P-ISSN: 2664-3685
E-ISSN: 2664-3693
www.paediatricjournal.com
IJPG 2018; 1(1): 20-22
Received: 05-01-2018
Accepted: 10-02-2018

Dr. Dilbag Singh
Department of Paediatrics, Govt. Dental College and Hospital, Jaipur, Rajasthan India

Corresponding Author:
Dr. Dilbag Singh
Department of Paediatrics, Govt. Dental College and Hospital, Jaipur, Rajasthan India

Assessment of clinical profile of cases of Sarcoidosis in children

Dr. Dilbag Singh

DOI: https://doi.org/10.33545/26643685.2018.v1.i1a.8

Abstract
Background: Sarcoidosis affects people of all racial and ethnic groups and occurs at all ages. The present study was conducted to assess clinical profile of sarcoidosis in children.
Materials & Methods: The present study was conducted on 23 children suffering from sarcoidosis of both genders. Diagnosis of sarcoidosis was based on clinical features, documentation of non-caseating granuloma from various body tissues, raised serum Angiotensin converting enzyme (ACE) levels and raised urinary calcium to creatinine ratio. In all children, clinical features and laboratory findings were recorded.
Results: Out of 23, males were 11 and females were 12. Common features were fever seen in 17, uveitis in 10, splenomegaly in 7, Hepatomegaly in 8, weight loss in 4, skin rashes in 6, difficult respiration in 4, cough in 2 and seizures in 1. The difference was significant ($P< 0.05$). TLC count was 10500/ cumm of blood, ESR was 54, hemoglobin level was 9.2 g/dL and serum ACE was 148 U/mL.
Conclusion: Sarcoidosis is rare in children. Authors found 23 cases in children. Common clinical features were fever, difficult in respiration, weight loss etc.

Keywords: Children, Sarcoidosis, weight

Introduction
Sarcoidosis affects people of all racial and ethnic groups and occurs at all ages, although it usually develops before the age of 50 years, with the incidence peaking at 20 to 39 years.3 The incidence of sarcoidosis varies widely throughout the world, probably because of differences in environmental exposures, surveillance methods, and predisposing HLA alleles and other genetic factors [1].
Pediatric sarcoidosis is a chronic disease characterized by non-caseating granulomatous inflammation. It most commonly affects young adults, and is very rare in children. The exact cause of sarcoidosis is still not known. Many studies suggest that genetic susceptibility and environmental factors contribute to disease development [2]. immunologically, sarcoidosis is an exaggerated immune response to so far unidentified antigens. Data for the clinical heterogeneity of sarcoidosis strongly suggest that pathogen-associated molecular patterns of microbial antigens can trigger or amplify inflammation. There is no evidence that sarcoidosis is an infectious disease; rather, it is an exaggerated immune response to pathogen-associated molecular patterns of killed and partly degraded mycobacteria and Propionibacterium [3].
Although sarcoidosis usually presents with bilateral hilar lymphadenopathy and lung infiltration, multiple organ systems may be involved. The disease mainly affects people in the third and fourth decades of life, but may also occur in children and elderly subjects [4].
The present study was conducted to assess clinical profile of sarcoidosis in children.

Materials & Methods
The present study was conducted in the department of Pediatrics. It comprised of 23 children suffering from sarcoidosis of both genders. Diagnosis of sarcoidosis was based on clinical features, documentation of non-caseating granuloma from various body tissues, raised serum Angiotensin converting enzyme (ACE) levels and raised urinary calcium to creatinine ratio.
All were informed regarding the study and written consent was obtained. Ethical approval for the study was obtained prior starting the study.
Data pertaining to children such as name, age, gender etc. was recorded. In all children, clinical features and laboratory findings were recorded. Results were tabulated and subjected to statistical analysis. P value less than 0.05 was considered significant.

Results

Table 1: Distribution of patients

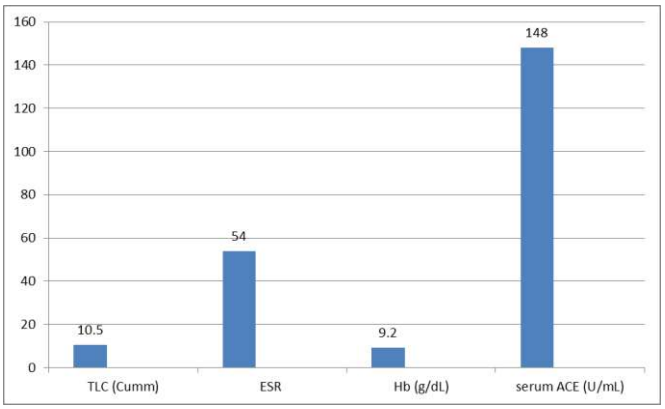
Total- 23		
Gender	**Males**	**Females**
Number	11	12

Table I shows that out of 23, males were 11 and females were 12.

Table 2: Clinical profile of patients

Clinical profile	Number	P value
Fever	17	0.01
Uveitis	10	
Splenomegaly	7	
Hepatomegaly	8	
Weight loss	4	
Skin rash	6	
Difficult respiration	4	
Cough	2	
Seizures	1	

Table I shows that common features were fever seen in 17, uveitis in 10, splenomegaly in 7, Hepatomegaly in 8, weight loss in 4, skin rashes in 6, difficult respiration in 4, cough in 2 and seizures in 1. The difference was significant (P< 0.05).



Graph I: Laboratory findings in patients

Graph I shows that TLC count was 10500/ cumm of blood, ESR was 54, hemoglobin level was 9.2 g/dL and serum ACE was 148 U/mL.

Discussion

Sarcoidosis is a multisystem disorder; however, mediastinal lymphadenopathy, pulmonary parenchymal infiltration, and cutaneous and ophthalmic disease are common [5]. Moreover, it may involve liver, central nervous system and kidneys. In young children (<5 year old), a triad of skin rash, uveitis and arthritis is more common than pulmonary and mediastinal involvement. Information on course of illness, effective treatment and outcome on childhood sarcoidosis is limited to few reports. Diagnostic criteria are well defined but monitoring and treatment regimen are not well defined [6].

The real incidence and prevalence of sarcoidosis worldwide is difficult to determine as many patients are asymptomatic. A higher incidence of the disease is reported in northern (about 60 per 100,000) than in southern European countries, including Italy (< 10 per 100,000) [7]. The present study was conducted to assess clinical profile of sarcoidosis in children.

In present study, out of 23, males were 11 and females were 12. We found that common features were fever seen in 17, uveitis in 10, splenomegaly in 7, Hepatomegaly in 8, weight loss in 4, skin rashes in 6, difficult respiration in 4, cough in 2 and seizures in 1. The difference was significant (P< 0.05).

Hoffmann *et al.* [8] found that clinical features at the time of diagnosis were fever (83%), uveitis (50%), difficulty in breathing (44%), hepatosplenomegaly, weight loss, arthritis and peripheral adenopathy. Imaging findings included: hilar adenopathy (94%), abdominal nodes (50%) and pulmonary infiltrates (44%). All children were treated with steroids (range 6-12 months) and weekly low dose oral methotrexate. All patients showed significant improvement over a mean (SD) duration of follow-up of 3.1 (0.9) years, as assessed by resolution of clinical symptoms, and improvement in spirometry parameters, erythrocyte sedimentation rate, and serum angiotensin converting enzyme levels.

We observed that TLC count was 10500/ cumm of blood, ESR was 54, hemoglobin level was 9.2 g/dL and serum ACE was 148 U/mL. Endobronchial ultrasound-guided Tran's bronchial needle aspiration is a highly effective investigation for mediastinal and hilar lymphadenopathy, and can prevent the need for 87% of mediastinoscopies [9]. Moreover, rapid on-site assessment by well-trained cytologists provides sufficient diagnostic information for the bronchoscopes about the need for additional lymph node passes or Tran's bronchial lung biopsy samples, and might be an alternative to doing endobronchial and Trans bronchial lung biopsy first. F-FDG PET can be used to accurately assess inflammatory activity in patients with unexplained, persistent, disabling symptoms without serological inflammatory activity, and can help to predict pulmonary deterioration at 1 year, and the pulmonary improvement expected after treatment [10].

Conclusion

Sarcoidosis is rare in children. Authors found 23 cases in children. Common clinical features were fever, difficult in respiration, weight loss etc.

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