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Research Article

Nasal sarcoidosis revealing systemic lupus erythematosus and Sjögren syndrome

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Abstract:

OBJECTIVE:
Describe an original case of a rare association of sarcoidosis with systemic lupus erythematosus and Sjögren's syndrome in its nasal location.
OBSERVATION:
A 52-year-old patient with a history of type 2 diabetes and high blood pressure consulted for bilateral nasal obstruction and epistaxis progressing for 1 year. Nasal endoscopy revealed polyploid formations. A nasal biopsy with histological study which demonstrated epithelioid and gigantocellular granulomatosis without caseous necrosis.The dosage of antibodies anti nuclear, anti SSA, anti DNA and anti Sm antibodies is positive.
CONCLUSION:
Chronic rhinosinusitis are common ailments, but they can also be symptoms of one or more diseases of the system. Their management requires a multidisciplinary approach which often includes otolaryngologists and internists.

Keywords: Chronic rhinosinusitis, sarcoidosis, systemic lupus erythematosus, Sjögren syndrome..

Supplementary information The online version of this article (https://doi.org/xx.xxx/xxx.xx) contains supplementary material,which is available to autho-rized users.

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Introduction:

Sarcoidosis is an inflammatory disease of undetermined origin, characterised by granulomatous deposits that can affect all tissues of the body [1-2]. More than 90% of patients are characterised by pulmonary involvement [3-4]. Sinonasal involvement is rare and occurs in approximately 1% of all sarcoidosis patients [2, 5, 6]. Similarly, the association of sarcoidosis with Sjögren's syndrome (SS) has been rarely described [7]. The coexistence of sarcoidosis and systemic lupus erythematosus (SLE) has been reported only rarely [8-9]. In this work we report a rare case of nasal obstruction that revealed the coexistence of sarcoidosis, SLE and SS. Despite the theoretical association, to our knowledge this is the first case reported in the literature.

Observation :

Patient G.S, 52 years old, with a history of type 2 diabetes and arterial hypertension consulted for bilateral nasal obstruction and epistaxis evolving for 1 year, not improved by symptomatic treatment. The interview revealed inflammatory polyarthralgias. The physical examination noted a left nasal swelling. Nasal endoscopy revealed polyploid formations in the nasal cavity with an appearance of crusty rhinitis and ophthalmological examination revealed a dry eye syndrome and left dacryocystitis.

A nasal biopsy was performed which revealed a mononuclear inflammatory infiltrate and an epithelioid and gigantocellular granuloma, well limited without caseous necrosis, associated with hyaline fibrosis (Figure 1).

The etiological investigation of this nasal granulomatosis showed: lymphopenia at 1000/mm3, a biological inflammatory syndrome, polyclonal hypergammaglobulinemia at 19.3g/l and positive anti-nuclear, anti-SSA, anti-DNA and anti-Sm antibodies. The renal and phosphocalcic balance and the angiotensin converting enzyme assay were normal. Bacilloscopy , sputum culture and tuberculin skin test were negative.

A maxillofacial CT scan showed partial filling of the ethmoidal cells, the left frontal sinus and the left sphenoidal hemi-sinus with mucosal thickening in the maxillary sinuses (Figure 2, 3 and 4). The thoraco-abdomino-pelvic CT scan showed bilateral laterotracheal adenomegaly of the left anterior mediastinal chain associated with subpleural parenchymal nodules, averaging 6mm in size, and a 9mm left adrenal mass (Figure 5 and 6). Investigation of the adrenal incidentaloma concluded that it was a benign non-secretory lesion. Bronchial fibroscopy with bronchoalveolar lavage showed inflammation of the bronchial mucosa and lymphocytic alveolitis with a high CD4/CD8 ratio. Accessory salivary gland biopsy revealed grade 4 lymphocytic sialadenitis from Chisholm and Mason without granuloma.

At the end of our aetiological investigation, the hypotheses of tuberculosis and lymphoma were invalidated and the diagnosis of nasal, pulmonary and adrenal sarcoidosis associated with SLE and secondary SS was retained. The patient was put on oral corticosteroid therapy at a dose of 0.5mg/Kg/d with a clear clinical improvement after 3 months.

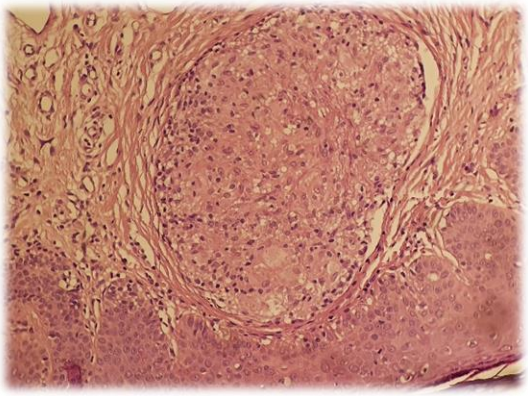
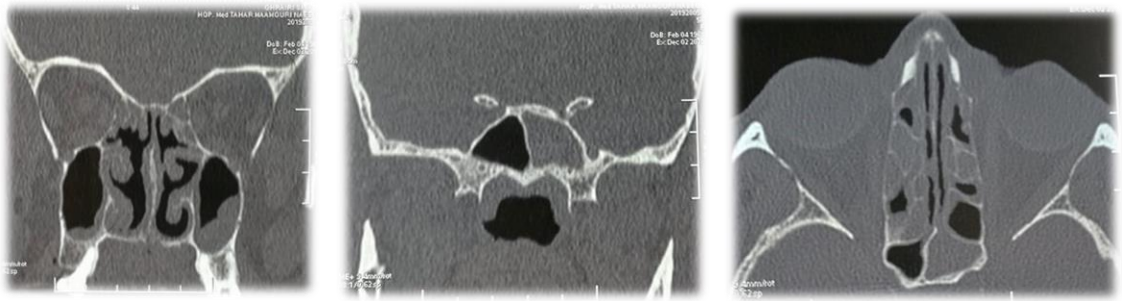
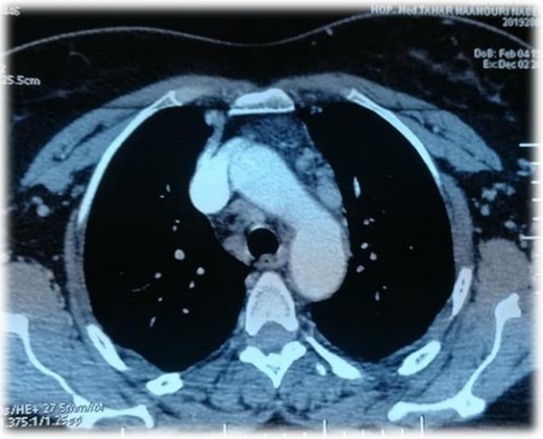


Figure 1: *Nasal mucosa with epithelioid and gigantocellular granuloma without caseous necrosis*



Figures 2, 3 and 4: CT images in coronal and axial sections showing framed thickening of the maxillary sinuses, filling of the left sphenoidal sinus and ethmoidal cells



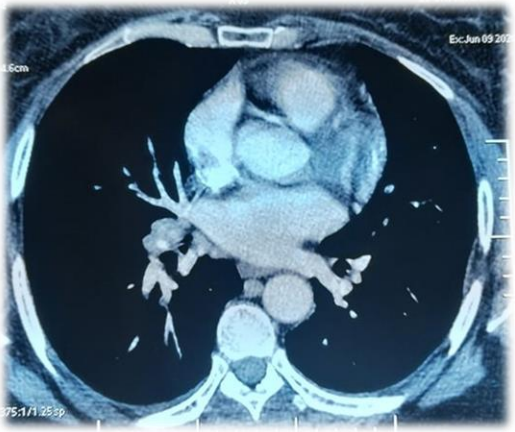


Figure 5 and 6: Axial CT images showing the aortic-pulmonary window and lateral-tracheal nodes

Discussion:

Sarcoidosis is a systemic granulomatosis that preferentially affects the lung and lymph nodes. Similarly, SLE is a multi-organ autoimmune disease. Although both sarcoidosis and SLE are considered to result from dysregulation of the immune system, their co-existence in the same patient is quite rare [10]. The exact prevalence of the sarcoidosis-autoimmune disease association is not known. The evidence comes mainly from case-control studies, retrospective cohort studies or case reports [9]. The first report on the coexistence of SLE and sarcoidosis was reported by Teilum in 1945, Begum, et al reported three patients with lupus and concomitant sarcoidosis in a series of about 300 patients followed for SLE over a period of 22 years, a percentage of 1%[8].This is considerably higher than the incidence of sarcoidosis in the general population which ranges from 1 to 40 per 100,000 population[11].

Coexistence of SS and sarcoidosis has been reported in only 1% of SS patients [12]. The association of the three conditions: sarcoidosis, SLE and SS in our patient, as well as the presence of numerous positive autoantibodies, could be related to complex immunological mechanisms. The etiology of this triple complex remains unknown, but studies using modern DNA technologies, combined with a comprehensive understanding of the entire human genome, are needed to clarify this mechanism.

Sinonasal manifestations of systemic diseases may involve the nose and paranasal sinuses and may be the initial reason for patients to consult, as in our case. For this reason, otolaryngologists must maintain a high index of suspicion to identify the underlying disease, make an early and accurate diagnosis by biopsy and histological examination, administer prompt treatment and refer the patient to the internist [1,2].

Nasal obstruction, rhinorrhoea, nasal crusting and epistaxis are very common symptoms in patients presenting to the ENT clinic. Although these symptoms are usually related to local causes, in some cases they represent manifestations of systemic conditions [1, 6, 13].

Nasal sarcoidosis is a diagnostic challenge because of the non-specific symptoms. These include nasal obstruction, mucosal congestion, rhinorrhoea, anosmia, nasal crusting and septal perforation [6]. Nasal endoscopy reveals mucosal changes such as small nodules or granulations, crusts, synechia, hypertrophy and a strawberry appearance. Yellowish nodules and granulation of the inferior turbinates and septum, mucosal thickening and lysis of the septum are also present on CT and MRI scans, as well as a corrugated iron-like image. Epiphora and anosmia occur when the lacrimal system or olfactory cleft, respectively, are involved [1, 6, 13].

In SLE, ulcers of the nasal mucosa and perforation of the cartilaginous septum may also be observed in less than 5% of patients. The nasal mucosa may be diffusely erythematous, edematous or atrophic. Epistaxis, nasal crusting, foul-smelling and mucopurulent discharge may also be seen [13].

Dryness, mucosal thickening, nasal crusting and epistaxis are also present in SS [13].

Nasal manifestations are treated with local corticosteroids combined with intermittent systemic corticosteroid therapy [5]. Endoscopic sinus drainage procedures may be necessary in the management of patients with acute or chronic

sinusitis not improved by well-conducted medical treatment [5]. However, local treatment is still necessary after surgical treatment to treat associated chronic inflammation [14].

Conclusion:

When faced with a sinonasal symptomatology that is atypical and unimproved by medical treatment, the most relevant examination is to perform a biopsy and anatomopathological examination. The contribution of otorhinolaryngologists in the detection of systemic diseases is essential. Close collaboration with internists is recommended for the etiological investigation, therapeutic management and appropriate follow-up of these patients**.**

References:

1. Send T., Tuleta I., Koppen T. et al. Sarcoidosis of the paranasalsinuses. Eur Arch Otorhinolaryngol. 2019; 276: 1969-74.
2. Catana I., Maniu A., Ilies R., Radeanu D., Catana A. A rare case of sarcoidosis affecting the nasopharynx. Romanian Journal of Rhinology. 10(37) :27-9.
3. Thorsten S., Peter K., Mattis B., Christoph B., Friedrich B., Dirk S. and Mark J. Clinical features of sarcoidosis patients presentingwithhead and neck manifestations - atwo-center retrospectivestudy and proposal of a diagnostic algorithm for the otorhinolaryngologist. Acta Oto-Laryngologica. 2020; 140(2): 144-8.
4. Korsten P., Tampe B., Konig M.F., Nikiphorou E. Sarcoidosis and autoimmunediseases: differences, similarities and overlaps. Current opinion in pulmonarymedicine. 2018; 24(5): 504-12.
5. Gulati S., Krossnes B., Olofsson J., Danielsen A. Sinonasalinvolvement in sarcoidosis: a report of seven cases and review of literature. Eur Arch Otorhinolaryngol. 2012; 269(3): 891-6.
6. Zeitlin J.F., Tami T.A., Baughman R., Winget D. Nasal and sinus manifestations of sarcoidosis. Am J Rhinol. 2000; 14(3): 157-61.
7. Ouarssani A., Rguibi M.I. Association syndrome de heerford et syndrome de GougerotSjogren : à propos d'un cas. Research. 2014; 1:957.
8. Begum S., Li C., Wedderburn L.R., Blackwell V., Isenberg D.A . Concurrence of sarcoidosis and systemic lupus erythematosus in three patients. Clin Exp Rheumatol. 2002; 20: 549-52.
9. Makary C.A., Gill B., Parman B., Unsal A., Holmes T., Reyes-Gelves C. and Kountakis S.E. Subjective and Objective Measurements of Sinonasal Manifestations in Patients With Autoimmune Disorders. The Laryngoscope. 2020; 00: 1-5.
10. Nakayama S., Mukae H., Morisaki T., Sakamoto N., Ohba K., Abe K., Kohno S. Sarcoidosis Accompanied by Systemic Lupus Erythematosus and Autoimmune Hepatitis. InternalMedicine. 2007; 46(19): 1657-61.
11. Wu C.H., Chung P.I., Wu C.Y., Chen Y.T., Chiu Y.W., Chang Y.T., Liu H.N. Comorbid autoimmune diseases in patients with sarcoidosis: A nationwide case-control study in Taiwan. The Journal of Dermatology. 2016; 44(4): 423-30.
12. Tokuyasu H., Harada T., Touge H., Kawasaki Y., Maeda R., Isowa N., Shimizu E. Primary Sjögren's syndrome complicated by sarcoidosis. InternalMedicine. 2008; 47(23): 2049-52.
13. Prokopakis E., Nikolaou V., Vardouniotis A., Jorissen M. Nasal manifestations of systemicdiseases. B-ENT. 2013; 9(3): 171-184.
14. Kohanski M.A., Reh D.D. Chapter 11: Granulomatousdiseases and chronicsinusitis. Am J RhinolAllergy. 2013; 27(3): 39-41.