Case Report

An extremely unusual presentation of isolated extrathoracic sarcoidosis of submandibular lymph node in a child

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ABSTRACT

A 12-year-old male child presented with left submandibular lymphadenopathy; excision biopsy revealed noncaseating granuloma with numerous Schaumann bodies in histopathology, suggestive of isolated extrathoracic sarcoidosis, which is an extremely rare entity in the pediatric age group.

KEY WORDS: Extrathoracic, granuloma, lymphadenitis, sarcoidosis

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INTRODUCTION

A Norwegian dermatologist Caesar Boeck coined the term "sarcoidosis" in 1899.[1] Although this disease affects all age groups, it usually develops before the age of 50 years with a peak incidence during the third and fourth decade of life[2] and a predilection for the female sex. It is a multisystem disease and is under-reported in our country on account of its similarity to tuberculosis, which is further compounded by the lack of awareness among physicians and pathologists. The frequency of detection of this disorder varies between 61.2/100,000 (Delhi) and 150/100,000 (Kolkata),[3] but isolated extrathoracic sarcoidosis in the pediatric age group is rarely reported, which we report here.

CASE REPORT

A 12-year-old boy reported to the outpatient department with the chief complaint of a painless swelling in the left submandibular region since four years. It was a lymph node swelling which had gradually progressed from its initial size of a pea around 1 cm in diameter to its current size of a small lemon around 5 cm in diameter. The swelling was firm, nontender, nonadherent to skin, cold to the touch, and showed no fluctuation or discharging sinus *in situ*. There was no associated history of fever, weight loss, or any other comorbidity. There was no significant family history of a similar illness or tuberculosis. Systemic examination was unremarkable except for the solitary lymph node swelling. Blood counts were within normal range [hemoglobin (Hb): 12 g%; total leukocyte count (TLC): 6,000; differential leukocyte count (DLC): Polymorphs (P): 67%, lymphocytes (L): 31%, eosinophils (E): 2%, monocytes (M): 1%] with an erythrocyte sedimentation rate (ESR) of 5 mm in the first hour. Renal function tests and liver function tests were within normal range. Serum calcium was within the normal limit, and angiotensin-converting enzyme (ACE) was 52 IU/L just at the higher end. Mantoux test showed an induration of 9 mm diameter after 72 hours (the patient had a history of BCG vaccination). Ultrasonography (USG) of the abdomen showed no abnormality; other systems were also normal. Skiagram of the chest in posteroanterior (PA) view and computed tomography (CT) of the thorax was also normal for the age [Figure 1]. Systemic examination was unremarkable. Fine needle aspiration cytology (FNAC) of the lymph node showed granulomatous inflammation. The lymph node was excised under general anesthesia, and histopathologic study revealed noncaseating granulomatous inflammation [Figure 2] with numerous Schaumann bodies present in the background suggestive of sarcoidosis [Figure 3]. The site of excision was completely healed one week after the excision, and the patient was put on systemic steroid therapy.

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DOI: 10.4103/0970-2113.142154

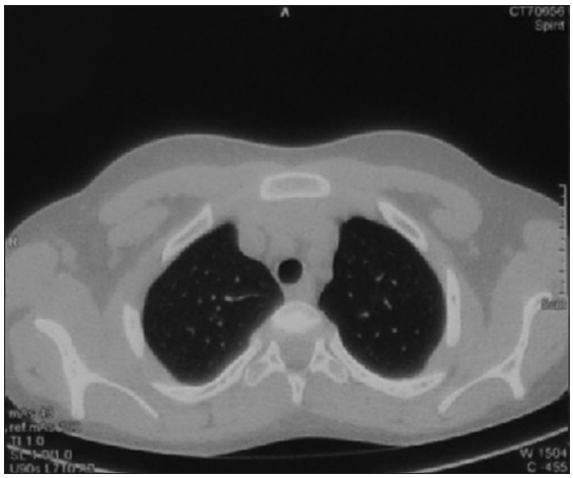

Figure 1: Computed tomography of thorax: Normal for age

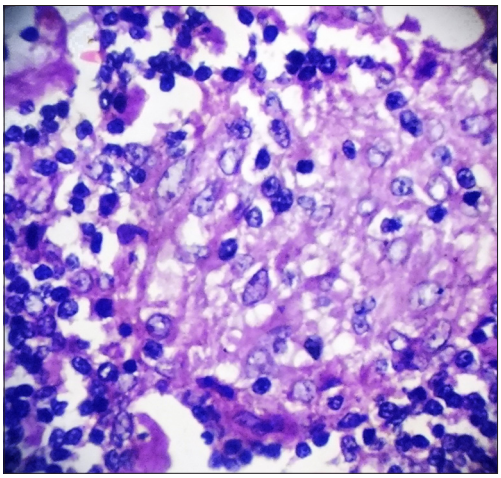

Figure 2: Giant cell noncaseating granuloma

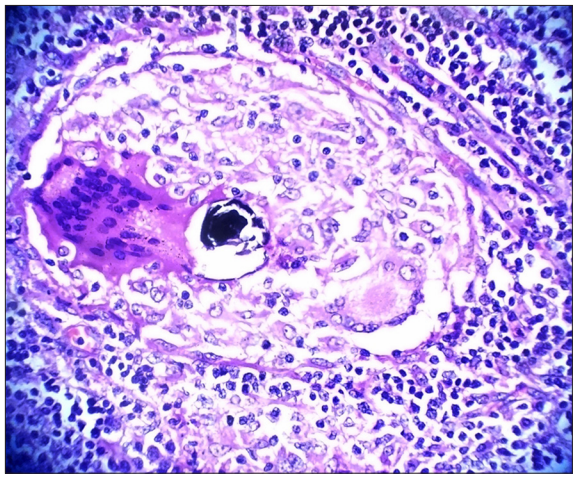

Figure 3: Granuloma with Schaumann bodies in multinucleated giant cell: ×400 magnification

DISCUSSION

Sarcoidosis was first identified by two dermatologists working independently, Dr. Jonathan Hutchinson in England and Dr. Caesar Boeck in Norway. Hence, it was originally called Hutchinson's disease or Boeck's disease.[4] Sarcoidosis is less common in children than in adults with an incidence of 0.06 cases per 100,000 in children below four years of age, gradually increasing to 1.02 cases per 100,000 in adults in a Danish study.[1] It commonly involves the lung, eyes, and skin, but in 2% cases, isolated extrathoracic sarcoidosis has been observed. It is also reported that extrathoracic sarcoidosis is more common in females than in males (36.7% vs. 28.6%).[5,6] Although the etiology of sarcoidosis is unknown, various studies have proven its association with exposure to irritants found in the rural setting like wood-burning, tree pollen, inorganic particles, insecticides, and moldy environment. *Mycobacterium*[7] and *Propionibacterium*[8] have also been reported as etiological factors. In recent studies, association has also been found between class 1 HLA-B8 (HLA: Human Leukocyte Antigen) and acute sarcoidosis.[1]

Sarcoidosis is characterized by its pathological hallmark: The noncaseating granuloma. A variety of inclusions like crystalline inclusions and colorless refractile crystals composed predominantly of calcium oxalate are frequently found in the giant cells of the granuloma of sarcoidosis. These many serve as nidi for deposition of calcium leading to the formation of Schaumann (conchoidal) bodies.[5] Typical laboratory findings of sarcoidosis like ACE, lysozyme, and calcium were nondiagnostic in this patient. USG of the abdomen and CT of the thorax are also important modalities in diagnosis, but the diagnosis was established only by histopathological examination of the excised lesion. The present case study is extremely rare in the context of its isolated extrapulmonary location and involvement of the pediatric age group. Although a few case reports of isolated extrapulmonary sarcoidosis have been reported by clinicians, in the pediatric age group, such a unique case has not been reported to the best of the authors' knowledge.

Being a systemic disease, sarcoidosis can affect any organ in the body, but isolated sarcoidosis may sometimes create a diagnostic dilemma. The high prevalence of tuberculosis in our country prevents the specialist from confirming the cause of lymphadenitis by excision biopsy, and the same is responsible for reporting fewer cases of sarcoidosis in this part of the world. The key message delivered by the present case is to perform excision biopsy to rule out other causes of lymphadenitis and to confirm the diagnosis before starting antitubercular therapy on clinical suspicion alone.

REFERENCES

1. Michael CL, Benjamin AR, Alvin ST. Sarcoidosis. N Engl J Med 2007;357:2153-65.
2. Weisinger H. Sarcoidosis: Case report and review. Clin Exp Ophtom 2006;89:361-7.

3. Joshi JM, Saxena S. Sarcoidosis in India. Med Update 2012;22:408-12.
4. Gupta N, Rajwanshi A, Gupta D. Schaumann body in a case of sarcoidosis diagnosed as transbronchial FNAC. J Cytol 2011;28:88-9.
5. Marc AJ. Extrapulmonary sarcoidosis. Semin Respir Crit Care Med 2007;28:83-101.
6. Giovinale M, Fonnesu C, Soriano A, Cerquaglia C, Curigliano V, Verrecchia E, *et al*. Atypical sarcoidosis: Case reports and review of the literature. Eur Rev Med Pharmacol Sci 2009;13 Suppl 1:37-44.
7. Shah JR. Tuberculosis Sarcoidosis. Lung India 2007;24:83-6.
8. Eishi Y. Etiologic link between Sarcoidosis and Propionibacterium acnes. Respir Investig 2013;51:56-68.

How to cite this article: Agrawal A, Singh K, Parihar D, C. An extremely unusual presentation of isolated extrathoracic sarcoidosis of submandibular lymph node in a child. Lung India 2014;31:413-5.

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