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CORRESPONDENCE

A case of disseminated extranodal Rosai–Dorfman disease diagnosed by skin manifestations

Abstract

Rosai–Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis that was first described as sinus histiocytosis with massive lymphadenopathy by Rosai and Dorfman in 1969. While classical/nodal RDD is typical, extranodal involvement is seen in up to 43% of all cases, with 23% showing only extranodal lesions. Here, we present a case of disseminated extranodal RDD, where the initial symptoms were cutaneous manifestations.

Rosai–Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis (LCH) that was first described as sinus histiocytosis with massive lymphadenopathy by Rosai and Dorfman in 1969.[1] While classical/nodal RDD is typical, extranodal involvement is seen in up to 43% of all cases, with 23% showing only extranodal lesions.[2] Here, we present a case of disseminated extranodal RDD, where the initial symptoms were cutaneous manifestations.

A 46-year-old Japanese man was referred to our department with a 5-month history of skin nodules on his occiput and right thigh. Physical examination revealed a flattened dark-reddish tumor (23×18 mm) on the right side of occiput (Figure 1A) and a red-brown tumor (45×35 mm) surrounded by multiple red-brown papules on the right thigh (Figure 1B). Positron emission tomography/computed tomography displayed 18F-fluorodeoxyglucose uptake in the tumor of occiput, right thigh, ileum, and right humerus (Figure 1C). Neither lymphadenopathy nor intracranial tumors were observed. Histopathological analysis of a skin biopsy from his right thigh revealed infiltration of numerous inflammatory cells, including macrophages, neutrophils, plasma cells, and monocytes, along with emperipolesis, from the upper dermis to the subcutaneous tissue (Figure 1D–F). Immunohistochemistry revealed that the infiltrating macrophages were positive for CD68, S100-protein, Cyclin D1, OCT2, and phospho-ERK, but negative for CD1a and langerin (Figure 1G–M). Based on these clinicopathological results, a diagnosis of disseminated extranodal RDD was established. As the patient suffered from only mild fever and bone pain, a conservative approach of observation without treatment was pursued. His skin lesions spontaneously decreased in size by 4 months after presentation, and no systemic symptoms appeared 18 months after the initial diagnosis.

Recently, the classification of RDD was revised and it is now part of the R group that includes familial and sporadic RDD.[2] Sporadic RDD is often self-limiting and is associated with good outcomes because spontaneous remission has been reported in approximately 50% of all cases.[3] Extranodal locations were observed in various organs including skin, central nervous system, bone, and nasal cavity. Although the diagnosis of systemic RDD from skin biopsies is relatively rare compared to cutaneous RDD, systemic examination is required.[4,5] Several literatures reported the efficacy of systemic corticosteroid treatment in both nodal and extranodal RDD.[6,7] Since our patient did not show serious symptoms, neither systemic therapy nor surgical resection was performed.

Emperipolesis is defined as the presence of an intact cell within another cell and is a hallmark feature of RDD. However, it is not specific for the diagnosis of RDD because it is seen in other histiocyte diseases such as LCH, Erdheim-Chester disease, and hematolymphoid disorders.[8] Recent studies have described immunohistochemical characteristics of RDD based on the expression of CD68, CD163, Cyclin D1, S-100 protein, OCT2, and phospho-ERK,[9,10] and immunostaining of skin tumor in our case showed the typical pattern of RDD.

To summarize, we present a case of disseminated extranodal RDD that was diagnosed based on immunohistochemical analysis from skin tumor. While the disease could resolve spontaneously, *systemic examination and* long-term observation are required.

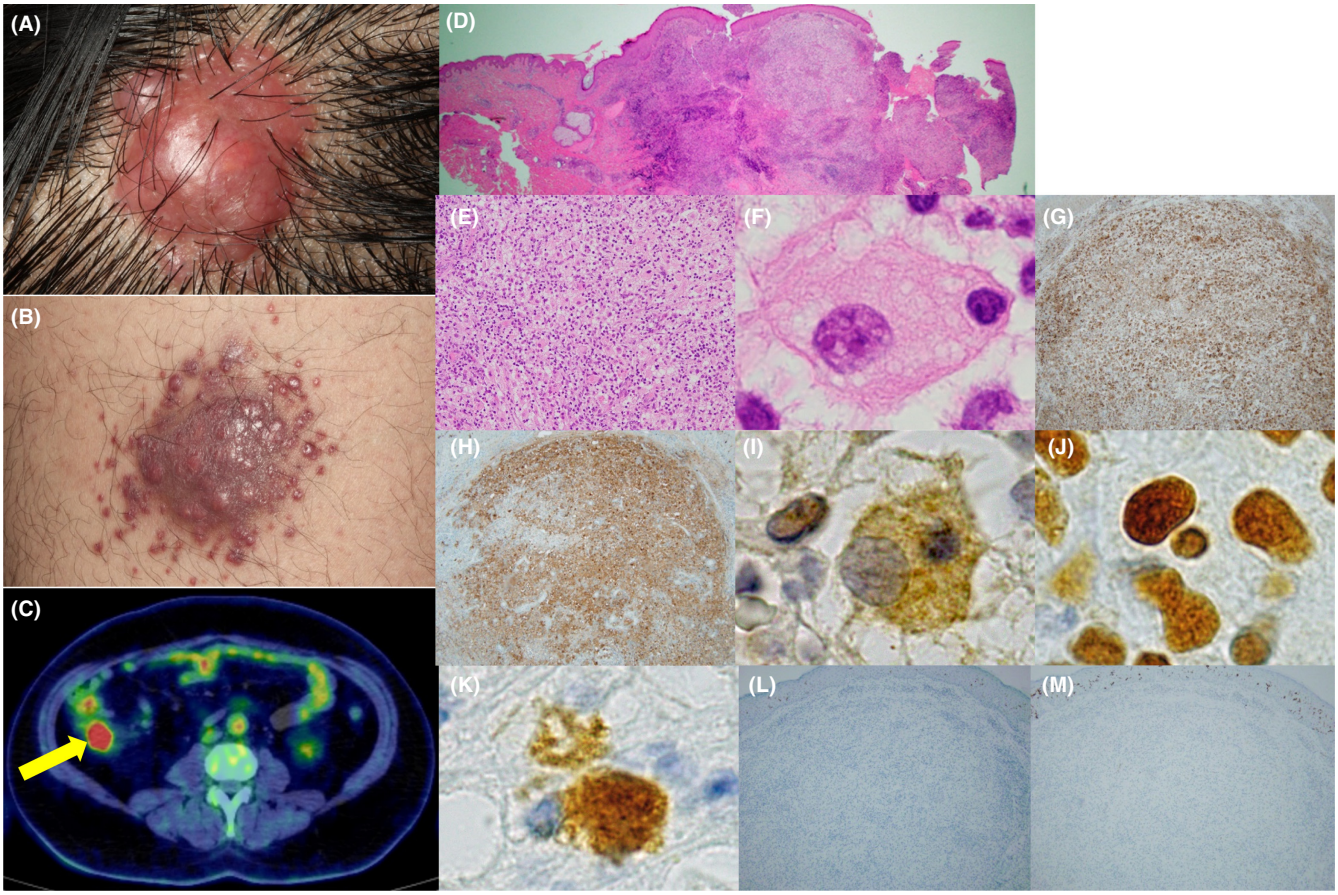

FIGURE 1 (A, B) physical examination. (A) Dark-reddish nodule on the right side of the occiput, (B) a nodule on the right thigh, surrounded by multiple reddish papules. (C) 18F-fluorodeoxyglucose positron emission tomography/computed tomography revealing accumulation of standardized uptake value in the mass of the terminal ileum (yellow arrow indicates mass) (D) histopathological findings showing dense cellular infiltration from the papillary layer to the subcutaneous tissue (hematoxylin and eosin [H and E] stain, original magnification ×12.5). (E) Infiltration of various inflammatory cells such as macrophages, neutrophils, plasma cells, and leukocytes (H and E stain: Original magnification ×200). (F) Large-sized histiocytes with foamy cytoplasm and emperipolesis (H and E stain original magnification ×1000). Immunohistochemical staining for (G) CD68, (H) S-100 protein, (I) cyclin D1, (J) OCT2, (K) phospho-ERK, (L) CD1a and (M) langerin for infiltrating large-sized histiocytes (original magnification ×100 and ×1000).

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KEYWORDS

extranodal, Japanese, Rosai–Dorfman disease

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

DECLARATION SECTION

Approval of the research protocol: Not applicable.

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