

Review

A false adenolymphoma revealing the disease of rosai-dorfman and destombes: About one observation and literature review

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We present one observation of the Rosai-Dorfman and Destombes disease (RDDD), particularly under its revealing form and by its favorable evolution under corticosteroid therapy. It was a patient 29 years old, who presented multiple cervical and abdominal adenopathy. All operating in the alteration of the general condition and fever. The diagnosis is based on histological evidences after ganglionic biopsy which showed a sinus lymphohistiocytosis. The evolution has been favorable under corticoid treatment. The Rosai-Dorfman and Destombes disease is a rare benign affection, thus difficult to make diagnosis.

Key words: RDDD, adenopathy, histology, evolution, corticoid, University Hospital Center (UHC)-Treichville.

INTRODUCTION

The Rosai-Dorfman and Destombes disease often called a sinus histiocytosis with massive lymphadenopathy is a benign and rare affection whose causes are unknown (Bernard and Sarran, 1999; Boman-Ferrand et al., 1990). It is linked with lympho-histiocytic disorder which creates a benign histiocytic proliferation (Davidson and Kelly, 1999; Destombes et al., 1965). It touches cervical ganglionic area preferentially (Bernard and Sarran, 1999; Foucar and Rosai, 1991; Franklin et al., 2006). Its diagnosis is based on histological study associated most of the time with the immunohistochemistry test. Its treatment is poorly codified but the prognostic is the most often favorable. We propose through an observation of the Rosai-Dorfman and Destombes (RDD) disease revealed on a ganglionic lymphoma form, to review this disease on diagnosis plan and make the literature review.

OBSERVATION

Sir G.A, 29 years old, consults our office for a cervical large mass that has been growing for several months and it is associated with the alteration of the general condition, an intermittent fever without figures. We noted in his antecedents a tuberculous contagion, and from our estimation, he smoked 6 packs per year.

During clinical examination, it was multiples bilateral cervical adenopathy with a size that varies between 2 to 5 cm, little painful, mobile with a red skin pre-suppurative in front (picture 1.a and picture 1.b).

The others superficial ganglionic areas were free, in the same way the ORL exam. Otherwise, we note a type II splenomegaly according to the Hackett's classification with hepatomegaly associated. The liver had a regular and smooth surface. The other organs especially lungs, endocrine and neurological were without particularity.

Biological assessment revealed a microcytic anemia (Hemoglobin rate was 8.7g/dl) in the blood formula with a bicytopenia associated (white blood cell were 3591 elements/mm³ and red cells were 3.7 10⁶) the



Picture 1a. Adenopathy with red skin in front

reticulocyte rate was 77 400/ mm³. The ferritin was 823 µg/l. sedimentation speed was 90 mm during the first hour and 120 mm during the second hour. The lactate dehydrogenase was 548 UI/l, the blood electrolytes and kidneys were normal. The HIV serology, the B hepatic serology, the C hepatic serology were negative. The tuberculin skin reaction (10 unities) was 12 mm. the research of the tuberculosis bacteria in the sputum was negative (3 series).

The cervical ultrasound has shown a bilateral polyadenopathy (jugular-carotid and under mandibular) with abdominal adenopathy associated (para-aortic and mesenteric). The thorax picture was normal.

The ganglionic puncture made was little contributive. Cervical lymphadenectomy with histological study revealed the ganglionic localization of the RDDD (picture 2).

A corticosteroid therapy with prednisone 20 mg tablets has been prescribed, 2,5 mg loading dose per day during 2 weeks to reach the degressive dose of 5 mg at the end of 3 weeks. The evolution has been favorably proved by the disappearance of cervical adenopathy (picture 3) and abdominal adenopathy after 6 months.

DISCUSSION

The lymphohistiocytosis or the Rosai-Dorfman and Destombes disease belongs to the family of histiocytosis non langerhans and has been described for the first time by Destombes (1965), and defined as a clinical-anatomical entity, by Rosai and Dorfman (1969-1972).

It is a benign affection and rare which touch any age with a frequency top at 20 years. A predilection to the male sex and the black people (Bernard and Sarran, 1999; Laboudi et al.,1990; Rosai and Dorfman in 1965 and 1972). In our observation this disease appeared to a person who is 29 years old, masculine sex and black.

The clinical signs are essentially dominated by the presence of a bilateral cervical lymphadenopathy which can be associated or be alone (Sardana et al., 2014; Boman-Ferrand et al., 1990; Rangwala et al., 1990). This ganglionic infringement can be unique or multiple and can be through the superficial ganglionic area (Kharrat and Sahtout 2008; Wenig et al.,1993).

As it was in our patient where we found multiple cervical adenopathy with abdominal adenopathy associated. In fact, Harmouche and Amrani (1999), Rangwala and Zinterhofer (1990) and Hicke et al., (1995) revealed that the cervical ganglionic infringement are observed in 87,3% cases during the RDDD. The intermittent fever, the alteration of the general condition, splenomegaly and hepatomegaly has been the clinic signs found in our patient.

Biologically we found an accelerated sedimentation speed, microcytic anemia, a bicytopenia. All this elements are found in the RDDD (Laboudi and Haouazine., 2001; Kharrat and Sahtout, 2008; Harmouche et al.,1999).

The myelogram could see an exceptional histiocytic infiltrate but it was not realized in our case. However the inflammatory aspect of the adenopathy, the high rate of the lactate-dehydrogenase with positive tuberculin skin



Picture 1.b. Bilateral adenopathy.

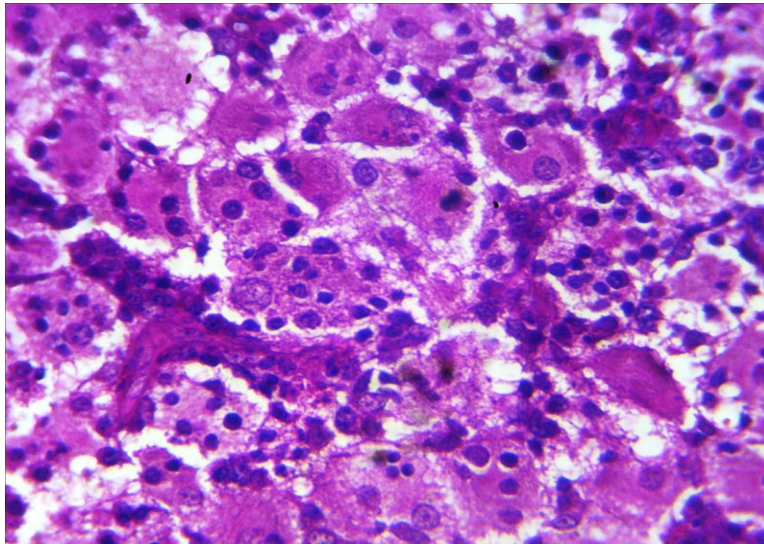
reaction associated, which are unusual, allowed us to evoke a systemic disease, a lymphoma or ganglionic tuberculosis.

The ganglionic puncture having been little contributive, ganglionic biopsy was needed. The histological exam has been done on the piece of adenopathy and confirms the diagnosis of the Rosai-Dorfman and Destombes disease. In fact, the optical microscopic aspect of the adenopathy, showed sinus dilatation characterized by the presence of the histiocytes, voluminous, sometime several cores with abundant cytoplasm, acidophil, vacuole or foamy containing lymphocytes, plasmocytes, polymorphonuclear or red cells (emperipolesis) (Rosai and Dorfman in 1965 and 1972).

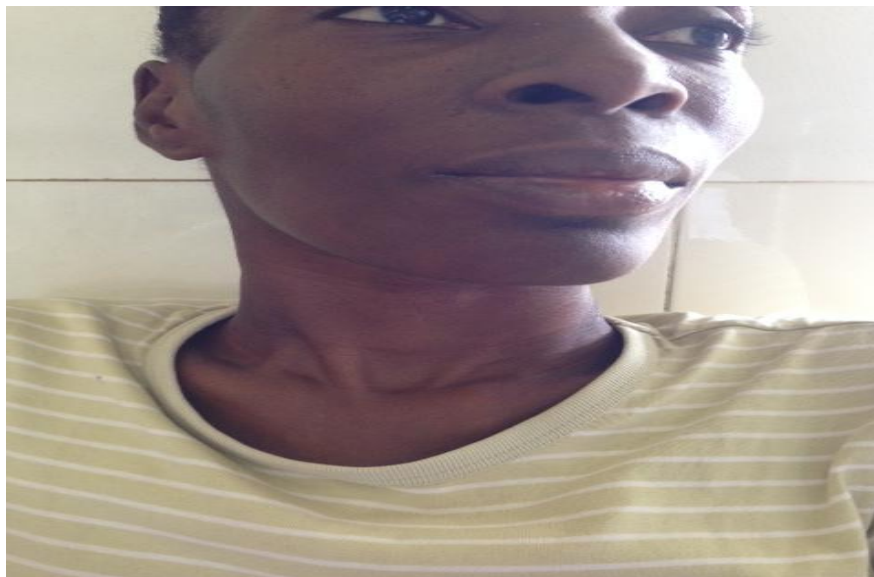
The RDDD's differential diagnosis is made with other disease like Kikuchi Fujimoto disease or the subacute necrotizing lymphadenitis, which is a rare disease. It has no cause and presents itself like cervical adenopathy with fever. This disease occurs specially in the young girl with an important prevalence in Asia continent (Tsang and Chan, 1994; Turner et al., 1983).

In our observation, the patient is young and a male sex. In biology data, a moderate leucopenia is frequent as in our case. The evolution is benign with complete remission in couple of weeks (Kuo, 1995). This has not been observed in our case. As differential diagnosis, we have the Castleman disease which is an atypical lymphoproliferative syndrome resulting to the polyclonal proliferation of B cells into lymphoid organs (Guillem et al., 2000). Its signs are characterized by the anemia, the fever, the sweat and the weight loss, frequently with a tumor syndrome associated (polyadenopathy), it can be associated with hepatomegaly and splenomegaly

(Waterson et al., 2004). The clinic signs such as polyseritis, skin rash, central neurological manifestations or peripheral associated with biology signs like inflammatory syndrome, an anemia, a thrombocytopenia, a hypo-albuminemia and a polyclonal hypergammaglobulinemia are often found like in the RDD disease (Farruggia et al., 2011). Only the histology can make the difference (Waterson et al., 2004). We can also discuss systemic diseases such as the lupus erythematosus in front of the lupic adenopathy characterized by variable degrees of hyperplasia follicular said reactional and necrosis with presence of specific hematoxylic bodies but little sensible (Osborne et al., 1982). In the Gougerot-Sjogren syndrome, systemic disease during which peripheral adenopathy present only an hyperplasia follicular reactional (Smedby et al. 2006). The problematic between the Gougerot –Sjogren syndrome and peripheral adenopathy is the fact that basically, patient having the Gougerot-Sjogren, has 4 or 5 time more risk than general population to develop a Non-Hodgkin lymphoma (Rangwala et al., 1990; Kassan et al., 1978). Contrariwise, the risk to develop a lymphoma is 2 time superior in the general population on people with rheumatoid arthritis with peripheral adenopathy (Rosai and Dorfman, 1972). We mention also the adult's still disease, systemic affection, rare which touches the young adult (Franklin et al., 2006; Ohta and et al., 1987). The clinic signs are fever, arthralgia, skin rash and an important biological inflammatory syndrome, an elevation of the transaminase in the blood and a neutrophilia frank. More than half of patients present, cervical peripheral adenopathy in general, often multiple sensitive and non-inflammatory, (Smedby et al., 2006),



Picture 2. Histiocytic engulfing blood elements. Microphotography - HExG40.



Picture 3. Regression of the cervical adenopathy after corticoid treatment.

which is not the case in our observation. The other affections can be accompanied by histiocytes cytophagic, particular macrophage activation syndrome, the familial lymphohistiocytosis and the malignant histiocytosis could be discussed (Konsem et al., 2014; Harmouche et al. 1999; Hicke et al.,1995). That's why it is important to make the immunostaining which observes in case of the RDD disease, histiocytes with S-protein 100 positive, and negativity of CD1 (Kuo,1995). Like in the sarcoidosis and systemic granulomatosis, the cause of the RDD disease, is unknown. Several theories have been proposed:

Infectious cause, especially Virus infection with EBV, HHV (Harmouche et al., 1999; Foucar et al.1991). Immunological proof by humoral disruption is almost constant and decreasing of the immunity cellular (Boman-Ferrand et al., 1990; Harmouche et al.,1999; Wenig et al.,1993). Both associations like an immunological answer is exaggerated (Boman-Ferrand et al., 1990; Foucar et al. 1991). It is true that the treatment is not well codified but the corticoid treatment with cytotoxic agent associated is practiced (Komp,1990). However it is not necessary

in the majority cases due to its spontaneous healing trend.

It is reserved most of the time to the form with compressive adenopathy or extra-adenopathy's form which threat or with the alteration of the general condition (Harmouche et al.,1999; Komp,1990). In our case the use of the corticoid allowed the full regression of these adenopathy.

CONCLUSION

The RDD disease affection is rare and benign in a young person often of black race. The association of clinic case and histological allows making precocious diagnosis. However, the corticoid treatment can improve the disease transiently .

Conflict of interest

The authors do not signal any conflict of interest.

REFERENCES

- Bernard F, Sarran N, Serre I, Baldet P, Callamand P, Margueritte G (1999). Sinus histiocytosis (Destombes-Rosai-Dorfman disease) revealed by extranodal spinal involvement. *Arch Pediatr*. 6:173-177.
- Boman-Ferrand F, Floquet J (1990). Histiocytose sinusale avec lymphadénopathie massive: syndrome de Destombes-Rosai-Dorfman. *Ann Pathol*. 10: 152-160.
- Davidson BKS, Kelly CA, Griffiths ID (1999). Primary Sjögren's syndrome in the north east of England: a long term follow-up study. *Rheumatology*. 38:245-253.
- Destombes P (1965). Adénites avec surcharge lipidique de l'enfant ou de l'adulte jeune observées aux Antilles et au Mali (4 observations). *Bull Soc Path Ex*. 6: 1169-1175.
- Farruggia P, Trizzino A, Scibetta N, *et al* (2011). Castleman's disease in childhood: report of three cases and review of the literature. *Ital J Pediatr*. 37: 50
- Foucar E, Rosai J, Dorfman R (1991). Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfmandisease): Review of the entity. *Semin Diag Pathol*. 7: 19-73.
- Franklin J, Lunt M, Bunn D, Symmons A, Silman A (2006). Incidence of lymphoma in a large primary care derived cohort of cases of inflammatory polyarthritis. *Am Rheum Dis*. 65:617-622.
- Galicier L, Fieschi C, Meignin V, Clauvel J, Oksenhendler E (2007). Rosai-Dorfman disease. *Presse Med*. 36:1669-1675.
- Guillem P, Karoui M, Deroide G, *et al* (2000). Maladie de Castleman de localisation mésentérique. *Gastroentero Clin Biol*. 24: 116 120.
- Harmouche H, Amrani L, Tazi Z, Aouni M, Adnaoui M, Mohattane A, Maaouni A, Bellabas M, Berbich A (1999). Forme particulière de la maladie de Destombes-Rosai-Dorfman (à propos de 2 cas). *Maroc Médical*. 21: 21-25.
- Hicke A, Wiczorek M, Olejnik I (1995). Rosai-Dorfman syndrome in a 17 year old boy as a diagnostic problem of lymphadenopathy. *Acta Haematol Pol*. 26: 425-428.
- Kassan SS, Thomas TL, Moutsopoulos HM, Hoover R, Kimberly RB, Budman DR (1978). Increased risk of lymphoma in Sjögren's syndrome. *Ann Intern Med*. 89:888-892.
- Kharrat S, Sahtout S, Oukhai M, Mekni E, Trabelsi S, Hauet S, Beltaief N, Besbes G (2008). Maladie de Rosai Dorfman à localisation multifocale : à propos d'un cas. *Fr ORL*. 94 : 395-398.
- Komp DM (1990). The treatment of sinus histiocytosis with massive lymphadenopathy (Rosai -Dorfman disease). *Semin Diagn Pathol*. 7: 83-86.
- Konsem T, Wend-Lasida Ouedraogo R, Millogo M, Gyebre Y, Bouaichi A, Zwetyenga N (2014). Two cases of sinus histiocytosis with massive cervical lymphadenopathy (Destombes-Rosai-Dorfman disease). *Rev Stomatol Chir Maxillofac Chir Orale* novembre; 115 (5): 313-7.
- Kuo TT (1995). Kikuchi'S Disease (Histiocytic Nevrotizing Lymphadenitis). A clinicopathologic study of 79 cases with an analysis of histologic subtypes, immunohistology, and DNA ploidy. *Am J Surg Pathol*. 19:798-809.
- Laboudi A, Haouazine N, Benabdallah L, Arzouk N, Cherradi N, Rhou H (2001). Rosai-Dorfman disease revealed by renal failure: case report. *Nephrologie*. 22:53-56.
- Ohta A, Yamaguchi M, Kaneoka H, Nagayoshi T, Hiida M (1987). Adult Still's disease: Review of 228 cases from the literature. *J Rheumathol*. 14:1139-1146.
- Osborne BM, Buttler JJ, Viriakotis D, Kott M (1982). Reactive lymph node hyperplasia with giant follicles. *A J Clin Pathol*. 78:493-499.
- Rangwala AF, Zinterhofer LJ, Nyi KM, Ferreira PP (1990). Sinus histiocytosis with massive lymphadenopathy and malignant lymphoma. *Cancer*. 65: 999-1002.
- Rosai J, Dorfman R (1969). Sinus histiocytosis with massive lymphadenopathy. A newly recognised benign clinicopathological entity. *Arch Pathol*. 87: 63-70.
- Rosai J, Dorfman R (1972). Sinus histiocytosis with massive lymphadenopathy: A pseudo-lymphomatous benign disorder: Analysis of 34 cases. *Cancer*. 30: 1174-1188.
- Sardana D, Goyal A, Gauba K (2015). Sinus histiocytosis with massive lymphadenopathy: A "massive" misnomer. *Diagn Cytopathol*. 43(4):315-319.
- Smedby KE, Baecklund E, Askling J (2006). Malignant lymphomas in auto immunity and inflammation: A review of risks, risk factors, and lymphoma characteristics. *Cancer Epidemiol Biomarkers Prew*. 15:2069-2077.

- Tsang William YW, Chan JKC. Kikuchi's lymphadenitis: a morphologic analysis of 75 cases with special reference to unusual features. *Am J Surg Pathol* 1994; 18: 219–31.
- Turner RR, Martin J, Dorfman RF. Necrotizing lymphadenitis. A study of 30 cases. *Am J Surg Pathol* 1983; 7: 115–23.
- Waterson A, Bower M (2004). Fifty years of multicentric Castleman's disease. *Acta Oncol.* 43:698-704.
- Wenig BM, Abbonolanzo SL, Childers EL, Kapadia SB, Heffner DR (1993). Extra-nodal sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease) of the head and neck. *Hum Pathol.* 24: 483-492.