Rosai-Dorfman disease affecting the nasal cavities and paranasal sinuses

Dear Editor,

Here, we report the case of a 17-year-old male who presented with a three-month history of nasal obstruction, asthenia, and febrile episodes. Physical examination revealed bilateral enlargement of cervical and axillary lymph nodes, all of which were painless on palpation. Laboratory tests showed mild leukocytosis, an elevated increased C-reactive protein level, and a high erythrocyte sedimentation rate. The venereal disease research laboratory test and monospot test were both negative, as was serology for HIV, toxoplasmosis, and cytomegalovirus. Computed tomography (CT) of the sinuses showed multiple, homogeneous, hypointense, rounded polypoid masses, which effectively narrowed the nasal passages, together with opacification of the ethmoid cells and sphenoid sinuses, with no evidence of bone erosion (Figure 1). Biopsies of a cervical lymph node and nasal lesions were negative for neoplasia and acid-fast bacilli, showing diffuse lymphoplasmacytic infiltration, foamy histiocytes, and emperipolesis. Immunohistochemistry showed positivity for S-100 protein, positivity for CD68, and negativity for CD1a. A diagnosis of Rosai-Dorfman disease was made, and corticosteroid therapy was started, resulting in slow, progressive improvement.

Recent studies in the radiology literature of Brazil have stressed the importance of CT and magnetic resonance imaging (MRI) in improving the diagnosis of head and neck masses\(^1\)–\(^8\). Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, is a rare, benign lymphoproliferative, usually self-limiting, condition characterized by bilateral, painless cervical passages, together with opacification of the ethmoid cells and sphenoid sinuses, with no evidence of bone erosion (Figure 1). Biopsies of a cervical lymph node and nasal lesions were negative for neoplasia and acid-fast bacilli, showing diffuse lymphoplasmacytic infiltration, foamy histiocytes, and emperipolesis. Immunohistochemistry showed positivity for S-100 protein, positivity for CD68, and negativity for CD1a. A diagnosis of Rosai-Dorfman disease was made, and corticosteroid therapy was started, resulting in slow, progressive improvement.

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lymphadenopathy\textsuperscript{(6–11)}, with spontaneous resolution in approximately half of all cases\textsuperscript{(57)}. The disease has a slight predilection for males and primarily affects children, adolescents, and young adults, 80\% of cases occurring in individuals under 20 years of age\textsuperscript{(6,8,9)}, being most common in immunocompromised individuals and preferentially affecting the skin, respiratory tract, reticuloendothelial system, genitourinary tract, or bones\textsuperscript{(6,8,9)}. Although uncommon, enlargement of the mediastinal, hilar, axillary, and inguinal lymph nodes can occur.

The etiology of Rosai-Dorfman disease is unclear, although it could be related to changes in the immune response or to infections caused by agents such as varicella-zoster virus and other herpes viruses, as well as Epstein-Barr virus, cytomegalovirus, Brucella spp., and Klebsiella spp.\textsuperscript{(6,7,5,11)}

Imaging tests such as CT and MRI are useful for evaluating the extent of Rosai-Dorfman disease, although there are no specific characteristics. When it affects the paranasal sinuses, it typically manifests as polypoid masses, mucosal thickening, with or without bone erosion, with preferential involvement of the maxillary sinuses and ethmoid cells\textsuperscript{(9,10)}. The diagnosis is established by histopathology\textsuperscript{(8)}.

The differential diagnoses include several types of lymphoreticular malignancy, such as lymphoma, malignant histiocytosis, and monocytic leukemia, which have histopathological features similar to those of Rosai-Dorfman disease but present atypia and a rapid, aggressive evolution. Another major differential diagnosis is Kikuchi-Fujimoto disease (histiocytic necrotizing lymphadenitis), the clinical profile of which resembles that of Rosai-Dorfman disease but with a more indolent course\textsuperscript{(12,13)}. The diagnosis is established by histopathology\textsuperscript{(12,13)}.

In Rosai-Dorfman disease, the treatment modality of choice depends on the extent of Rosai-Dorfman disease and the timing of treatment. The choice of treatment strategies depends on the severity of the disease, with corticosteroids, chemotherapy, radiotherapy, or surgery\textsuperscript{(6–11)}.

In conclusion, although Rosai-Dorfman disease does not present specific imaging characteristics, it should be considered among the diagnostic possibilities in cases of painless bilateral cervical lymphadenopathy, particularly in children and adolescents.

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