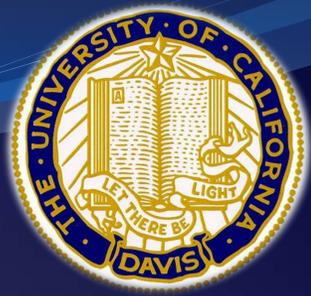




# STROMAL RICH VARIANT OF HYALINE-VASCULAR CASTLEMAN DISEASE A CASE REPORT OF AN UNUSUAL TUMOR WITH REVIEW OF THE LITERATURE



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## Introduction:

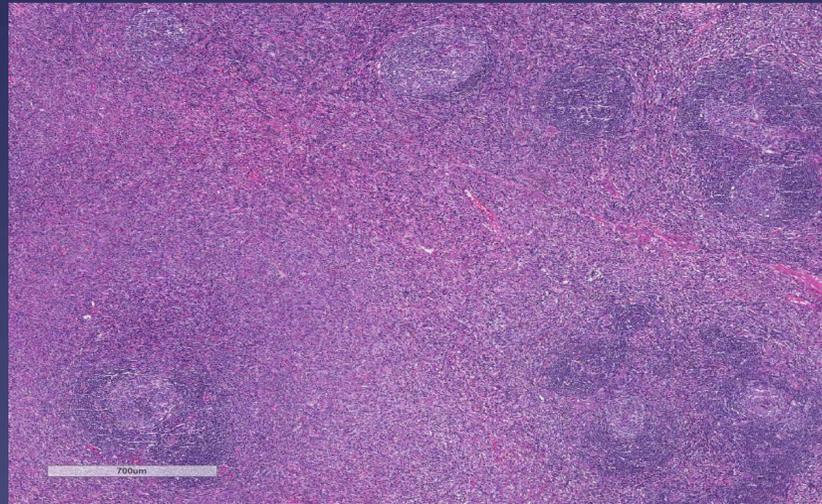
Castleman disease is an uncommon form of lymph node hyperplasia. It commonly involves the mediastinum but can also involve extrathoracic sites, including the neck, pelvis, and retroperitoneum. The stroma-rich variant of Castleman disease of the hyaline-vascular type (CDHV) is a rare entity that shows overgrowth of a variety of stromal cells in the expanded interfollicular area.

## Case History:

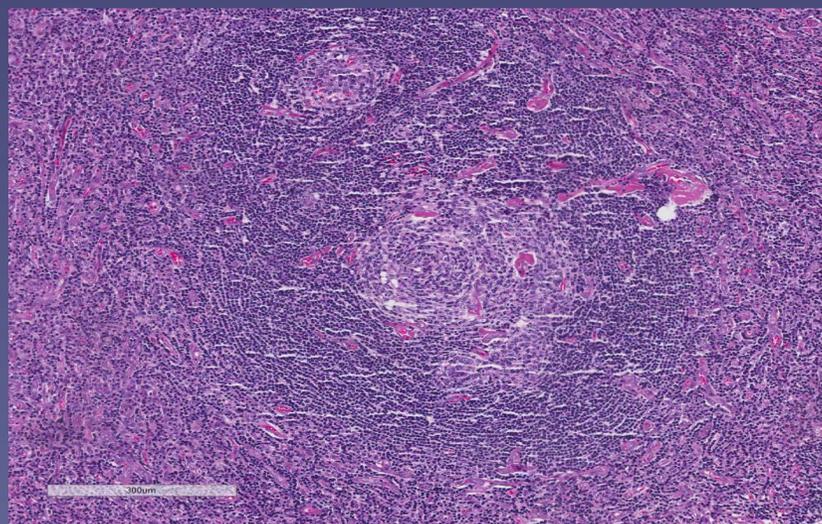
A 20-year-old female presented with nausea, vomiting, and a syncopal event. An ultrasound revealed a 5.6 cm right retroperitoneal mass. An excisional biopsy showed a lobulated lymph node architecture effaced by sheets of spindled and ovoid cells forming fascicles in a storiform pattern. At the periphery and interspersed between the spindle cells were residual lymphoid follicles with regressive changes and multi-germinal center pattern. Sclerotic vessels with hyaline walls radially penetrate the germinal centers, reminiscent of the features of hyaline-vascular Castleman disease. The interfollicular spindle cells were positive for smooth muscle actin, and they were negative for CD21, CD34, CD117, CD163, ALK-1, HHV-8, and S-100 proteins.

## Case discussion:

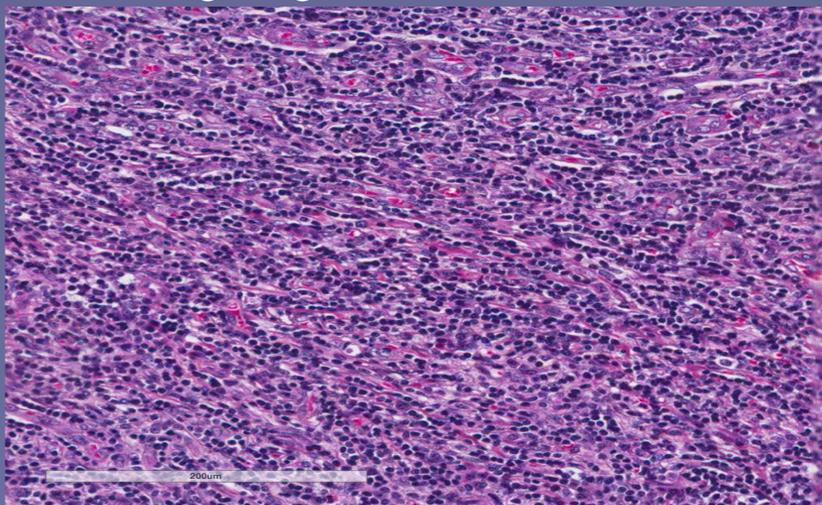
This case was diagnosed as a stromal-rich variant of Castleman disease. This is a rare entity that shows overgrowth of stromal cells in the widened interfollicular area. Clinically, it usually presents as an asymptomatic, solitary nodule that predominantly develops in the retroperitoneum. This stromal-rich lesion is typically hyperplastic and clinically benign, and it must be distinguished from



Expanded interfollicular area, 2X

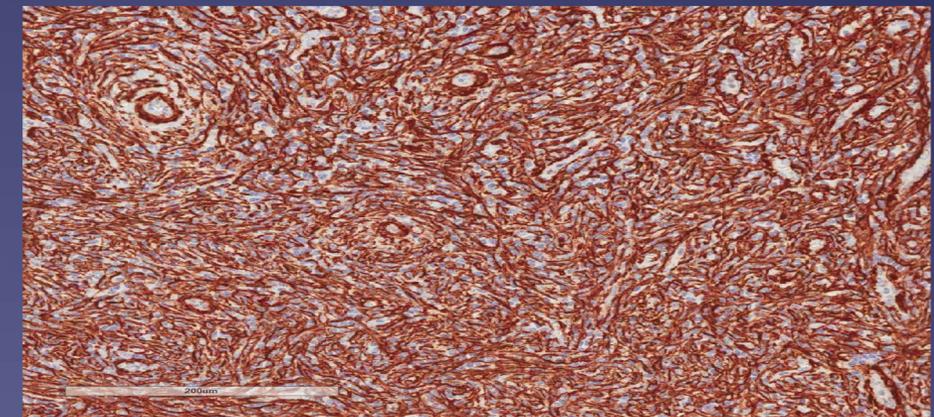


Sclerotic vessels with hyaline walls radially Penetrating the germinal centers, 10X

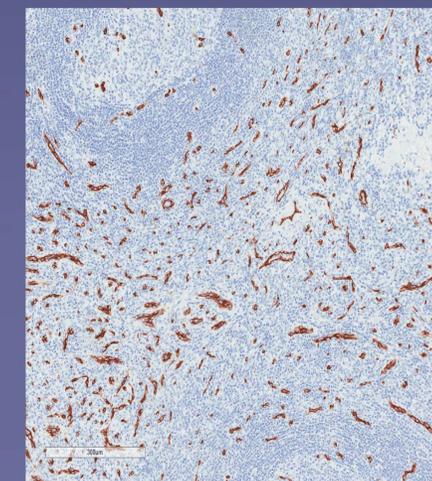


Interfollicular area, 16X

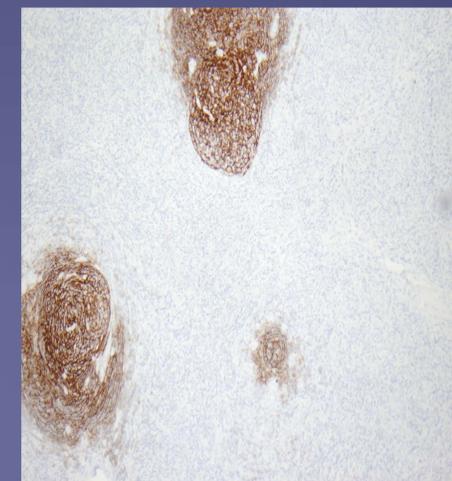
neoplastic stromal proliferation such as follicular dendritic cell (FDC) tumor, which will show an expansion of dendritic cells in the interfollicular area staining for CD21, CD23 and CD35 or vascular neoplasm associated with Castleman disease (CD) because of its potential for recurrence and metastasis. The biologic behavior of FDC proliferations arising in CD is variable and is not predictable with certainty. Whereas surgical excision appears to be the treatment of choice for angiomoid overgrowths.



Smooth muscle actin + interfollicular cells, 16X



CD34 highlights the vessels in the interfollicular area, 8X



CD21, highlights the follicular dendritic cells

Reference: Miki Izumi, et al., Angiomoid proliferative lesion: an unusual stroma-rich variant of Castleman's disease of hyaline-vascular type, Virchows Arch (2002) 441:400-405