin, no definitive evidence of tubercular or fungal infection and unsatisfactory response to therapy with antitubercular drugs, a diagnosis of sarcoidosis was made. The

child was treated with oral prednisolone in a dose of 2 mg/kg/day. Antitubercular drugs were continued for four more weeks and then stopped. Within one week of starting corticosteroids the child was afebrile and eating well. The skin rashes and joint swelling resolved and the cough reduced within the next 2 weeks. A repeat X-ray chest 8 weeks later showed clearance of parenchymal lesions with persistent hilar lymphadenopathy. The size of the liver and spleen regressed and the patient gained 2.4 kg weight over 8 weeks. The dose of prednisolone was tapered to 0.5 mg/kg/day over next 2 months. Within a week however, the maculopapular rash, high grade fever and features of uveitis recurred. The dose of prednisolone was increased to 1 mg/kg/day for one month and gradually tapered to 0.75 mg/kg/day. The patient is free of symptoms on this dose for the last 6 months.

Case 2: An 8 year-old girl presented with fever and mild cough of two months duration. The girl was pale and sick looking. The height and weight were at the 50th centile for the age. The liver and spleen were enlarged 5 cm and 4 cm, respectively below the costal margin. The level of hemoglobin was 9 g/dl, total leucocyte count 8900/cu mm with 70% polymorphonuclear cells and 30% lymphocytes. The ESR was 54 mm at the end of first hour. The Mantoux test was negative and blood and urine cultures sterile. The chest radiograph was normal. Ultrasonography of abdomen showed hepatosplenomegaly. There were no retroperitoneal lymph nodes and the renal outlines were normal. The liver biopsy showed non caseating

granulomatous lesions. A diagnosis of disseminated tuberculosis was made and the patient treated with antitubercular drugs, as in the first case. At the end of 2 months the patient did not show resolution of fever and cough and developed dyspnea on mild exertion. Auscultation of the chest showed coarse crepitations bilaterally. Slit lamp examination showed posterior synechiae in both eyes. The sputum was negative for AFB and a repeat chest radiograph now showed diffuse infiltration and hilar lymph nodes on both sides. The blood level of angiotensin converting enzyme activity was 39 IU/dl (normal 20-40 IU/dl).

A diagnosis of sarcoidosis was made and the child was started on oral prednisolone in a dose of 2 mg/kg/day. Antitubercular drugs were continued for an additional 8 weeks and then stopped. Following therapy with steroids the patient became afebrile in 8 days, and the intensity of cough and dyspnea on exertion decreased. The liver and spleen regressed in size and a chest radiograph taken 4 weeks later showed clearance of parenchymal lesions with persistence of hilar nodes. The prednisolone dose was reduced to 1 mg/kg/day at the end of 4 weeks and then gradually tapered and stopped over 20 weeks without reappearance of any symptoms.

Discussion

Both patients showed systemic symptoms and non-caseating granulomatous lesions involving one or more sites. The diagnosis of sarcoidosis was not considered initially and despite absence of definitive evidence suggestive

of tuberculosis, they received 8-12 weeks of antitubercular therapy before the diagnosis was reviewed. The prompt remission of symptoms on corticosteroid therapy alone excludes the diagnosis of typical or atypical mycobacterial infection. The clinical features of sarcoidosis are protean and may involve a variety of organs. *Table 1* shows the common clinical and laboratory features of sarcoidosis in children. Almost 75% of children with sarcoidosis have involvement of 5 or more organ systems(2). Older children usually show symptoms related to the involvement of lungs, lymph nodes and eyes. However, in children below 4 years of age, the chief clinical features are limited to those of the skin, joints and eyes with infrequent involvement of the lungs(3).

The diagnosis of sarcoidosis should be suspected clinically in patients with evidence of involvement of multiple organs. Demonstration of non-caseating granulomatous lesions on histopathology is a strong supportive evidence for the diagnosis. The various sites where granulomas may be seen on histopathology include lymphnodes, skin, liver, conjunctiva, salivary glands, bones, nasal mucosa and muscles(4,5). Fine needle aspiration cytology from the affected organ has been used as an alternative to the biopsy with satisfactory results(6). A positive Kveim test using 0.15 to 0.20 ml of emulsified sarcoid tissue administered intradermally supports the diagnosis of sarcoidosis(7). In patients with sarcoidosis, a nodule may form at the injection site after 28 to 42 days; biopsy of the nodule shows typical noncaseating granulomas. The emulsified sarcoid antigen is not easily available, limiting the use of this test.

TABLE I—Clinical and Laboratory Features of Sarcoidosis in Children

Clinical features	Laboratory findings
Fever	Blood
Weight loss	Raised ESR, eosinophilia, leucopenia
Cough	
Fatigue/lethargy	Renal
Bone and joint pain	Decreased creatinine clearance
Arthritis	Hypercalcemia, hypercalciuria
Red eye, photophobia	
Parotid enlargement	Liver
Maculopapular erythematous rash	Increased SGOT, SGPT, alkaline phosphatase
Uveitis acute or healed	Increased total proteins
Lymphadenopathy	Decreased serum albumin
Hepatomegaly	
Splenomegaly	Radiology
Dyspnea, crepitations	Chest: Parenchymal lesions, adenopathy
Cyanosis, clubbing	Lytic lesions long bones
	Others
	Increased serum angiotensin converting enzyme activity

Adapted from Edward *et al.*(2) and Marciller *et al.*(10).

The activity of disease can be monitored by the levels of angiotensin converting enzyme in the blood. The levels are higher in patients with active disease and return to normal levels with treatment(8). Recent studies have suggested that total lymphocyte count and ratio of lymphocyte subpopulation (CD 4/CD 8), and alveolar macrophage activity in the bronchoalveolar lavage fluid can be used to judge the activity of the disease(9). The lymphocyte subpopulation ratio in peripheral blood cannot be used to judge the disease activity because there is compartmentalization of the immune system in sarcoidosis. There are increased number of T helper cells at the site of lesion but the same is decreased in peripheral blood(9).

Treatment of patients with systemic corticosteroids relieves symptoms in most cases. Treatment with steroids is indicated in patients having pulmonary symptoms, hypercalciuria and hypercalcemia or evidence of uveitis which is not controlled by topical therapy alone. The initial dose of prednisolone may vary between 1-2 mg/kg/day, which is then gradually tapered to a small maintenance dose. In general there is considerable improvement in clinical manifestations, chest radiographs pulmonary function tests and the number of areas of involvement. However, up to one third of patients may show subnormal pulmonary functions, cor pulmonale, cardiac dysrhythmias, and bony and joint abnormalities on follow up(10).

The experience with our two cases shows that patients with sarcoidosis may often not be properly diagnosed. The condition must be suspected clinically in patients with involvement of multiple organ systems. Therapy with corticosteroids in selected patients leads to rapid relief of symptoms and possibly decreases the incidence of long term complications.

REFERENCES

- Chetty A, Mathur M, Verma IC, Ray D. Sarcoidosis. *Indian J Pediatr* 1987, 54: 785-788.
- Edward DM, Strobe GL, Starley SM, Flay DW. Childhood sarcoidosis. *J Pediatr* 1986,108:169-177.
- Hetherington S. Sarcoidosis in young children. *Am J Dis Child* 1982,136:13-15.
- Nessan VJ, Jacoway JR. Biopsy of minor salivary glands in the diagnosis of sarcoidosis. *N Eng J Med* 1979, 301: 922.
- Samer SK. Random biopsy in the diagnosis of sarcoidosis. *Ear Nose Throat* 1973, 52: 54.
- Jakely PE Jr, Silverman JF, Holbrook CT, Fairman RD, Daeschner CW, Joshi VV. Fine needle aspiration cytology as an adjunct in the diagnosis of childhood sarcoidosis. *Pediatr Pulmonol* 1992,13:117-120.
- Kendig EL. Sarcoidosis. *In: Disorders of the Respiratory Tract in Children*, TV ed. Eds. Kendig EL Jr, Chernick V. London, W.B. Saunders Company 1983, pp 710-721.
- Beneteau-Burnat B, Baudin B, Morgant C, Baumann FC, Giboudeau J. Serum angiotensin converting enzyme in healthy and sarcoidotic children: Comparison with the reference interval for adults. *Clin Chem* 1990, 36: 344-346.
- Chadelat K, Baculard A, Grimfeld A. Pulmonary sarcoidosis in children: Serial evaluation of bronchoalveolar lavage cells during corticosteroid treatment. *Pediatr Pulmonol* 1993, 16: 41-47.
- Marciller, McCarthy M, Barton JW, Merten D, Spock A. Long term outcome of pediatric sarcoidosis with emphasis on pulmonary status. *Chest* 1992,102:1444-1449.

Calcification in Renal Tuberculosis

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Genito-urinary tuberculosis consti-

tutes the single largest group of extrapulmonary tuberculosis in all age groups. It is estimated that 15-20% of

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