

Images of Interest / Imagens de Interesse

Castleman Disease: Atypical Cause of Pneumonectomy

*Doença de Castleman: Causa Atípica de Pneumectomia*Mariana Conceição¹, Tito Abrantes¹, Sofia Campelos^{2,3}¹Pulmonology Department, Centro Hospitalar Tondela-Viseu, Viseu, Portugal²Laboratory of Pathology, IPATMUP - Institute of Molecular Pathology and Immunology of University of Porto, Porto, Portugal³Institute for Research and Innovation in Health (I3S), University of Porto, Porto, Portugal

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Abstract

Castleman Disease is a lymphoproliferative disorder that is classified clinically as unicentric or multicentric, and histologically as hyaline vascular variant or plasma cell variant. The unicentric Castleman Disease often presents as an incidental mediastinal mass, but the involvement of the pulmonary hilum is rare.

Keywords

Castleman disease; Computed tomography; Pneumonectomy; Thoracic surgery.

Resumo

A Doença de Castleman é uma doença linfoproliferativa, podendo classificar-se clinicamente como unicêntrica ou multicêntrica, e histologicamente como variante hialino-vascular ou variante de células plasmáticas. A Doença de Castleman unicêntrica apresenta-se frequentemente como uma massa mediastínica incidental, contudo o envolvimento pulmonar hilar é raro.

Palavras-chave

Doença de castleman; Tomografia computadorizada; Pneumectomia; Cirurgia torácica.

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A previously healthy 24-year-old male, smoker (4 pack-year), presented with a traumatic fracture of the right humerus. Preoperative chest radiography revealed a left perihilar lesion (Figure 1A). He had no respiratory or constitutional symptoms. The physical examination was unremarkable. Laboratory data, including neuron-specific enolase and chromogranin, were normal. Chest computed tomography showed a rounded left perihilar lung mass, well-defined, with 45x40mm and slight contrast uptake (Figure 1B and C). Positron emission tomography revealed increased fluorodeoxyglucose-F18 uptake (SUVmax:5,4) in the left hilar lesion. 68Ga-DOTA-NOC PET-CT also showed an abnormal uptake from the nodular formation in the left pulmonary hilum, suggesting a neuroendocrine tumor. Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) was performed with puncture of the left mass; TBNA samples were negative for malignancy. Lung function was normal. He was therefore referred for evaluation of thoracic surgery.

Given the tumor's central location with major pulmonary vessels and main bronchus involvement, he underwent left pneumonectomy. Hematoxylin and eosin staining showed regressed germinal centers with follicular dendritic cell prominence, surrounded by mantle zones containing small lymphocytes arranged in a concentric pattern (Figure 1D). Microscopic features and immunostaining were consistent with Castleman Disease-Hyaline Vascular Variant. The patient received no further therapy, maintaining regular surveillance.

Unicentric Castleman Disease (UCD) frequently presents as an incidental solitary mediastinal mass, however, intrapulmonary location with the hilum involvement is rare. The preoperative diagnosis can be challenging as clinical and radiological findings are nonspecific.^{1,2} The standard treatment for UCD is complete surgical resection.³ This case emphasizes that although UCD with hilar-presentation is a rare and benign condition, anatomic resection and even a pneumonectomy may be required for diagnostic and therapeutic purposes.

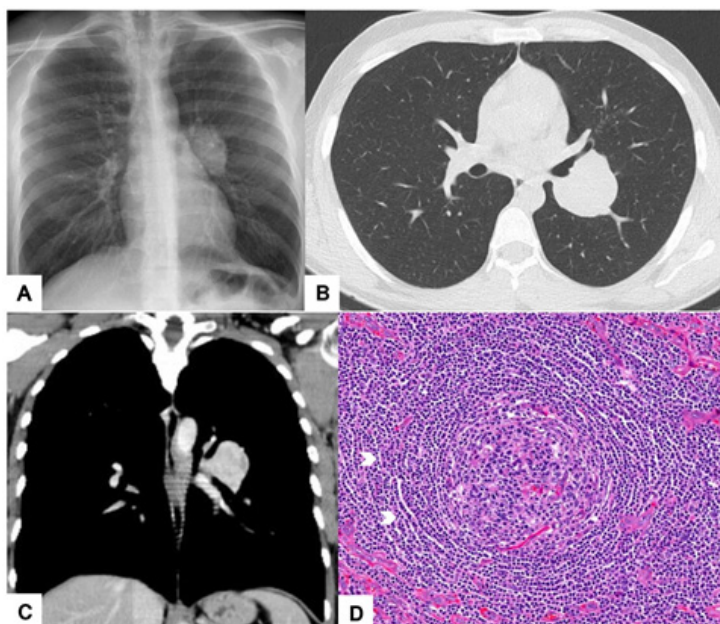


Figure 1. A: PA Chest radiograph showing a left hilar lesion. B and C: Axial (lung window) and coronal (mediastinal window) chest CT, respectively, revealing a rounded and well-defined left perihilar mass with mild contrast uptake. D: Hematoxylin and Eosin (H-E) staining revealing an enlarged follicle with concentric layering of mantle zone lymphocytes (arrows) encircling an atretic germinal center (x200).

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Proteção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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