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D S Katz, L B Poe and R J Corona, Jr

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Sinus Histiocytosis with Massive Lymphadenopathy: A Case of Simultaneous Upper Respiratory Tract and CNS Disease without Lymphadenopathy

Douglas S. Katz,¹ Larry B. Poe,^{1,3} and Robert J. Corona, Jr.²

Summary: The authors describe a 20-year-old man who initially developed an intradural mass in the upper cervical region and subsequently presented with nasal/paranasal sinus and posterior fossa masses, well demonstrated by CT and MR. Histopathology demonstrated dense fibrous tissue, aggregates of histiocytes with round to oval vesicular nuclei, and other features diagnostic of Rosai-Dorfman disease; a right nasal biopsy prior to surgery showed similar microscopic findings.

Index terms: Histiocytosis; Paranasal sinuses, neoplasms; Posterior fossa, neoplasms; Rosai-Dorfman disease

In 1969, Rosai and Dorfman (1) defined a clinicopathologic entity they termed sinus histiocytosis with massive lymphadenopathy (SHML), which was distinct from the other histiocytoses. This entity was characterized by bilateral, painless, massive lymphadenopathy, especially of the cervical region in which the sinuses of affected lymph nodes were dilated with numerous histiocytes. The eponym Rosai-Dorfman disease has been used for this disorder, however, involvement of extranodal sites is common and may occur without lymphadenopathy (1–3). We report a case of SHML with recurrent dural-based disease of the posterior fossa and extensive upper respiratory tract involvement without lymphadenopathy.

Case Report

A 20-year-old man presented with a 3-month history of intermittent nausea, vomiting, anorexia, and occipital headaches, and a 4-year history of nasal obstruction and rhinorrhea. At age 12, he developed progressive weakness in the lower extremities and gait disturbance. Physical and routine laboratory examination was otherwise normal. A well-circumscribed, dorsal, extradural soft-tissue mass was

discovered on computed tomography (CT) in the upper cervical spine and was resected. It extended from the foramen magnum to C3, compressing the spinal cord. The mass was surgically resected. Microscopic examination revealed sinus histiocytosis and a diagnosis of Rosai-Dorfman disease was made. Subsequent CT examinations showed no residual or recurrent disease. A full recovery followed, and he remained asymptomatic for 4 years.

At age 20, he presented to our institution with flesh-colored masses completely obstructing both nasal passages. Neurologic examination was remarkable for bilateral wasting, sustained clonus of the left ankle, a left Hoffman sign, and mild midline cervical spinal tenderness with marked paraspinal muscle spasm. There was no lymphadenopathy.

Axial CT showed a large, homogeneous soft-tissue mass completely filling and expanding the maxillary and ethmoid sinuses, completely filling the nasal cavity and the nasopharynx and extending into the oropharynx. The nasal bones were splayed and the turbinates eroded (Fig. 1D). The interorbital distance was slightly widened. The CT scan disclosed an ill-defined, minimally hyperdense midline posterior fossa mass compressing the caudal fourth ventricle and mild supratentorial ventriculomegaly. Magnetic resonance (MR) revealed the posterior fossa mass to be dural based and arising from the region of the foramen magnum (Fig. 1A). The dural-based mass and the upper respiratory masses were predominantly isointense to white matter on short TR/short TE, long TR/short and long TE sequences. After Gd-DTPA was administered intravenously, the upper respiratory tract masses enhanced prominently but heterogeneously while the lobulated posterior fossa extraaxial mass enhanced more homogeneously (Figs. 1B and 1C).

Two adjacent encapsulated masses that were adherent to dura were resected by a suboccipital craniectomy 2 months later. Histopathologic features suggested a diag-

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¹ Department of Radiology, Division of Neuroradiology, and ² Department of Pathology, SUNY Health Science Center at Syracuse, Syracuse, NY 13210.

³ Address reprint requests to Larry B. Poe, MD, SUNY Health Science Center, Department of Radiology, 750 E. Adams Street, Syracuse, NY 13210.

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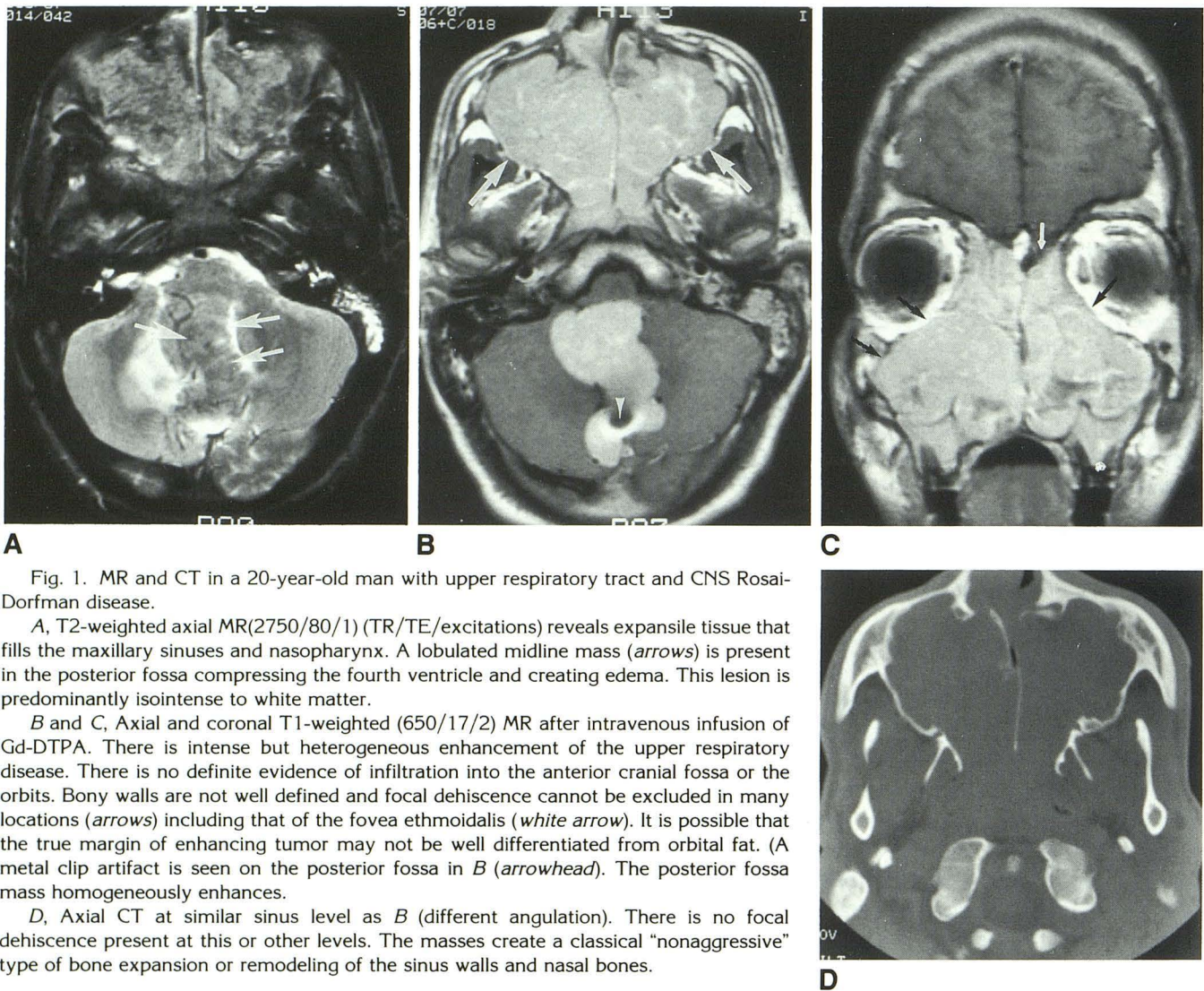


Fig. 1. MR and CT in a 20-year-old man with upper respiratory tract and CNS Rosai-Dorfman disease.

A, T2-weighted axial MR(2750/80/1) (TR/TE/excitations) reveals expansile tissue that fills the maxillary sinuses and nasopharynx. A lobulated midline mass (arrows) is present in the posterior fossa compressing the fourth ventricle and creating edema. This lesion is predominantly isointense to white matter.

B and C, Axial and coronal T1-weighted (650/17/2) MR after intravenous infusion of Gd-DTPA. There is intense but heterogeneous enhancement of the upper respiratory tract. There is no definite evidence of infiltration into the anterior cranial fossa or the orbits. Bony walls are not well defined and focal dehiscence cannot be excluded in many locations (arrows) including that of the fovea ethmoidalis (white arrow). It is possible that the true margin of enhancing tumor may not be well differentiated from orbital fat. (A metal clip artifact is seen on the posterior fossa in B (arrowhead). The posterior fossa mass homogeneously enhances.

D, Axial CT at similar sinus level as B (different angulation). There is no focal dehiscence present at this or other levels. The masses create a classical "nonaggressive" type of bone expansion or remodeling of the sinus walls and nasal bones.

nosis of Rosai-Dorfman disease: there was dense fibrous tissue with aggregates of histiocytes with round to oval, vesicular nuclei and small nucleoli, numerous plasma cells, and lymphocytes and plasma cells phagocytized by an occasional histiocyte (Fig. 2) (2, 3). A right nasal biopsy preceding the neurosurgery showed similar microscopic findings.

Discussion

Since 1969, over 423 cases of SHML (Rosai-Dorfman disease) have been reported (3). It is a disorder of unknown etiology, usually with a benign, self-limited course. A predominance of cases (83.7%) have cervical lymphadenopathy, which is characteristically bilateral, painless, and massive (3). Axillary, inguinal, or mediastinal lymphadenopathy is less frequent. Forty-three

percent of patients have at least one extranodal site of involvement, most commonly in the skin, the upper respiratory tract, the orbit, bone, salivary glands, and the central nervous system (CNS) (3, 4). Microscopic examination of lymph nodes reveals pericapsular fibrosis and marked dilatation of subcapsular and medullary sinuses with nonneoplastic histiocytes, numerous plasma cells, and many lymphocytes that have been engulfed by histiocytes (1, 2). Individual lymph nodes may measure up to 6 centimeters in diameter. Not all involved nodes are necessarily enlarged. While various imaging modalities may show the lymphadenopathy, there are no reported distinguishing imaging features (5).

Fever may occur at the time of presentation, and the sedimentation rate is frequently elevated. There may be leukocytosis with neutrophilia,

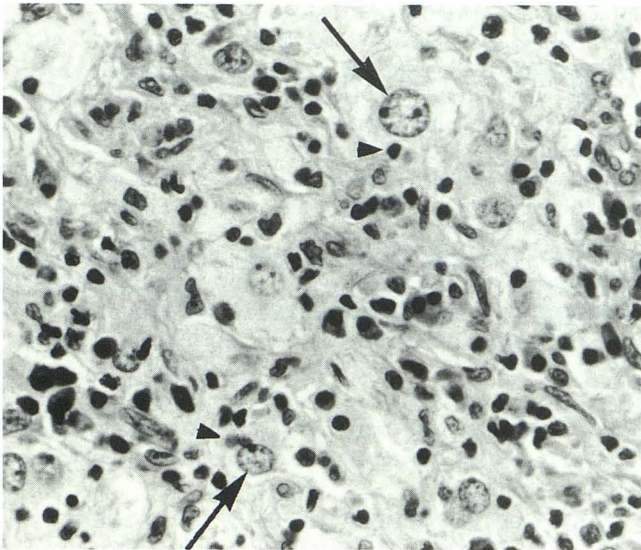


Fig. 2. Microscopy of the biopsy specimen from the posterior fossa. High-power view of the lesions demonstrating the histiocytes with round to oval vesicular nuclei and small nucleoli (arrows). Identification of phagocytized lymphocytes (arrowheads) and plasma cells are noted in an occasional histiocyte (hematoxylineosin stain; $\times 40$).

hypergammaglobulinemia, and a mild anemia. The presentation is often insidious and the clinical course may be protracted, with total recovery occurring spontaneously in most cases. There is no specific therapy (1–3), with most patients treated, if necessary, in a similar manner as Langerhans cell histiocytosis or hematopoietic malignancy with immunosuppressive chemotherapy (6). The mean age of onset on Rosai-Dorfman disease is 20.6 years. All races and ages are affected, with a slight male predominance (58% of all cases). Fifty-six patients have had various associated immune disorders.

Forty-eight cases of Rosai-Dorfman disease involving the upper respiratory tract have been reported, typically with polyps or mass lesions in the nasal cavity or paranasal sinuses. The nasal cavity may be totally obstructed. Extensive disease, as occurred with our case, has been described sparingly. Seventy percent of these cases have additional extranodal sites of disease (3). Radiographically paranasal sinus disease may be expressed as mucosal thickening, or polypoid masses that enhance and may erode adjacent bone.

A total of 22 cases of CNS involvement in Rosai-Dorfman disease have been reported (3, 7). Approximately one half had no lymph node disease, and more than half had another site of extranodal disease. (3, 7). The CNS findings in

our case are fairly representative of the few such cases that have been described.

The characteristic histologic picture of SHML consists of lymphocytes, plasma cells, and a predominance of histiocytes with a large vesicular nucleus and abundant clear cytoplasm. Emperipolesis (lymphophagocytosis) is a constant, though nonspecific, feature (8). SHML may involve nodal and extranodal sites making pathologic distinction from malignant lymphoma difficult. The lesions in the CNS classically show dense fibrous tissue containing nodules and cords of histiocytes with the previously described cytologic features, lymphocytes, and plasma cells (3). "Histiocytosis X" may produce meningeal or dural lesions and can involve bone and adjacent neural structures but, in contrast to SHML, the histiocytic nuclei are folded or lobulated and by electron microscopy the cytoplasm contains Langerhans granules (9).

In our patient, a CT scan of the paranasal sinuses was first performed to evaluate the nearly homogeneous soft-tissue masses filling the nasal passages, ethmoid, sphenoid, and maxillary sinuses. The expanded bone walls were "remodeled" in a manner typical of a "nonaggressive" tumor of the paranasal sinuses (10). The large lobulated S-shaped enhancing mass in the midline of the posterior fossa was noted at this time. It is very unusual for even benign tumors of the paranasal sinuses be so extensive at presentation. Large lesions that could conceivably, but rarely, mimic the pattern of paranasal sinus disease found in our patient include a variety of conditions leading to "benign" expansion of the bony walls, such as lymphoma, other histiocytoses, inflammatory polyps, inverting papilloma, minor salivary gland tumors, extramedullary plasmacytoma, and hemangiopericytoma (11). Certainly when the upper respiratory disease and the mass in the CNS is considered, lymphoma, or one of various histiocytoses, would be a major consideration in the radiographic differential diagnosis.

References

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a newly recognized benign clinicopathological entity. *Arch Pathol* 1969;87:63–70
2. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a pseudolymphomatous benign disorder—analysis of 34 cases. *Cancer* 1972;30:1174–1187
3. Foucar E, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): review of the entity. *Semin Diagn Pathol* 1990;7:19–73

4. Nardu RK, Urken ML, Som PM, Danon A, Biller HF. Extranodal head and neck sinus histiocytosis with massive lymphadenopathy. *Otol Head Neck Surg* 1990;102:764-767
5. McAlister WH, Herman T, Dehner LP. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). *Pediatr Radiol* 1990;20:425-432
6. Komp DM. The treatment of sinus histiocytosis with massive lymphadenopathy. *Semin Diagn Pathol* 1990;7:83-86
7. Foucar E, Rosai J, Dorfman RF, Brynes RK. The neurologic manifestations of sinus histiocytosis with massive lymphadenopathy. *Neurology* 1982;32:365-371
8. Rosai J. Sinus histiocytosis with massive lymphadenopathy. In: *Ackerman's surgical pathology*. Vol II. 7th ed. St. Louis: Mosby, 1989:1297
9. Kepes JJ. "Xanthomatous" lesions of the central nervous system: definition, classification and some recent observations. *Prog Neuropathol* 1979;4:179-213
10. Som PM, Shugar JMA. The significance of bone expansion associated with the diagnosis of malignant tumors of the paranasal sinuses. *Radiology* 1980;136:97-100
11. Som PM. Tumors and tumor-like conditions of the sinonasal cavity. In: Som PM, Bergeron RT, eds. *Head and neck imaging*. 2nd ed. Philadelphia: Saunders, 1990:169-224