

# Rosai-Dorfman Disease: Two Cases Report and Literature Review

Atoumane Faye\*, Nafissatou Diagne Sakho, Fatou Samba D. Ndiaye, Seynabou Fall, Awa Cheikh Ndao Mbengue, Boundia Djiba, Baidy Sy Kane, Souhaibou Ndongo, Abdoulaye Pouye

Medical Clinic 1 University Teaching Hospital Aristide Le Dantec/Cheikh Anta Diop University of Dakar, Dakar, Sénégal

Email: [\\*atoufayemi@yahoo.fr](mailto:atoufayemi@yahoo.fr)

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## Abstract

The lymph node is the revelation mode of several pathology. In tropical area, their etiology are dominated by tuberculosis and hemopathy. Some etiology such as histiocytosis are rarely mentioned. We report two cases of Rosai-Dorfman-Destombes disease. The first observation concerned a patient of 45 years without pathological antecedent, who had a inguinal and cervical tumoral lymph nodes. This evolved in a feverish poor general condition. Infection research was negative. Morphological explorations found deep lymph nodes. The excision biopsy examination concluded to Rosai-Dorfman-Destombes disease. The second observation concerned a patient of 40 years without pathological antecedent, who had a chronic non-inflammatory left supra-collarbone lymph node, associated with poor general condition and fever. X-ray showed mediastinal lymph nodes and ultra-sonography showed mesenteric lymph nodes, and latero-aortic lymph nodes. The examination of the excision biopsy was for Rosai Dorfman Destombes disease. The difficulty of diagnosis in our regions based on technical tray lack and our patients financial limits. Also this disease is rarely mentioned first. This is often the source of considerable diagnostic delay noted in our two patients and therefore the initiation of an untimely anti-tuberculosis treatment.

## Keywords

Histiocytosis, Rosai-Dorfman-Destombes Disease, Sub-Saharan Africa

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## 1. Introduction

Destombes-Rosai-Dorfman-Disease (DRD) also known as sinus histiocytosis with massive lymphadenopathy or

\*Corresponding author.

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sinus histiocytosis cytophagic is a rare disease of unknown aetiology. This is a rare and benign disorder; the evolution is often self-limiting after a few months. Sometimes it can induce permanent damages some of which can be dramatic. The prognosis depends on the extranodal disease.

Described for the first time in 1965 by P. Destombes, clinically it manifested as a massive lymphadenopathy mainly in cervical region but it could involve other sites. Affecting other lymph node is however, not exceptional [1].

The delay in diagnosis is partly due on one hand to the rarity of the disease, and on the other hand to the higher prevalence of infectious aetiology which is caused by hard socio-economic conditions of our patients. This often leads us to report two observations.

### 1.1. Observation 1

A 45 years old patient without significant past medical history presented with asymmetric bilateral cervical lymphadenopathy predominating on the right, with the longest axis measured 8.2 cm, associated submental, left axillary and bilateral inguinal; the lymph nodes were non tender hard and fixed in deep tissue, the patient reported a mild fever, weight loss and loss of appetite. Giving the higher frequency of tuberculosis in our region, an anti-tuberculosis treatment was started. Anti TB treatment stopped after one month in the lack of clinical improvement and absence of *Mycobacterium tuberculosis* in culture. The patient readmitted and Clinical examination showed a non-icteric pale conjunctiva and a mild splenomegaly, ENT examination by specialist yielded nothing and there was no fever. Laboratory analysis revealed normocytic normochromic anaemia of 6.8 g/dl. Haptoglobin was not analysed; serum inflammatory markers were in normal ranges. Serum protein electrophoresis showed a polyclonal hypergammaglobulinemia of 27.1 g/l and LDH was 4 times higher than normal.

The PPD and HIV serology was negative. The CXR revealed right hilar and pretracheal lymphadenopathy. Abdominal ultrasound showed superior mesenteric and hepatic hilum lymphadenopathy and a 14 cm spleen. The excisional biopsy of cervical lymphadenopathy revealed architectural disruption and populated with a polymorphic infiltrate composed of mainly foamy histiocytes and cytophagocytes, lymphoplasmacytes associated with neutrophils suggestive aspect of Destombes-Rosai-Dorfman-Disease (DRD). Immunohistochemistry could not be done. Because of persistence and the slow progression of the lymphadenopathy without local inflammation and compression signs, corticosteroids at a dose of 1 mg/kg/day had been started and continued for 6 weeks.

The outcome was favourable with clinically significant decrease in the size of lymph nodes; spleen and a weight gain of 8 kg in 2 months without evidence of steroid impregnation.

Biologically, the haemoglobin increased from 6.8 to 13 g/dl without transfusion or iron supplementation. Morphologically, ultrasound noted the persistence of hilar lymphadenopathy. Thus, given the clinical and laboratory improvement, a maintenance dose was maintained with gradual dose reduction; thereafter and for professional reasons, the patient was lost of view.

### 1.2. Observation 2

A 40 years old black man, with unremarkable past medical history consulted for a painless slowly progressing left sub-clavicular swelling since one month ago. A few weeks later, he noted similar swellings in the left axillary and right inguinal region

These symptoms evolved in a context of a moderately poor general health status, fever and night sweating. Giving the clinical data and epidemiologic context an anti TB treatment started. Despite the Anti TB treatment general status worsened and the swellings increased so the patient was referred to our department. On admission physical examination revealed an average general condition, palor of the mucous membranes and a low-grade fever. He also revealed a painless mobile left supraclavicular lymphadenopathy, measuring 4 cm in diameter, rubbery consistency, without periadenitis or fistulisation tendency and as well as right axillary and inguinal lymphadenopathy measuring between 1.5 and 2.5 cm in large axis with the same characters. There was no hepatosplenomegaly.

The remainder of the physical examination including a specialised ENT examination was normal.

Hematologic examination revealed microcytic hypochromic anaemia with a haemoglobin of 7.9 g/dl, and an ESR of 125 mm/1 hour and a serum protein of 61 g/l. CRP and protein electrophoresis were not done.

Blood biochemistry, serum calcium as well as liver and renal function tests were in normal ranges, PPD and retroviral serology was negative.

The chest radiograph showed mediastinal lymphadenopathy. Abdominal ultrasound found voluminous peritoneal and retroperitoneal lymph nodes enlargement. There was no splenomegaly.

Lymph node biopsy noted homogeneous lymph node architecture with many histiocytes sheet and some eosinophil. The cells showed no atypical nuclear features. There was no giant cell and epithelioid granuloma.

Corticosteroid therapy was instituted at a dose of 1 mg/kg/day. The outcome was favourable after few weeks with a good regression of cervical and inguinal lymphadenopathy. After few months unfortunately, the patient was lost to follow-up due to a false address and the lack of a phone number on her file that made vain any attempt to touch her.

## 2. Discussion

The sinus histiocytosis or Destombes Rosai Dorfman disease is a chronic condition characterized by benign proliferation of non-Langerhans histiocytosis cells.

Its aetiology remains obscure although its infectious nature is very often mentioned including infections EBV, CMV, measles, rubella, HHV8, HHV6 and toxoplasmosis infections [2] but also immunological disorders with abnormal histiocytes reaction.

This is a rare condition. And until 1990, Rosai established an international registry and Dorfman and it contained 423 observations. This register is currently the most comprehensive source of data [3]. However hundreds of isolated cases have been reported worldwide, but only 1 - 3 cases is reported in Africa [4]-[6].

The sinus histiocytosis is a worldwide disease occurring preferentially among black patients especially children and young adults. No racial group is spared [7] [8] even if the unfavourable socio-economic condition, as so was the case for one of our patients have been widely highlighted by some authors [6] [9].

It is a disease of adolescents and young subjects. The average age of onset is 20 years [3] [7] [10], but the condition may appear at any age [9]-[11]. In our case, our patients were aged 45 years for the first case and 40 years for the second case. Diebold [9] reports the case of Martinique aged 58 and Essalki [5], two cases Senegalese children aged 12 and 7 years. Forms the extreme ages were reported in particularly by Sacchi [12] who reported a case of congenital histiocytosis and Galicier [3] with a patient aged 74-year by time of diagnosis. A slight male predominance was noted [10] [13] [14] with a sex ratio of 1.4/1 in favour of men. The lymph nodes constitute a classical mode of revelation of the disease. They are chronic, non-inflammatory often localised on the cervical spine. Indeed, this location is suggestive of the disease in more than 90% of cases [1] [3] [12] [13] as is the case in our observations where the cervical lymph nodes were the presenting feature.

These are typical voluminous cervical lymphadenopathy that can measure up to 7 cm long axis [13] [14] often bilateral, asymmetric, firm, painless, although with limited mobility and of a pseudo-tumour aspect without necrosis or tendency to suppuration and fistulisation.

They can last for months or years and are generally non-compressive. Other axillary and inguinal lymph nodes most often, may be interested. Sometimes the disease is diffuse interesting even deep area as is the case with our patients with diffuse superficial and deep lymph nodes clinically consistent with that of literature.

In our first observation, the most voluminous lymphadenopathy was 8.2 cm of long axis with the same characteristics described above, non-compressive. Slaoui [15] in Morocco, reported a case of a 3 years girl who presented with a compressive lateral cervical lymphadenopathy originating from superior vena cava syndrome and from Claude Bernard Horner syndrome.

Hepatosplenomegaly is rare [10] [11]. For other authors, liver and spleen are consistently normal. In one of our cases, type 1 splenomegaly was found. This chronic splenomegaly may fall under our various aetiologies including tropics infection (scalable, visceral malaria, hepatosplenic schistosomiasis, visceral leishmaniasis), haematological (haematological malignancies, hereditary haemolytic anaemia such as sickle cell disease) or portal hypertension by intrahepatic block especially post hepatitis B cirrhosis. The poor general condition often described by Rosai [11] and Boman [8] was found in our patients but for most authors [3] [15] [16], the general condition is constantly maintained.

These multiple lymph nodes in this context of poor general condition had lead to suspected tuberculosis as an aetiology, which had justified the introduction of anti TB treatment before admission to our department.

Indeed, the early initiation of anti TB treatment is explained by the prevalence of this aetiology in our region where the disease endemic, favoured by promiscuity, lacks of vaccination with BCG at birth and non systematic pasteurizing cow's milk. Extra-nodal affections noted in 40% - 43% of cases depending on the series [10] [13] [14]. The most common sites are skin, head and neck, bone or involving the central nervous system. Biology is

indeed less contributive to the diagnosis of disease Dorfman Rosai Dorfman. However it reveals a non specific inflammatory syndrome with hypochromic rarely microcytic anaemia of inflammatory type, accelerated SR and sometimes major polyclonal hypergammaglobulinemia which was noted in one of our patients of 27.1 g/l; the other could not do protidogramme. This hypergammaglobulinemia was found more than 9 times out of 10 [3] and often more than 20 g/l. Biological data observed in our patients are consistent with those of the literature.

Histological examination revealed regardless of location, conserved nodal architecture with a significant expansion of the sinuses that are infiltrated by histiocytes with round or oval roughly nucleolus nucleus, abundant cytoplasm and little basophilic and presence in these histiocytes of many intact cells.

The latter completed by immunohistochemistry revealed positive CD64, and CD68 strong expression of S100 protein and negativity of CD1a.

Unfortunately for our two patients, immunohistochemical assessment could not be carried out because of its limited accesses that lead to a prominent place in lymph node biopsy associated with epidemiological, clinical and biological accessibility.

The evolution of the disease is chronic and can last several years with spontaneous remissions and exacerbations. For Foucar [7], healing would be complete in 23% of cases. Other authors [2] [3] [8] [11] argue that the tumour syndrome is spontaneously regressive in a substantial proportion of patients within an average of 5 years. In our cases, this period was short enough to judge the spontaneous evolution given “early” start of treatment.

To date, there is no consensus on the initiation of a systematic treatment of the disease [4] [11] except in cases where the life is threatened such as spinal cord compression or a respiratory disorder that affect ENT.

In a review by Pulsoni recent recovery [17] and compiling all publications from 1969 to 2000, half of the patients had not received any treatment and 80% of them had more than one clinical illness, which justifies this fact that, no treatment in most cases. For some authors, steroids, radiation therapy and chemotherapy have been used but without any effect. However corticosteroids remains important in the management, which is confirmed with the work of Lampert [18].

According to him, promising results were obtained with prednisone or a combination of prednisonevinblastine or chlorambucil. We used corticosteroid at a dose of 1 mg/kg/day for 6 to 8 weeks with spectacular clinical and laboratory improvement. However, due to the relatively short follow-up time (less than 6 months), it would be difficult to assess the long-term effect corticosteroid treatment.

Giving the self-healing possibility, therapeutic effect assessment remains quite delicate.

### 3. Conclusions

Our two observations show, despite its rarity, the reality of Destombes Rosai Dorfman disease in our regions. The main presenting feature of the disease is cervical lymph nodes enlargement. Although the treatment is not well classified, corticosteroids remain the backbone which caused a significant reduction in the size of lymph nodes in our patients. However, sufficient data are needed to assess the long-term evolution.

The prevalence of this condition is not well known and few reported cases are limited to the lymph nodes; looking for other locations is not done routinely.

Thus, to clarify the actual frequency of the disease in its nodal and extra nodal form, it would be relevant for a better follow-up to make a comprehensive assessment of extension of the disease and to establish a register of chronic diseases and orphan.

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