Case Report
Extranodal Rosai-Dorfman disease involving appendix and mesenteric nodes with a protracted course: report of a rare case lacking relationship to IgG4-related disease and review of the literature

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Abstract: Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare disease of unknown etiology that typically presents as nodal disease in young children. However, it also can present in various extranodal sites and can be difficult to recognize if not considered in the differential diagnosis. Here, we report a case of appendix involvement by extranodal RDD, which occurred in a 69-year-old woman with a long duration of 12 years for intermittent right lower quadrant pain. The patient underwent a right hemicolectomy for a clinical diagnosis of appendiceal cancer. A mixed inflammatory infiltration of mature lymphocytes, plasma cells and histiocytes exhibiting emperipolesis were indentified. Other areas had storiform fibrosis and sclerosis admixed with numerous plasma cells. These histologic features combination with immunoreactivity for CD68 and S100 protein were indicative of a diagnosis of extranodal RDD. We discuss the clinical, pathologic findings as well as differential diagnoses and consideration of a possible relationship of this entity to IgG4-related lesion.

Keywords: Rosai-Dorfman disease, extranodal disease, gastrointestinal tract, appendix, IgG4-related disease, differential diagnosis

Introduction

Rosai-Dorfman disease (RDD), also referred to as sinus histiocytosis with massive lymphadenopathy, is a rare, idiopathic, non-Langerhans-cell, histiocytic proliferative disorder of unknown etiology [1-3]. RDD is histologically characterized by a nonmalignant proliferation of distinctive histiocytic/phagocytic cells within lymph node sinuses and lymphatics in extranodal sites. The proliferative histiocytes have abundant pale to slightly eosinophilic cytoplasm often containing lymphocytes and plasma cells within, a phenomenon variously known as emperipolesis or lymphocytophagocytosis. Although classical RDD usually presents as massive, painless, bilateral neck lymph node enlargement, approximate 40% of RDD cases

documented to date present in extranodal organs or tissues, in some without associated lymphadenopathy which may or may not develop later in the disease course [1-3]. The most often affected extranodal sites include skin and soft tissues, upper respiratory tract and bone followed by genitourinary tract, lower respiratory tract and the oral cavity. Gastrointestinal tract involvement in RDD is exceptionally rare, which occurs in less than 1% of all extranodal cases, according to the data in the RDD Registry [1, 2]. RDD presenting in extranodal sites can be difficult to diagnose as these often appear as a nonspecific fibro-inflammatory lesion with stromal sclerosis of variable degree and emperipolesis can be quite subtle and difficult to recognize. Immunoglobulin (Ig) G4-related disease is a recently characterized
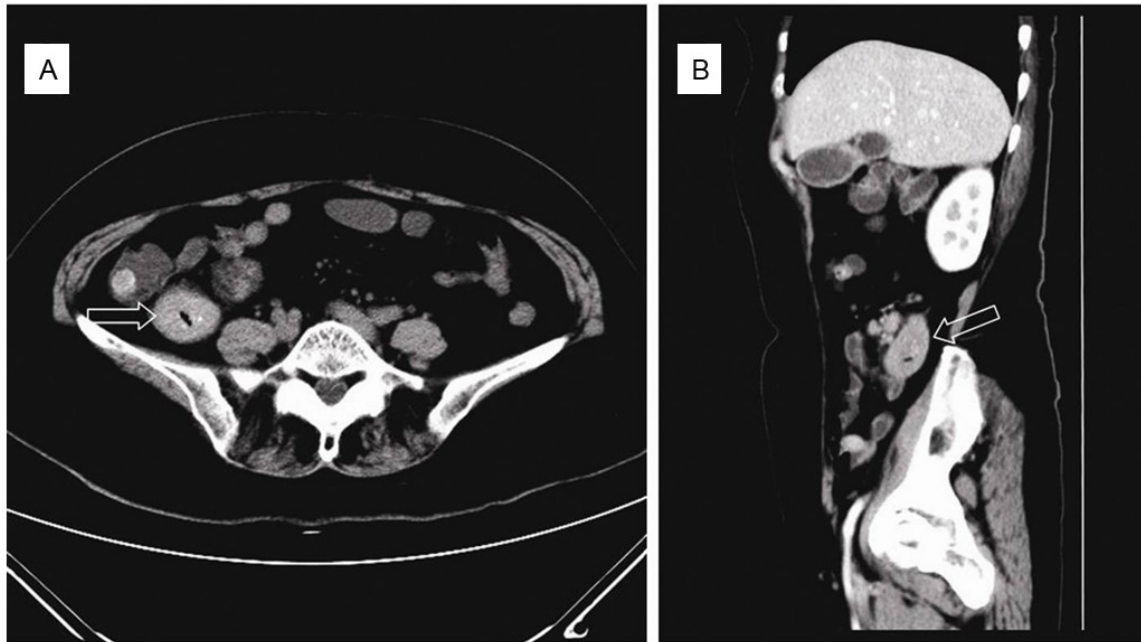
Figure 1. Abdominal computed tomography scan demonstrated an ileocecal mass intramurally involved the appendix and caused marked thickening of the wall of the appendix (hollowed arrow), with multiple mesenteric nodes enlargement. (A: cross plane; B: sagittal plane).

entity that occurs in a variety of anatomic locations and organ systems. It is histologically characterized by storiform fibrosis with an intense IgG4-positive plasma cells infiltration, with or without obliterating phlebitis [4-6]. Recent emerging evidence suggests that a subset of RDD exhibits features of IgG4-related disease and indicates an overlap between certain aspects of these two diseases [7, 8]. This report describes a unique case of extranodal RDD involving appendix and mesenteric nodes in an elder woman with 12-year duration. We also investigate a possible relationship to the IgG4-related disease and review the pertinent literature.

Case presentation

A 67-year-old Chinese woman presented with intermittent right lower quadrant pain for approximate 12 years, the abdominal pain was dull that slightly aggregated when doing physical work and released after rest. The patient's medical history was unremarkable and she denied fever, nausea, vomiting, hematochezia, appetite changes, or weight loss. Her physical exam including abdominal tenderness and rebound was negative and no lymphadenopathy or abnormal mass was noted. Laboratory examination demonstrated a normal complete

blood count, metabolic profile, and tumor markers including cancer antigen 19–9, cancer antigen 125, and alpha-fetoprotein. Colonoscopy was performed and demonstrated relatively normal-appearing mucosa without evidence of massive or ulcerative lesion present. Subsequent imaging evaluation including chest/abdomen computed tomography scan revealed an ileocecal mass with intramural involvement of the appendical wall, as well as multiple mesenteric lymphadenectasis which are suspicious for involvement by the ileocecal tumor (**Figure 1A**, **1B**). On the basis of a presumptive clinical diagnosis of appendical cancer, the patient underwent a right hemicolectomy; operative impression included a firm, ill-defined, serosal-based appendical mass adhesive to the surrounding cecum with multiple mesenteric lymphadenopathies. The patient's recovery was uneventful and the symptom of abdominal pain was persistently relieved, and there was no evidence of local recurrence of the disease or development of lymphadenopathy elsewhere of the body 10 months after the surgery.

Methods

The resection specimen was fixed in 10% buffered formalin, and then was processed routine-
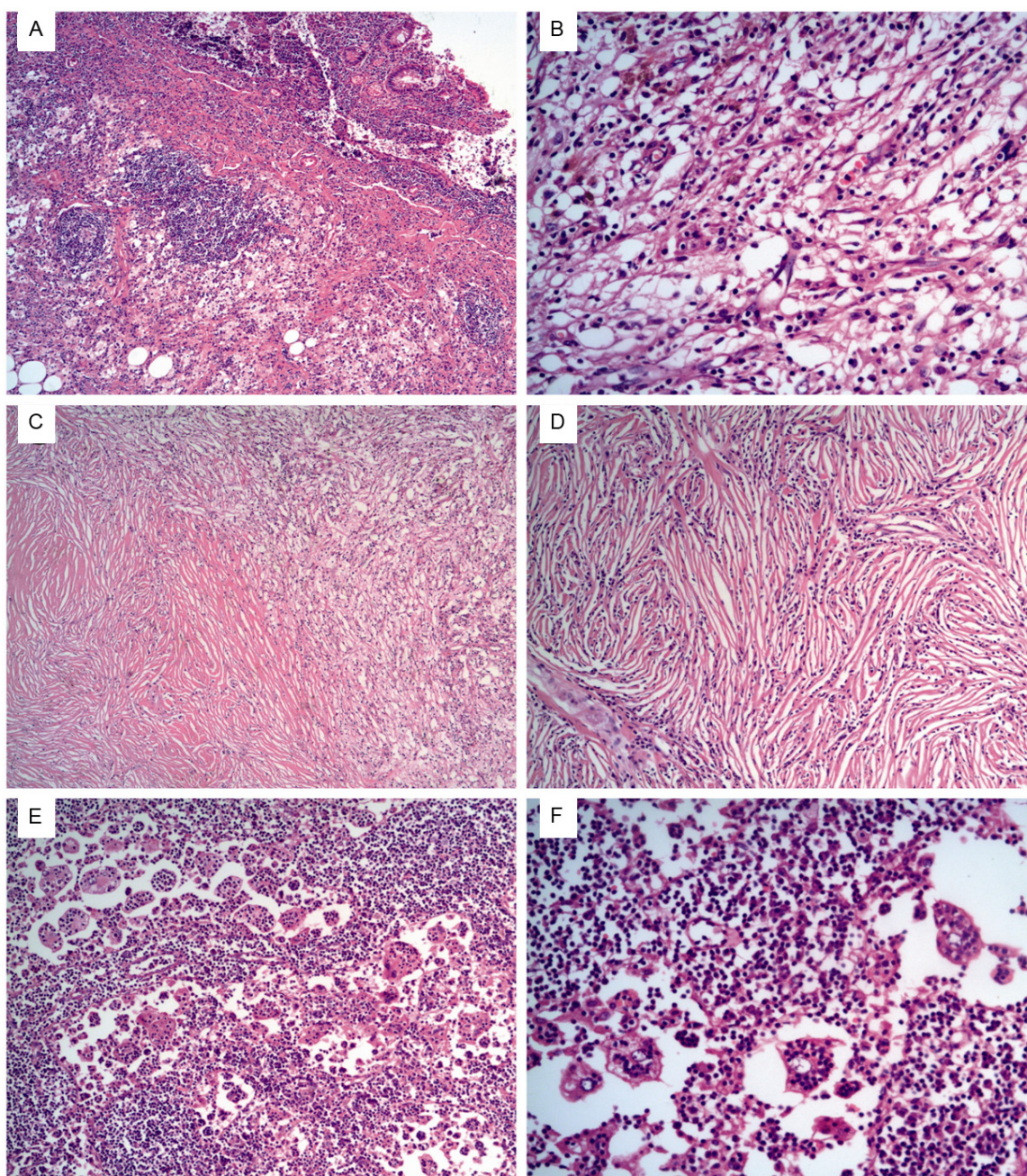
Figure 2. Microscopic features of the appendiceal mass. A: Low power view showed an intramural inflammatory infiltration comprising pale areas of histiocytes and dark areas of lymphocytes and plasma cells, which focally extended into the overlying mucosa. B: High power view showed the histiocytes with abundant pale to granular cytoplasm and small bland nuclei. Some of the lymphocytes and plasma cells appeared to be within the cytoplasm of the histiocytes that characterized the disease. C, D: Areas showed stroriform fibrosis associated with an intensive plasma cells infiltrate mimicking inflammatory myofibroblastic tumor or IgG4-related disease. E, F: Mesenteric nodes exhibited the characteristic features of nodal RDD with ectatic lymphatic sinus filled with numerous histiocytes showing extensive cytoplasmic emperipolesis.

ly. The sections were stained with hematoxylin and eosin. For immunohistochemistry, the following primary antibodies were used: CK AE1/AE3 (AE1/3; DAKO, Glostrump, Denmark), CD68 (KP1; DAKO), CD1a (polyclonal; DAKO), S100 protein (polyclonal; DAKO), CD34 (QBEnd/10; DAKO), CD21 (2G9; DAKO), CD35 (Ber-MAC-DRC; DAKO), c-kit (polyclonal; DAKO),
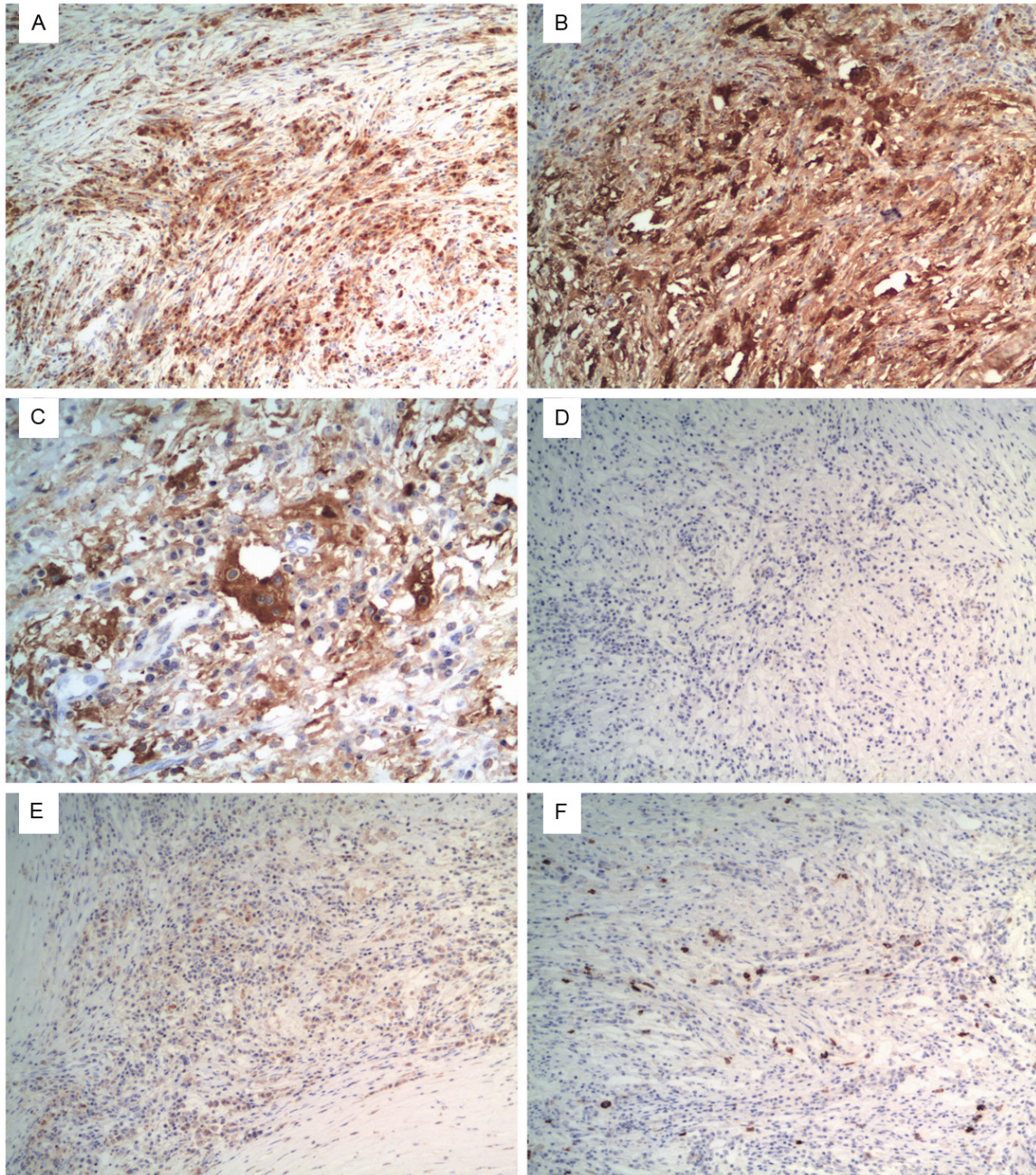
Figure 3. Immunohistochemical staining features of appendiceal RDD. The histiocytes showed strong and diffuse reactivity to antibodies against CD68 (A) and S100 protein (B, C) but not CD1a (D). Note that S100 protein staining highlighted the lymphocytophagocytosis of the histiocytes (C). There were only a few IgG-positive (E) and IgG4-positive (F) plasma cells present within the lesion.

smooth muscle actin (SMA; 1A4, DAKO), desmin (D33; DAKO), IgG (polyclonal; DAKO), IgG4 (HP6025, Invitrogen, Carlsbad, USA), and Ki67 (MIB-1, DAKO). Immunostaining was performed according to standard protocols using advin-biotin complex labeled with peroxidase. Appropriate positive and negative controls were run concurrently for all the markers tested.

Pathologic findings

The surgical specimen consisted of a 25.0 cm segment of ileum and cecum and an obviously enlarged vermiform appendix measuring 8.0×3.0×3.0 cm. On cut surface, an ill-circumscribed, grayish white to yellow, firm mass measuring 7.0×5.0×3.5 cm was observed, located

mainly at the serosa of the appendix and producing marked intramural thickening of the appendix and narrowing of the lumen. No hemorrhage or necrosis was seen. The surface mucosa of both the ileum and cecum were flat, intact, and normal-appearing without evidence of ulceration or massive occupation. The serosal surface of the appendix revealed multiple fibrous adhesions, and multiple, isolated lymph nodes measuring 1.2 cm to 2.5 cm were identified in the mesenteric fat.

Microscopic evaluation of the appendix revealed a serosa-based transmural infiltrate composed of abundant chronic inflammatory cells and large pale histiocytes focally extending into the overlying mucosa with focal ulceration (**Figure 2A**). The histiocytes demonstrated ample pale to slightly eosinophilic cytoplasm, or sometimes showed foamy or granular cytoplasm. A few numbers of histiocytes showed occasionally phagocytosis of intact lymphocytes and plasmocytes (**Figure 2B**). The nuclei of histiocytes were round to oval and vesicular to hyperchromatic, no evidence of elevated mitotic figures and apoptotic activities was present. Additionally, areas of dense stroiform fibrosis and sclerosis were identified in the appendiceal wall in association with an extensive plasma cells and lymphocytes infiltration and only a few eosinophils (**Figure 2C**, **2D**). The largest nodule within the mesenteric adipose tissue was an enlarged lymph node demonstrating partial architectural distortion, ectatic lymphatic sinuses filled with numerous histiocytes with prominent emperipolesis (**Figure 2E**, **2F**). A total of seventeen benign lymph nodes were identified, of which 15 lymph nodes exhibited at least focal RDD involvement, with numerous histiocytes engulfing lymphocytes and plasma cells in their cytoplasm. Immunohistochemical studies revealed strong reactivity in most of the histiocytes in both the appendic lesion and the lymph nodes using CD68 (**Figure 3A**) and S100 protein (**Figure 3B**, **3C**) antibodies; they were negative for CD1a (**Figure 3D**). The sclerosing stroma showed immunoreactivity to SMA but not c-kit, desmin, CD34, CD21, or CD35. Immunostaining for IgG (**Figure 3E**) and IgG4 (**Figure 3F**) demonstrated no increased numbers of either IgG-positive plasma cells (up to 109 plasma cells/high power field) or IgG4-positive plasma cells (up to 21 plasma cells/high power field).

Discussion

RDD is firstly recognized as a distinctive clinico-pathologic entity by Rosai and Dorfman in 1969 under the term sinus histiocytosis with massive lymphadenopathy. At the same time, they established a registry to collect and catalogue cases of RDD worldwide so this uncommon disorder may be better studied [1-3]. Clinically, RDD is typically a disease of childhood and early adulthood, painless lymphadenopathy is the most common presenting symptom and involes the cervical region in up to 90% of patients. Axillary, para-aortic, inguinal, and mediastinal lymph nodes are also frequently affected. Otherwise patients may present as systemic symptoms including fever, night sweat, malaise, and weight loss which may be related to enhanced production of monokines by the distinctive histiocytes. Although initially thought to be rare, extranodal involvement by RDD has been recognized with increasing frequency, with approximately 40% of the RDD patients having at least one extranodal site of disease, judging from the data in the RDD registry [1-3]. Practically every organ system can be affected by RDD, but the most common affected extranodal locations include skin and soft tissue, the upper respiratory tract, orbit and eye, bone, and salivary gland. Simultaneous involvement of multiple extranodal sites is not unusual. Involvement of the kidney, lower respiratory tract and liver are associated with a worse clinical outcome as is the number of extranodal sites involved [1-3].

Gastrointestinal tract seems to be the least commonly involved site by RDD and represents less than 1% of extranodal presentations [1-3]. In 2000, Lauwers et al [9] reported to date the largest series of eleven cases of RDD involving the digestive system that included five cases arising within the intestinal tract, and to our knowledge, including the current one, only 15 cases of gastrointestinal RDD have been reported in the English literature to date [9-18].

Based on the published cases, gastrointestinal RDD appears to affect mainly middle-aged female patients and have a predilection for ileocecal area and distal colon with a whelming majority of the cases being located beyond the pylorus [9-18]. The gastrointestinal tract involved by RDD may be solitary or segmental. Three cases, including the current one, involved

the appendix [9]. The reported other involved sites in the gastrointestinal tract include ileocecum (n=2) [11, 18], jejunum (n=1) [9], descending colon (n=1) [13], sigmoid colon (n=2) [10, 16], rectum (n=3) [9, 12, 14], colon not other specified (n=2) [9, 15], and the total gastrointestinal tract including oesophagus, stomach, duodenum, ileum, colon, and rectum (n=1) [17]. A majority of these patients had either multiple extranodal or nodal lesions simultaneously. Most patients were symptomatic and the documented presenting symptoms included fever, hematochezia and constipation, intestinal occlusion, abdominal pain, and abdominal mass with extensive growth [9-18]. A few patients may be asymptomatic and the diseases were incidentally discovered at colonoscopy, at autopsy, or in an appendectomy specimen [9]. Of the patients for whom follow-up data were available for more than 12 months, approximate 20% were ultimately succumbed to the complications from the disease [9, 12, 17], and the remaining patients were alive with continuing disease. This, and our case, which had a as long as 12-year duration before surgery, lend additional support to the impression that RDD involving extranodal sites can have a protracted, unfavorable course in contrast to cases limited to lymph nodes regions in which spontaneous regression often occurs within months [1-3].

Histologically, the hallmark of the RDD is the presence within the histiocytic cytoplasm of variable numbers of intact lymphocytes, a phenomenon referred to as lymphocytophagocytosis or emperipolesis, defined as lymphocytic penetration of and movement into the histiocyte [1-3]. These lymphocytes are often housed within vacuoles and escape degradation during their transit through the histiocyte. Associated with the histiocytic proliferation are numerous, mature plasma cells often aggregated around post-capillary venules. Collections of neutrophils, eosinophilic microabscesses and reactive germinal centers are occasionally seen but are not prominent findings in this disorder. Although RDD involving extranodal sites shows essentially similar morphologic features to its nodal counterpart, more fibrosis and fewer histiocytes with emperipolesis are encountered that can elicit a variety of differential diagnosis [1-3].

The histologic differential diagnosis of RDD involving gastrointestinal tract is centered on a variety of other histiocytic lesions including langerhans cell histiocytosis (LCH), Erdheim-Chester disease (ECD), xanthomatosis; and follicular dendritic cell sarcoma [19]. LCH can occur at any age and has been documented in practically every organ system including the digestive system [20]. In contrast to the large histiocytes with round nuclei and vesicular chromatin noticed in RDD, histiocytes in LCH have smaller, usually folded or groove nuclei, and are associated with eosinophilic microabscesses, and lack of the emperipolesis characteristic of RDD. Moreover, langerhans cells stain for S100 protein and CD1a, but only rare CD1a-positive cells are found in RDD [19, 20]. Erdheim-Chester disease with manifestation in the gastrointestinal tract seems to be extremely rare [21]. This disease is often associated with considerable amounts of fibrosis and histiocytes infiltration that can be confused with RDD. However, the histiocytes in ECD typically have small, hyperchromatic nuclei, and the cytoplasm appears foamy, and occasionally are admixed Touton-type giant cells [22]. The histiocytes express CD68 and factor XIIIa, whereas the expression of S100 is typically negative [22]. Xanthomatosis of the gastrointestinal tract is characterized by the accumulation of lipid-laden, foamy histiocytes in the lamina propria, submucosa, muscularis propria, and/or subserosa [23]. Typically, there is only mild to moderate inflammation without fibrosis. The histiocytes express CD68 but usually not S100 protein [23]. Follicular dendritic cell sarcoma is an uncommon histiocytic sarcoma and generally is a lesion of lymph node. However, extranodal involvement, among others in the gastrointestinal tract, can occur [24]. The neoplastic cells are often spindle shaped, with elongated nuclei, and arranged in a storiform or swirling pattern. One or more of the follicular dendritic markers CD21, CD35 or CD23 are usually positive, whereas CD68 and S100 protein are typically negative [24].

Several non-histiocytic lesions should also be considered in the differential diagnosis of gastrointestinal RDD. As previous has mentioned, although the histopathology of extranodal RDD simulates nodal disease, the histologic hallmarks of the disease are less obvious than those seen in the lymph nodes. Emperipolesis may be less conspicuous, the histiocytes being frequently spindled, arranged in a storiform pattern, and associated with fibrosis [1-3]. In

this scenario, inflammatory pseudotumor, now is known for inflammatory myofibroblastic tumor (IMT), may enter into the diagnosis of RDD [25, 26]. IMT is a heterogenous group of spindle myofibroblasts proliferations with admixed lymphocytes, histiocytes, and plasma cells that exhibit histologic overlapping with RDD. The omentum and mesentery are the most common extra-pulmonary sites for IMT but bowel wall can also be affected by tumor direct extention [27]. Some authors have suggested that these two lesions are part of a spectrum of inflammatory or reactive conditions and postulated that the morphological and immunophenotypic features of their lesions could be secondary to aberrant cytokine expression in an infammatory pseudotumor, resulting in transformation case of histiocytes to resemble those seen in RDD [28]. Nevertheless, the presence of morphologic similarities between RDD and inflammatory pseudotumor does not necessarily represent evidence that these entities should be grouped together [26, 29]. Gastrointestinal lesions of RDD are often multiple and almost always accompanied with lymphadenopathy, in contrast to IMT which is usually a solitary lesion [27]. Positivity for S100 protein confirms the diagnosis of RDD and facilitates the search for emperipolesis, as it allows the outline of the individual histiocytes with the negative image of the phagocytized cells in IMT. IMT is a myofibroblastic lesion with tumor cells often expressing SMA and less commonly desmin, and approximate 60% cases of IMT express anaplastic lymphoma kinase (ALK), a protein expressed by the chimeric gene resulted from the t (2; 5) translocation [30]. Although S100 stain has not been performed in many cases of IMT, in a few reported cases of S100-positive IMT, the histiocytes did not show the characteristic morphology of those seen in RDD [31].

Inflammatory fibroid polyp (IFP) is another rare, benign mesenchymal tumor that can arise throughout the gastrointestinal tract [32]. IFP is mainly centered within the submucosa layer and histologically is characteristic of presence of perivascular onion skinning distribution of spindle-shaped fibroblastic cells, set in a myxoid to collagenous strom and associated with prominent eosinophils infiltration. However, it may alternatively present with a short fascicular growth pattern, a sparse number of eosinophils, and prominent hyalinization that cause diagnostic confuse with RDD. In contrast to RDD, IFP contains less predominance of plasma cells and the spindle-shaped lesional cells in IFP often show diffuse immunoreactivity for CD34 but not S100 protein [32, 33].

The etiology of RDD remains unknown and is considered an idiopathic histiocytosis by most investigators, although some evidence exists to suggest that immune dysfunction and viral infections such as Epstein Barr virus and human herpes virus play a role in its pathogenesis [1-3]. IgG4-related disease is a recently described entity of uncertain pathogenesis characterized by the presence of abundant IgG4+ plasma cells in the lesion, with tissue sclerosis and elevated serum IgG4 concentration [4-6]. This disease can occur in a variety of anatomic locations all over the body including the gastrointestinal tract location [34-36]. Because abundant plasma cells infiltration and stromal sclerosis characteristic of IgG4-related disease are also histological features shared by many cases of RDD (as the current case has shown), and multiple organs system involvement can be seen in both lesions, several researchers have presumed a possible relationship between RDD and IgG4-related disease [7, 8]. In a most recent study by Zhang et al [8], about 30% RDD cases showed various degrees of sclerosis and increased number/ percentage of IgG4+ plasma cells with IgG4+ cells accounting for more than 40% of IgG+ plasma cells, and the IgG4+ plasma cells exhibited a wide spectrum of morphology and distribution in the lesions, these findings led the authors to indicate that a subset of RDD overlapped with IgG4-related disease, or it was during certain phases of the disease when RDD may demonstrated features of IgG4-related disease. Most recently, Wimmer et al [18] reported a case of cecal RDD which showed some histologic features of IgG4-related disease including areas of storiform fibrosis and numerous IgG4 positive plasma cells with elevated ratio of IgG4 to IgG. The patient's postoperative IgG and IgG4 serum levels were both within normal limits; however, preoperative serum examination for the production of IgG4 was performed in this patient. Although there are indications that a close relationship between RDD and IgG4-related disease may exist, the definitive link between these two disorders needs further research to clarity, particularly given that a significant subset of RDD

does not demonstrate histologic features that fulfill with the diagnostic criteria of IgG4-related disease, and RDD is frequently a self-limited disease, steroid is generally not used for the treatment of RDD [37]; in contrast, IgG4-related disease typically responds dramatically to corticosteroid treatment [4-6]. As to the current case, we considered the possibility that this disease may fall within the spectrum of IgG4-related disease. However, immunostained sections with IgG4 failed to reveal a significant increase in the number of IgG4 plasma cells or the ratio of IgG4 to IgG, although serum study of the production of IgG4 was not preoperatively performed.

In summary, we have presented a unique case of extranodal RDD presenting as an ileocecal mass and involving the appendix and multiple mesenteric nodes with a 12-year of duration in an elder Chinese woman. This diagnosis should be considered in the differential diagnosis of a variety of tumor or tumor-like lesions featuring of storiform fibrosis with admixed infiltration of mature plasma cells, lymphocytes, and pale pink histiocytes. We did not find evidence that this disease may be part of the spectrum of IgG4-related lesion.

Disclosure of conflict of interest

The authors have disclosed that they have no significant relationships with, or financial interest in, any commercial companies pertaining to this article.

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