

A rare presentation of extra nodal rosai-dorfman disease (case report)



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ABSTRACT

Rosai-Dorfman disease is considered to be one of the rare idiopathic disorders characterized by non-painful lymphadenopathy.

We reported a case of Rosai Dorfman disease that was presented with an isolated temporoparietal mass in an old Somalian patient.

It is a very rare condition that Rosai-Dorfman disease might show a temporoparietal area involvement can be seen from a review of all literature. The presentation of the disease, differential diagnosis and treatment were discussed.

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1. Introduction

Rosai-Dorfman disease characterized by sinus histiocytosis with massive lymphadenopathy (SHML). Rosai and Dorfman first described sinus histiocytosis with massive lymphadenopathy in 1969 [1]. The disease appears as a massive painless cervical adenopathy, and mainly affects the children or in young adults of African ancestry.

Some variants of this disease may occur in extra-nodal sites, often without any involvement of lymph nodes. 43% of cases occur in extra-nodal disease, which may be widespread and most frequently involves the respiratory tract, paranasal sinuses, visceral organs, skin, bones, central nervous system, genitourinary tract and orbits [2].

In some extra-nodal manifestations, patients can present soft tissue swelling/masses.

This pathology is very rare and the involvement of temporoparietal parotid gland area is exceptional. In most cases this involvement presents lympho-proliferation in the soft tissues. Approximately 10% of all cases of Rosai-Dorfman disease (RDD) are associated with soft tissue involvement, but some may show a sole manifestation of the disorder [2,3]. The cause of RDD is not yet clear. Epstein-Barr virus [2] and human herpes virus have been

isolated in a few patients, but clear association can't be verified. Autoimmune disease, immunocompromised, and neoplastic cell disease may be a cause but this remains unclear [2].

Diagnosing Rosai-Dorfman disease is considered to be dependent on its pathognomonic characteristics.

2. Case report

A 70year-old previously healthy female was seen with complaint of painless swelling over the left temporoparietal-eye area for the past four weeks. Rather than age related arthritis and carpal tunnel syndrome, her systemic history was not significant.

On examination there was an erythematous undulant nodular mass in the left temporal area [Figs. 1 and 2]. The lesion had a smooth surface with normal surrounding skin and was not tender. The mass appeared to be adherent to the underlying tissue.

Systemic evaluation of blood profile showed shifting from normal values. The patient underwent incisional biopsy and the mass was subjected to histopathological examination. EM examination with Haematoxylin and eosin stained sections showed a mixed cellular infiltration, predominantly composed of histiocytes that was mixed with lymphocytes including plasma cells and polymorphous nuclear leucocytes. emperipolesis was shown within several histiocytes, (displaying phagocytized lymphocytes). The histiocytes were filled with pink cytoplasm and contained lymphocytes, which is a pathognomonic finding of rosai dorfman disease [Figs. 3 and 4]. Stains for bacteria, fungus and acid-fast bacilli were negative.

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Fig. 1. Clinical photograph demonstrating erythematous indurated mass in the left temporoparietal area (A side view).



Fig. 2. Clinical photograph demonstrating erythematous indurated mass in the left temporoparietal area (face front).

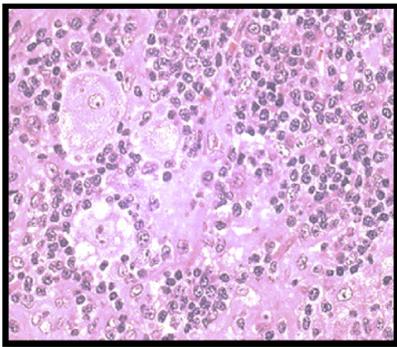


Fig. 3. EM, showing chronic inflammatory cell infiltrate with lymphocytes, plasma cells and histiocytes containing lymphocytes in their pink cytoplasm and demonstrating emperipolesis.

The patient was started on systemic prednisolone (1 mg/kg body weight per day) on a tapering dose for a period of four weeks. After six weeks of follow-up, there were notable regression of the lesion and the patient was advised a follow-up after three months.

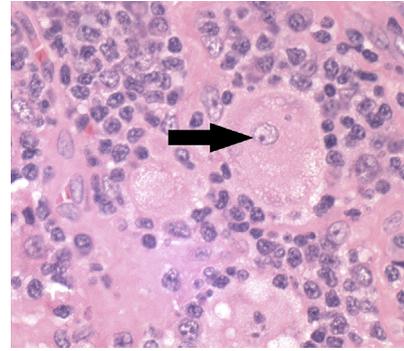


Fig. 4. EM showing emperipolesis (black arrow).

3. Discussion

The onset of disease is deceptive; its active phase seems to be prolonged and its remission may be spontaneous with possibility recurrence [3]. The factors that determine recurrences are not well understood.

The predominant clinical manifestation of the disease is tumoral cervical adenopathy (87.3% of cases) that, in most cases, is painless and bilateral, affecting one or all cervical chains. The neck lymph nodes are the most frequently involved, followed by inguinal, axillary and mediastinal lymph nodes [4].

Lymph nodes are isolated, mobile, and small during the initial stages but become adherent with disease progression, forming a voluminous multinodular mass. The most common extra nodal sites are the skin, the upper respiratory tract and bones. Head and neck involvement - about 22% of extra nodal disease [5] include involvement of the nasal cavity, the paranasal sinuses, the nasopharynx, submandibular glands, the parotid, the larynx, the temporal bone, the intratemporal fossa, the pterygoid fossa, the meninges and the orbit [6]. The skin is commonly affected; half of these patients have another associated extra nodal site. Orbit and ocular globe involvement have been reported, usually as a retro arbitrary mass and proptosis [7], although nodal involvement may have occurred during an earlier phase. Fever occurs in up to 30% of cases but was absent in our patient.

In 85% of cases, patients with Rosai-Dorfman disease are in good general health without significant symptoms of the disease [8].

Our patient was complaining of soft tissue mass in the temporal region, in addition to age related polyarthritis and radial tunnel syndrome.

Laboratory alterations are frequent and include anaemia (65.7%), leucocytosis (59.1%), neutrophilia (68.4%), increased ESR (88.5%), and hyper-gamma-globulinemia (90%) [1,2], which was similar to the laboratory results in our case.

The cause of the disease has not yet been established [9], but two theories exist. In the first theory, SHML is caused by a specific infectious process as Epstein Bar Virus and human Herpes viruses, which have been isolated in some patients [11], based on the generally infectious process seen at the onset of the disease (localized adenopathy, fever, leucocytosis with neutrophilia, increased erythrocyte sedimentation rate, and hyper-gamma-globulinemia), which tends to spontaneously regress after some time. However, no laboratory evidence points to an etiologic agent). In the second theory, the disease is attributed to an abnormal immunologic response, because depression of immunologic cells can be observed.

A histopathological diagnosis was made and showed histiocytic

infiltration mixed with lymphocytes and other inflammatory cells. One typical feature of this entity has been emperipolesis, which is the presence of cells of one type within the cytoplasm of cells of another lineage.

Rosai-Dorfman disease is an entity that is part of a group of systemic disorders, which share numerous anatomic, histological and pathophysiologic features. Therefore, a differential diagnosis of RDD should be excluded from xantho-granulomatous disease. B-cell lymphoma and benign lymphoid hyperplasia [1]. Other disorders that may superficially resemble RDD include Burkitt's lymphoma, rhabdomyosarcoma, granulocytic sarcoma, neuroblastoma and Langerhans cell histiocytosis.

The differential diagnosis of extra nodal SHML may be a challenge, and is based on the clinical and histological examination.

The differential diagnosis of Rosai-Dorfman is made following a biopsy of the affected tissue. A small piece of the tissue is obtained so that it can be viewed under a microscope by a pathologist, if the cells in the tissue have certain specific characteristics, the diagnosis of RDD can be made. This procedure can be performed of the lymph nodes, skin, bone, liver, lung, or bone marrow. RDD cells stain with S100 protein and CD68 [3,10].

In histological examination the histiocytes in RDD, Langerhans cell histiocytosis, and other histiocytosis express the S-100 protein, (a neural tissue-specific protein) however; the pathophysiology of this S-100 expression remains obscure. Although positive staining for S-100 strongly suggests RDD, it is not absolutely required to make the diagnosis in the presence of typical histology of RDD [7]. The disease is classically described to have an indolent, self-limiting course however; this is not always the case.

Other differential diagnosis was made with lymphoreticular malignancies such as lymphomas, Hodgkin's disease, malignant histiocytosis and monocytic leukaemia, all of which have similar histopathological features. Atypia in cytology and the aggressive clinical course establish the diagnosis in most cases. Other histiocytosis, such as rhinoscleromas, Wegener's granulomatosis, may also be included in the differential diagnosis [7].

Serology for HIV, toxoplasmosis and syphilis was done since these conditions are not rare in our context. Extra nodal manifestations in the head and neck are significantly more common in SHML patients with immunological abnormalities [4,5].

The therapy for RDD is yet to be determined because of the rarity of the disease and its propensity for spontaneous remission. Excision, radiation therapy, chemotherapy and systemic steroids have all been tried by different authors [10]. The first line of interventional therapy to be considered is gross total excision along with adjuvant steroids or more aggressive chemotherapy or radiotherapy when necessary [8].

In our patient the disease was stable at six weeks however a close follow-up is needed to watch for any increase in size of the lesion or systemic spread.

Treatment depends upon the patient individually and is planned after thorough testing to determine the extent of disease. Ideal treatment, however, has not been established, and there is no ongoing clinical trial [11].

It is believed that 70%–80% of patients recovered from symptoms without treatment, although they may have alternating episodes of exacerbation and remission of symptoms for a long period of time, some patients with severe or persistent disease or cases with risk of organ dysfunction may require treatment with surgery, steroids, and/or chemotherapy. radiation therapy is rare to be used.

Chemotherapy may include vinblastine, 6-MP, and methotrexate. Thalidomide or Gleevec. The ultimate goal of an overall treatment plan is to use as little treatment as possible to keep the disease under control and preserve quality of life [12].

This disease not usually threats life or induces organ dysfunction. [13] 5%–10% of patients show progress of the disease that may damage tissue. However, for most patients, the disease is self-limited, and the outcome is good [13,14].

3. Conclusion

Extra nodal manifestations of Rosai-Dorfman disease are rare and it ranges from confined to a highly extensive form. The ideal treatment has not yet been established, with surgical excision showing a poor outcome. as the head is the preferred site of the extra nodal form of the disease, then this disease should be considered during differential diagnosis.

The disease considered to be a diagnostic challenge, especially in extra nodal sites. With soft tissue involvement, the characteristic features of large histiocytic cells with emperipolesis may be overshadowed by a fibro inflammatory component. In the presented case it is easy to confuse this lesion with reactive nodules and benign neoplasms since painless enlargement of the cervical lymph nodes have followed the soft tissues lesions, because cutaneous sinus histiocytosis may be unfamiliar to general pathologists, cases limited to the skin may be under recognized. In the absence of lymphadenopathy, a high index of diagnostic awareness is required to recognize the disease, Histological and immunological confirmation is essential for definitive diagnosis.

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