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CASE REPORT

# Clinically unsuspected tonsillar sarcoidosis in a child revealed by routine histologic examination

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**Summary** In most hospitals, making a microscopic examination to tonsil and adenoid specimens from pediatric patients who present recurrent infections has become a standard practice. However, studies have shown that systematic examination of tonsils and adenoids was not needed for individual aged 21 or less. In this context, we describe the third report of an unsuspected tonsillar sarcoidosis revealed by histologic examination. A 5-year-old white girl was referred to our department because of frequent nasal and pharyngeal infections. Both tonsillectomy and adenoïdectomy were performed. Histologic evaluation revealed noncaseous epithelioid granulomas morphologically compatible with a diagnosis of sarcoidosis. Our observation illustrates the benefit of histological analysis in every tonsillectomy and adenoïdectomy specimen.


## 1. Introduction

Sarcoidosis is a systemic granulomatous disease of unknown etiology, generally affecting young or middle-aged adults. Most frequently, it presents with bilateral hilar lymphadenopathy, pulmonary infiltration, skin or ocular lesions [1]. It is quite uncommon in children [2]. Involvement of the Waldeyer's ring alone is very rare as well. Moreover, only two reports of unsuspected sarcoidosis revealed by routine histologic examination of tonsillectomy specimens can be found in the literature [3,4]. The case

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we report here is that of a 5-year-old girl who developed a sarcoidosis involving both tonsils and adenoids.

## 2. Case report

This young patient was referred to the E.N.T. department of our hospital because of frequent nasal and pharyngeal infections. She was born in Columbia but has been living in Belgium for the last 3 years. She had a history of snoring at night, chronic mouth breathing and had many documented episodes of tonsillitis over a 2-year period, often requiring antibiotic therapy. Examination of the oropharynx revealed enlarged cryptic tonsils without asymmetry. Rhinopharynx X-ray films showed a noticeable hypertrophy of the adenoid pad. She was scheduled for both tonsillectomy and adenoïdectomy. The results of preoperative blood tests were within normal limits. She went through a classic tonsillectomy and adenoïdectomy and recovered normally.

## 3. Results

Histologic evaluation of the surgical specimens revealed the presence of multiple noncaseous granulomas morphologically consistent with a diagnosis of sarcoidosis (Fig. 1). No acid-fast bacilli were discovered using the Ziehl—Neelsen stain. Special stains for fungi were also negative. No palpable adenopathy was found after repeated physical examination. Chest X-ray films did not show any anomaly. Complementary serologic investigations (*Borrelia burgdorferi*, the agent of *toxoplasmosis*, *Bartonella hensellae*) as well as the tuberculosis skin test were also negative. Iontophoresis was normal. Elevated serum titers of angiotensin converting enzyme (ACE) were found: 89 U/L (normal range: 8—52 U/L). Due to the absence of systemic involvement, no further therapy was given. There was no sign of clinical disease 6 months after surgery, but until now the ACE level remained slightly elevated (74 U/L).

## 4. Discussion

Sarcoidosis is a chronic and systemic granulomatous disorder of unknown etiology, histologically characterized by noncaseating granulomas [1]. It has been assumed that a yet unidentified antigenic stimulus and a genetic predisposition might produce such a granulomatous reaction [5]. The clinical and

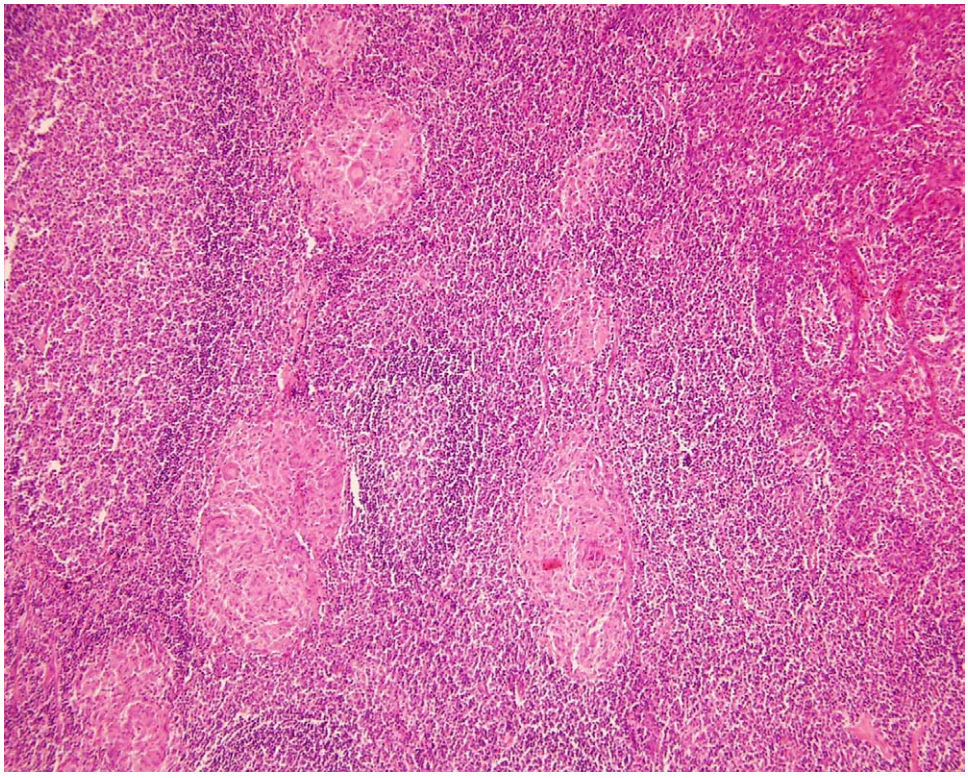

**Fig. 1** This low power view shows a few granulomas. These are located in the interfollicular area between the germinal center (on the left) and the lymphoepithelium (on the right).

histological features of tuberculosis and sarcoidosis are more or less similar and it has been suggested that *Mycobacterium tuberculosis* might be the etiologic agent of sarcoidosis. Isolation of *M. tuberculosis* DNA from sarcoid granulomas first sustained such hypothesis [6]. However, more sophisticated DNA extraction techniques did not confirm this finding [7,8]. In a recent review of 22 cases of tonsillar granulomas, Kardon and Thompson stated that isolated tonsillar sarcoidosis may represent a peculiar granulomatous response to the usual pathogens responsible for chronic tonsillitis. In this context, the history of recurrent tonsillitis in our patient may support this last proposal [9].

In the United States, the prevalence of sarcoidosis is approximately 5 per 100,000 in Caucasians and 40 per 100,000 in African—Americans [10]. A study of pediatric cases found no difference in sex ratio [2,10].

The diagnosis of sarcoidosis is based on the histologic demonstration of noncaseating granulomas but also on the results of several laboratory tests [12]. Noncaseating granulomas may contain multinucleated giant cells, and central necrosis is occasionally seen. The Kveim's reaction has been the test traditionally used for the diagnosis of sarcoidosis. This test requires the intradermal injection of an antigenic extract prepared from spleens of patients with known sarcoidosis. A positive test is characterized by the development of a nodule at the site of the injection within 4—6 weeks. Histologic analysis of this nodule shows the presence of typical noncaseating granulomas. This diagnostic procedure is positive in 80% of patients with sarcoidosis and there are approximately 2% false-positive cases [4]. Abnormal laboratory findings may include high erythrocyte sedimentation rate (ESR), anemia, leukopenia or eosinophilia. Hypercalcemia or hypercalciuria may also be found [2,13]. Serum titers of angiotensin converting enzyme are typically high in up to 80% of children with late-onset sarcoidosis [14]. Epithelioid histiocytes in granulomas are thought to be the source of serum ACE in sarcoidosis. Study of ACE titers is not only useful for the diagnosis of sarcoidosis but it also allows a precise evaluation of the course of the disease and its response to therapy in older children and adults [15]. In early-onset sarcoidosis, Gedalia et al. have shown that serum ACE is linked with disease activity in children [16]. Such a test is not entirely specific for sarcoidosis and many other disorders may be associated with increased serum ACE activity, like miliary tuberculosis, leprosy or Gaucher disease.

The clinical evolution of sarcoidosis is highly variable and spontaneous recovery may be observed in a significant number of patients. The lung is the organ most commonly affected. At least 60% of patients have abnormal chest radiograph showing the classical features of bilateral hilar lymphadenopathy with or without parenchymal involvement [17]. The most common physical sign of reticuloendothelial involvement is peripheral lymph node enlargement, which is also the most accessible site for diagnostic biopsy [10]. In children, ocular involvement is extremely frequent and conjonctival biopsy is sometimes required for the diagnosis of this disorder [10].

Sarcoidosis of the head and neck area is relatively rare, accounting for 2—18% of generalized cases [18]. It can affect the ear, sinonasal region, salivary glands, pharynx, larynx and tonsils [18]. In this regard, tonsillar involvement in systemic sarcoidosis is well described to the extent that some authors have suggested tonsillar biopsies as a useful diagnostic procedure [19].

The fortuitous detection of isolated sarcoidosis of the Waldeyer's ring by histological examination is a very rare event [4]. We only found two other similar case reports in our literature review [3,4]. A point of controversy also exists about the opportunity of systematically examining every tonsil resected. Some authors state that this is useless and a waste of time and resources, considering the low yield of significant histologic findings in routine tonsillectomy specimens (22 of 13,700 cases: 0.16%) [9—20]. At the contrary, our observation further illustrates the interest of such a systematic examination, as previously proposed by Yarington et al., and Erwin [3,4]. Those cases of granulomatous disease in addition to the two cases of non-Hodgkin's lymphoma diagnosed by Garavello et al. in a series of 1123 patients are strong support for routine histopathologic examination of all tonsil and adenoid specimens [21]. In this context, the suction diathermy adenoïdectomy technique which has gained in popularity over recent years [22] presents the disadvantage that little or no pathologic specimens are retrieved for routine histologic examination. The possibility of missing an occult neoplasm or a granulomatous disease and the related medicolegal implications that such a situation may involve certainly argue in favor of systematic histologic examination of all tonsil and adenoid samples.

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