Journal of Dermatology
Manuscript ID: JDE-2018-0382 2nd Revised Version
Letter to the editor

Hyaline vascular-type unicentric Castleman disease presenting as a subcutaneous nodule in a child

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Text word count and number of references, tables, and figures:
493 words in the main text, 5 references, 1 figure

Funding statement: None

Conflicts of interest: None

***Short title*:** Castleman disease as a subcutaneous nodule

***Key words*:** (1) Castleman disease, (2) subcutaneous nodule, (3) unicentric type, (4) hyaline-vascular type, (5) infantile

Main Text

Castleman disease (CD)[1] is a rare lymphoproliferative disorder of unknown etiology that typically manifests in the mediastinum.[2] Unicentric CD (UCD) and multicentric CD (MCD) are fundamentally distinct clinical entities: MCD is a systemic disease; UCD is a localized, non-systemic disease. Here we report a juvenile case of UCD that presented as a subcutaneous nodule.

A 9-year-old Japanese girl presented with a subcutaneous nodule in her back. At the age of 3, a small nodule of 1 cm in diameter was noticed. She visited a pediatrician at her neighborhood hospital. During the 2-year follow-up, the tumor expanded gradually, and then magnetic resonance imaging (MRI) was performed. The imaging revealed a mass of 3 × 2 cm in size, showing iso-intensity on T1-weighted images in the subcutaneous tissue. It showed slight hyper-intensity on T2-weighted images. Hemangioma, nodular fasciitis and lymphoma were considered as diagnoses by MRI. At three years later, the nodule had enlarged to 4 cm in diameter. At the age of 8, she was referred to a dermatologist in the hospital and a punch biopsy was performed.

Histopathologically, the biopsy specimen showed the proliferation of lymphoid follicles with hyalinization in the subcutaneous fat. Poorly formed germinal centers were seen in the intrafollicular areas. A concentric arrangement of lymphocytes and vascular proliferation were observed at the periphery of the mantle zone. The histopathological diagnosis was the hyaline-vascular type (HV) of CD (HVCD). We differentiated the present case from cutaneous pseudolymphoma based on the histopathological features of lymphoid follicles with hyalinized vessels in the present tumor.[3] These findings are

characteristic of HVCD.

She was referred to our hospital for treatment. At our initial examination, a firm, painless nodule of 5 cm in diameter was palpable. Her lymph nodes were not palpable. Laboratory studies revealed no abnormal findings. A well-circumscribed tumor was excised with a minimal margin. Macroscopic examination revealed an oval mass of 4 cm on the major axis, and the half-split face was yellowish.

The histopathological features of the resected mass were similar to those seen in the biopsy sample. Neither monoclonal *IGH* gene rearrangement nor *TRG* gene rearrangement was detected.

The patient's condition was finally diagnosed as HV of UCD. There has been no recurrence for the 7 months since the resection operation.

Extranodular CD lesions, particularly subcutaneous ones, are very rare. Only 13 subcutaneous cases of CD have been reported, including ours.[4, 5] Of these 13 cases, 12 were UCD and one was multicentric CD. As for the histopathological types, one of the UCD cases showed mixed histopathological features of HV and the plasma-cell type, although the other cases are all classified as HV. The subcutaneous CD patients' ages were from 3 to 69 years, with the present case being the youngest. As the present case had a very early-onset CD lesion at an uncommon site, it was 6 years before she was diagnosed with UCD.

We should add UCD as a differential diagnosis for subcutaneous tumors on the trunk of

children.

ACKNOWLEDGMENTS

We would like to thank Prof. Emer. Masanobu Kumakiri for his helpful comments.

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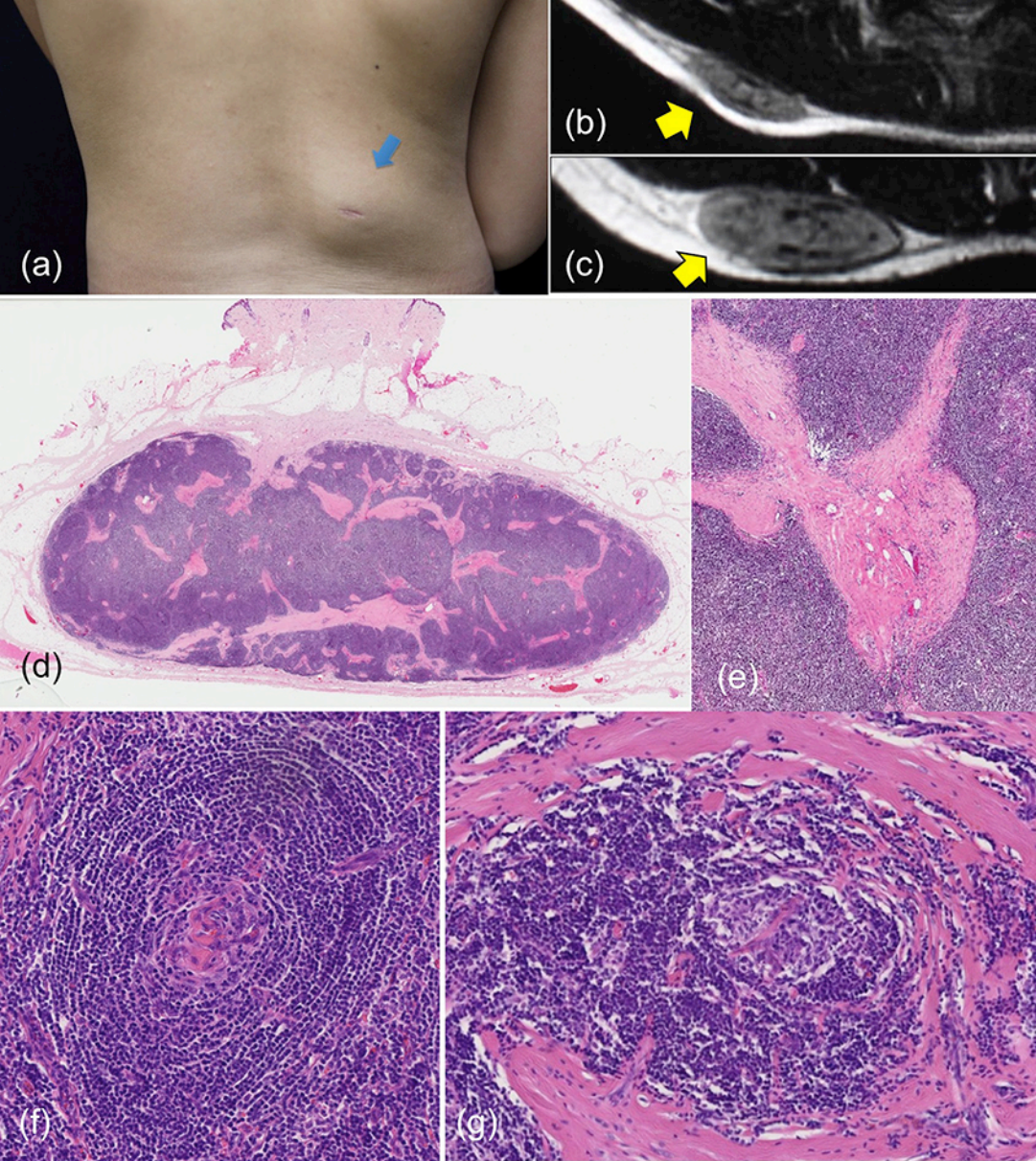
FIGURE LEGEND

Figure 1. The clinical, MRI and histopathological features of the present patient. (a) The clinical features of the subcutaneous nodule on the back. (b, c) A T2-weighted MRI image shows a subcutaneous mass (arrow) of 2.0 × 0.7 × 1.8 cm in size at age 5 (b), which has enlarged to a mass (arrow) of 3.8 × 1.6 × 3.9 cm in size at age 9 (c). (d-g) The histopathological findings of the resected subcutaneous nodule. (d) The margin of the subcutaneous nodule is sharp, and the nodule consists of lymph node-like structures with interstitial hyalinization. (e) Hyalinized blood vessels in the interfollicular region. (f) The broad mantle zone, showing an “onion skin” pattern. (g) A depleted germinal center with increased vasculature showing a “lollipop” appearance. Hematoxylin-eosin staining (d-g), original magnifications ×1.25 (d), ×5 (e), ×20 (f), ×10 (g).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(b)
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