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Extranodal Soft Tissue Rosai Dorfman Disease with Floret Cells Involving the Inguinal Region, Masquerading as Inflammatory Liposarcoma. Report of a Case Featuring Increased IgG4 Positive Plasma Cells

Aseel Qassem Al-Omari[1], Zain Qudah[2], Majdi Barakat[2], Samir Sami Amr[1*]

[1]Department of Pathology and Laboratory Medicine, Istishari Hospital, Amman, Jordan
[2]Department of Oncology, Istishari Hospital, Amman, Jordan
Email: *samir.amr48@gmail.com, aseel.q.omari@gmail.com, Zainqudah96@gmail.com, majdiabd@msn.com

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Abstract

Rosai Dorfman disease (RDD) is a rare histiocytic proliferative disorder of unknown pathogenesis that had been originally described in lymph nodes, but was found to occur in different visceral organs and anatomic sites as well. We report herein a patient who presented with a mass in the inguinal region which was initially diagnosed on core biopsy as inflammatory pseudotumor, then as inflammatory liposarcoma on excision of the mass. The patient was referred to our hospital for further management. On review of the histopathological material, we made the diagnosis of extranodal soft tissue Rosai Dorfman disease (ENST-RDD) by the identification of the characteristic S100-positive histiocytes demonstrating emperipolesis. We noted the presence of floret cells, which probably prompted the earlier diagnosis of inflammatory liposarcoma. The presence of these cells had not been documented in earlier reports on ENST-RDD. In addition, there was heavy IgG4 positive plasma cell infiltration associated with storiform fibrosis in the same lesion, probably pointing to a link between ENST-RDD and IgG4-related disorders.

Keywords

Extranodal Soft Tissue Rosai-Dorfman Disease (ENST-RDD), Inguinal Mass, Inflammatory Pseudotumor, Inflammatory Liposarcoma, Floret Cells, IgG4-Related Disorders

1. Introduction

Rosai-Dorfman disease (RDD) is an uncommon distinct entity described initially by Rosai and Dorfman in 1969, with a prevalence of only 1 in 200,000 in the United States. They reported pediatric and young adult patients who presented with painless bilateral cervical lymphadenopathy, characterized by a proliferation of large histiocytes with distinctive intact intracytoplasmic leukocytes (emperipolesis) and a variably mixed inflammatory infiltrate [1] [2].

Although RDD usually involves lymph nodes, the disease can have extranodal manifestations in 43% of all cases [3]-[5]. In rare cases, only extranodal involvement is seen, including central nervous system, meninges, eyes, gastrointestinal tract, head and neck, heart, kidney, thyroid, liver, pancreas, skin, breast, lung, mediastinum, mesentery, retroperitoneum, bone, and deep soft tissue [3] [6]-[8].

RDD of soft tissue most commonly involves the subcutaneous adipose tissue and presents as a slowly growing, painless, well demarcated mass; sometimes with ill-defined margins, and reportedly without lymph node involvement [6] [7].

RDD can be diagnostically difficult and often misleading in extranodal sites, especially when it presents without associated lymphadenopathy, and may be easily confused with reactive nodules or benign and malignant neoplasms, including soft tissue tumors and lymphomas [6].

Our case illustrates the difficulty in diagnosing ENST-RDD, since key diagnostic features of the disease were markedly overshadowed by the admixture of reactive inflammatory cells and scattered floret cells in the lesion. Accordingly, it was not considered initially in the differential diagnosis, and consequently was misinterpreted as inflammatory pseudotumor on core biopsy, and later as inflammatory liposarcoma on excision of the mass.

In addition, numerous IgG positive plasma cells were observed in our case associated with storiform fibrosis, 30% of them expressing IgG4, as highlighted by immunohistochemical stain. An undetermined relationship between RDD and IgG4-related disease had been recently postulated in few limited studies, while other studies suggested that RDD does not belong in the spectrum of IgG4-related diseases [9].

2. Case Presentation

A 32-year-old male; diabetic and smoker, presented with a one-year history of slowly growing painless subcutaneous mass arising in the lower left abdominal wall and extending to the left inguinal region.

Laboratory results revealed normal blood film, and normal complete blood count. Fasting blood glucose was elevated at 272 mg/dl (normal range: 70 - 150). Liver function tests were normal. Creatinine serum level was 0.79 mg/dl (normal range: 0.70 - 1.20), Lactate dehydrogenase was elevated at 479 IU/L (normal range: 105 - 333), and calcium serum level was slightly elevated at 11.1 mg/dl (normal range: 8.90 - 10.70). In addition, INR was 0.9 (normal value: 1.1 or below), PT was 12.3 seconds (normal range: 11 - 13.5), PTT was 26.3 seconds (normal value: 25 -

35), and ESR was 12 mm/hour (normal range: 0 - 22).

A computerized tomography (CT) scan of the thorax, abdomen and pelvis showed an ill-defined, irregular mass measuring 9.5 × 5.5 cm within the subcutaneous fat of the left side of lower abdominal wall. The mass had a lobulated contour with stranding of the surrounding fat, with other smaller subcutaneous nodules noticed, free of calcifications or cystic changes. The mass was infiltrating the deep fascia, but the abdominal wall muscles were preserved. The CT scan report suggested multiple differential diagnoses, including desmoid tumor, soft tissue sarcoma, chronic granulomas and chronic fat necrosis. Tru-cut biopsy was advised.

The initial needle core biopsy done at an outside hospital demonstrated markedly sclerotic tissue with scattered lymphoid aggregates composed of small mature lymphocytes, many plasma cells and few foamy macrophages in the background. Ziehl Neelsen stain for acid fast bacilli was negative. Grocott-Gomori methenamine silver stain for fungus was negative. These findings were interpreted as "Consistent with inflammatory pseudotumor".

The patient then underwent wide local excision of the left inguinal mass at another hospital. The mass measured 17 × 11 × 6.5 cm and had irregular grey yellow cut surface and firm consistency. Pathological examination revealed a mixture of mature adipocytes, spindle cells with collagenized stroma, plump pleomorphic epithelioid cells, multinucleated floret-like giant cells and scattered vacuolated lipoblast-like cells. A significant inflammatory component is seen composed of plasma cells, lymphocytes and reactive lymphoid follicles. Few satellite nodules were seen with margin of resection involved. These findings were interpreted as "Inflammatory liposarcoma".

The tumor recurred at the same site six months later, and resection of the recurrent tumor with overlying skin was done.

The patient was referred to our hospital. Our review of the slide of the Tru-Cut biopsy revealed dense fibrous tissue harboring scattered lymphoid aggregates, clusters of plasma cell and foamy macrophages, and spindly pale staining cells, with thick collagen fibers. No granulomas are seen. Few entrapped mature fat cells are seen.

Review of the slides of the first resection of the left inguinal mass revealed areas of dense fibrosis with entrapped clusters of fat cells (**Figure 1(A)** and **Figure 1(B)**). Infiltration by many histiocytes with several scattered floret multinucleated cells and Touton type cells was also present (**Figure 2(A)** and **Figure 2(B)**). No granulomas were seen. In addition, there were several clusters of large histiocytic cells with abundant eosinophilic or granular cytoplasm with large pale rounded nuclei, some of them were phagocytosing lymphoid cells or plasma cells; a finding known as emperipolesis (**Figure 3(A)** and **Figure 3(B)**).

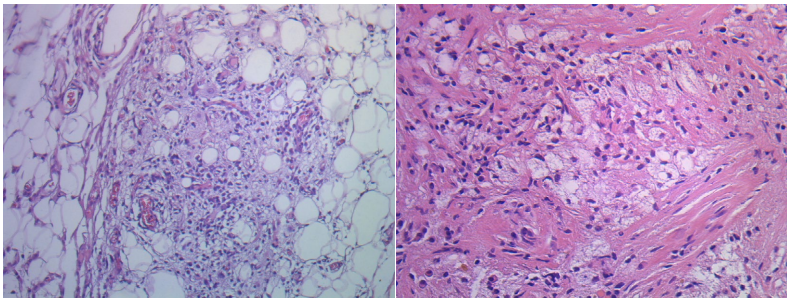
We performed S100-protien immunostain and it showed strong positive staining in these histiocytes, including those with emperipolesis (**Figure 3(C)** and **Figure 3(D)**).

Review of the slides of the second resection of the left inguinal mass recurrence showed similar histological findings to those observed in the previous resection

specimen. There were lobules of normal fat surrounding the inflammatory process. One section shows skin with underlying thick dermis, possibly due to scar formation related to the previous surgery.

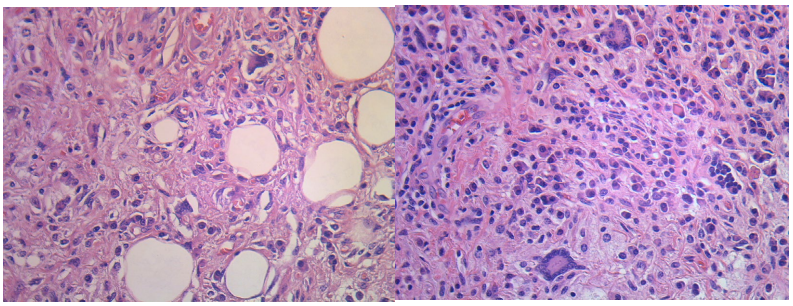
In addition, there was also heavy plasma cell rich-lymphoplasmacytic infiltration (**Figure 4(A)**) associated with dense fibrosis, which is focally storiform in pattern (**Figure 4(B)**). Numerous IgG4 positive plasma cells averaging 30% of the total IgG positive plasma cells were highlighted by immunohistochemistry (**Figure 4(C)** and **Figure 4(D)**).

The patient had no recurrent masses during a 1-year follow-up time period and no further treatment was required.



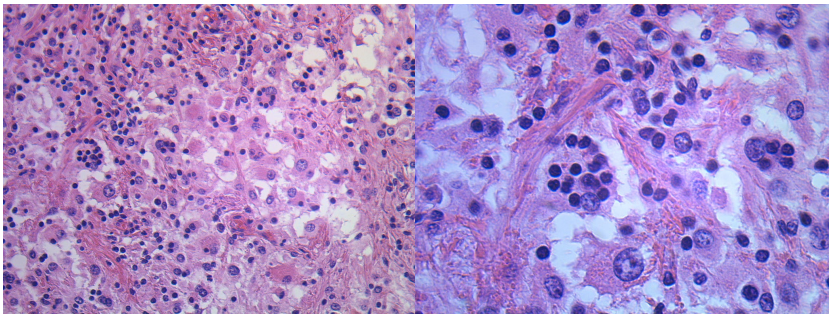
(A) (B)

Figure 1. (A): Lobules of fatty tissue infiltrated by lymphoplasmacytic cells (H&E × 20); (B): Areas of fibrosis with entrapped fat cells, histiocytes and plasma cells (H&E × 40).

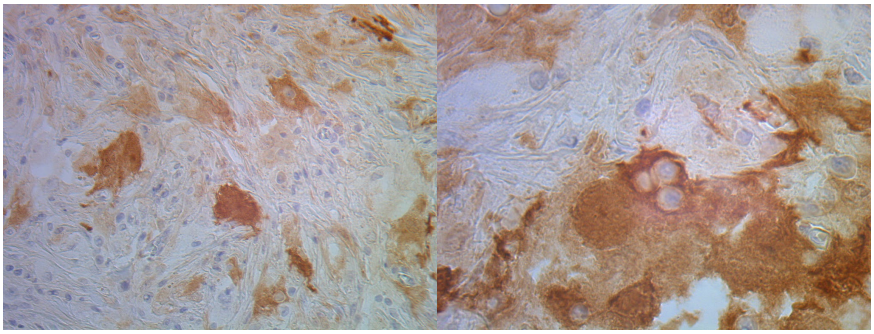


(A) (B)

Figure 2. (A): Infiltration of fibrofatty tissue by occasional floret cells (H&E × 40); (B): Several scattered Touton multinucleated giant cells with mononuclear inflammatory cell infiltrate (H&E × 40).

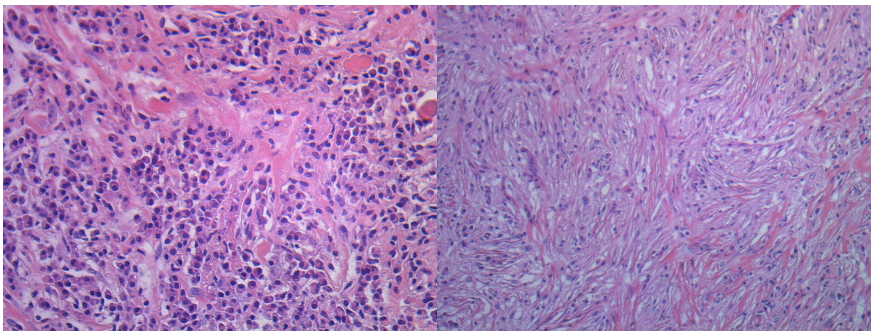


(A) (B)

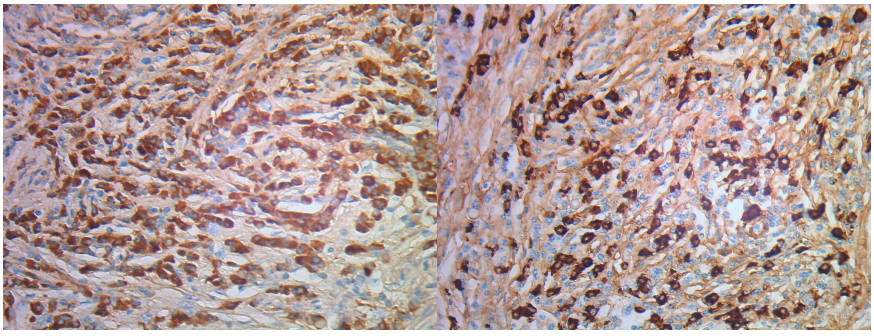


(C) (D)

Figure 3. (A): Large histiocytic cells with abundant eosinophilic granular cytoplasm and large pale round nuclei, some of them phagocytosing lymphoid cells (emperipolesis) (H&E × 40); (B): Large histiocyte with emperipolesis, phagocytosing up to 11 lymphocytes (H&E × 100); (C): The histiocytes in Rosai Dorfman disease are strongly positive for S100-protein immunostain (×40); (D): Higher magnification for S100-protein positive histiocytes. Note phagocytosed lymphocytes (×100).



(A) (B)



(C) (D)

Figure 4. (A): Heavy plasma cell rich-lymphoplasmacytic infiltration (H&E × 40); (B): Dense fibrosis with focal storiform pattern (H&E × 20); (C): Numerous IgG positive plasma cells were highlighted by immunohistochemical stain for IgG (×40); (D): Many IgG4 positive plasma cells averaging 30% of the total IgG positive plasma cells were highlighted by immunohistochemical stain for IgG4 (×40).

3. Discussion

Rosai-Dorfman disease (RDD) is an uncommon idiopathic disorder characterized by a proliferation of large histiocytes with distinctive intact intracytoplasmic

leukocytes (emperipolesis) and a variably mixed inflammatory infiltrate.

The lesion was not recognized to be a distinct entity until 1969, when it was described in lymph nodes of pediatric and young adult patients who presented with painless bilateral massively enlarged cervical lymph nodes by Rosai and Dorfman [1]-[3].

They described the characteristic presence of extremely large histiocytic cells, with abundant, clear, granular or finely vacuolated cytoplasm and large, round, vesicular nuclei containing distinct nucleoli [3]. These large cells showed a peculiar finding, namely the presence of many lymphocytes within cytoplasmic vacuoles, most probably representing the ability of these lymphoid cells to enter and leave the cytoplasm of these histiocytic cells without undergoing degenerative changes, a phenomenon known as emperipolesis [3]. They further emphasized the essentially benign nature of the disease and the capacity of this disorder to clinically simulate a malignant process [2].

Later, immunohistochemical studies were able to support these large cells as having the features of histiocytes and activated macrophages. They differed from reactive sinus histiocytes by virtue of their strong staining for S100 protein. In addition, they differed from Langerhans cell histiocytes in that they failed to express CD1a antigen [4] [5].

ENST-RDD tends to arise in older age than classic nodal disease, with a mean age of 42.5 to 46 years, and demonstrates female predilection [6] [7].

RDD may be diagnostically difficult and often misleading in extranodal sites, especially when it presents without associated lymphadenopathy. With soft tissue involvement, the characteristic diagnostic features of large histiocytic cells with emperipolesis may be overshadowed by a fibro-inflammatory component. Rare cases of ENST-RDD present with unusual features such as pseudovascular spaces and asteroid bodies in histiocytes [10]. In these cases, it is easy to confuse this lesion with reactive nodules or benign and malignant neoplasms. The usual misdiagnoses are soft tissue tumors (benign and malignant), lymphomas (especially Hodgkin disease) [6], and inflammatory pseudotumors [11] [12].

In another study from the Armed Forces Institute of Pathology (AFIP), of 29 lesions in 18 patients of ENST-RDD, the initial diagnosis of the referring pathologist was RDD, inflammatory pseudotumor, and inflammatory malignant fibrous histiocytoma [12]. In that series, five patients had 2 or more multifocal lesions. There were 4 males and 14 females. Mean and median age at diagnosis was 42.5 years (range, 8 - 81 years). Anatomic locations included trunk and proximal extremity (19), followed by distal extremity (5), abdominal wall (2), intra-abdominal (1), face (1), and unknown subcutis site (1). Sizes ranged from 0.5 to 13.7 cm (median, 2.4 cm) [12].

Immunohistochemical stain for S100 protein is helpful in confirming the diagnosis of RDD, particularly in cases with inconspicuous emperipolesis. It should also be recognized that emperipolesis in isolation is non-specific, occurring in other disorders, including leukemia, lymphomas and myeloproliferative disorders [13].

ENST-RDD is a difficult diagnosis and may be missed if the disease is not considered in the differential diagnosis of soft tissue masses. The imaging characteristics of RDD of soft tissue are not specific and may be suggestive of a sarcoma [4] [14].

Thus, distinguishing ENST-RDD from its mimics is essential since RDD generally pursues a benign course and is usually self-limiting or responds well to therapy; nevertheless, it occasionally behaves aggressively when associated with immunologic abnormalities and rare fatal cases with multiorgan involvement have been reported [15].

There had been reports of recurrence of ENST-RDD after initial excision, as in the current case. Guedes *et al.* reported a 27-year-old woman from Brazil who developed four recurrences of ENST-RDD in different sites of the posterior right leg within the span of few months [16]. Hindermann and Katenkamp reported five cases from Germany, one of them arising in the soft tissues of the left knee area with one recurrence taking place four years after initial debulking [17]. Betini *et al.* reported a case of ENST-RDD that presented as a slow-growing mass located the lower back of an 18-year-old female of one year duration. On physical and radiological examination, a firm, non-tender large superficial soft tissue mass that was approximately 25 centimeters long and 12 centimeters wide overlying the lumbar spine was noted. The mass was surgically excised. Nine months following her operation, an MRI of the lumbar spine demonstrated a large T1 hypointense, T2 heterogeneously hyperintense, and enhancing lesion with irregular borders and central area of scarring, most likely representing a recurrence of the lesion [18]. Recurrent ENST-RDD had also been reported in 7 out of 17 cases in an earlier series from AFIP [6]. An additional case with locally recurrent lesion in soft tissues was reported by Levine and Landry following multiple resections [19].

Multiple lesions of RDD involving soft tissue and bone mimicking sarcoma were reported in a patient who had left thigh lesion with associated distal right and left femur lesions, left proximal tibia and a pulmonary nodule [20]. In a series of 10 cases of extranodal RDD, 5 lesions were located in subcutaneous or soft tissues sites. One patient, a 28-year-old female, had multifocal metachronous lesions; one arose in the colon as a polyp, and the other was found in the soft tissue of the thigh [21]. Young et al. reported an additional case of RDD presenting as multiple soft tissue masses [22].

Giant cell formation was rarely reported in ENST-RDD, which was described as either multinucleated RDD histiocytes or foreign-body-type giant cells associated with poorly formed granulomas [3].

Yet, floret-like multinucleated giant cells (FMGC) were not previously reported as an unusual finding in ENST-RDD. The presence of FMGC was described in a growing list of soft tissue tumors, the most significant of which is pleomorphic lipoma, a mimic of liposarcoma, that was described initially in 1981 by Shmookler and Enzinger in a series of 48 cases encountered at the Armed Forces Institute of Pathology (AFIP). 31 of these 48 cases (65%) were submitted to the AFIP with a definitive or presumptive diagnosis of liposarcoma due to the presence of the

FMGCs as in the current case. The authors stated that pseudosarcomatous or sarcoma-like lesions of soft tissue, such as pleomorphic lipoma, presented more of a diagnostic dilemma to the reviewing pathologist [23]. Azzopardi et al. from UK reported in 1983 additional 9 cases of pleomorphic lipoma, most were initially misdiagnosed as liposarcoma [24].

Other lesions that showed FMGCs include adult gynecomastia [25], sporadic and NF1-associated neurofibroma [26], collagenoma [27], giant cell fibroblastoma [28] and giant-cell-rich variant of solitary fibrous tumour also known as giant cell angiofibroma [29].

Although the etiology of RDD is still unknown, but a subset of RDD exhibited features of IgG4-related disease [30]. Zhang *et al.* studied the distribution of IgG4 positive plasma cells and regulatory T (TREG) cells, a major regulator of IgG4 production, in twenty-six specimens of RDD, including 15 nodal, and 11 extranodal; 3 cases of which involved soft tissues [9]. Overall, 84.6% (22/26) of the specimens showed various degrees of sclerosis. Nineteen cases (73.1%) exhibited more than 10 IgG positive cells/0.060 mm^2, and 8 cases (30.8%) showed more than 40% of IgG positive cells being IgG4. The authors concluded that a subset of RDD shows features of IgG4-related disease, which indicates an overlap between certain aspects of the two diseases. [9]. The current case fulfills such features with numerous IgG4 positive plasma cells, averaging 30% of the total IgG positive plasma cells as highlighted by immunohistochemical stain.

IgG4-related diseases are recently described entities that can affect many organs, including the pancreas, salivary glands, orbit, hepatobiliary tract, retroperitoneum, thyroid, mediastinum, pleura, gastrointestinal tract, and lymph nodes. They are usually suspected as neoplastic processes because they often present as mass lesions. However, clinically, the IgG4 diseases are characterized by a good outcome and with response to steroid therapy. Histologically, there is dense lymphoplasmacytic infiltration, numerous IgG4 positive cells, fibrosis which is at least focally storiform, as in the current case, and obliterative phlebitis. An international consensus statement published in 2012 proposed that at least two of these characteristics must be seen for a histological diagnosis in the appropriate clinical setting [31].

To emphasize further the relationship between RDD and IgG4-related disease, Menon *et al.* reported in 2014 additional 28 cases of RDD associated with the presence of plasma cells expressing IgG4. 15 of these cases were involving lymph nodes, while the remaining 13 involved extranodal sites, including two cases in soft tissues (gluteal and arm masses), one retroperitoneal, one mesenteric, and one peri-hilar renal mass. IgG4 percentage of the total IgG positive plasma cells ranged from 5% up to 97%, with 12 cases having a ratio above 40% [32]. There are a few reports on RDD with increased IgG4-positive plasma cells; one involving the meninges [33], the orbit [34], and the colon [35].

On the other hand, Liu et al. studied the number of IgG4 positive plasma cells and the IgG4/IgG ratio in 32 biopsy specimens (13 nodal, 19 extranodal) from 29 patients with RDD and compared the findings with those in IgG4-related disease

of the pancreas and reactive lymph nodes. They found that RDD cases had much lower numbers of IgG4 positive plasma cells and lower IgG4/IgG ratios compared with IgG4-related disease, but were similar to reactive lymph nodes. They concluded that their study suggests that RDD does not belong in the spectrum of IgG4-related diseases [36].

Wang et al. presented another limited prospective study from China, which included 7 cases of RDD mimicking IgG4-related disease, mainly in the CNS [37]. IgG4 positive plasma cells were quantified in all RDD mimickers of IgG4-related disease patients, and the proportion of IgG4/IgG in tissues was 10 - 40% in 4 patients and more than 40% in 2 patients. However, none of those patients displayed obliterative phlebitis or storiform fibrosis. Most of those patients were treated with glucocorticoids combined with immunosuppressants, and a good prognosis was obtained following treatment.

Accordingly, the exact relationship and clinical significance of the relationship of RDD to IgG4-related diseases remains undetermined, and further large cohort studies to clarify it should be pursued.

4. Conclusions

Our case illustrates the difficulty in diagnosing extranodal soft tissue Rosai Dorfman disease (ENST-RDD). Clinically, the disease may mimic a number of different entities. As outlined above, the histopathological diagnosis was missed twice upon review of the case at two different histopathology labs. The presence of infiltrative large histiocytes with emperipolesis, that showed strong positive staining with S100-protien immunostain, were the key diagnostic features. However, these key findings may be markedly overshadowed by the admixture of reactive inflammatory cells in the lesions.

ENST-RDD is not uncommonly mistaken for inflammatory pseudotumor, inflammatory malignant fibrous histiocytoma, and inflammatory liposarcoma as in the current case; as well as lymphomas, especially Hodgkin disease. This case depicts the presence of many floret cells in the same lesion, a feature not reported previously with RDD, which led to the misdiagnosis of inflammatory liposarcoma.

In addition, heavy IgG4 positive plasma cell infiltration associated with storiform fibrosis in the same lesion points to a possible relation between ENST-RDD and IgG4-related disorders.

Future large cohort research is encouraged to define the undetermined relationship between RDD, floret cells and IgG4-related diseases.

Data Availability

The case report data used to support the findings of this study are included within the article.

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The authors did not receive any funding for this work.

Consent

Written informed consent was obtained from the patient for publication of this case report.

Author Contribution

All the authors read and approved the final version of the manuscript.

Aseel Al-Omari: conception, acquisition of data, literature research and preparing the manuscript.

Zaina Qudah: acquisition of clinical data.

Majdi Barakat: acquisition of clinical data, revising of the manuscript.

Samir Amr: conception, acquisition of data, preparing and revising the manuscript critically.

Disclosure

This work was presented as a poster presentation at the 34th International congress of the International Academy of Pathology (IAP), held in Sydney, Australia on 11 - 15 October 2022.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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