ACTA RADIOLÓGICA PORTUGUESA
May-August 2018 | Vol 30 nº 2|31-33

Radiological Case Report / Caso Clínico

Rosai-Dorfman Disease

Doença de Rosai-Dorfman

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Abstract

Rosai-Dorfman disease is a rare, usually benign, entity that mostly affects children and adolescents. We report a clinical case in which the most common clinical and imaging manifestation is present: cervical lymphadenopathy. Histological confirmation is usually needed to make a confident diagnosis.

Keywords

Lymphadenopathy; Sinus histiocytosis; Rosai-Dorfman disease.

Resumo

A doença de Rosai-Dorfman é uma entidade rara e geralmente benigna, que surge mais frequentemente em crianças e adolescentes. Descreve-se um caso clínico com a manifestação clínica e imagiológica mais comum: adenomegalias cervicais. A confirmação histológica é geralmente necessária para estabelecer o diagnóstico.

Palavras-chave

Linfadenopatia; Histiocitose sinusal; Doença de Rosai-Dorfman.

Introduction

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, is a rare and usually benign clinical-pathological entity. It occurs more frequently in children and adolescents, the most common clinical presentation being bilateral painless cervical lymphadenopathy.[1,2] The authors present a clinical case of this disease, including imaging and histological findings, reviewing its main characteristics.

Clinical history

A 6-month male infant, with no relevant medical history, is transported to the emergency department due to fever with 4 days of evolution (maximum rectal temperature of 39ºC) and left cervical swelling with ipsilateral flexion of the neck. At physical examination the swelling was flushed, with elastic and adherent consistency. The ultrasound evaluation (Fig. 1) showed a left cervical adenopathic conglomerate measuring approximately 40 mm at greater axis, with adenomegalies of round morphology and heterogeneous echoestructure, without areas of necrosis or abscess. The analytical study revealed leukocytosis (19.21 x 109/L), with a slight neutrophilia of 54% and an increase in C-reactive protein (46.5 mg/L, to a normal value <3 mg/L).
Intravenous antibiotic therapy and clinical surveillance were decided. During hospitalisation serologies for toxoplasmosis, rubella, syphilis, cytomegalovirus, Epstein-Barr virus, simple herpes virus 1 and 2 and parvovirus B19 were negative. After 12 days of antibiotic treatment, the infant was discharged from hospital, without fever or local inflammatory signs and with a standardised laboratory study, and was advised to reassess the patient in an outpatient clinic due to the persistence of left cervical swelling with overlapping dimensions.

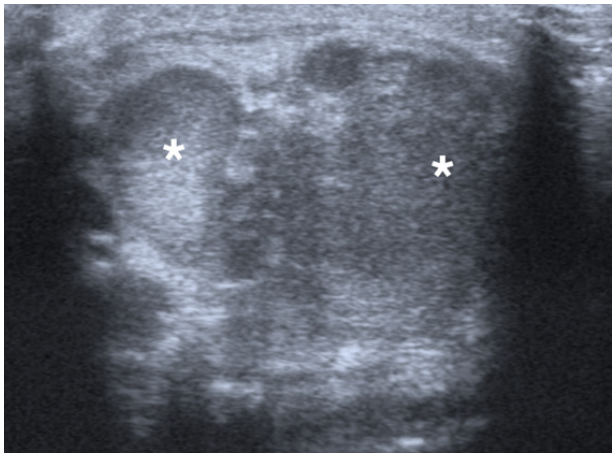

Figure 1 – Cervical ultrasound shows a left cervical adenopathic conglomerate, with multiple adenomegalies (asterisks) of rounded morphology and heterogeneous echoestructure, with no discernible echogenic hilum.

Persistent swelling one month later led to an MRI of the neck (Fig. 2), which showed bilateral cervical adenomegalies, forming an adenopathic conglomerate on the left with a greater axis of 48 mm. These adenomegalies were intensely enhanced after administration of intravenous contrast and were mostly hypersignal in the T2-weighted sequences, but some T2 hyposignal areas were evident.
A biopsy guided by ultrasound of left adenopathic conglomerate (Fig. 3) was made whose histological and immunohistochemical study (Fig. 4) revealed the presence of numerous bulky histiocytes with immunostaining S100

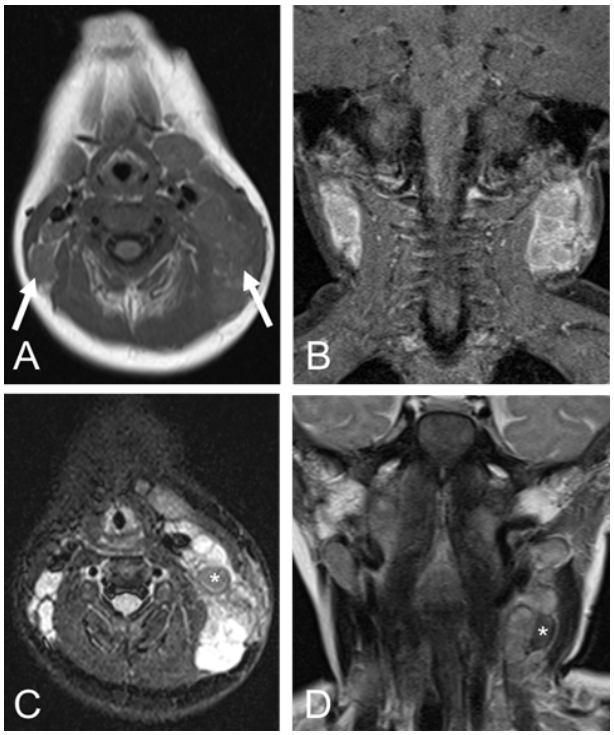

Figure 2 – Magnetic resonance imaging of the neck. (A) Axial T1 demonstrates bilateral cervical adenomegalies (arrows) practically isointense in comparison with the muscular structures, configuring adenopathic conglomerate on the left. (B) Coronal T1 with fat saturation and after intravenous contrast reveals intense and diffuse enhancement of adenomegalies. (C) T2 Axial with fat saturation and (D) T2 Coronal show adenomegalies mostly with T2 hypersignal, and some evident hyposinal areas (asterisks).

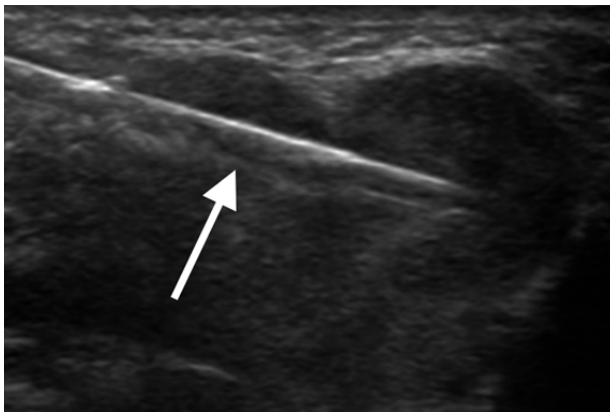

Figure 3 – Guided biopsy by ultrasound to left cervical adenopathic conglomerate, with a clear image of biopsy needle (arrow).

and CD68 exhibiting pictures lymphocytophagocytosis/emperipolesis and erythrophagocytosis.

The clinical-pathological diagnosis of Rosai-Dorfman disease was established. An expectant conservative treatment was chosen, with a gradual dimensional reduction of cervical adenomegalies, absent at the previous ultrasound reassessment performed approximately one year after the initial clinical presentation.

Discussion

Rosai-Dorfman disease is considered a rare histiocytic proliferation of aetiology not yet established, which appears more frequently in children and adolescents and has a higher incidence in the male gender.[1,2]

Patients usually present fever, leukocytosis and bilateral

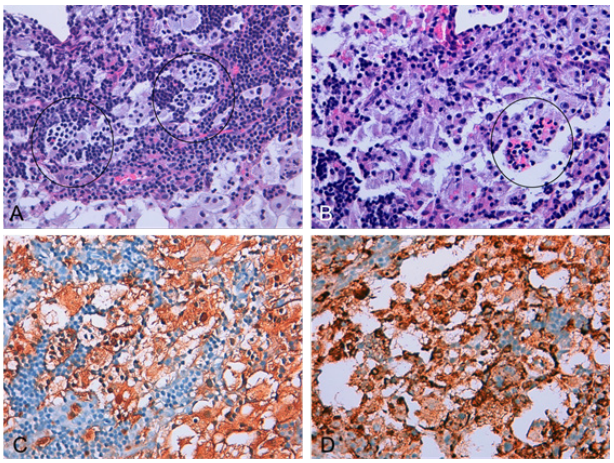

Figure 4 – Photomicrography of cervical ganglion biopsy with lesions of hemophagocytic lymphohistiocytosis in the context of Rosai Dorfman disease. Numerous large histiocytes are observed, exhibiting images of lymphocytophagocytosis/emperipolesis (figure A, H & E, 400x) and erythrophagocitosis (figure B, H & E, 400x). There is immunoexpression of S100 (Figure C, 400x) and CD68 (Figure D, 400x) in the bulky histiocytes.

cervical adenomegalies, which appear on physical examination as palpable cervical swellings. However, other ganglion territories can be hit.[1]

It is estimated that 43% of patients develop extra-ganglion manifestations of the disease, with 75% of these cases occurring in the head and neck region. Possibilities include cutaneous involvement of the nasal cavity, perineal sinuses, salivary glands, orbit and central nervous system. The prognosis of these patients is worse than those with only lymph node attainment.[1,2]

The imaging expression of Rosai-Dorfman disease is diverse and varies depending on the organs affected by the disease. The most common finding consists of cervical adenomegalies, which appear as enlarged lymph nodes in numbers and dimensions in different imaging techniques. Computed tomography (CT) and MRI scans usually show diffuse contrast enhancement after intravenous contrast administration.[1,2]

In MRI in particular, the adenomegalies present isosinal compared to the muscle in the T1 sequences prior to the administration of intravenous contrast, the T2 signal being variable according to different bibliographical references. Some publications refer hypersignal in T2,[2] while others mention hyposinal[1]. In the present case, the cervical adenomegalies appear predominantly hyperintense in T2, surprisingly, with some areas of hyposignal.

The most frequently affected perineal sinuses are the maxillary and ethmoidal sinuses, with opacification and thickening of the mucosa. In cases of involvement of the salivary or lachrymal glands, the increase in glandular volume is the most common imaging finding, with an intermediate intensity signal in T1 and T2 in MR evaluation.[1,2]

The imaging features previously described are highly non-specific for the diagnosis of Rosai-Dorfman disease. However, this entity should be weighed, especially in the presence of bilateral and painless cervical adenomegaly in children and adolescents.[1,2]

In this age group, cervical adenomegalies are mostly related to acute or chronic infections of the upper airway, and the differential diagnosis also includes tuberculosis, lymphoma, lymph node metastasis and other less prevalent entities such as Langerhans cell histiocytosis and Castleman.[2,3]

The typical histological feature of Rosai-Dorfman disease is the presence of emperipolesis, which consists more frequently in phagocytosis of intact lymphocytes, but also of plasmocytes, polymorphonuclear erythrocytes or leukocytes by histiocytes, Although emperipolesis is not unique to this entity, the immunoexpression of the S-100 and CD68 protein supports the diagnosis.[2,4]

When the diagnosis of Rosai-Dorfman disease is established, the potential involvement of other organs should be weighed.[2] In the clinical case described, chest radiography and abdominal, renal and pelvic ultrasound evaluation revealed no alterations.

The treatment is preferably conservative, with regular clinical surveillance. The clinical course is usually benign, with exacerbations and remissions, which usually cease spontaneously.[1,5] In the case of vital organs being affected, therapies such as corticosteroids, chemotherapy, radiotherapy or surgical interventions may be necessary.[2,3] There is no evidence of cases with malignant transformation of this entity.[1]

In summary, the Rosai-Dorfman disease hypothesis should be taken into account for painless cervical adenomegalias in children and adolescents. Since the clinical and imaging manifestations are non-specific, histological characterisation is fundamental in establishing the diagnosis and in the orientation and follow-up of these patients.

Received / Recebido 01/11/2017
Acceptance / Aceite 23/03/2018

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.

Financing Support: This work has not received any contribution, grant or scholarship.

Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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