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Rosai-Dorfman Disease with Brain Damage in One Case at the National Hospital of Zinder

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Abstract

Introduction: Rosai-Dorfman-Destombes disease (MRDD), or non-Langerhansian histiocytosis, is characterized by the presence of multiple polyadenopathies. It is a rare myeloproliferative disorder of histiocytes with a broad spectrum of clinical manifestations and distinctive morphological features, with abnormal accumulation of histiocytes (in emperipolesis) in lymph nodes. **Observation**: A 16-year-old Black male, orphaned by his father and his mother, a housewife, from a sibling group of six (06) children, of which he is the eldest, with no particular pathological history, was admitted to our clinical haematology department at the Zinder/Niger National Hospital for chronic painless bilateral cervical polyadenopathy associated with persistent fever resistant to the usual analgesics. The diagnosis of lymph node tuberculosis was considered. The patient received standard anti-tuberculosis treatment with good compliance for six (06) months. The course was marked by persistent night sweats and fever, which remained at a plateau despite antipyretics. Repeated transfusions were observed. **Conclusion**: Rosai-Dorfman-Destombes disease (RDDD) is a histioproliferative disorder characterized by lymphadenopathy. It is a type of histiocytosis that usually appears in children as bilateral cervical lymphadenopathy. It can manifest itself in different ways, depending on the location of the lesion. It remains a major diagnostic challenge for clinicians. Cases involving the central nervous system are rare.

Keywords

Rosai-Dorfman Disease, Polyadenopathy, Histiocytosis, Langerhans,

Emperipolesis

1. Introduction

Rosai-Dorfman-Destombes disease (MRDD), or non-Langerhansian histiocytosis, described in 1965 [1], is characterized by the presence of multiple polyadenopathies. It is a rare myeloproliferative disorder of histiocytes with a broad spectrum of clinical manifestations and distinctive morphological features, with abnormal accumulation of histiocytes (in emperipolesis) in lymph nodes [2]. It occurs more frequently in subjects under 20 years of age. The cause is not yet well defined. Once considered a benign, self-limiting condition, current evidence points to mutations in the MAPK/ERK pathway (MAPK: Mitogen-activated protein kinases/ERK: Extracellular signal-regulated kinases is a chain of proteins in the cell that communicates a signal from a receptor on the cell surface to DNA in the cell nucleus) in 30% - 50% of cases [3]. In the fifth edition of the World Health Organization's classification of hematopoietic tumors and in the International Consensus Classification, Rosai-Dorfman-Destombes disease has been incorporated as a histiocytic neoplasm [2]. Today, therefore, it is a clonal neoplastic process. Localizations are usually cervical, axillary, inguinal, thoracic, or abdominal lymph nodes. Rarer extra-nodal localizations have been described. Non-Langerhansian histiocytosis remains a major diagnostic challenge for clinicians, particularly when it involves the Central Nervous System (CNS). We discuss the diagnosis of Rosai-Dorfman-Destombes disease using a case report in a 16-year-old patient with superficial cervical adenopathy and motor deficit of the left hemicircle that may simulate rare cerebral involvement associated with bilateral cervical lymph node localization.

2. Observation

A 16-year-old black male, orphaned by his father and a housewife, from a sibling group of six (06) children of which he was the eldest, with no particular pathological history, was admitted to our clinical haematology department at the Zinder/Niger national hospital for chronic painless bilateral cervical polyadenopathy associated with persistent fever resistant to the usual analgesics. The diagnosis of lymph node tuberculosis was considered. The patient received standard anti-tuberculosis treatment with good compliance for six (06) months. The course was marked by persistent night sweats and fever, which plateaued despite antipyretics. Significant weight loss in excess of 10% of body weight over six (06) months, persistent and enlarged cervical adenopathies, and hemolytic anemia requiring repeated transfusions were observed. In this context, the patient was referred to the national hospital in Zinder for treatment.

On physical examination, the patient was conscious, presented with an altered general condition (bedridden), and cervical lymph nodes approximately 5 cm in

diameter, bilaterally firm, painless, and with no tendency to fistulize. Figure 1 illustrates the cervical lymphadenopathy in our clinical case.

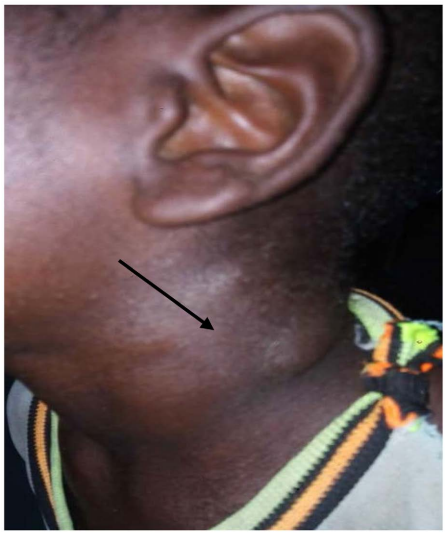
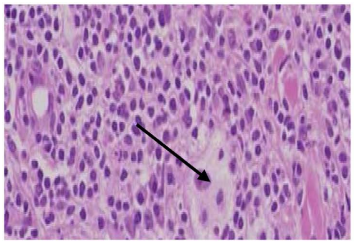


Figure 1. Cervical adenopathy.

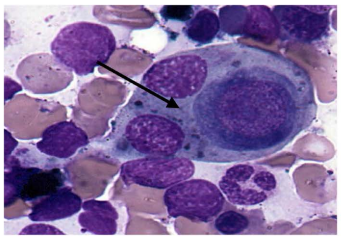
The conjunctivae were pale, with hemodynamic and ventilatory disturbances. The symptomatology had evolved over time with headaches and visual disturbances, and the presence of a left neurological deficit.

3 Paraclinical Findings

- **Biological tests:** Showed hyperleukocytosis, normocytic anemia, elevated erythrocyte sedimentation rate, and triple-digit quantitative CRP. Serologies for HIV and hepatitis B and C were negative.
  - **On lymph node cytology:** On fine-needle lymph node smear, after staining with May-Grünwald Giemsa (MGG), there was a cytomorphology of large histiocytes in emperipolesis. Figure 2(A) and Figure 2(B) illustrate the histiocytic cell undergoing empipolesis in cytology after staining with MGG.



A: low magnification × 10



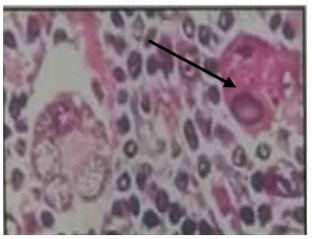
B: high magnification × 100

Figure 2. Histiocytosis on lymph node smear.

  - **Ganglion histopathology:** High magnification shows ganglion tissue with distorted and affected architecture. The distortion is due to diffuse layers

of histiocytes extending into the paracortex and surrounding the preserved cortical follicles. The histiocytes show focal emperipolesis and are mixed with a large number of plasma cells. No evidence of tuberculosis was observed on lymph node histology. Microscopy shows sections of lymph node with histiocytic cells showing focal emperipolesis and focal multinucleated giant cells. These are surrounded by lymphoplasmacytic cells and arranged in an amyloid-like stroma. **Figure 3(A)** and **Figure 3(B)** show the macroscopic and microscopic histological architectures of the lymph node.

A: macroscope × 40



B. H and E: microscopy × 40

Figure 3. Histopathology of lymph nodes.

A-ganglion with distorted and affected architecture.

B-histiocyte infiltration and presence of emperipolesis

- Immunohistochemistry: Histiocytic cells show diffuse positive staining for S100 and focal positive staining for cyclin. **Figure 4(A)** and **Figure 4(B)** illustrate diffuse staining of the S100 protein and focal positive staining for cyclin in immunohistochemistry.

A x 40

B. x 40

Figure 4. A-Diffuse positive staining for S100; B-focal staining is positive for cyclin.

The entire panel (CD68, CD163, CD1a, CD207, Kappa/Lambda, etc.) was not tested due to insufficient financial resources. The immunohistochemical study revealed diffuse expression of the S100 protein within large histiocytic cells, which strongly suggests non-Langerhans cell histiocytosis. Although not specific, S100 positivity is a sensitive marker for Rosai-Dorfman cells. Furthermore, focal expression of cyclin D1, although sometimes observed in other contexts, has been reported nonspecifically in Rosai-Dorfman disease, without this reflecting neoplastic proliferation. The presence of empyema on histological examination strongly

supports the diagnosis.

- **Medical imaging**
  - Brain scan

The brain scan showed a double focus of right hemispheric supratentorial porencephalic cavity, with cortico-subcortical atrophy. Area of frank hypodensity in the right cerebral hemisphere, with widening of the right cortical sulci and the right sylvian valley. Appearance consistent with ischemic stroke sequelae in the right carotid territory. Figure 5 illustrates this situation.

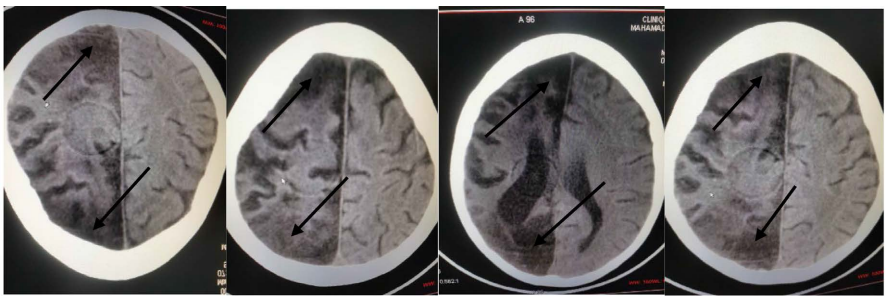


Figure 5. Cerebral CT scan.

The brain scan showed an extra-axial lesion, well circumscribed and often in contact with the dura mater, mimicking a meningioma.

- MRI, Angiography, and Brain biopsy

These were not performed due to insufficient technical facilities and financial resources. At the end of the investigations within our scope, the diagnosis of Rosai-Dorfman lymph node disease with sequelae of ischemic stroke in the right carotid territory, with no real link established to this effect, was retained. The epidemiological, clinical, and paraclinical markers involved in the differential diagnosis allowed us to rule out tuberculosis (unilateral lymphadenopathy, giant cell granuloma, caseous necrosis, negative S100), lymphoma (persistent lymphadenopathy, B signs, monoclonal lymphoid proliferation, negative S100), Langerhans cell histiocytosis (bone lesions, skin lesions, histiocytes with reniform nuclei, granulomas, eosinophils, high S100), and autoimmune adenitis (painful cervical lymphadenopathy, fever, sometimes arthralgia, areas of necrosis, apoptosis, immunoblastic cells, often S100 negative).

The therapeutic landscape for Rosai-Dorfman disease is limited to the immediate use of glucocorticosteroids in our resource-limited countries. After diagnostic confirmation, our patient received a five-day bolus of high-dose intravenous solumedrol, which improved the clinical status.

4. Discussion

Patients with Rosai-Dorfman-Destombes disease typically present with bilateral, painless, massive cervical lymphadenopathy associated with B-type symptoms. In a 2022 study in the USA, entitled Between proliferation and neoplasia in MRDD, around 43% of patients presented with extra nodal involvement [2]. We report

the case of a young Black Nigerian adult with brain involvement associated with MRDD. It is more common in young adults of African descent (mean age: 20.6 years) [4] [5]. When MRDD affects the central nervous system (CNS), it is referred to as CNS-RDD [6]. Neurological presentation in the form of an isolated brain tumour is rare [7]. Most patients with Rosai-Dorfman disease present with multiple tumour lesions [8]. These extra-ganglionic lesions mainly affect the skin and nasal cavity, and are diagnosed histopathologically [9]. The common radiographic appearance of intracranial MDDR is a solitary, homogeneously enlarging, extra-axial dural mass mimicking a meningioma [10] [11]. Fine needle aspiration cytology (FNAC) in our patient showed a cytomorphology of large histiocytes in emperipolesis. This cytological feature of the disease can be used as a first-line test. It is a useful finding, but is not necessary for diagnosis, as it may be focal [12] [13]. Histologically, infiltration of histiocytes was observed, as well as the presence of emperipolesis in the specimen. The section showed ganglion tissue with distorted and affected architecture. Distortion due to diffuse layers of histiocytes extends into the paracortex and surrounds the preserved cortical follicles. Histiocytes show focal emperipolesis and are mixed with large numbers of plasma cells. Sections of the lymph node show histiocytic cells with focal emperipolesis and focal multinucleated giant cells. These are surrounded by lymphoplasmacytic cells and arranged in an amyloid-like stroma. Histiocytic cells show diffuse positive staining for S100. Histiocytic cells show focal positive staining for cyclin. MRDD is a histiocytic, S100-positive proliferation that can cause ganglionic and extranodal disease [14]. Characteristic lesional histiocytes are S100+, CD68+, and CD1a- and exhibit a variable frequency of emperipolesis [15]. Pathology and immunohistochemistry have been of crucial importance, showing a specific pattern of histiocytosis [16]. Studies published in the Egyptian Journal of Radiology and Nuclear Medicine in 2024 reported a rare case of intraventricular Rosai-Dorfman disease in a 34-year-old female patient (imaging: T2 hypointense, homogeneous dense enhancement, no calcification or hemorrhage, and diagnosis confirmed on surgical specimen: histiocytes with empyema, S100 positive, CD68 positive, CD1a negative) [17]. In 2025, the Egyptian Journal of Neurosurgery reported seven cases of intracranial Rosai-Dorfman disease in a retrospective study, providing a set of clinical, radiological, and histological data from a larger sample [18].

5. Conclusion

Rosai-Dorfman-Destombes disease (RDDD) is a histioproliferative disorder characterized by lymphadenopathy. It is a type of histiocytosis that usually appears in children as bilateral cervical lymphadenopathy. It can manifest itself in different ways, depending on the location of the lesion. It remains a major diagnostic challenge for clinicians. Cases involving the central nervous system are rare.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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