# A Rare Incidental Diagnosis After Adenoidectomy: Sarcoidosis

Case Report

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**Abstract**

Sarcoidosis is a systemic disease of unknown etiology. It is characterized by non-caseating granulomatous inflammation. It most commonly affects the pulmonary and intrathoracic lymph nodes. Isolated nasopharyngeal involvement is very rare. Pediatric sarcoidosis and isolated nasopharyngeal involvement are rare entities. Symptoms of nasopharyngeal involvement can mimic adenoid hypertrophy. In this case report, we present a nine-year-old female who was diagnosed coincidentally with sarcoidosis with the adenoidectomy specimen.

**Keywords:** Sarcoidosis, nasopharynx, adenoidectomy, case report

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## Introduction

Sarcoidosis is a systemic disease of unknown etiology, characterized by non-caseating granulomatous inflammation (1, 2). Its incidence has been reported to range from 0.1 to 109 per 100,000 and to be more common in females (1, 2). Sarcoidosis is most commonly encountered between the ages of 20 and 40 years (2). Approximately 2% of the patients with sarcoidosis are younger than 10 years old. The disease may be asymptomatic or can represent varying symptoms depending on the organs involved. It most commonly involves the pulmonary and intrathoracic lymph nodes. Extrapulmonary involvement has been reported in approximately 35% (3). The most common extrapulmonary involvements are skin lesions such as erythema nodosum. Eye, skin, liver, bone, joint, heart, and brain involvements are other examples of extrapulmonary lesions (2). Approximately 9% of the cases are observed in the head and neck. Only 1% of the cases are encountered in the sinonasal region (4). Isolated nasopharyngeal localization is extremely rare and only a few cases have been reported in the literature (5). In this case report, we

present a pediatric patient who was diagnosed coincidentally with sarcoidosis after adenoidectomy.

## Case Presentation

A nine-year-old female patient was admitted to our tertiary ear nose and throat clinic with complaints of mouth breathing, snoring, and nasal blockage. The patient did not have a cough or dyspnea. These symptoms were present for several years. The patient did not have any history of recurrent tonsillitis or allergic rhinitis; she had no history of chronic diseases, surgery, or chronic drug use. She had no family history of chronic disease. The patient's tonsils were grade 2 according to the Brodsky scale. She had no tonsillar asymmetry. Anterior rhinoscopy and bilateral ear examinations were normal. The fiberoptic endoscopic nasopharyngeal examination was consistent with adenoid vegetation. The lesion almost completely blocked the nasopharyngeal airway. Neck examination was normal, there were no palpable lymph nodes. There were no skin eruptions. Transoral adenoidectomy was performed with curettes and angled cup forceps under general anesthesia. The patient was discharged one day after the operation with oral antibiotics and analgesics. Histopathological examination reported non-caseating granulomatous inflammation areas and reactive lymphoid hyperplasia (Figure 1). Decision was made to investigate the patient in terms of granulomatous diseases because of the non-caseating granulomatous lesion reported in the histopathological examination. The patient was referred to the pediatric rheumatology department. The rheumatology department investigated the patient for serum angiotensin-converting enzyme (ACE), immunoglobulin G (IgG) and blood calcium levels and found ACE: 72.2 U/L, IgG: 15.26 g/L, and blood calcium level 9.52 mg/dL (reference values: 8.0-52.0 U/L, 5.72-14.74 g/L, 8.5-10.4 mg/dL, respectively). Anti-cytomegalovirus IgG, Epstein–Barr virus (EBV)-EBNA IgG, and EBV-VCA IgG were positive. Complete blood tests, complete urinalysis, other serological tests, and routine biochemical tests were normal. Tuberculin skin test was negative. No pathology was observed in the chest X-ray and the patient was accepted as Stage 0 according to the Siltzbach Classification. The patient was diagnosed with sarcoidosis based on tissue diagnosis and blood parameters after other possible pathologies were eliminated. In the pediatric rheumatology department, treatment was not initiated, and follow-up was recommended because the patient had no systemic involvement and no active complaints. The patient's airway obstruction had improved in the 6$^{th}$ month of the follow-up. She had no signs of recurrence in the nasopharynx and had no systemic symptoms related to sarcoidosis.

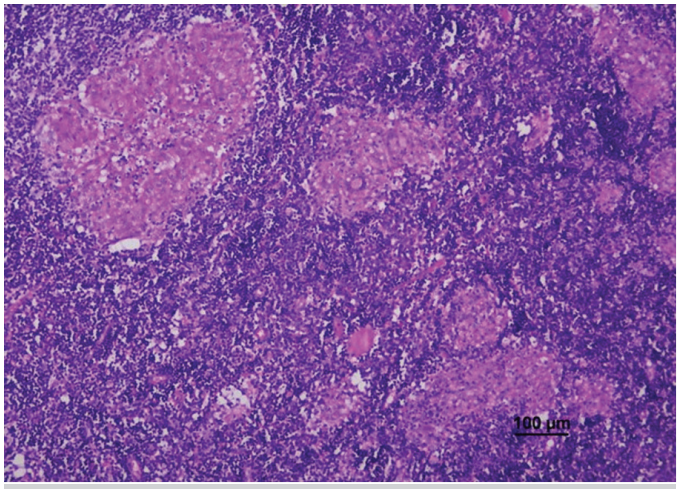


**Figure 1.** Image of histopathological examination of the adenoidectomy specimen, granulomas are seen (100x, Hematoxylin & Eosin)

Informed consent was obtained from the parents of the patient for this case report.

## Discussion

While the symptoms of sarcoidosis are quite variable depending on the organs involved, shortness of breath, chest pain, and cough are the most common. Systemic symptoms may include weakness, fever, weight loss, and arthralgia. Approximately 16% of the cases are asymptomatic at diagnosis (2). Since sinonasal involvement presents non-specific rhinitis symptoms, the diagnosis of sarcoidosis can be missed due to possible pathologies such as septum deviation, allergic rhinitis, turbinate hypertrophy in the same patients (6). In their 2022 article Benettini et al. (7) presented two cases of nasopharyngeal sarcoidosis and reviewed the relevant literature. They identified 27 cases of nasopharyngeal sarcoidosis reported between 1952 and 2020. Nasal obstruction was the most frequently reported symptom among these cases. There was no significant gender difference, and mean age at diagnosis was 35 years. There were lung and/or intrathoracic lymph node involvement in 16 of the cases. Four of these cases were in the pediatric age group, ages were 5, 12, 13 and 15 years, and the gender distribution was equal. Two of the pediatric cases had lung and/or intrathoracic lymph node involvement. Systemic treatment was required in two of the four cases (7). Also in our case, sarcoidosis caused nasal obstruction by creating adenoid hypertrophy, which is a very rare clinical condition for this disease.

The diagnosis of sarcoidosis is defined by the presence of appropriate clinical and radiological findings, histological demonstration of non-caseating granulomatous inflammation, and elimination of other granulomatous diseases (2, 8). Chest radiography is sufficient for radiological diagnosis, and follow-up. High serum ACE activity is present in 40–90% of the active sarcoidosis cases. ACE activity can also reflect the total burden of granulomas (8). The tuberculin skin test is negative in 30–70% of the cases. In

their study, Güngör et al. (2) found negative tuberculin skin tests in 51.6% of the patients. Serum calcium concentrations have been reported to be elevated in 2% to 10% of patients with sarcoidosis.

In a study conducted with 27 pediatric sarcoidosis patients, the most common blood parameters were ACE elevation (74%), IgG elevation (64%), anemia (54%), high erythrocyte sedimentation rate (51%), and hypercalcemia (12%) (9). The chest X-ray of our patient was normal. She did not have lower respiratory tract symptoms, so thorax computed tomography was not done. She had high ACE and IgG values which supported the diagnosis of sarcoidosis. She did not have anemia, increased erythrocyte sedimentation rate, or hypercalcemia. Her tuberculin skin test was also negative.

One of the diagnostic criteria for sarcoidosis is the demonstration of non-caseating granulomatous inflammation in tissue samples (2, 8). Biopsy should be taken from the most easily accessible tissue (2). In our case, sarcoidosis was detected coincidentally during the histopathological examination of the adenoidectomy specimen. While pathology reports of the adenoidectomy samples mostly indicate lymphoid hyperplasia, different pathologies can also be rarely detected, as in our patient. Current studies on adenoidectomy specimens recommend histopathological examination when atypical pathology is suspected. Some authors suggested that histopathological examination would not be necessary if there is no preoperative risk factor (10). But we believe that every surgical specimen should pathologically be examined to be able to diagnose any rare and differentiated conditions as was the case in our patient. Otherwise, the diagnosis of some rare systemic diseases involving lymphoid tissue can be missed, and this would cause delays in the diagnosis and the treatment of the patient.

Corticosteroids are the first choice of treatment in patients with symptomatic sarcoidosis (8). Sarcoidosis has high spontaneous remission rates. Given the possible side effects of the treatment, asymptomatic or mild symptomatic cases should be followed closely before initiating the treatment. Medical treatment should be considered for symptomatic or progressive patients (2, 8). There are also local treatment options with corticosteroids, such as intralesional administrations. Nasal steroids are a treatment option for localized nasal disease (5). The symptoms of our patient regressed after adenoidectomy. Therefore, systemic and topical treatment was not used and the patient was followed closely.

Sarcoidosis generally has a good prognosis (2). In the study of Tunçay et al. (8), 57% of 338 patients were followed without treatment, and spontaneous remission was observed in 30%. During the six-month follow-up of our case nasal obstruction resolved and no recurrence was observed in the nasopharynx.

## Conclusion

Pediatric sarcoidosis and isolated nasopharyngeal involvement are rare entities. Symptoms of nasopharyngeal involvement can mimic adenoid hypertrophy. Histopathological examination of adenoidectomy specimens is important, and we believe that every surgical specimen should be examined to avoid diagnostic delays.

**Informed Consent:** Informed consent was obtained from the parents of the patient for this case report.

**Peer-review:** Externally peer-reviewed.

**Authorship Contributions**

Surgical and Medical Practices: M.E.S., V.A., M.D., Concept: M.E.S., V.A., M.D., Design: M.E.S., V.A., M.D., Data Collection and/or Processing: M.E.S., V.A., M.D., Analysis and/or Interpretation: M.E.S., V.A., M.D., Literature Search: M.E.S., V.A., M.D., Writing: M.E.S., V.A., M.D.

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**Main Points**

- Sarcoidosis is a systemic disease of unknown etiology and characterized by non-caseating granulomatous inflammation.
- Approximately 2% of the patients with sarcoidosis are younger than 10 years old.
- The rate of involvement of the head and neck region is approximately 9%. Isolated nasopharyngeal involvement is extremely rare and only a few cases have been reported to date.
- Symptoms of nasopharyngeal involvement can mimic adenoid hypertrophy.

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