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Case Study

Multisystemic Sarcoidosis Revealed by Bilateral Hearing Loss: A Case Study

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Abstract

Sarcoidosis is a multisystem inflammatory disease of unknown cause, characterized by the formation of non-caseating granulomas in various organs, particularly the lungs and the lymphatic system. Although pulmonary involvement is the most common, sarcoidosis can also affect the central and peripheral nervous systems, leading to a wide range of manifestations. Among these manifestations, bilateral hypoacusis, that is, a decrease in hearing on both sides, can occur due to the involvement of cranial nerves, particularly the vestibulocochlear nerve (cranial nerve VIII). Although rare this impairment can be a revealing feature of the disease, although it is rare. Patients may experience progressive or sudden hearing loss, sometimes accompanied by vertigo or tinnitus. The diagnosis is based on a combination of clinical, radiological, and histopathological criteria, including the biopsy of suspected lesions to confirmation the characteristic granulomas.The treatment of sarcoidosis with neurological involvement generally includes corticosteroids to reduce granulomatous inflammation. In some cases, immunosuppressants may be used in combination with or as an alternative to corticosteroids, depending on the therapeutic response and side effects. Multidisciplinary management is essential to optimize the treatment and follow-up of patients with multisystem sarcoidosis with neurological manifestations. We report the case of a patient presenting with bilateral hearing loss revealed multisystemic sarcoidosis.

Keywords: *Multi-System Sarcoidosis; Hearing Loss; Vestibulocochlear Nerve; Diffuse Interstitial Lung Disease*

Introduction

Sarcoidosis is a systemic inflammatory disease with still poorly understood origins, which can affect a wide variety of organs. If pulmonary and lymph node involvement represent the most common manifestations, this pathology can present in atypical forms, making its diagnosis particularly complex. Neurological involvement, although rare, is one of these presentations, grouped under the term neurosarcoidosis. Among these neurological forms, bilateral hypoacusis, resulting from damage to the vestibulocochlear nerve, constitutes a rare but clinically significant entity. This auditory disorder, often associated with symptoms such as vertigo and tinnitus, can be the first revealing sign of multisystem sarcoidosis. This atypical clinical picture requires a thorough evaluation based on clinical, radiological, and histopathological examinations to establish an accurate diagnosis. This article proposes to examine a case of multisystem sarcoidosis revealed by bilateral hearing loss, highlighting the clinical specifics and the challenges posed by its therapeutic management.

Patient and Observation

This is a 52-year-old patient, originally from and residing in El Haouz, a housewife, with no toxic habits and no particular exposure, who has been experiencing a progressive onset of a dry cough associated with exertional dyspnea for the past 6 months, along with a gradual decline in visual acuity and xerostomia. The evolution was marked by the sudden onset of bilateral deafness, which prompted the patient to consult an otolaryngologist, all occurring in a context of apyrexia and general deterioration (asthenia, anorexia, and weight loss). The pleuropulmonary examination revealed some bilateral basal crackles. Clinical examination ENT: no particularities Ophthalmic examination: Papillitis and multifocal choroiditis in the right and left eye A chest X-ray was performed: Reticulo-micronodular images in both pulmonary fields. From a biological standpoint, the complete blood count (CBC) showed no abnormalities: WBC: 6030, neutrophils: 4280, lymphocytes: 1549, Hb: 15.6, an increase in ESR to 31 was noted, the rest of the liver, renal, and electrolyte tests were unremarkable. Hypercalcemia at 110 mg/l, urinary phosphocalcic assessment; without abnormalities Elevation of angiotensin-converting enzyme to: 81.2 Serologies: HIV, HBV, HCV, Syphilis: negative Immunological assessment: ANA, anti-native DNA antibodies, anti-CCP antibodies, RF, anti-ANCA antibodies: normal Gene x pert in the sputum: Negative BGSA: Focal lymphocytic sialadenitis grade 3 Brain MRI: no abnormalities Pure tone and speech audiometry: Sensorineural hearing loss TTE: without abnormalities Bronchoscopy: Grade 1 inflammatory state, absence of polyp or granuloma, staged biopsies were performed: subacute inflammatory remodeling of the bronchial mucosa, without specific characteristics, bronchoalveolar lavage (BAL) with lympho-histiocytic predominance, inflammatory cytodiagnosis, search for BK and gene x pert in bronchial aspirations: negative.

Chest CT: Subpleural and bilateral fissural pulmonary nodules and micronodules, absence of mediastinal lymphadenopathy.

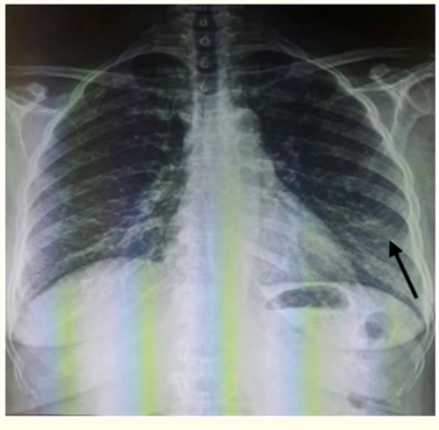

Figure 1: *Bilateral reticulo-micronodular opacities.*

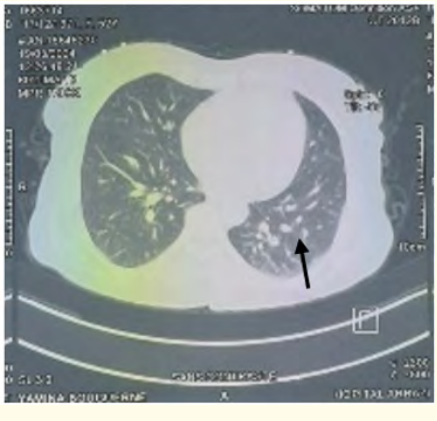

Figure 2: *Bilateral and fissural subpleural nodules and micronodules.*

The diagnosis of multifocal sarcoidosis was made based on:

- Ocular involvement: Papillitis with bilateral multifocal choroiditis.
- Pulmonary involvement: Subpleural and bilateral fissural pulmonary nodules and micronodules.
- Salivary gland involvement: with a BGSA showing focal lymphocytic sialadenitis grade 3.
- Ocular impairment: Sensorineural hearing loss on audiometry.
- ECA raised to 81.2.
- Hypercalcemia at 110 mg/l.

The patient received 3 boluses of methylprednisolone 1 g/day, then switched to oral administration 1 mg/kg/day (due to ocular involvement) with boluses of cyclophosphamide (Endoxan) each month for 6 months.

Discussion and Conclusion

Sarcoidosis is a systemic inflammatory disease of unknown cause, which can involve multiple organs. Although pulmonary involvement is predominant, about 5 to 10% of patients develop neurosarcoidosis, which can include involvement of the vestibulocochlear nerve [1,2]. This rare presentation, manifesting as bilateral hearing loss, vertigo, or tinnitus, reflects the inflammatory granulomatous infiltration of this nerve [3]. Bilateral hypoacusis, often a sensorineural in nature, can be the first revealing sign. A recent review reports that cranial nerve involvement is common in neurosarcoidosis, with the facial nerve (VII) and the vestibulocochlear nerve (VIII) being the most frequently affected [4]. These manifestations can be transient or persistent, depending on the severity of the inflammation and the promptness of treatment [5]. To confirm the diagnosis, imaging plays a key role. Brain MRI with gadolinium is the reference examination for detecting contrast uptake at the level of cranial nerves or adjacent structures, indicating active inflammation [6]. In addition, tonal audiometry, auditory evoked potentials (AEP), and vestibular tests help evaluate auditory function and locate the lesion [7,8]. A tissue biopsy remains crucial to confirm the non-caseating granulomatous nature and exclude other etiologies, such as neoplasms or infections [9]. From a therapeutic standpoint, corticosteroids are the first-line treatment. Their effectiveness in reducing inflammation and improving auditory symptoms is well documented [10]. However, in case of insufficient response or side effects, immunosuppressants such as methotrexate, azathioprine, or infliximab may be necessary [8,10]. Studies have also explored the effectiveness of biological agents targeting TNF-α in resistant forms [9]. The prognosis of neurosarcoidosis depends on prompt and multidisciplinary management. Regular follow-up with audiological evaluation and imaging allows for treatment adjustment based on clinical progression [9,10].

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