

COINCIDENCE OF RHINOSCLEROMA AND ROSAI-DORFMAN DISEASE (SINUS HISTIOCYTOSIS WITH MASSIVE LYMPHADENOPATHY): REPORT OF A CASE

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Abstract: A 35-year-old man was admitted because of a 2-year history of nasal obstruction, a nasopharyngeal mass, malaise, and massive cervical lymphadenopathy. Nasal symptoms and nasopharyngeal mass were due to rhinoscleroma that was confirmed by an incisional biopsy and culture. The patient underwent cervical lymph node biopsy that revealed the tissue compatible with Rosai-Dorfman disease, also called sinus histiocytosis with massive lymphadenopathy. Rhinoscleroma was treated by a combination of excision and antibiotics. Culture became negative for *Klebsiella rhinoscleromatis*. Then prednisolone administered for one month and gradually tapered. Lymph nodes became small at the end of treatment. Twelve months follow-up showed no evidence of recurrence of both diseases. To the best of our knowledge, this is the first report of association of rhinoscleroma with Rosai-Dorfman disease. Association of these two rare diseases may be due to just a coincidence, or a common immunologic or microbiologic underlying factor.

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Key words: Rhinoscleroma, Rosai-Dorfman disease, *Klebsiella Rhinoscleromatis*, Emperipolesis.

INTRODUCTION

Rhinoscleroma, coined by von Hebra in 1870, is a chronic granulomatous disease involving the mucosa of the upper respiratory system (1,2). Rhinoscleroma, caused by *Klebsiella rhinoscleromatis*, is an unusual infection.

Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy; SHML), first described by Juan Rosai and Ronald F. Dorfman in 1969, has

an unknown etiology (3-4). Rosai-Dorfman disease presents in its most typical form as massive, painless, bilateral lymph node enlargement in the neck (4).

We report in this paper a case of documented rhinoscleroma and Rosai-Dorfman disease.

Case Report

A 35-year-old man, weaving worker, was seen in May 1999, because of a progressive 2-year history of nasal obstruction. Additionally, the patient complained of moderate left-sided hearing loss, difficulty in dealing with his job, and multiple swellings in his neck.

There was no history of smoking or alcohol use. Examination revealed bilateral multiple lymph node enlargements located at the upper portion of anterior and posterior cervical triangles, particularly at the left; firm in consistency, with no erythema, tenderness, or adhesion to skin. Rhinologic examination showed hypertrophied inferior turbinate and a firm polypoid mass in the left nostril. This mass had extended to choana and nasopharynx. In the oral cavity, a distinct mass was easily visible suspending from the nasopharynx and extending to posterior pillar of palatine tonsil. Other examinations were unremarkable.

The patient's laboratory assessment depicted the following data: Hb 15.2 g/dL, Hct 46%, and WBC 13000/mm³ (PMN=88%). Serum Ca, P, alkaline phosphatase, and erythrocyte sedimentation rate like liver function tests were normal. Blood smear was normal.

The patient underwent an incisional biopsy of the intranasal lesion. Culture of the tissue yielded *Klebsiella rhinoscleromatis*. Microscopically, fibrosis, eosin-staining Russell bodies (Fig. 1 A), and vacuolated Mikulicz's cells (Fig. 1 B) were seen. Thus, the diagnosis of rhinoscleroma was established and substantiated in the next biopsy.

Since the lymphadenopathy in rhinoscleroma is very rare, an excisional biopsy was employed for the

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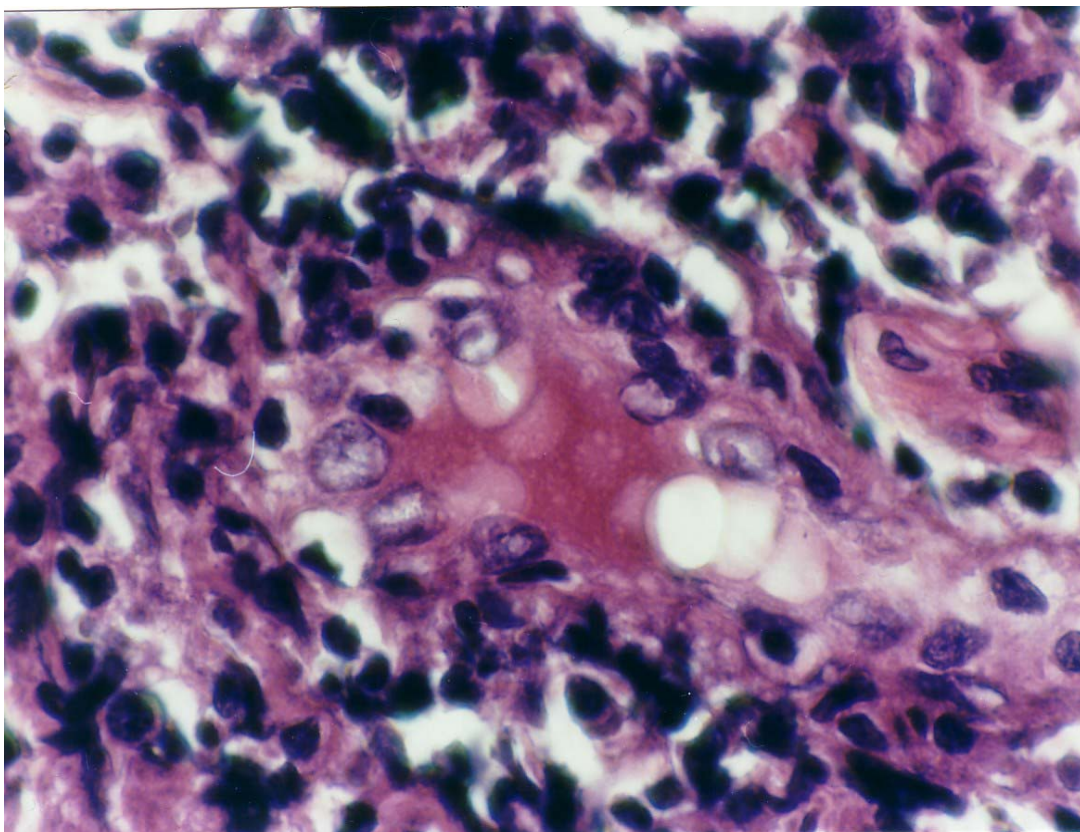
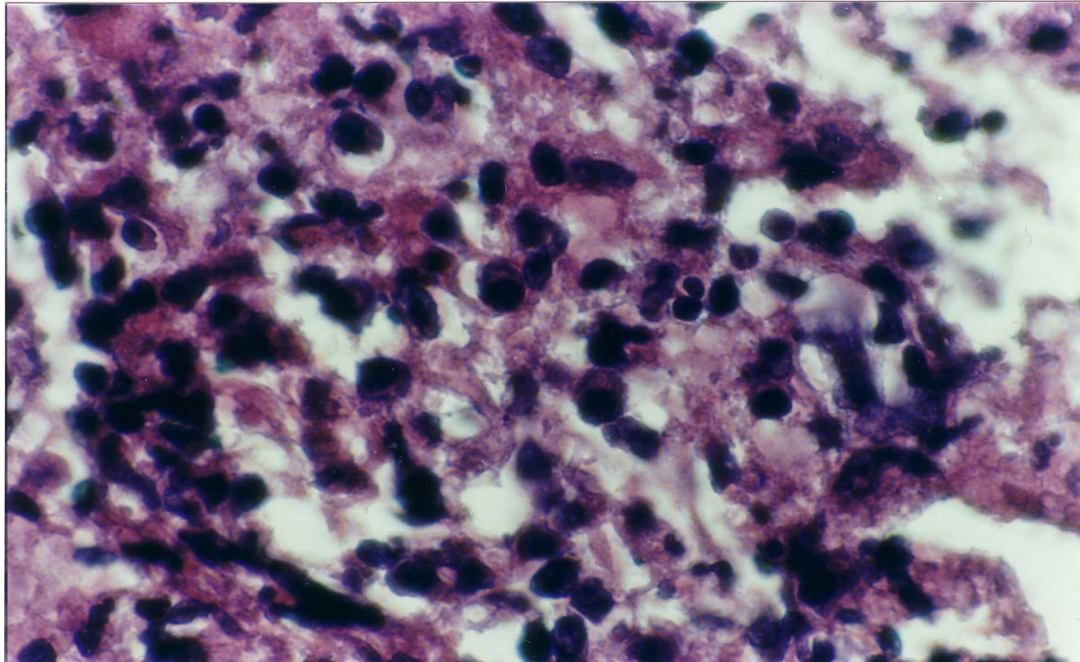


Fig. 1A. High-power view showing eosin-staining Russell bodies in the cytoplasm of some plasma cells. **B.** Vacuolated Mikulicz's cell.

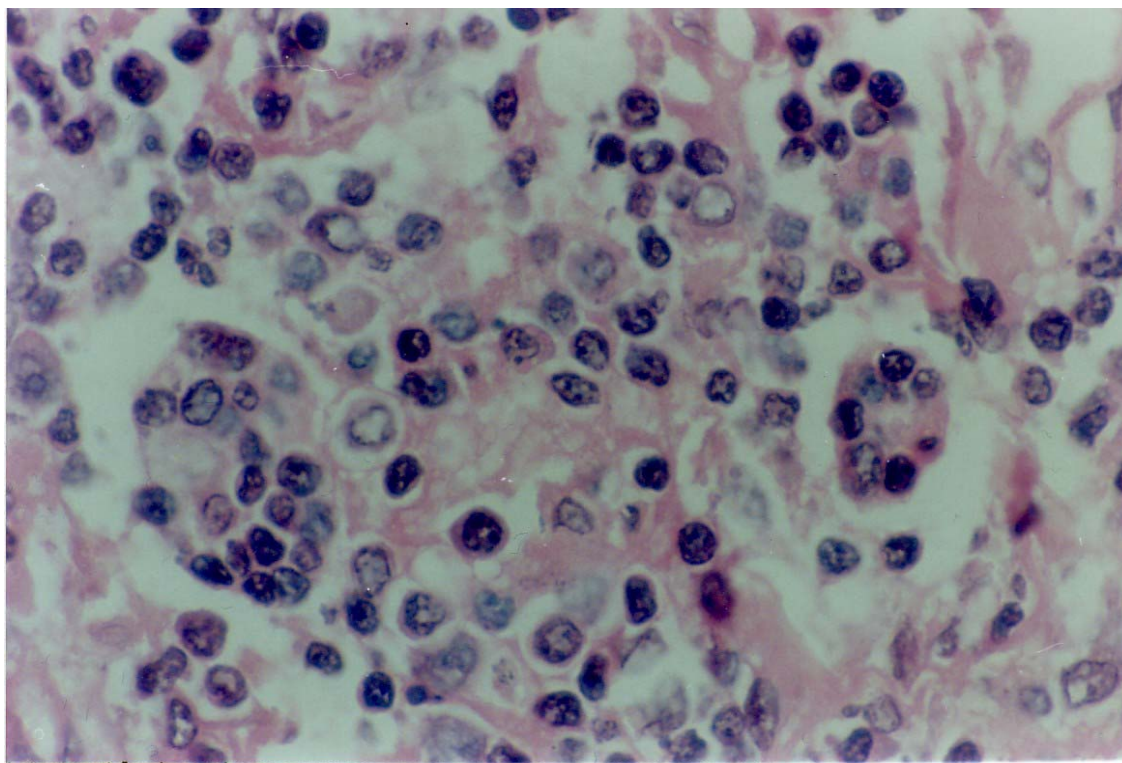


Fig. 2. High-power view showing lymphophagocytosis by the sinus histiocytes (emperipolesis).

coalescent lymph nodes. Grossly, the nodes were woven together and there was prominent perinodal fibrosis. Microscopically, there was dilatation of the lymph sinuses filled with lymphocytes, plasma cells, and histiocytes. Many of these histiocytes showed lymphophagocytosis or “emperipolesis” (Fig. 2), pathognomonic for sinus histiocytosis with massive lymphadenopathy (SHML; Rosai-Dorfman disease).

To rule out other diagnoses, another blood smear was obtained and bone marrow aspiration/biopsy accomplished, without any abnormality.

Rhinoscleroma was treated by excision and streptomycin (1 g/day) plus tetracycline (2 g/day) for one month. Then the sclerotic lesions responded to ciprofloxacin (500 mg bid for 3 weeks). The culture became negative for *K. rhinoscleromatis*, and then prednisolone (1mg/kg/day) was administered for one month, when the drug gradually tapered. Lymph nodes became small, at the end of treatment.

After twelve months of follow-up, there was no evidence of recurrence of both diseases.

DISCUSSION

Rhinoscleroma is a rare specific granulomatous disease of the respiratory tract with chronic evolution (1,5-7). The specific organism has been identified as *Klebsiella rhinoscleromatis* (1-2). It was initially

described involving the nose (so justified the term *rhinoscleroma*) but now it is reported to also involve the larynx, trachea, and bronchi (1,7).

In the nose, rhinoscleroma has distinct stages of development, including: [1] catarrhal stage (foul-smelling, purulent rhinorrhea for weeks or months); [2] atrophic stage (large plaques or crusts that are foul-smelling); [3] granulomatous stage (multiple granulomatous nodules throughout the nose, pharynx, larynx, trachea, or bronchi); and [4] fibrotic and stenotic stage (1,8). On admission, the patient was in the stage 4.

Diagnosis is suspected by finding coalescent granulomatous nodules near the nasal vestibule. In general, the diagnosis is based on the microbiologic identification of *K. rhinoscleromatis* and histological features (9). Identification of causative agent is diagnostic (1,10). Finding of vacuolated Mikulicz's cell and transformed plasma cells with Russell bodies are suggestive but not pathognomonic to confirm the diagnosis (1). The differential diagnosis includes malignancy, fungal infections, and numerous granulomatous diseases (6).

Various antibiotics have been used for treatment of this infection. Long-term streptomycin plus tetracycline, and trimethoprim-sulfamethoxazole are the recommended treatments (1-2). The sclerotic lesions respond well to ciprofloxacin (11). Corticosteroids and radiotherapy are not effective (1).

Local spread and cicatricial complications mandate surgical manipulation e.g. excision (12) or carbon dioxide laser vaporization (13).

Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy; SHML) is an uncommon benign proliferation of hematopoietic and fibrous tissue (14) that usually presents with massive, painless, bilateral lymph node enlargement in the neck associated with fever, leukocytosis, elevated erythrocytation rate, and polyclonal hypergammaglobulinemia (3-4). These sinuses are occupied by lymphocytes, plasma cells, and histiocytes. The key histologic feature of Rosai-Dorfman disease is the presence of large, pale histiocytes that contain within their cellular borders apparently engulfed lymphocytes (lymphophagocytosis or emperipolesis) (4,14,15). These histiocytes are S-100 protein positive by immuno-staining (4,14).

The etiology of Rosai-Dorfman disease is unknown (4,15,16). The two most likely possibilities are infection by a virus or some other microorganism and the manifestation of a subtle undefined immunologic defect (4,17).

About 30-40% of affected individuals have extranodal manifestation (4,17). The most common sites include eyes and ocular adnexae, head and neck, upper respiratory tract, skin and subcutaneous tissue, and central nervous system (4). Some other more unusual sites are thyroid (18), pituitary (19), and gastrointestinal tract (4).

The differential diagnosis of Rosai-Dorfman disease includes nonspecific sinus hyperplasia, tuberculosis, Langerhans' cell granulomatosis, leprosy, Hodgkin's disease, non-Hodgkin's lymphoma, metastatic carcinoma, and metastatic malignant melanoma (3,4,14). It is important to distinguish Rosai-Dorfman disease from other causes of histiocytosis because of the different treatment modalities.

Rosai-Dorfman disease is usually benign and self-limited whose treatment is aimed predominantly at controlling its local manifestations (20,21). Multiple treatment modalities have been used in more severe progressive forms with variable success. Some instances include excision, various medications (e.g. prednisolone, cyclophosphamide, vincristine sulfate, and methotrexate), and radiation (3,19,21). Since our patient had a slowly progressive course, we successfully treated him with prednisolone.

Association of rhinoscleroma with Rosai-Dorfman disease is a rare occurrence and to the best of our knowledge, no association between them has been reported so far in the literature. This association

may be due to coincidence of these two rare diseases or due to a common immunologic or micro-biologic underlying factor.

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