Pathologic Quiz Case

Bilateral Apical Lung Masses in an Autopsy Patient

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A 68-year-old white man was brought to the emergency department after being found down in his home by a neighbor. The patient was intubated in the field and, on admission, was comatose, hypotensive, and in atrial fibrillation. A computed tomographic scan of the head was negative for an acute process. A subsequent review of the patient's medical records revealed that he had been previously seen 4 months earlier in the pulmonary clinic for a chief complaint of increasing shortness of breath. Prior to that encounter, the patient had not seen a doctor for more than 30 years. At the time, the patient was noted to have difficulty remembering past events in his life. He was, however, accompanied by his niece, who was only able to relate that her uncle was an occasional smoker and had been in the construction business. She did not have any definitive knowledge of specific exposures, such as asbestos or tuberculosis, or anything else unusual. Chest radiographs showed a focal fibrosing process concentrated in the bilateral lung apices. A transbronchial biopsy was performed on the right upper lobe that showed only nonspecific interstitial thickening with fibrosis and organizing pneumonia. No granulomas were identified. Stains for fungal and acid-fast organisms were negative.

Three days after the most recent admission, the patient was extubated but, hours later, became acutely unresponsive and apneic. A computed tomographic scan of the chest showed multiple pulmonary emboli of the lower lobes. The patient died shortly thereafter. Consent for an autopsy was granted by the niece.

At autopsy, the most significant finding was an extensive but focal area of fibrosis located in the bilateral lung apices (Figure, A). Both the apices of the right lower and left lower lobes also showed smaller but identical processes. The right lung was further remarkable for the presence of a large pleural plaque extending from the apex of the upper lobe to the peripheral edge of the lower lobe. Grossly, the uninvolved lung parenchyma showed evidence of mild bronchiectasis, bronchopneumonia, and multiple, small pulmonary emboli. The nonfibrous parenchyma did not show evidence of interstitial lung disease; however, pulmonary emboli and acute bronchopneumonia were microscopically confirmed. The apical areas showed paucicellular fibrous obliteration of the normal lung parenchyma (Figure, B, low magnification; Figure, C, high magnification), as confirmed by a trichrome stain (Figure, D). The adjacent pleural plaque consisted of acellular, hyalinized, eosinophilic material. In the fibrous caps, a few scattered patches of chronic inflammation were present, but no cellular atypia was noted. No granulomas were identified in multiple sections. Stains for acid-fast and fungal organisms were negative. An iron stain did not show ferruginous bodies or hemosiderin macrophages. A mesothelioma panel was noncontributory.

What is your diagnosis?

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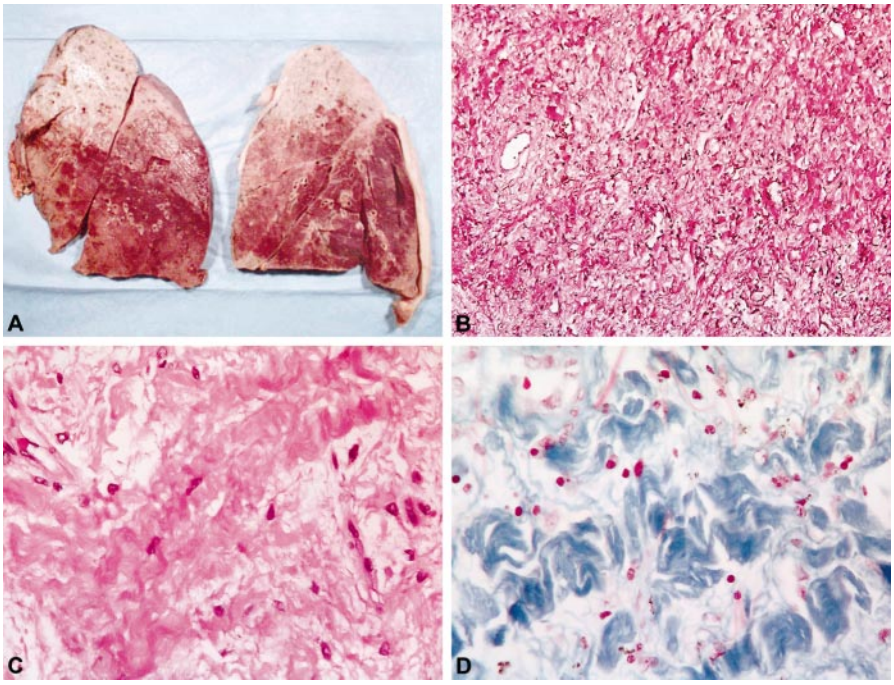

Pathologic Diagnosis: Pulmonary Apical Cap

Pulmonary apical cap is a term applied to a distinct pattern of focal pulmonary fibrosis. Pulmonary apical cap presents as a well-delineated area of fibrosis located in the lung apices.[1] The process is typically bilateral and occurs predominantly in elderly men.[2,3] Grossly, the fibrosis is gray-white with scattered anthracotic pigmentation. The affected area is typically defined by a pyramidal configuration. On palpation, the fibrosis is expectedly firm but retains a slight softness that is not typical of a malignancy with associated desmoplasia. Microscopically, at low magnification, a slight basophilia of the fibrosed area may be noted.[3] The apical cap is distinctly paucicellular, but anthracotic pigment, macrophages, and scattered chronic inflammatory cells may be found among the bland fibrosis. In his series of 13 cases, Yousem[3] reports that an elastic stain will show cords of preserved elastic tissue, suggesting that the framework of the pulmonary lobule essentially collapses upon itself due to ''contraction of mature intra-air space collagen.'' Abnormalities of the vessels immediately adjacent to the fibrous area may also be present. These changes may consist of sclerosis and mural thickening.[1]

A multitude of specific disease processes and exposures will result in pulmonary fibrosis. The unique apical concentration, however, may be seen with higher frequency in tuberculous infection, asbestosis, or histoplasmosis or as a complication of ankylosing spondylitis, among other entities.[4–7] In the early 1900s, most pulmonary cases with bilateral apical fibrosis were originally thought to be a consequence of tuberculous infection due to both the affinity of mycobacterium for this area and the prevalence of the disease at the time.[3,4] Tissue studies, however, have continually failed to demonstrate granulomatous inflammation.[1–3] In the largest series consisting of 123 cases, no association between pulmonary apical cap and tuberculosis was found.[2] A defining feature of pulmonary apical cap, therefore, is that there is a distinct absence of granulomatous inflammation.[1–3] In addition, other clues that would point to a specific etiology, such as polarizable material and ferruginous bodies, are absent.[1–3] Occult malignancy, particularly sarcomatous mesothelioma, must always be in the differential, but the distinct blandness and paucicellular nature of pulmonary apical cap should be evident.[3]

Many theories have been proposed for the pathogenesis of pulmonary apical cap. Butler and Kleinerman[1] felt that the relative, physiologic ischemia of the lung apices was critical to the process. They believed that pulmonary apical cap was probably a consequence of a chronic, low-grade, nontuberculous infection, the clearing of which was impaired by the ischemia. In their case series, they also noted a preponderance of mural thickening in the small muscular arteries immediately subadjacent to the apical cap areas. This finding was noted in only 25 of their 48 cases but was thought to be significant.[1] Renner et al[2] drew a similar conclusion, hypothesizing that pulmonary apical cap was related to the healing of pulmonary disease in the presence of chronic apical ischemia, a belief that is shared by Yousem.[3] The patient presented in this case ultimately died from multiple acute pulmonary emboli. The contribution that pulmonary embolism may provide in the development of chronic ischemia leading to pulmonary apical cap, though logically compelling, is not yet clearly defined. Although abnormalities in the vasculature underlying the apical fibrogenic areas have been noted in several series, the author is not aware of any studies citing chronic pulmonary embolism as the leading cause of apical ischemia leading to pulmonary apical cap.[1,3] In their examination of the reasons for apical localization of certain pulmonary diseases, Goodwin and Des Prez[6] further asserted that the lung apices are subject to a degree of physiologic lymph stasis that compounds the relative apical ischemia and impairs the clearance of antigenic substances. In a letter commenting on Yousem's study, Dail[8] suggests that the weight of the lung itself also plays a part in the pathogenesis of pulmonary apical cap by causing ''microscopic tears/stresses in tissue substructure,'' which may result in ''repetitive small scarring events.''

The limitations of these theories, however, are that they do not explain the sex predilection of this entity or its relative rareness given the prevalence of both pulmonary infection and, with the exception of children, the universal, relative apical ischemia of adults. Some of the theories also do not adequately explain the usually bilateral nature of pulmonary apical cap. To the author's knowledge, no literature exists that provides a proven pathogenesis. Pulmonary apical cap, however, is a distinct entity that should be recognized in the differential of focal pulmonary fibrosis in an apical location.

References

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