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Case Report

A rare stroma-rich variant of hyaline-vascular Castleman's disease associated with calcifying fibrous pseudotumor

Haifen Ma, Maofen Jiang, Weihua Xiao

Department of Pathology, Beilun People's Hospital, Ningbo 315800, Zhejiang Province, China

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Abstract: Objectives: The stroma-rich variant of hyaline-vascular type of Castleman's disease (SR-HVCD) should be differentiated from vascular or follicular dendritic reticulum cell neoplasms. In this paper, we present a rare case of HVCD. We also suspect a possible association between SR-HVCD and calcifying fibrous pseudotumor. Methods: A 34-year-old man was found an abdominal mass by computed tomography (CT) in a general health checkup. The mass was resected from the mesenteric root. The specimens were evaluated for detailed characterizations through gross examination, microscopy and immunohistochemistry. Results: The mass showed histologic patterns and immunohistochemical results of HVCD with significant angiomyoid proliferations, collagenation and focal calcification. Histologically, stromal elements of HVCD in our case were similar to those of a calcifying fibrous pseudotumour. Conclusions: A possible association was suspected between SR-HVCD and calcifying fibrous pseudotumor. To the best of our knowledge, this is the fourth report of describing an association between the two diseases in the English literature.

Keywords: Castleman's disease, hyaline-vascular variant, stroma-rich, calcifying fibrous pseudotumor, association

Introduction

The hyaline-vascular type of Castleman's disease (HVCD) is a rare disorder of the lymphatic system characterized by both angiomyoid and follicular dendritic cell proliferations. Occasionally, HVCD shows overgrowth of a variety of stromal cells with predominantly vascular components and fibrosis (more than 50% of the lesion), which is called stroma-rich variant of HVCD (SR-HVCD) by Danon et al [1]. The SR-HVCD should be differentiated from vascular or follicular dendritic reticulum cell neoplasms. In this paper, we present a rare case of HVCD with significant angiomyoid proliferations, collagenation and focal calcification. We also suspect a possible association between SR-HVCD and calcifying fibrous pseudotumor. To the best of our knowledge, this is the fourth report of describing an association between the two diseases in the English literature.

Case report

A 34-year-old man was found an abdominal mass by computed tomography (CT) in a general health checkup six months ago. The mass was considered to be a neurogenic tumor from the CT findings. Six-month follow-up by CT revealed no grow. Subsequently, the mass was resected from the mesenteric root.

Pathological findings

The specimen was sent completely for pathologic analysis.

Grossly, the mass was measured 3 × 3 × 2.5 cm in size and was well circumscribed. An irregular circumferential flesh-colored area was seen on the cut surface with a width of approximately 0.5 cm, in addition to a whitish and interlaced area measuring 2.5 × 2 cm in the center with focal calcification (**Figure 1A-C**). No bleeding or necrosis was detected inside the tumor.

Histologically, the surrounding area of the mass showed all typical histological changes that are usually seen in HVCD. Dystrophic follicles showed hyperplasia of the mantle layer with a typical onion-skin pattern, atrophic germinal centers and signs of vascular invasion. In addi-

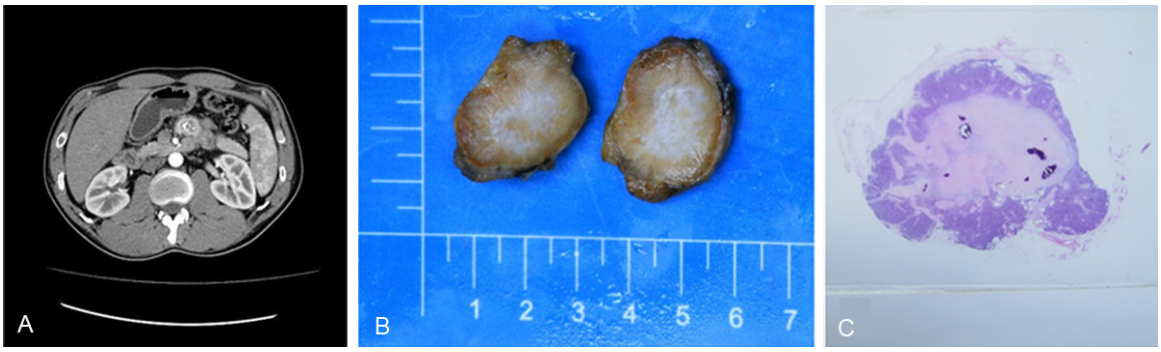

Figure 1. CT reveals an abdominal mass (A). Gross aspect of the lymph node. Noted a whitish and interlaced area in the center (B). The large pathologic slice. Noted distinct nodular growths with focal calcification in the central area (C).

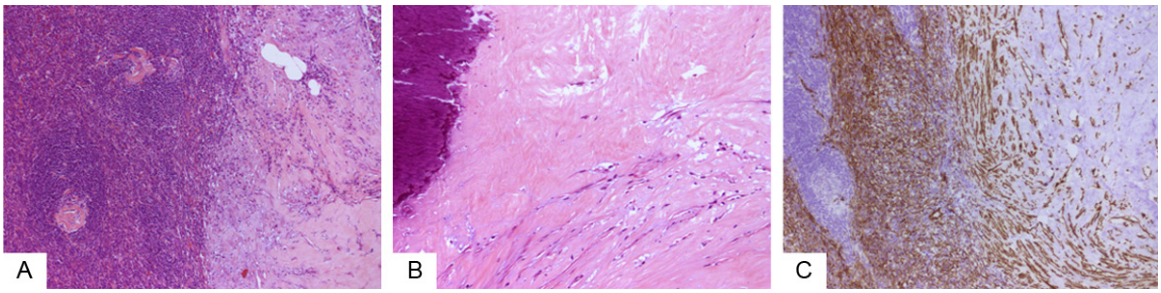

Figure 2. H&E stained sections showing the interface between the lymph tissue and the stromal nodule. The lymphatic tissue showed dystrophic follicles and signs of vascular invasion that are usually seen in HVCD (A). A paucicellular and heavily collagenized tissue that contained focal calcification was seen in the central area (B). Immunohistochemical findings showing the stromal cells strongly stained for SMA, but not the collagenized areas (C).

tion, hypervascular interfollicular tissue and lack of sinuses were found. The central area was composed of significant angiomyoid proliferations and a paucicellular and heavily collagenized tissue which contained focal calcification (**Figure 2A**, **2B**). In the border area, lymphoid tissue mixed with myofibroblast was detected.

On immunohistochemical analysis, the immune structure of dystrophic follicles was delineated by CD20, CD3, bcl-2, and Ki67, while CD21 clearly outlined the networks of follicular dendritic cells. The stromal cells were diffusely positive for SMA, except for the collagenized areas (**Figure 2C**). Immunostaining of CD34 showed proliferation of the vascular endothelial cells. Occasional staining of S100 and Desmin were also seen.

Follow-up

No recurrence was found 6 months after the surgery.

Discussion

The SR-HVCD is a new pathological entity which shows overgrowth of stromal cells. Danon et al found obvious nodular proliferation of reticulum cells and increased vascularity in 5 cases in a histologic review of 102 cases of HVCD [1]. Lin et al reported 10 cases of HVCD with angiomyoid, follicular dendritic cell, or fibrohistiocytic proliferations, as well as various vascular tumors [2].

In our case, significant nodular proliferation of the spindle cells and heavily collagenized tissue that contained focal calcification was seen in HVCD. There is no doubt that the lesion is classified as SR-HVCD. Histologically, stromal elements were similar to those of a calcifying fibrous pseudotumor (CFT), although immunostaining of the spindle cells with strong expression of SMA did not support.

Few reports about association between HVCD and CFT have been published. A CFT was

reported in a HVCD in a 9-year-old boy who had undergone a fine-needle aspiration [3]. Azam et al reported a CFT arising from the gastric wall with areas focally mimicking HVCD and the surrounding lymph nodes showing HVCD [4]. Recently, another case was reported of a 25-year-old woman underwent surgery for a small bowel mass at the site of an ileocolic intussusception with mesenteric lymphadenopathy. Pathologic analysis revealed that the mass was a calcifying fibrous tumor associated with mesenteric Castleman-like adenopathy [5].

From these cases, it may be speculated that HVCD may represent an incipient presentation of CFT. With the proliferation of stromal cells, SR-HVCD gradually develops tumor-like appearance. In the present case, significant angiomyoid proliferations with paucicellular area and heavy collagenation that contained focal calcification in HVCD may represent a mid-stage process. Based on this, it is reasonable for us to assume that CFT possibly represents an abnormal healing response and the end stage of HVCD.

The pathogenesis of the two diseases remains unknown. Initially, CFT was thought to be a re-active inflammatory process that resulted from an abnormal healing response [6]. And there are examples of CFT that have followed trauma [7]. But it is unclear whether CFT occasionally occurs in HVCD in response to a tissue injury or likely secondary to a similar etiology. However, there is no history of injury or inflammation in our case. Further investigation with a larger st-udy is needed to confirm these assumptions.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Haifen Ma, Department of Pathology, Beilun People's Hospital, Ningbo 315800, Zhejiang Province, China. Tel: +86-13958311234; Fax: +86-574-86776335; E-mail: mhfen@163.com

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