CASE REPORT

Extensive upper respiratory tract sarcoidosis

Mafalda Trindade Soares,¹ Carolina Sousa,² Luísa Garanito,¹ Filipe Freire¹¹Otorhinolaryngology Department, Prof. Doutor Fernando Fonseca Hospital, Amadora, Portugal²Pneumology Department, Prof. Doutor Fernando Fonseca Hospital, Amadora, Portugal**Correspondence to**
Dr Mafalda Trindade Soares,
mafaldamctsoares@gmail.com

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SUMMARY

Sarcoidosis is a chronic granulomatous disease of unknown aetiology. It can affect any part of the organism, although the lung is the most frequently affected organ. Upper airway involvement is rare, particularly if isolated. Sarcoidosis is a diagnosis of exclusion, established by histological evidence of non-caseating granulomas and the absence of other granulomatous diseases. The authors report a case of a man with sarcoidosis manifesting as a chronic inflammatory stenotic condition of the upper respiratory tract and trachea.

BACKGROUND

Upper airway sarcoidosis is a rare condition. It took nearly 5 months to achieve the diagnosis for this patient. The authors believe that physicians should be alerted to this possibility in order to obtain diagnosis as early as possible in future cases.

CASE PRESENTATION**Introduction**

Sarcoidosis is a chronic granulomatous disease of unknown aetiology that primarily affects adults between 25 and 45 years of age.^{1 2} It was first described by John Hutchinson, a dermatologist, in 1875, and was later named Besnier-Boeck-Schaumann disease, for the contribution of these doctors to the investigation.³ Sarcoidosis occurs more often in females, with an overall estimated incidence of approximately 16.5/100 000 in males and 19/100 000 in females.⁴ Sarcoidosis is considered to be the consequence of a chronic immunological response, which is associated with a genetic susceptibility and specific infectious or environmental factors.⁴ Sarcoidosis can affect any organ or system, but the lung and intrathoracic lymph nodes are the most commonly affected.¹ Systemic sarcoidosis presents with otolaryngological manifestations in 2–15% of patients, with laryngeal involvement occurring in only 1–5%.^{1 5} Thus, upper airway involvement is rare, particularly if isolated. A sarcoidosis diagnosis requires a high level of suspicion and is performed by histological examination of non-caseating granulomas of the sarcoid type and exclusion of other granulomatous diseases.⁴

The authors report a case of a man with sarcoidosis manifesting as a chronic inflammatory stenotic condition of the upper respiratory tract and trachea.

Case report

We report a case of a 38-year-old man, originally from Brazil, residing in Portugal for 11 years, who presented to our emergency department, with

dysphonia, odynophagia and progressive dyspnoea for 6 months. Stridor was present on observation. An infiltrative and stenotic soft palate lesion was also detected. It had extension to the lateral and posterior oropharynx, hypopharynx and larynx, with epiglottis involvement as well as involvement of ventricular bands and vocal cords, with subsequent glottic lumen reduction (figures 1 and 2). Fibrosis was present in the transition between the nasopharynx and oropharynx (figure 3), narrowing the communication between them. The patient underwent a tracheotomy and was admitted for investigation.

CT showed obliteration of the upper airway, including subglottic and tracheal invasion. Bilateral cervical lymphadenopathies were present, but mediastinal and hilar lymphadenopathies were absent as were parenchymal lung and pleura disease (figures 4–6). ACE was high (85.49 U/L). The complete blood count was normal and ANA, anti-dsDNA, c-ANCA, p-ANCA, venereal disease research laboratory test (VDRL) and HIV screening tests were negative. Bacteriological and mycobacteriological direct examination and cultural sputum were also negative.

Figure 1 Infiltrative and stenotic soft palate lesion with extension to the lateral and posterior oropharynx walls (arrow).

Figure 2 Vocal fold scarring and fibrosis.

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Figure 3 Fibrosis in the transition between the nasopharynx and oropharynx (arrow).

Rigid bronchoscopy confirmed extension of the infiltrating inflammatory process in the trachea for its entire length and circumference, reducing its lumen without bronchial involvement (figure 7). Multiple pharyngeal-laryngeal-tracheal biopsies were performed, demonstrating intense inflammation with sarcoid type non-caseating granulomas (figure 8). Bacteriological, mycological and mycobacteriological bronchial secretion examinations were negative. Bronchoalveolar lavage showed a CD4/CD8 ratio of 1.6. Cytology was negative for neoplastic cells. Pulmonary function tests revealed a flow/volume loop with extrathoracic obstruction and decreased diffusing capacity of the lungs for carbon monoxide (68%; figure 9). Arterial blood gases were normal.

Extensive upper respiratory tract sarcoidosis was suspected based on the clinical history, physical examination and histological results.

Intravenous methylprednisolone (1 mg/kg/day) was started with significant clinical improvement. The patient was then subsequently treated with prednisolone (40 mg/day) for 45 days, with progressive reduction (reduction of 5 mg/week to 20 mg/day maintained for 1 month, reduced to 5 mg/day maintained until 1 year of therapy was completed). Immunosuppressants, such as

Figure 5 CT: obliteration of the upper airway with tracheal lumen narrowing.

methotrexate, were reserved for eventual relapse. The tracheotomy was removed at 6 months, after stabilisation of the respiratory distress. Currently, the patient is asymptomatic and the disease has stabilised.

DISCUSSION

Upper airway sarcoidosis is a rare condition, particularly if isolated without systemic involvement. This case report concerns a young patient who presented with a chronic stenotic inflammation of the upper respiratory tract and trachea. Sarcoidosis affects the larynx in approximately 1–5% of cases, predominantly in the supraglottic region. Glottic and subglottic involvement is less common than supraglottic, occurring in only 15–20% of laryngeal sarcoidosis.¹ Clinical manifestations are dependent on the location and severity of the disease. In laryngeal sarcoidosis, airway obstruction can be present in 8–10% of cases and manifests with stridor, as observed in this case.^{1 4 6} Tracheal invasion occurs in 1–3% of cases.⁷

Establishing a diagnosis of sarcoidosis is challenging, as there are neither pathognomonic clinical findings nor tests. ACE is high in many other diseases and its increase only occurs in 40–60% of patients with acute illness.¹ When sarcoidosis is suspected, it is essential to exclude other diseases, such as tuberculosis, granulomatosis with polyangiitis, histoplasmosis, blastomycosis, coccidioidomycosis, actinomycosis, amyloidosis, syphilis and neoplastic disease.^{4 8 9} Our patient did not show

Figure 4 CT: obliteration of the nasopharynx.

Figure 6 CT: absence of mediastinal lymphadenopathies as well as parenchymal lung and pleura disease.

Figure 7 Rigid bronchoscopy confirmed the extension of the infiltrating inflammatory process in the trachea for its entire length and circumference (arrow).

any systemic manifestations of those diseases, and laboratory tests and histological results were also negative.

Corticosteroid therapy is the main treatment and should be started as soon as possible in symptomatic patients.² Prednisone (1 mg/kg/day) is recommended for 4–6 weeks, followed by progressive reduction during the next 4–6 months. If recurrence is suspected, then the cycle must be restarted or the use of alternatives, such as immunosuppressants and antimalarial medication, is recommended.¹ Topical corticosteroid injections and CO₂ LASER resection were also used in laryngeal sarcoidosis treatment.^{6 10}

The prognosis of sarcoidosis is usually good, depending on the affected site and the aggressiveness of the disease.⁴ Given the chronic and sometimes relapsing nature of sarcoidosis, the patient's surveillance is essential.

CONCLUSION

Upper respiratory tract sarcoidosis is a rare, multifaceted entity in its presentation. Sarcoidosis diagnosis, which is achieved through histological examination, requires a high level of suspicion. This article highlights the wide range of the sarcoidosis

Figure 8 Histology: intense inflammatory infiltrate with two non-caseating granulomas of the sarcoid type (arrows).

Figure 9 Flow/volume (F/V) loop with extrathoracic obstruction. ITGV, Intrathoracic Gas Volume; RV, residual volume; TLC, total lung capacity.

clinical spectrum, by presenting a life-threatening pharyngolaryngotracheal sarcoidosis with severe stenosis of the upper respiratory tract.

Learning points

- Pharyngolaryngotracheal sarcoidosis without pulmonary involvement is a very rare presentation of sarcoidosis.
- A high level of suspicion is required for the diagnosis.
- Histological examination is crucial for achieving diagnosis.
- Sarcoidosis must integrate the differential diagnosis of stenosis of the upper respiratory tract and trachea.

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