Pulmonary Apical Cap—What’s Old Is New Again

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The pulmonary apical cap (PAC) is a morphologically distinct type of unilateral or bilateral fibroelastotic scar involving the lung apices. Despite being relatively common and having been described more than a hundred years ago, it remains underappreciated as a unique diagnostic entity by clinicians, radiologists, and pathologists alike. Given the centrality of modern chest imaging in the workup of diseases of the lungs, it may be expected that the PAC will be biopsied with increasing frequency. As such, pathologists should be familiar with its presentation, appearance, and differential diagnosis. This article serves as a short overview of PAC and as a practical aid in its diagnosis for surgical pathologists.

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The pulmonary apical cap (PAC) has been recognized for more than a hundred years as an anatomically and microscopically distinct form of localized scarring, although its etiology still remains a matter of speculation. Despite its “age,” it seems to have been overlooked as a unique diagnostic consideration in current times and it remains underappreciated by clinicians, radiologists, and pathologists alike. Indeed, with modern techniques in thoracic imaging and their centrality in the evaluation of diseases of the chest, it stands to reason that the PAC, being a focal (or multifocal) space-occupying lesion, is more likely than ever to be biopsied in the evaluation of suspected neoplasms. The aim of this article is to provide a short overview of the PAC and serve as a practical aid in its diagnosis for the general surgical pathologist.

BRIEF HISTORY AND ETIOLOGIC CONSIDERATIONS

Unilateral or bilateral fibrosis of the lung has been recognized as a unique finding since the heyday of morbid anatomy and was believed to be largely a consequence of pulmonary tuberculosis. However, subsequent studies failed to reveal evidence of granulomas in these scars. An early challenge to the tuberculosis theory came in the 1930s when the fibrosis was posited to be a consequence of silica-containing (“siliceous”) inhaled dust, yet in the absence of well-developed silicosis. In the 1940s, MacMillan noted branches of the pulmonary artery narrowed by “endarteritis obliterans” within the scars and suggested the possibility of “low-grade inflammation” and collapse secondary to poor respiratory movement as possible explanations.

This theory received significant support in an important study by Butler and Kieferman in 1970. They reviewed 183 consecutive lungs from 183 patients at autopsy and noted mural thickening of small muscular pulmonary arteries immediately underneath the lesion in 25 of 48 PACs, while noting this change in the subapical vessels in only 2 of 10 lungs without apical cap. They also observed more frequent evidence of chronic bronchitis in association with PAC, which was especially common in their younger cohort where this finding achieved a high degree of statistical significance. Although the observed vascular changes did not achieve statistical significance, they drew attention to the association of chronic bronchitis with (1) viral and bacterial infections and (2) increased pulmonary vascular resistance. They offered a plausible explanation for the consistent localization of the apical cap, which is enshrined in its name: “The clearly unique feature of the apex...is the high ratio between ventilation and perfusion, the major factor in which is decreased perfusion. We submit that relative apical ischemia may play a critical part in the pathogenesis...and offer in support the high proportion of involved lungs showing significant subjacent small arterial disease.”

They therefore hypothesized that PACs are due to repeated or smoldering nontuberculous pulmonary infections, the healing of which is impaired secondary to chronic ischemia intrinsic at the lung apices. This theory also was felt to help explain the increased prevalence of PAC in older patients. They found no evidence of granulomas in PACs and no evidence of furrinous bodies including asbestos or any other inorganic dust.

More recently, Yousen studied 13 cases of PAC in a series of patients who had undergone wedge resection for a presumed diagnosis of pulmonary carcinoma. He too noted the presence of subjacent small muscular arteries showing mural sclerosis and recanalization in 11 of 13 cases. He also remarked on the histologic resemblance of PAC with old pulmonary infarcts and endorsed the notion that PACs are likely a consequence of a combination of repeated low-grade infection and chronic ischemia, with the latter favored as the major factor. In a letter to the editor, Butnor and colleagues made an interesting connection between radiation pneumonitis and PAC, noting their histologic similarity and association with oblitative vascular changes,
lending support to the role of ischemia in the pathogenesis of PAC. Dail, also commenting in a letter to the editor, suggested the possibility of the weight of the lung itself as causing "microscopic tears/stresses in tissue substructure, which may result in repetitive small scarring events" and that these stresses are greatest at the apices.

**EPIDEMIOLOGY**

The PAC is relatively common, and its prevalence increases with advancing age. In the study of Butler and Kleinerman, the ages ranged from 16 to 94 years, with increasing prevalence in older patients. There was no sex predilection. In an autopsy series in which both lungs were evaluated, Renner and colleagues found "apical pleural scar" in 47% of 57 patients. In patients aged 66 to 75 years, the prevalence reached 64%, while below the age of 45 years only 20% had scars. It was unilateral in 60% of cases, without predilection for either side. When bilateral, PACs may be asymmetric (see Figure 1). In the same study, Renner and colleagues also evaluated 258 routine chest radiographs, and identified a unilateral "cap shadow" in 29 cases (11.2%) and a bilateral cap shadow in 28 cases (10.9%). In patients up to 45 years of age, 6.2% had such a shadow, which increased to 15.9% of patients above the age of 45 years.

**SYMPTOMATOLOGY AND NATURAL HISTORY**

The PAC is regarded as essentially asymptomatic. It is usually identified incidentally during chest imaging, where it may be mistaken for neoplasm. Butler and Kleinerman observed that PACs, in addition to being more prevalent in older patients, tended to also be larger than those in younger patients. On this basis, they reasoned that the PAC may slowly enlarge over time. It is also the author's experience that a PAC may enlarge over time in an individual patient, in one case, growing from 10 mm to 14 mm over the course of 3 years (unpublished
observation). In addition, a PAC may show mild fluoro-
deoxyglucose avidity on positron emission tomography
scan. It is possible that the low degree of metabolic activity
may be due to the relatively common presence of
lymphocytic inflammation, often present as follicular
aggregates dispersed around the PAC (see next section).

**MORPHOLOGY AND DISTRIBUTION**

Grossly, the PAC is a wedge-shaped, triangular, or
crescentic fibrous subpleural parenchymal scar with discrete,
well-defined margins. This appearance lends credence to the
theory of vascular ischemia, since this is evocative of the
classic pulmonary arterial infarct. The “base” of the pyramidal
shape is along the pleural aspect. The size can range from
microscopic to multiple centimeters (up to 6.0 cm), as
measured along this “base.” The thickness is much less than
this, the “apex” usually projecting less than 1 cm into the
parenchyma. This gross appearance is recapitulated on chest
computed tomography scan, where wedge and triangular-
shaped opacities with broad pleural contact may be seen
(Figure 1). The radiologic margins can be irregular and
spiculated, reinforcing the potential confusion with primary
bronchogenic carcinoma. Although the PAC is most common
at the apex of the upper lobes, it can be found in the apical
segments of lower lobes as well, presumably owing to their
similar physiologic predisposition to reduced perfusion.

At low power, the PAC has discrete margins and a
characteristic “stuck on” appearance (Figure 2). At higher
power, the most striking and helpful feature of PAC is a
unique combination of fibroelastotic scarring composed of
an admixture of mature collagen and elastin fibers (Figure 3,
A and B). The abundance of elastin fibers endows the lesion
with a basophilic hue on hematoxylin-eosin staining. The
fibrosis involves the subpleural pulmonary parenchyma, and
larger lesions frequently entrap the airways (Figure 4). It can
be easily surmised that this propensity helps explain the
finding of “air bronchograms” on chest imaging in some
cases (Figure 1). The fibrosis is sharply marginated from the
adjacent uninvolved lung. The overlying pleura frequently
displays mild to occasionally dense fibrosis (Figure 5). In
In some cases, there may be an overlying true pleural plaque with its characteristic hyalinized, basket-weave pattern of fibrosis; however, since there is no other evidence to suggest that PACs are caused by or associated with asbestos exposure, this is presumably an unrelated finding. Occasional subepithelial fibroblast foci (Figure 6) can be seen in PAC, although they are usually not numerous and are typically found in the center of the lesion. Plump, hyperplastic pneumocytes (Figure 7), of the sort commonly associated with fibrosis of any etiology, are often encountered entrapped within the lesion and should not be confused for well-differentiated adenocarcinoma.

Additional features that can be seen include dystrophic calcification, metaplastic ossification, and lymphocytic inflammation, usually of mild degree, which often present as scattered lymphoid aggregates. Although the presence of frank infarct-type necrosis has not been well described in previous reports, in my experience this is not an uncommon finding in PAC, and is compatible with its putative pathogenesis via chronic ischemia.

CAVEATS AND DIFFERENTIAL DIAGNOSIS

It is important to remember that the PAC is both (1) common and (2) not mutually exclusive with other types of pathologic entities. Therefore, it is entirely possible and not uncommon that a PAC may coexist with other pathologic processes. Potential coexistent pathologic entities run the gamut of possibilities, from granulomatous inflammation to malignancy (Figure 8). On a wedge resection, the diagnosis of a “pure” PAC is straightforward, since the entire lesion can be sectioned and examined. More commonly, however, a PAC will be biopsied by core needle via a transthoracic approach. This of course yields far less tissue than a wedge resection. As such, on needle biopsy, as on other small biopsy samples, the problem of sampling error must be kept in mind. The finding of PAC tissue on core needle biopsy may not necessarily explain the clinical and radiologic suspicions in every case. For example, the presence of only PAC tissue on core needle biopsy would not explain a highly metabolically active mass, or one with very rapid growth. Therefore, with small biopsy samples, it is this author’s preference to use the term fibroelastotic scar with pertinent negative findings (“negative for granulomatous inflammation and neoplasm”) rather than “apical cap,” since (1) the apical cap remains an unfamiliar entity with many clinicians and radiologists and (2) it may result in a false sense of assurance given the potential for sampling error.

There are very few other lesions that enter into the differential diagnosis of PAC (see also next section). Chief among these is so-called rounded atelectasis, also termed folded lung or shrinking pleuritis. It is similar to PAC in being a localized fibrotic lesion. However, rounded atelectasis comprises intrapleural fibrosis above the elastic layer of the visceral pleura, whereas PAC is subpleural parenchymal fibrosis. In rounded atelectasis, the elastic layer is wrinkled deceptively similar to pulmonary apical cap (PAC) and, by extension, pleuroparenchymal fibroelastosis (PPFE), and UIP may be misdiagnosed as the latter. However, other areas in this biopsy specimen (not shown) showed the characteristic findings of UIP, including honeycomb fibrosis and spatial and temporal variegation, findings which—in combination—do not occur in PAC or PPFE (hematoxylin-eosin, original magnification ×20).
and infolded. In addition, in contrast to PAC, rounded atelectasis has a predilection for the lower lobe, especially the posterior surface.

**FUTURE DIRECTIONS**

The presence of paucicellular fibrosis rich in basophilic elastic fibrils, microscopically identical to PAC, is shared by another, more recently named process, so-called pleuroparenchymal fibroelastosis (PPFE). However, unlike PAC, PPFE is not localized fibrosis but diffuse, and is currently regarded as a rare form of idiopathic interstitial pneumonia. As such, it comes to clinical attention in a manner similar to other interstitial lung diseases, namely, prominent chest symptoms (such as dyspnea and cough), abnormalities on pulmonary function testing, and evidence of diffuse parenchymal disease on chest imaging. As mentioned, the histologic findings in PPFE are strikingly similar to PAC, and differ only in extent (Figure 9). An additional similarity is the reported preferential involvement of the upper lobes in PPFE. Despite their similarity, it is unclear whether there is any relationship between PAC and PPFE, given the chasm in their clinical presentation. Additional research on this interesting entity is clearly needed. However, whatever relationship the PAC and PPFE turn out to have (if any), I would like to emphasize that much caution should be exercised in the diagnosis of PPFE, especially if it is based on a small or upper lobe–only biopsy specimen. Indeed, in addition to their similar histologic profile, it must be recalled that the PAC is very common and may coexist with other more common interstitial lung diseases than PPFE. In particular, usual interstitial pneumonia may involve the upper lobes and possess abundant elastosis as well as its more typical collagen fibrosis (Figure 10).

**SUMMARY**

First described well over a hundred years ago, the PAC is an anatomically and microscopically distinct form of localized parenchymal scarring. It is believed to be a consequence of intrinsic physiologic underperfusion to which the apical segments of the upper and lower lobes are subject, perhaps compounded by repeated inflammation and low-grade infection, but its pathogenesis remains unproven to this day. Microscopically, its hallmark is paucicellular fibrosis rich in elastosis. It is a common finding in both sexes and its prevalence increases with age. It may be unilateral or bilateral; when bilateral, it is often asymmetric. Pathologists should be familiar with the PAC, since it is occasionally biopsied during the course of the evaluation of lung nodules.

**References**