Case Report

Isolated facial cutaneous sarcoidosis

Sumir Kumar, Ravinder Garg[1], Simmi Aggarwal[2], Jaskanwal Kaur

Departments of Dermatology, [1]Medicine and [2]Radiodiagnosis, GGS Medical College and Hospital, Faridkot, Punjab, India

Address for correspondence:
Dr. Ravinder Garg, Department of Medicine, GGS Medical College and Hospital, Faridkot 151203, Punjab, India E-mail: drravindergarg@yahoo.co.in

Abstract

Isolated cutaneous sarcoidosis is a rare multisystemic granulomatous disorder of unknown etiology. Cutaneous lesions have been classified into specific and nonspecific depending on the presence of noncaseating granulomas on histopathologic studies. Macrophages most likely initiate the response of sarcoidosis by presenting unidentified antigens to CD4+ lymphocytes. A persistent poorly degradable antigen-driven CMI response leads to cytokine cascade, granulomaformation, and fibrosis. In the present study, we report a case of isolated cutaneous sarcoidosis, localized to the face, in an adolescent girl without systemic manifestations which is a rare entity.

Key words: Cutaneous, facial, sarcoidosis

INTRODUCTION

Sarcoidosisis is a rare inflammatory disorder with multisystem presentation which can involve any organ of the body. It is characterized by the formation of noncaseating granulomas primarily affecting the lungs but other organs like heart, central nervous system, and skin may be involved. Skin lesions form tip of an iceberg which may indicate more changes in other body organs. Cutaneous manifestations occur in 25–30% of the patients with sarcoidosis.[1] Cutaneous lesions have been classified into specific and nonspecific depending on the presence of noncaseating granulomas on histologic studies. The lesions are nonspecific if they are not characterized by granulomatous inflammation and specific if they have granulomas histologically.[2] Specific lesions include maculopapules, plaques, nodules, lupus pernio, scar infiltration, alopecia, ulcerative lesions, hypopigmentation, and many others. Most common nonspecific lesion is erythema nodosum. Others include calcifications, prurigo, erythema multiforme, nail clubbing, and sweet syndrome.[3] The potentiality of the body to react in different ways to single disease agent is not better illustrated than sarcoidosis. High degree of suspicion and precision are required for the diagnosis of cutaneous sarcoidosis as it might be overlooked for common conditions such as tinea, psoriasis, etc. The origin of the disease process is not fully understood. The diagnosis is established when clinical findings are supported by histological evidence of noncaseating epitheloid cell granulomas. The disease is rare in pediatric population[4,5] with an estimated incidence of 0.22–0.27 per 1,00,000 children per year.[6]

Access this article online	
Quick Response Code:	Website: www.jnsbm.org
	DOI: 10.4103/0976-9668.95980

CASE REPORT

An 11-year-old girl presented with two raised asymptomatic erythematous lesions over face with the duration of over 1 year. The lesions started as papules and gradually progressed to the present size. There was no history of dyspnea, prolonged fever, or other systemic complaints. The girl was fully immunized including BCG vaccination. There was no significant family history including that of tuberculosis. On examination, there were two nontender annular plaques of size 3 ´ 3 cm^2 each, one lesion on the chin and the other adjacent to the mandibular region [Figures 1 and 2]. On diascopy apple jelly color was present. Hematological and biochemical investigations including angiotensin converting enzyme (ACE) levels, ESR, CRP, ANA, S Ca were within normal limits, the Montoux

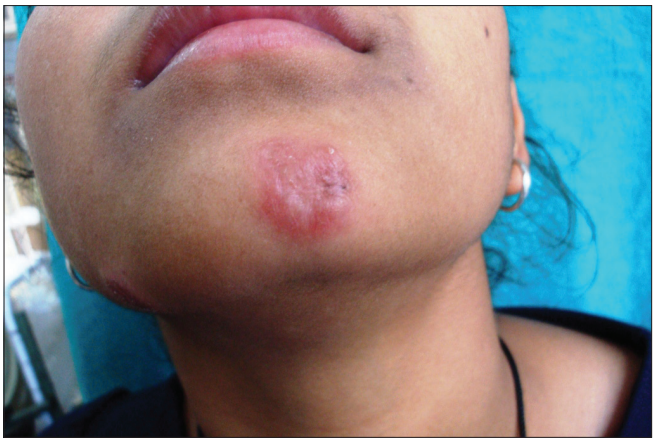

Figure 1: The lesion

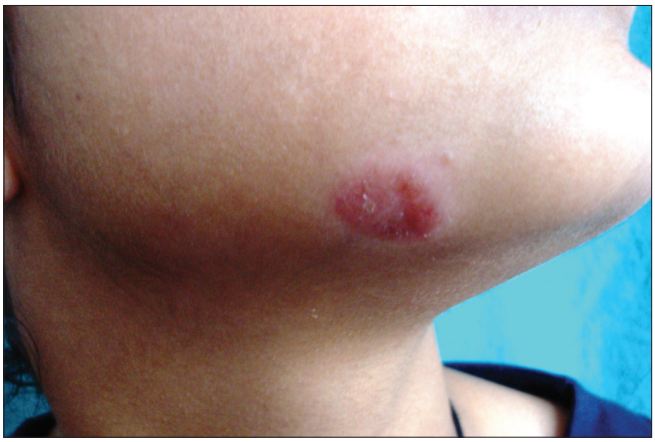

Figure 2: Other view of the lesion

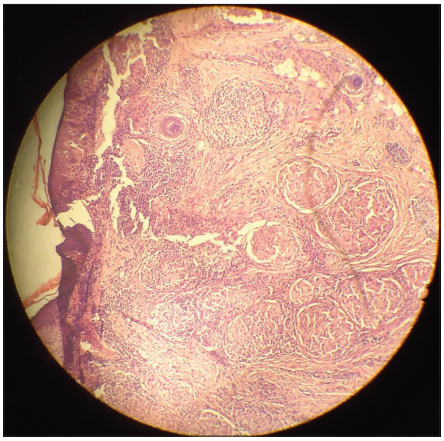

Figure 3: Histopathology

test was negative, histopathology was consistent with sarcoidosis showing noncaseating epitheloid cell granuloma [Figure 3]. X-ray chest, sonography, and CT scan were normal. Her parents denied further invasive procedure such as Fine needle aspiration cytology or bronchial lavage. Ophthalmological assessment was normal. The patient was put on potent topical corticosteroid locally and called for regular follow-up.

DISCUSSION

Sarcoidosis is a multisystem systemic granulomatous disease of unknown etiology that commonly affects young adults who frequently present with hilar lymphadenopathy, pulmonary infilteration, ocular, and cutaneous manifestations. The disease is relatively rare in pediatric population. There are two forms of juvenile sarcoidosis. First early-onset present in 1–5 years of age with the triad of arthritis, uveitis, and pulmonary disease with poor prognosis which is often confused with juvenile rheumatoid arthritis occurring in whites. Second occurring in older children in adolescence which is rare and the clinical course is similar to adult with lung and lymph node involvement. Most reported cases are between 13 and 15 years.

Prevalence of sarcoidosis in adult population ranges from 10 to 40 per 100,000 in USA and Europe.[7] The true incidence and prevalence of childhood sarcoidosis are unknown because of the rarity of the disease and small number of reported cases in childhood. A recent review reported in Danish children younger than 15 years was 0.22–0.27 per 1,00,000 children per year which corresponds to three new cases per year. As in adults, many children with sarcoidosis may be asymptomatic and the disease may remain undiagnosed. Most reported childhood cases have occurred in patients aged 13–15 years with no clear sex predominance. Between 20% and 35% patients of systemic sarcoidosis have skin lesions, but cutaneous sarcoidosis can occur without systemic disease. Cutaneous manifestations occur in 77% of younger children and 24–40% in older children.[8,9] Prevalence of cutaneous sarcoidosis in India is not exactly known because of paucity of recorded Indian/Asian data in the literature.

Various theories and factors are suggested that appear to be implicated in etiopathogenesis, as a cause of the disease is unknown.[10] Antigens which might be involved include infectious agents *Mycobacterium tuberculosis* and other atypical species, *Propionibacterium acnes* and *Chlamydia* species. Mineral dust such as silica and titanium are also implicated. Firefighters are at a great risk of developing sarcoidosis. Genetic factors are also important in defining the pattern of disease presentation, severity, and prognosis of the disease. There is association between class I HLA B 8 antigens and acute sarcoidosis. HLA–DRB I and DQB I have been associated with sarcoidosis. Sarcoidosis susceptible genes are present on chromosome 3 p and 5q 11.2 and protective genes on region of 5p 15.2.

The development and accumulation of granulomas is the main abnormality in sarcoidosis. Granulomas form to confine pathogens, restrict inflammation, and protect surrounding tissue. Granulomas are compact centrally

organized collections of macrophages and epitheloid cells encircled by lymphocytes. There is depression of cutaneous delayed type hypersensitivity and heightened helper T-cell type response at sites of disease. Circulating immune complexes along with signs of B-cell hyperactivity may be found. Most granuloma-associated lymphocytes produce high levels of tumor necrosis factor (TNF), Interleukin 12, IL-15, IL-18, MIP I, MCP I, GM-CSF. The CD4+ lymphocytes and immune effector cells such as macrophages, mast cells, and natural killer cells perpetuate inflammatory response by release of cytokines.

Skin lesions of sarcodosis can be psychologically devastating. Most frequent presentation is soft red to yellowish brown or violaceous flat-topped papules or plaques most frequently on the face. Larger lesions may be found on the trunk, extremities, and buttocks. Erythema nodosum was noted in 31% of patients in a study. Other cutaneous manifestations include lupus pernio which is indurated lumpy violaceous lesions on nose, cheeks, lips, and ears. Other cutaneous lesions include angiolupoid form, scar sarcoidosis, scarring alopecia, lichenoid form, nodular form, mannular form, and subcutaneous sarcoidosis also occur.

No lab test is diagnostic of sarcoidosis. Laboratory evaluation may reveal elevated ESR, anemia, leucopenia, hypercalcemia, or hypercalciuria. The serum level of ACE is elevated in over 50% of children with late-onset sarcoidosis. Chest radiograph may reveal bilateral hilar adenopathy. Bronchoalveolar lavage (BAL) demonstrates the increased number of lymphocytes which are activated helper inducer T-cells. However in children, BAL lymphocytosis does not correlate with disease activity, treatment response, or prognosis so it is not recommended in children.[11] The diagnosis of sarcoidosis is confirmed by demonstrating a typical noncaseating epitheloid cell granuloma on biopsy.[12]

The therapy of choice for cutaneous sarcoidosis with multisystem involvement is topical or systemic corticosteroids. Oral prednisolone is usually initiated at 1–2 mg/kg/day for 4–8 weeks. Asymptomatic patients with bilateral hilar adenopathy may not need systemic steroid therapy. Other drugs used in cutaneous sarcoidosis include hydroxychloroquin, methotrexate, thalidomide, minocycline, and doxycycline but these drugs are not preferred in children. Cutaneous sarcoidosis improves with prolonged application of more than 8 weeks of class 1 topical steroids. Intralesional injection of triamcinolone is more effective. Topical tacrolimus has been effective for cutaneous sarcoidosis in several cases. Electrodessication, pulse dye laser, carbon dioxide laser, and reconstructive surgical procedures have been used successfully to improve cosmetic disfigurement of cutaneous sarcoidosis, but these interventions do not have effect on disease progression.

The prognosis and natural history of sarcoidosis is unclear, because of the rarity of the disease and small number of cases reported. However, the overall prognosis is good as it is usually self-limiting, non-life-threatening disease.

CONCLUSIONS

Isolated cutaneous sarcoidosis is a rare phenomenon for any age group, the diagnosis of which is likely to be missed. The case is being reported for its requirement of precise diagnosis and its rarity as isolated cutaneous involvement localized to the face. The diagnosis should not be overlooked because the disease may have vicious roots underneath.

REFERENCES

1. Abu-Hilar M, Krotva J, Chichierchio L, Obeidat N, Madanat M. Dermatologic aspects and cutaneous manifestations of sarcoidosis G. Ital Dermatol Venrol 2010;145:733-45.
2. Marchell RM, Judson MA. Semin Respir Crit. Care Med 2010;31:442-51.
3. Fernandez-Faith E, McDonnell J. Cutaneous sarcoidosis: Differential diagnosis.J Clin Dermatol 2007;25:276-87.
4. Shetty AK, Gedalia A. Sarcoidosis in children. Curr Probl Pediatr 2000;30:149-76.
5. Hoffman AL, Milman N, Byg KE. Childhood sarcoidosis in Denmark 1979 1994 incidence, clinical features and laboratory results at presentation in 48 children. Acta Pediatr 2004;93:30-6.
6. Milman N, Hottman AL, Byg K-E. Sarcoidosis in children. Epidemiology by Danes, Clinical features, diagnosis, treatmentandprognosis. Acta Paediatr 1998;87:871-8.
7. Hunninghake GW, Costable U, Ando M, Baughman R, Cordier JF, du Bois R, *et al*: ATS/ERS/WASOG statement of sarcoidosis. American Thoracic Society/European Respiratory Society/World Association of Sarcoidosis and other Granulomatous Disorders. Sarcoidosis Vasc Diffuse Lung Dis 1999;16:149-73.
8. Hetherington S. Sarcoidosis in young children. Am J Dis Child 1982;136:13-5.
9. Pattishall EN, Strope GL, Spinola SM, Denny FW. Childhood Sarcoidossis. J Pediatr 1986;108:169-77.
10. Baculard A, Blanc N, Boule M, Fauroux B, Chadelat K, Boccon-Gibod L, *et al*. Pulmonary Sarcoidosis in Children: A follow up study. Eur Repir 2001;17L:628-35.
11. Chadelat K, Baculard A, Grimfield A, Tournier G, Boule M, Boccon-Gibod L, *et al*. Pulmonary Sarcoidosis in Children: Serial evaluation in bronchoalveolar lavage cells duringcorticosteroid treatment. Pediatr Pulmonol 1993;16:41-7.
12. Rosen Y. Pathology of Sarcoidosis. Semin Respir Crit Care Med 2007;28:36-52.

How to cite this article: Kumar S, Garg R, Aggarwal S, Kaur J. Isolated facial cutaneous sarcoidosis. J Nat Sc Biol Med 2012;3:87-9.

Source of Support: Nil. **Conflict of Interest:** None declared.