

Case Report

Childhood Sarcoidosis - Case Reports

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ABSTRACT

Childhood Sarcoidosis is a rare disease in pediatric practice. However, no case has been reported in the literature from the Arabian Gulf area. Although it is a multisystemic disease affecting the lungs, eyes and skin,

it usually has a good prognosis. The following case reports and discussion review the most recent knowledge about this disease.

KEYWORDS: granulomatous changes, multisystemic granulomatous disease, sarcoidosis

INTRODUCTION

Sarcoidosis is a multisystemic granulomatous disease of unknown etiology involving the lungs, lymph nodes, skin, eyes, bones and the parotid glands. The clinical picture is attributed to the presence of granulomas, which are composed of epithelioid and multinucleated cells with little or no necrosis. Sarcoidosis in children is rare, and apart from a single report from Saudi Arabia, there is no other report from the Arabian Gulf region^[1]. In this article, for the first time in Kuwait, we describe two children with sarcoidosis.

CASE REPORTS**Case 1**

A.B. is a 6-year-old Kuwaiti boy, who was admitted to the hospital because of productive cough with increasing effort and cyanosis of two weeks duration. In the previous year, he had suffered from dry eyes which were diagnosed as xerophthalmia and was given eye drops with minimal improvement. The patient had a history of nephrolithiasis which was not investigated. His birth history, past medical and developmental history was normal. His father and grandfather were diabetic. He had three other siblings in good health.

His physical examination on admission showed that he was in distress with a respiratory rate of 40 breaths per minute. His temperature was 36.6 °C, pulse rate 136/min, blood pressure 98/62 mmHg, weight 15.9 kg (10th percentile) and height 107 cm; (10th percentile). He looked pale with pigmented sclerae, normal tympanic membranes and pharynx. The neck was supple with multiple shotty generalized palpable lymph nodes. The parotid

glands were slightly enlarged. He had symmetrical chest expansion and clear to auscultation. The precordium was dynamic, the first heart sound was normal and the second heart sound was split with a grade I/VI systolic ejection murmur. The spleen and the liver edges were firm and palpable below the costal margin. Skin was dry, and the joints were normal.

His investigations included ESR: 20 mm first hour, WBC 6.2 (polymorphnuclear neutrophils 73%, lymphocytes 24%, eosinophils 3%), Hemoglobin 12 gm/dl, Platelets 243,000. Transcutaneous O₂ saturation was 82% on room air. His Angiotensin Converting Enzyme level (ACE) was 255 U (normal 12- 55 for age). The Anti Nuclear Antigen, Epstein Barr Virus and Cytomegalovirus serology were negative. Purified protein derivative (PPD) was non-reactive. Chest X-ray showed bilateral hilar lymphadenopathy, hyperinflation, air bronchogram and peribronchial infiltration. Conjunctival biopsy showed the presence of pea shaped non- caseating granuloma confirming the diagnosis of sarcoidosis. Ultrasound of the abdomen showed two separate stones in the left kidney and one stone in the right kidney. There was no evidence of hydronephrosis. A 24-hour urine collection ruled out hypercalciuria. His initial creatinine was 3 gm in 24 hours (normal limit = 1.0 to 1.8 gm)

He was treated with methylprednisolone 2 mg/kg per day every other day for three doses, followed by a maintenance dose of prednisolone of 30 mg/day with remarkable improvement. His appetite increased with less fatigability on respiratory exertion, and the resting heart rate decreased to 100/min. Oxygen saturation on room air was now 96-100%. Dryness of the skin also

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improved. Pulmonary Function Tests (PFT) performed before and after treatment showed reduction of the restrictive lung disease. He required salbutamol nebulization every eight hours. The eye signs also improved with steroid therapy.

On follow-up, the patient was found to remain stable for a long period except with one incidence of relapse which required another course of steroids. At present he is on a maintenance dose of prednisolone.

Case 2

A.S., presented at the age of four years with recurrent episodes of urticaria associated with arthralgia without any evidence of joint involvement. He later developed low grade fever, lymphadenopathy and skin rash. Parotid glands were bilaterally palpable. The eyes were normal and the lungs were clear to auscultation. There was no hepatosplenomegaly. A skin biopsy was done which showed "granulomatous changes". Eighteen months later, the child began to develop multiple dermal and subcutaneous nodules; the largest was on the right cheek measuring 4 x 5 cm. The others, about 30 in number, were on the arms and torso. Apart from a degree of hearing impairment that was managed by a hearing aid, the child had been well. A skin biopsy showed the appearance of non-caseating epithelioid cells granuloma specific for sarcoidosis. Culture was negative. Chest X-ray and CT scan of the head were normal. Because of the large skin lesion, it was decided to start systemic steroid therapy with prednisolone 1 mg/kg/day. This resulted in the reduction of the size of all skin nodules and steroids were then tapered over six months. Intralesional injection of triamcinolone acetonide mixed with 2% lignocaine was administered to the large nodule with subsequent reduction of its size. The child improved significantly and subsequent follow up was done by dermatology.

DISCUSSION

It is known that sarcoidosis is rare in children. The prevalence in USA is 1- 40 per 100,000. Its low incidence in this part of the world is attributed to lack of screening as well as the presence of other commonly recognized granulomatous diseases, especially tuberculosis, that mimic sarcoidosis.^[2]

The clinical manifestations depend on the organ involved. The symptoms may be relatively mild initially despite extensive tissue involvement. Progression is usually slow with relapses and remissions and eventual healing in most cases. Thus, the clinical picture is extremely variable and arises mainly from the effects of fibrosis and/or pressure exerted by the enlarged structures. It

commonly affects the lungs causing cough, dyspnea and wheezing with weight loss. Unilateral hilar adenopathy in the initial stage eventually becomes bilateral (case 1)^[3].

The radiological findings in the lungs are variable and may appear as flocculent densities, miliary nodules, focal streakings and pulmonary reticulations^[4]. On the other hand, abnormal PFT may show impaired diffusion, reduced compliance, decreased lung volumes and vital capacity with small airway resistance.

Skin lesions manifest as granulomata on the face and lateral aspects of the arms appearing as yellow waxy papules and nodules but may also take the form of large lichenoid conglomerates (case 2)^[5].

Ocular involvement is commonly manifested as uveitis requiring early referral for slit lamp examination. Decrease in visual acuity, blurring, irritation with photophobia and excessive lacrimation may occur. Iritis is the most frequent lesion followed by trabecular nodules and tent-like peripheral anterior synechia, retinal perivasculitis and spotty retinochoroidal exudates^[6].

Joint involvement resembles rheumatoid arthritis. The joints, however, have large boggy effusion with thickening of the joint membranes^[7].

Other systems involved include renal, cardiovascular, central nervous and endocrine systems. Upper respiratory tract, testicles and various other organs are also involved.

Once the diagnosis is suspected, the work up should include chest roentgenograms, PFT, electrocardiograms, echocardiograms, and angiotensin-converting enzyme (ACE) activity. Complete blood cell counts, erythrocyte sedimentation rates, serum calcium, immunoglobulin levels, B- and T-cell enumerations, and intradermal skin tests for delayed hypersensitivity are also important^[8].

However, the diagnosis of sarcoidosis rests mainly on tissue studies with biopsies taken from lymph nodes, conjunctivae, scalene fat pad, skin lesions, buccal mucosa or lung tissue.

The treatment of sarcoidosis is mainly supportive with the corticosteroids being the cornerstone of treatment when indicated. They decrease the extent of the organ involvement as well as improve the symptoms and signs. Prednisone is given as 1-2 mg/kg/day until symptoms improve then tapered to reduce the dose to 15 mg every other day for a total period of six months. Sequential oral and inhaled corticosteroid therapy may be an alternative treatment regimen for sarcoidosis in children^[9]. Diet low in calcium is advised.

The prognosis of sarcoidosis is usually good with eventual resolution except in the case of early onset of juvenile sarcoidosis. Mortality in childhood

sarcoidosis is about 5%, with long-term sequelae in 10 to 20%^[10].

The present case reports are intended to draw attention to the occurrence of sarcoidosis in Kuwait, as early recognition and proper management may prevent complications such as blindness, pulmonary insufficiency, and renal impairment.

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