

Nasopharyngeal Sarcoidosis: A Rare Location

Nazofaringeal Sarkoidoz: Nadir Bir Lokasyon

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ABSTRACT Sarcoidosis is a multisystemic disease of unknown etiology; which rarely involves the upper respiratory tract. 2-18% of generalized sarcoidosis cases hold the ear-nose-throat system. It can affect the ear and temporal bones, sinonasal region, salivary glands, pharynx, tonsils and larynx. It may lead to hoarseness, dysphagia, laryngeal paralysis or upper airway obstruction according to location of the disease. The nasopharyngeal involvement is also very rare in sarcoidosis. Localised nasal forms can be treated with topical corticosteroids and intranasal injections. Surgery may be an alternative in obstructive cases, or those resistant to medical treatment. In this article, we report a 31-year-old male with nasopharyngeal involvement of sarcoidosis.

Keywords: Nasal obstruction; nasopharynx; sarcoidosis

ÖZET Sarkoidoz nadiren üst solunum yolunu tutan etyolojisi bilinmeyen multisistemik bir hastalıktır. Tüm sarkoidoz vakalarının %2-18'i kulak burun boğaz bölgesini tutmaktadır. Sarkoidoz, kulagi ve temporal kemikleri, sinonazal bölgeyi, tükürük bezlerini, fareks, tonsilleri ve larinksi etkileyebilir. Tutulum yaptığı bölgeye göre bulgu veren sarkoidoz ses kısıklığı, yutma güçlüğü, larengeal paralizisi ve üst solunum yolunda tıkanıklığa yol açabilmektedir. Nazofaringeal tutulumu da sarkoidozda oldukça nadirdir. Lokalize nazal formlar topikal kortikosteroidlerle ya da intralezional enjeksiyonlar ile tedavi edilebilir. Cerrahi tedavi, obstrüktif vakalarda ve tıbbi tedaviye dirençli vakalarda alternatif olabilir. Biz bu yazıda, sarkoidoz nedeni ile nazofarenkste kitle tespit edilen 32 yaşında bir erkek hastayı rapor ettik.

Anahtar Kelimeler: Nazal tıkanıklık; nazofarinks; sarkoidoz

Sarcoidosis is a chronic, multisystemic and granulomatous disease. The etiology is unknown. Upper respiratory tract involvement is extremely rare, seen approximately in 6% of systemic sarcoidosis.^{1,2} Diagnosis requires exclusion of nasopharyngeal malignancy and other diseases with similar clinical manifestations and histologic properties such as amyloidosis, tuberculosis, histoplasmosis, and syphilis. This usually is a diagnosis of exclusion. Therefore the tissue samples should be taken from any suspicious region in the head and neck area and following a systematic scan. Treatment should initially be conservative and always individualized.³

In this article, we report the clinical features and imaging findings of nasopharyngeal sarcoidosis.

CASE REPORT

A 31-year-old male patient was admitted to our clinic with nasal obstruction. The patient had been well until about one year before admission, when he began complaining of poor appetite and weight-loss.

Bilateral inferior turbinate hypertrophy and obstructive mass in the nasopharynx are detected on endoscopic nasal examination. Magnetic resonance imaging (MRI) of the nasopharynx revealed an obstructive mass in the nasopharynx (Figure 1). Outfracturing of the inferior turbinates, nasopharyngeal biopsy as well as adenoidectomy is planned. Preoperative laboratory values were normal. Written informed consent for these procedures was obtained from the patient. Adenoidectomy and inferior turbinate outfracture was performed. Nasopharyngeal mass was sent for histopathologic examination, which showed granulomatous inflammation (Figure 2). Acid resistant bacteria (ARB) staining was negative for the specimen.

Further examination revealed normal blood and urine calcium levels. Serum angiotensin converting enzyme level was slightly elevated. Thoracic computed tomography (CT) showed presence of mediastinal lymphadenopathies. The patient was referred to the Pulmonology Department for further investigation. Prednisolone (40 mg/day) has been administered by pulmonology department as there are systemic symptoms. The patient's appetite was restored, and his weight is currently stable. Although he suffers occasional low-grade fever and malaise, his general condition has been good to date. At the 6th month follow-up, the nasopharyngeal region was open. The ground-glass attenuation

of the lung and the bilateral hilar lymphadenopathy greatly reduced.

DISCUSSION

Primary involvement of the nasopharynx is extremely rare. Very few cases were reported in the literature. Nasopharynx cancer, tuberculosis, amiloidosis, sarcoidosis, Tangier's disease are certain pathologies that may primarily involve nasopharynx. The nasopharynx have no afferent lymph vessels, so metastases to the nasopharynx are extremely rare. In particular, renal cell carcinomas are known to metastasize to the nasopharynx.^{1,4,5}

Sarcoidosis is a multisystem granulomatous disorder of unknown cause. Presenting features of sarcoidosis ranging from asymptomatic but abnormal findings on chest radiography in many patients to progressive multiorgan failure. On chest radiography, pulmonary involvement with enlargement of the hilar lymph nodes and swelling of the peripheral lymph nodes might be seen as early signs. However, virtually any organ or tissue maybe involved in cases with disseminated granulomas. Symptoms caused by pulmonary, cardiac, neural, gastrointestinal, hepatic, renal, cutaneous, ophthalmic, and endocrine involvement manifest in later stages of the disease.^{1,5}

Sarcoidosis of the ear-nose-throat system is relatively rare. It can affect the ear and temporal

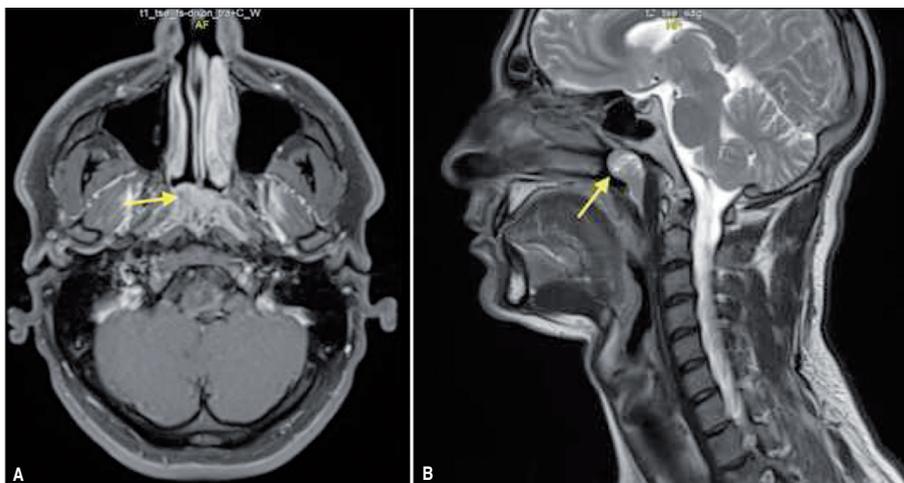


FIGURE 1: MRI axial (A) and sagittal (B) section showing nasopharyngeal mass (yellow arrow).

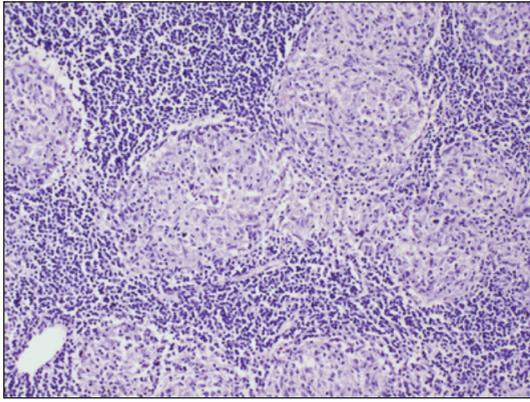


FIGURE 2: Histopathologic section, showing respiratory epithelium with granulomas in lymphoid stromae (HE, x100).

bones, sinonasal region, salivary glands, pharynx, tonsils and larynx. Among cases of ear-nose-throat sarcoidosis, nasal mucosal lesions are frequently detected (69%), while laryngeal and pharyngeal involvement are less frequent (15%).³ Nasal cavity involvement is present in all cases with laryngeal and pharyngeal disease. The present case did not have cutaneous involvement, in contrast to the high prevalence of skin invasion in reported cases (92%) of ear-nose-throat sarcoidosis.⁴

To date, nasopharyngeal involvement of sarcoidosis has been reported in a small number of cases. However, the actual number of such cases may be underestimated, because otorhinolaryngologic examination is not generally performed in patients with systemic sarcoidosis. On the other hand, patients with sarcoidosis often initially seek treatment from an otolaryngologist, because the earliest signs and symptoms of sarcoidosis may be identical to those of other forms of chronic sinonasal inflammation.^{1,5}

Although the exact etiology of sarcoidosis remains obscure, its development is attributed to an excessive, antigen-driven cellular immune response occurring within target organs, and promoting nonspecific systemic inflammation. Since the pharyngeal tonsil is exposed to antigens in inspired air, immunologic activation may be occurring at this site, subsequently leading to systemic sarcoidosis. In this respect, it merits emphasize that a careful and detailed otorhinolaryngologic examination including biopsy of the nasopharynx should

be performed routinely in any patient suspected of having systemic sarcoidosis. This way, the lesion can be biopsied, and even excised without difficulty via transnasal approach. The patients should be directed to related departments for the further investigation.^{1,5}

There are few reports in the literature of palatine tonsillar and nasopharyngeal involvement due to sarcoidosis. Saussez et al. and Yarrington et al. reported a cases of incidentally identified tonsillar and adenoidal sarcoidosis.^{6,7} Gil Galero et al., El Bousaadani et al., Akin et al., Wilson et al. and Tugrul et al. described sarcoidosis patients which were diagnosed after adenoidectomy.^{1,4,5,8,9}

In an epidemiologic study at the Mayo Clinic, 9% of all sarcoidosis patients had involvement of head and neck area. Another study reported the percentage of patients with had head and neck involvement of sarcoidosis as 3% (n=736).¹⁰

James et al. reported that 36 patients of 818 multisystem sarcoidosis patients exhibited involvement of the sinonasal mucosa.¹¹ In nasal examination may be seen yellow mucosal papules, crusting and granulomatous mass. In our patient the endoscopic nasal examination was normal except for the inferior turbinate hypertrophy.

Epiglottis is the most commonly involved structure in laryngeal sarcoidosis. Involvement of the vocal cords and subglottic region are rare. These involvements can lead to stridor, dyspnea, dysphonia, dysphagia.⁹

Treatment depends on the severity of symptoms associated with sinonasal disease and involvement of other organs. In topical treatment, nasal steroids, lubricant drops and nasal washing to reduce dryness and intralesional steroid injections in severe symptoms are applied. Systemic steroid applications should be preferred in cases of more severe and systemic diseases with destructive changes. Methotrexate, azothiopyrine, chlorokine, thalidomide, pentoxifylline or cyclophosphamide are the recommended drugs in cases where high dose steroid-resistant and high-dose steroids cannot be continued or where steroid doses should be reduced. Surgery may be an alternative in obstruc-

tive cases, or those resistant to medical treatment. Long-term and interdisciplinary follow-up is necessary because, despite long and aggressive treatment, relapse and chronicity are frequent after reduction or discontinuation of corticosteroid therapy.¹²

The patients with nasopharyngeal involvement of sarcoidosis have nonspecific symptoms such as nasal obstruction and snoring. In conclusion nasopharyngeal sarcoidosis is extremely rare, it should also be kept in mind in differential diagnosis of nasopharyngeal lesions.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Levent N. Özlüoğlu; **Design:** Ozan Erol, Ceren Baş; **Control/Supervision:** Levent N. Özlüoğlu; **Data Collection and/or Processing:** Ozan Erol; **Analysis and/or Interpretation:** Ozan Erol; **Literature Review:** Ceren Baş; **Writing the Article:** Ozan Erol, Ceren Baş; **Critical Review:** Levent N. Özlüoğlu; **References and Fundings:** Ozan Erol, Ceren Baş; **Materials:** Levent N. Özlüoğlu, Ozan Erol, Ceren Baş.

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