



Tumors of Uncertain Histogenesis

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This group of tumors, although rare, represents a challenge in diagnosis, especially when confronted with small biopsy specimens. In addition, despite the knowledge generated by immunohistochemistry and molecular techniques, the cell of origin in these tumors for the most part remain somewhat inconclusive. The group of tumors that will be presented in this section includes:

- Clear cell “sugar” tumor (Pecoma).
- Inflammatory myofibroblastic tumor (Inflammatory pseudotumor).
- Pneumocytoma (Sclerosing Hemangioma).
- Granular cell tumor.
- Hemangioblastoma-like clear cell stromal tumor of the tumor.

Clear Cell “Sugar” Tumor

Clear cell tumors of the lung are usually discovered incidentally on chest radiograph or CT. It commonly presents as one or multiple rounded lesions with smooth margins that show enhancement with administration of intravenous contrast. No cavitation

or calcification is seen. On imaging, this entity cannot be differentiated from primary lung cancer or metastasis [1]. However, clear cell tumor of the lung is considered a benign tumor and complete surgical resection appears to be the treatment of choice.

The tumor was originally described in 1963 in an abstract form [2] as a benign clear cell tumor, followed in 1971 with a series of 12 cases [3] in which the name “sugar” tumor was added to the description. Although it may be logical to assume that the name “sugar” came out of the color of the neoplasm, it is likely that the name “sugar” derived from the presence of hexose in one of the cases described. To date that description remains the largest case series and the one in which the histological features of the tumors were outlined. Interestingly, in early reports these particular tumors were found to share similar features with tumors in the uterus and stomach. Currently, this tumor and analogous tumors in other locations including stomach and uterus are considered to be part of the spectrum of Pecomias, which include other conditions with shared immunohistochemical features including positive staining for CD34 and HMB45 [4]. Several theories including Clara cells, melanocytic, and pericytic origin have been presented in the literature to account for this tumor [5–8].

Pathological Features

Grossly, these tumors are rarely over 3 cm in greatest dimension. However, reports document sizes ranging between 1 and 7 cm in greatest diameter. The tumor is well-circumscribed, solid, and varying in color from pink to gray or brown. The tumor in general does not show areas of necrosis or hemorrhage.

Histologically, clear cell tumors, as the name implies, are composed of cells with clear cytoplasm, small nuclei, and inconspicuous nucleoli. The cellular proliferation may be arranged in cords or sheets of cells with intervening thin bands of fibroconnective tissue and dilated vascular structures. The tumor in some areas may show a prominent heman-

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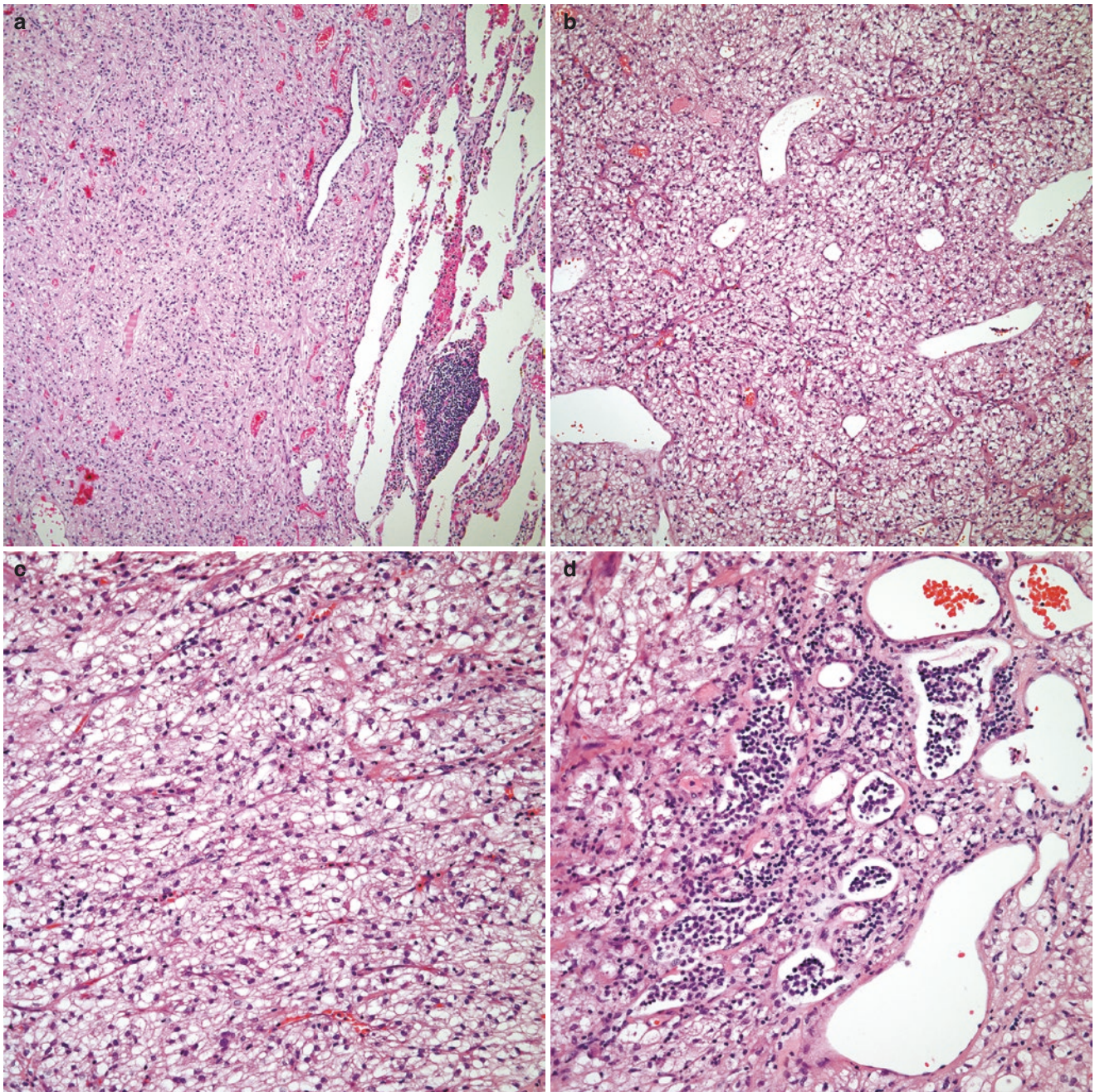


Fig. 16.1 (a) Low power view of clear cell tumor of the lung, (b) the tumor shows striking clear cell features and the presence of dilated vascular structures, (c) diffuse cellular proliferation composed of medium

size cells with clear cytoplasm, (d) focal areas with inflammatory reaction, (e) entrapped areas of airway, (f) focal areas of calcifications

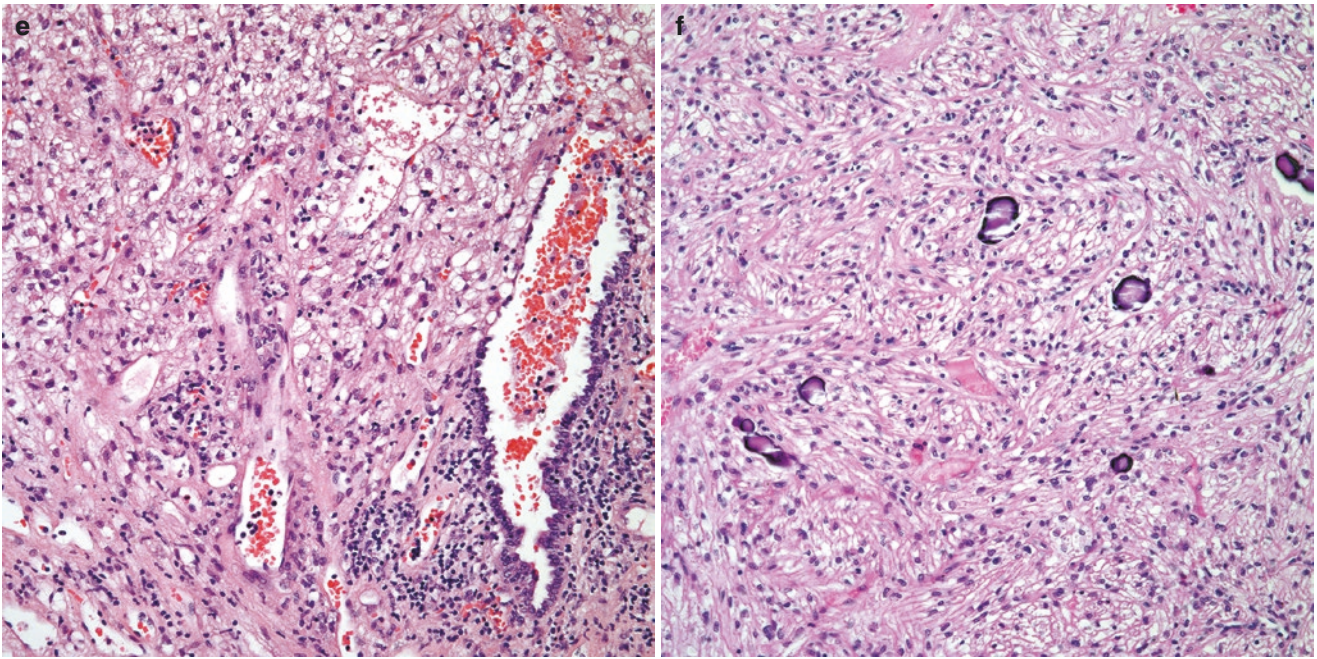


Fig. 16.1 (continued)

giopercytic growth pattern, while in other areas the tumor may show subtle spindle cell morphology (Fig. 16.1a–f). In general, the tumor does not show mitotic activity or cellular atypia.

Histochemical stains using mucicarmine and PAS show that tumor cells are negative for intracellular mucin and positive PAS (glycogen).

The immunohistochemical features of these tumors are rather characteristic in that the tumor may show positive staining for CD-34, CD1a, HMB45, MMB50, NKIC3, HAM-56, cathepsin B, factor XIIIa, neuron-specific enolase (Fig. 16.2a–c). Importantly, the tumor is negative for keratin, EMA, TTF1, and Napsin A.

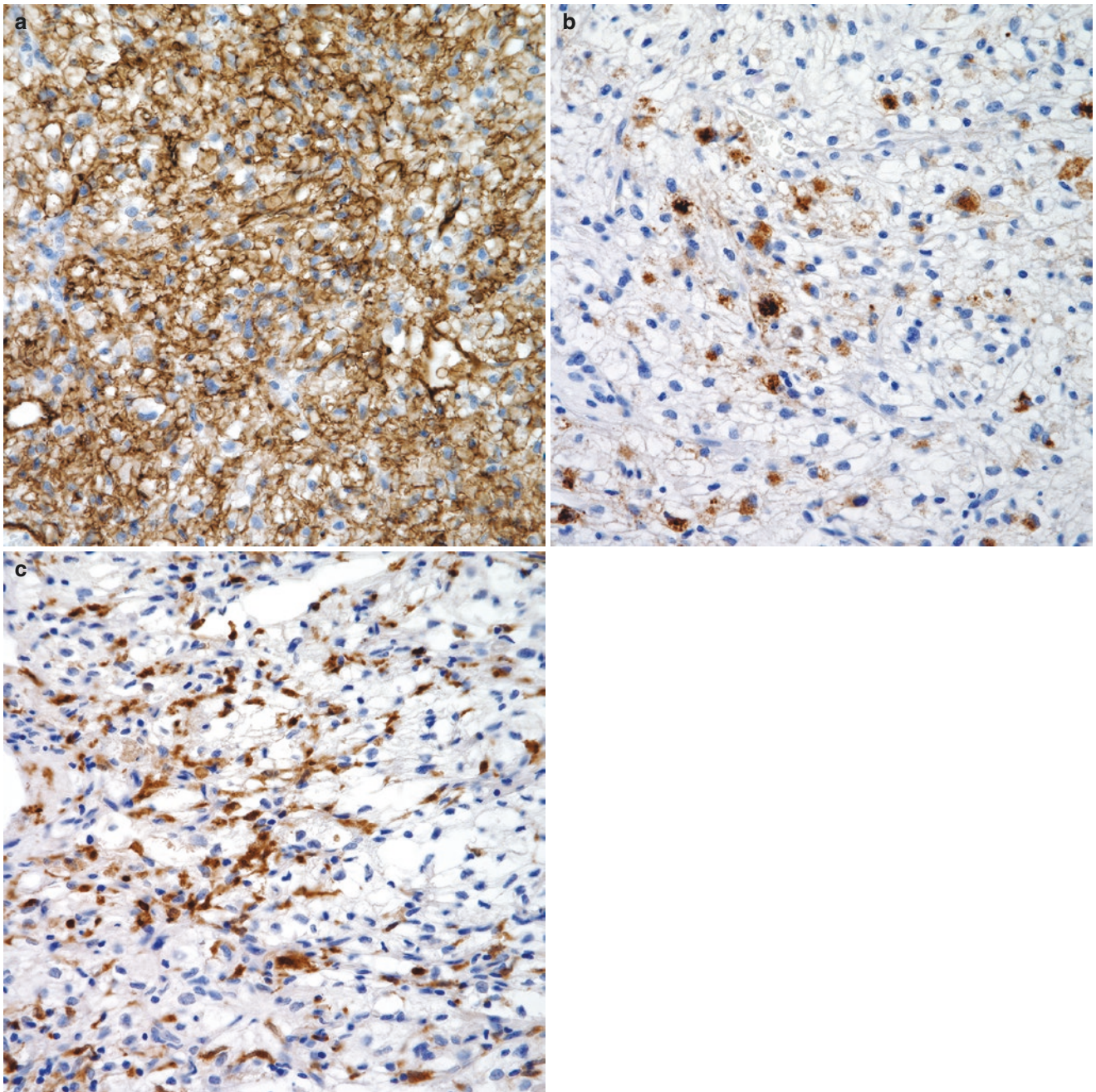


Fig. 16.2 Immunohistochemical positive stains in clear cell tumor of the lung, (a) CD-34, (b) HMB-45, and (c) factor XIIIa

Inflammatory Pseudotumor (Myofibroblastic Tumor)

The term inflammatory pseudotumor is rather controversial, with some authors preferring the designation of myofibroblastic tumor. Past reports of this entity have linked this tumor to possible infections with herpesvirus-8 [9, 10].

In general, these tumors represent no more than 1% of all lung neoplasms. The true incidence is difficult to estimate due to the differences in definition and classification in the past. For instance, other conditions that have been coded under the designation of “inflammatory pseudotumor” include organizing pneumonia and pseudolymphoma. Clinically, these patients do not present with specific symptomatology, and in many cases, the patients are completely asymptomatic. However, clinical symptoms that have been reported include hemoptysis [11]. In one of the largest series [12] of these tumors, (61 patients), the patients ranged from 17 to 61 years of age with more tumors in men than in women (36:25). The treatment of choice for these tumors is complete surgical resection and survivals at 5 years is 91%. The mortality estimates have not been directly linked to the lesion [13]. In cases with incomplete surgical resection, recurrence may occur.

Pulmonary inflammatory pseudotumor appears as a solitary well-circumscribed and lobulated nodule or mass usually

localized to the lower lobes on chest radiograph or CT [14]. The lesions are multiple in 5% of cases [15]. They show homogeneous or heterogeneous enhancement on contrast-enhanced CT and may be associated with atelectasis and/or pleural effusion [16] (Fig. 16.3). Amorphous or dystrophic calcifications are seen more frequently in children than adults [17]. These lesions can be FDG avid on PET/CT and this parameter can be used to monitor response to therapy [18].

Pathological Features

Grossly, these tumors may be central or peripheral, which likely determines the symptomatology in these patients. They are well circumscribed but not encapsulated, yellowish, and slightly lobulated (Fig. 16.4).

Histologically, based on the literature, it appears that the histology of these tumors in the lung can show two different features (1) predominantly plasma cells and (2) fibrohistiocytic [19–21]. In the plasma cell variant, the characteristic feature is the presence of plasma cells admixed with areas of hyalinization and calcification (Fig. 16.5a–e). However, these histological characteristics are unusual and the majority of inflammatory pseudotumors belong to the category of myofibroblastic tumors. They are characterized by the presence of a spindle cell

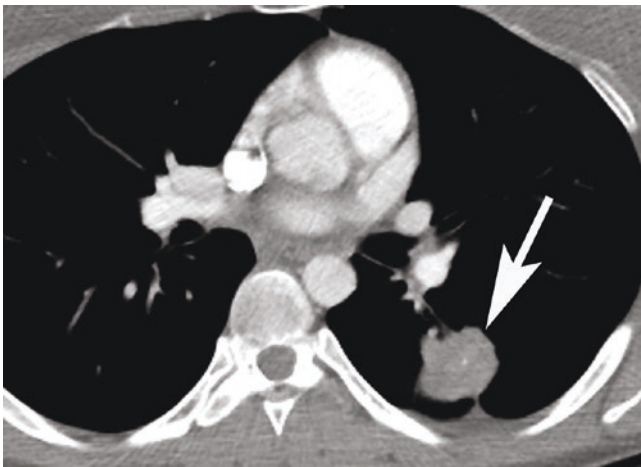


Fig. 16.3 Inflammatory myofibroblastic tumor. Contrast-enhanced CT showing soft tissue mass in the left lower lobe (arrow)



Fig. 16.4 Centrally located inflammatory pseudotumor showing a well-circumscribed tumor mass with yellowish color

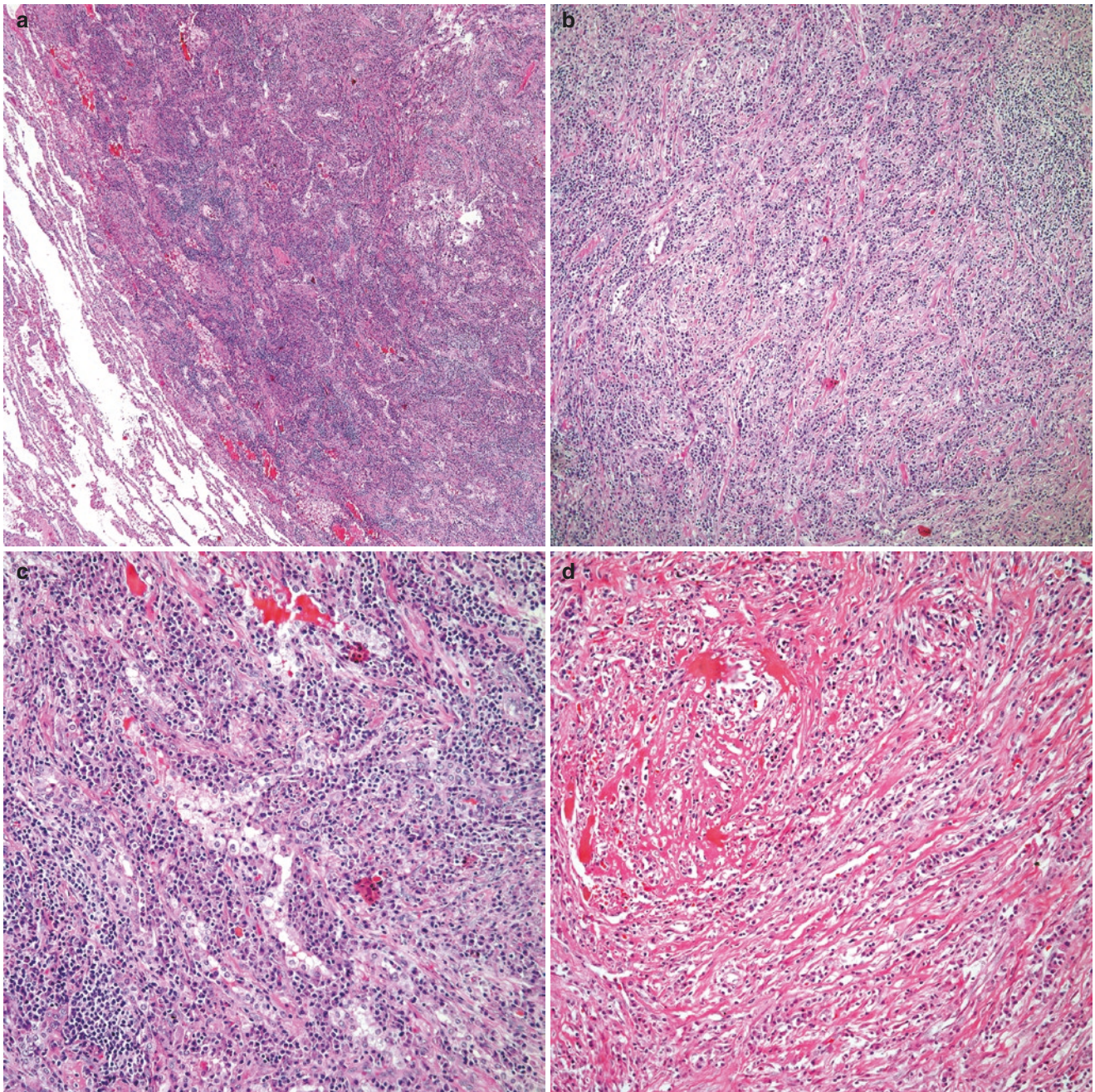


Fig. 16.5 (a) Low power view of an inflammatory pseudotumor—plasma cell variant, (b) plasma cell proliferation with intervening fibrocollagen, (c) plasma cells admixed with collections of xanthoma cells, (d) extensive hyalinization, (e) focal areas of ossification

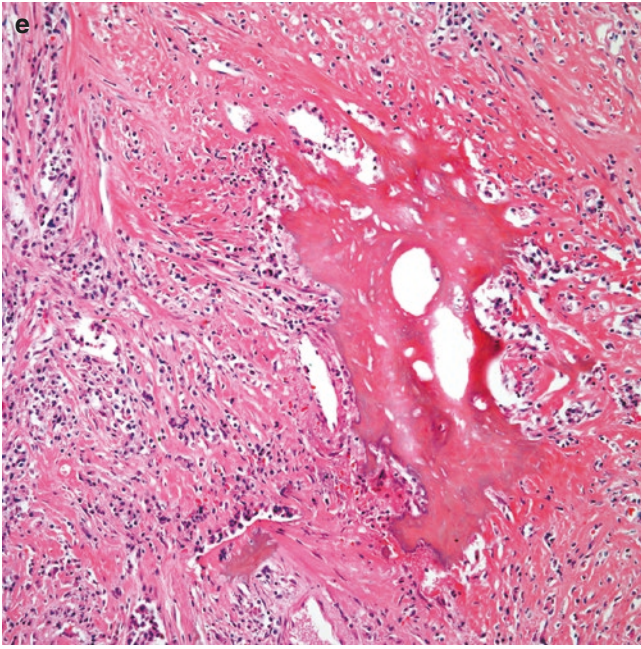


Fig. 16.5 (continued)

proliferation with a subtle storiform growth pattern. Admixed with this spindle cell proliferation, plasma cells are almost invariably encountered. In addition, the spindle cell proliferation alternates with clusters of xanthoma cells and Touton-type giant cells, characteristic features for the diagnosis of this tumor (Fig. 16.6a–g). In general, these tumors do not show increased mitotic activity or cellular atypia.

The immunohistochemical features of these tumors are also not completely specific, as the tumors may show positive staining for actin, vimentin, CD-68, CD-138 (plasma cells). More recently, the use of AKL-1 and p80 has become an important tool for the diagnosis of these tumors [22]. However, the positivity varies and has been stated as less than 40%, while the use of AKL rearrangement by FISH may be just slightly better with 45% of the cases. At least 50% of these tumors will show negativity for ALK and p80. In general, these tumors are negative for EMA, keratin (positive in entrapped alveolar cells) and neuroendocrine markers, TTF-1, p40, p63, keratin 5/6, and Napsin A.

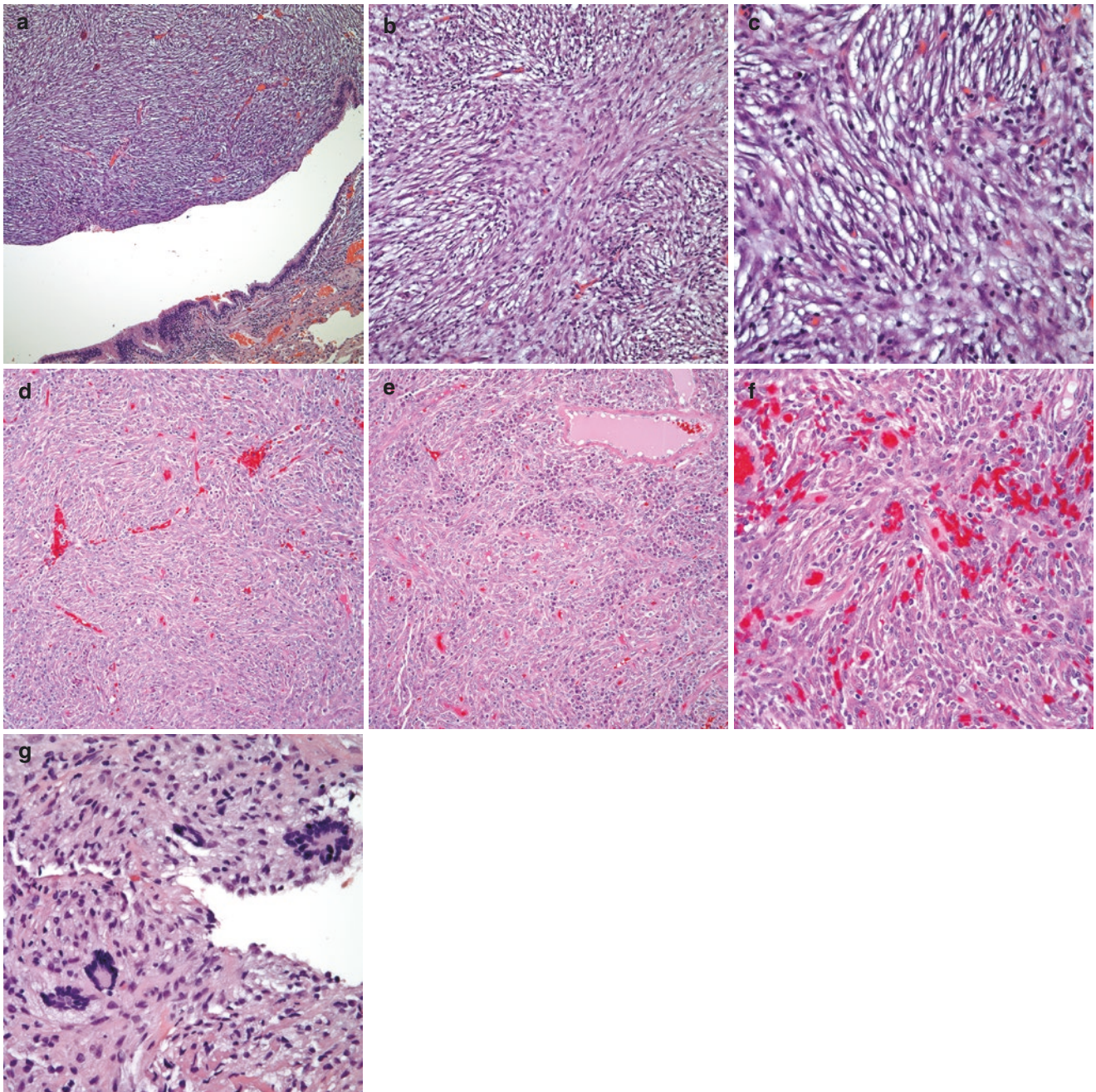


Fig. 16.6 (a) Centrally located inflammatory pseudotumor – myofibroblastic tumor variant, (b) spindle myofibroblastic proliferation, (c) spindle cells with lack of nuclear atypia or mitotic activity, (d) inflammatory pseudotumor with more fibrohistiocytic appearance, (e) spindle

cells admixed with plasma cells, (f) higher magnification of the spindle cells without mitotic activity or marked nuclear atypia, (g) fibrohistiocytic variant of inflammatory pseudotumor (inflammatory myofibroblastic tumor) with presence of Touton-type giant cells

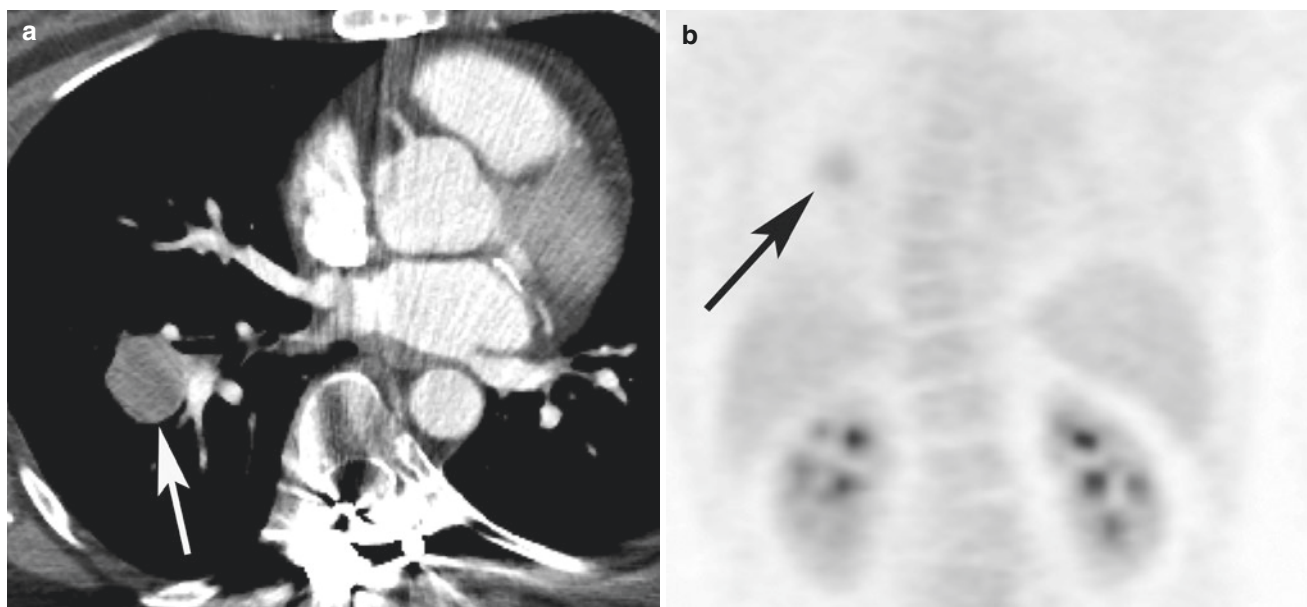


Fig. 16.7 Contrast-enhanced CT (a) showing a soft tissue nodule in the right lower lobe (arrow). PET MIP image (b) shows mild FDG uptake in the nodule (arrow)

Pneumocytoma (Sclerosing Hemangioma)

The original description of this tumor dates to 1956 when Liebow and Hubbell noted the similarities of this tumor with a similar skin tumor coined the term “sclerosing hemangioma” for this particular entity. Interestingly, since its first description, several manuscripts in the literature attempted to explain the origin of this neoplasm, positing mesothelial, histiocytic, and/or vascular origin. Currently, with the aid of immunohistochemical markers, this tumor is considered of pneumocytic origin, thus the current designation of Pneumocytoma [23–30].

The tumor appears to be most common in young female patients who may be completely asymptomatic; their tumor is found during routine chest films. On rare occasions, the tumor has been described in children, and in a few cases, the tumors have been reported as involving lymph nodes. Surgical resection is the treatment of choice and appears to be curative.

By imaging, usually Pneumocytoma (sclerosing hemangioma) is a solitary, well-defined mass on chest radiograph. CT is generally performed to further characterize the lesion. On CT, sclerosing hemangioma manifests as a round or ovoid shape nodule or mass with a smooth margin, commonly subpleural in location. Calcification is variable but may be present in a minority of tumors; cystic degeneration can be seen in large tumors although cavitation is uncommon. Marked contrast enhancement is typical, usually homogeneous in smaller tumors and heterogeneous in larger tumors, presumably due to cystic areas. There have been some reports of a lucent zone

around the lesion on chest radiographs (air meniscus sign), which have subsequently been confirmed on CT as a region of air trapping [31–33]. The MRI appearance is variable, depending upon cellular, fibrotic, hemangiomatic, and cystic components [32]. The tumor usually shows low-to-moderate uptake on FDG PET/CT imaging [33] (Fig. 16.7a, b).

Pathological Features

Grossly, these tumors are rarely more than 3 cm in greatest dimension; however, reported cases have varied in size from 1 to 5 cm. The tumor may appear hemorrhagic but not necrotic. It is well circumscribed but not encapsulated. The tumor most often is a single peripheral lung lesion.

Histologically, the presence of hemorrhagic areas as well as vascular-like structures mimicking a vascular neoplasm led to the initial report of pneumocytoma as a possible vascular neoplasm, as previously mentioned. However, the tumor may show a variety of growth patterns including (1) papillary, (2) sclerosing, (3) solid, and (4) vascular like (Fig. 16.8a–g). Some tumors may show all these patterns within the same lesion while in other tumors, one pattern may predominate over the others. In general, at higher magnification, the presence of two types of cells is apparent: (A) surface cuboidal cells that line papillary areas and (B) stromal cells composed of oval or spindle cells. Collections of foamy macrophages, scattered giant cells, or cholesterol clef granulomas may be seen in some cases.

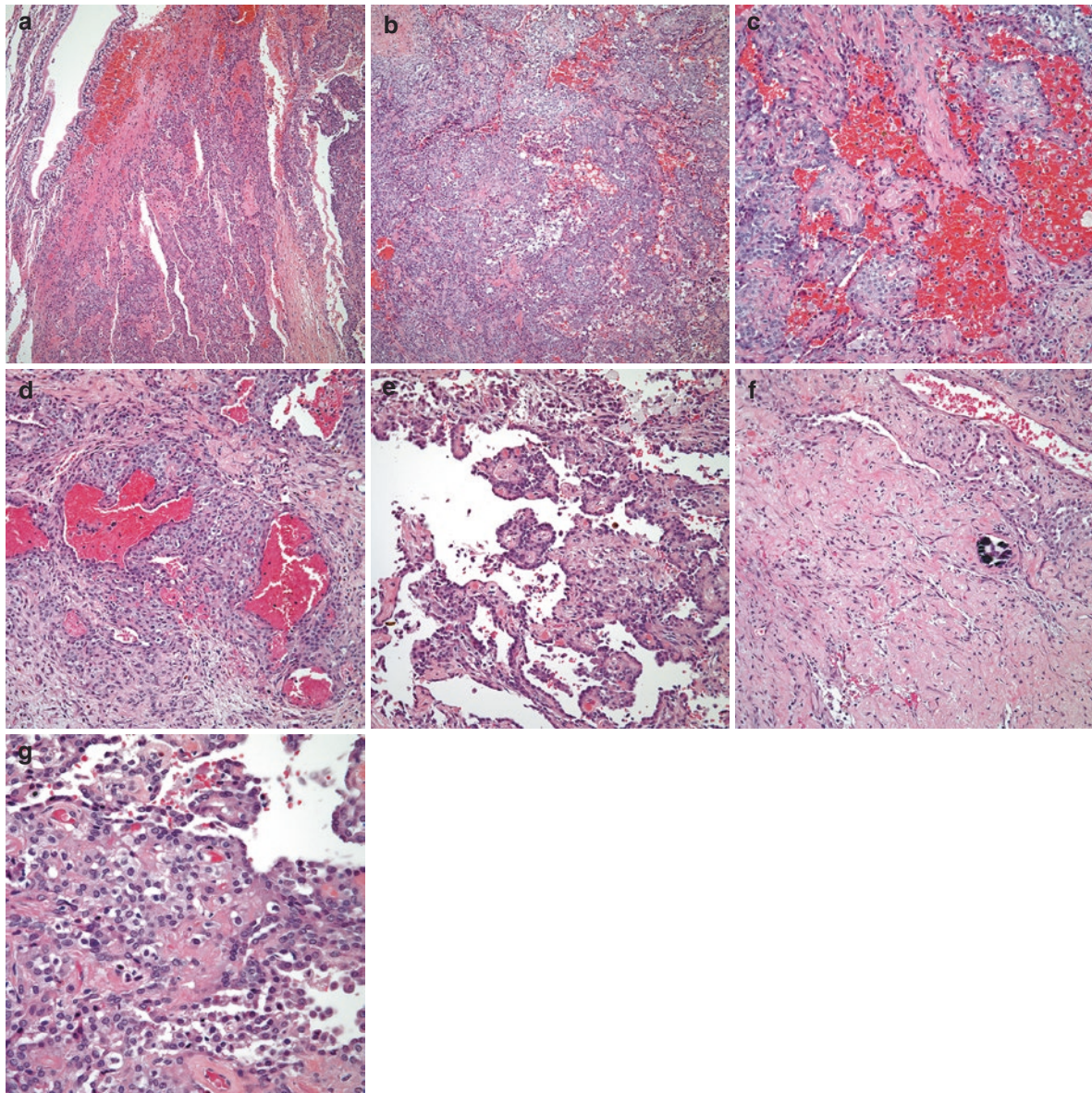


Fig. 16.8 (a) Low power view of a pneumocytoma, (b) solid component of the tumor admixed with foamy macrophages, (c) areas of hemorrhage, (d) vascular-like structures, (e) papillary proliferation, (f)

extensive areas of sclerosis with focal calcifications, (g) two types of cells—cuboidal and stromal cells

Immunohistochemical stains may aid in supporting the diagnosis of these tumors. Positive staining for low-weight keratin CAM 5.2 is seen mainly in the cuboidal cells, EMA mainly in the stromal cells, TTF-1 predominantly in stromal cells, and surfactant apoprotein predominantly in cuboidal cells (Fig. 16.9a–d). Other markers that may show positive staining include napsin

A, beta-catenin, clear cell antigen, S-100 protein and, in some unusual cases, p40 and p63. Due to the extensive positive immunoprofile that these tumors may show, it is important to correlate the immunohistochemical findings with the morphology of the tumor. Vascular markers such as CD31, CD34, and factor VIII are negative in these tumors.

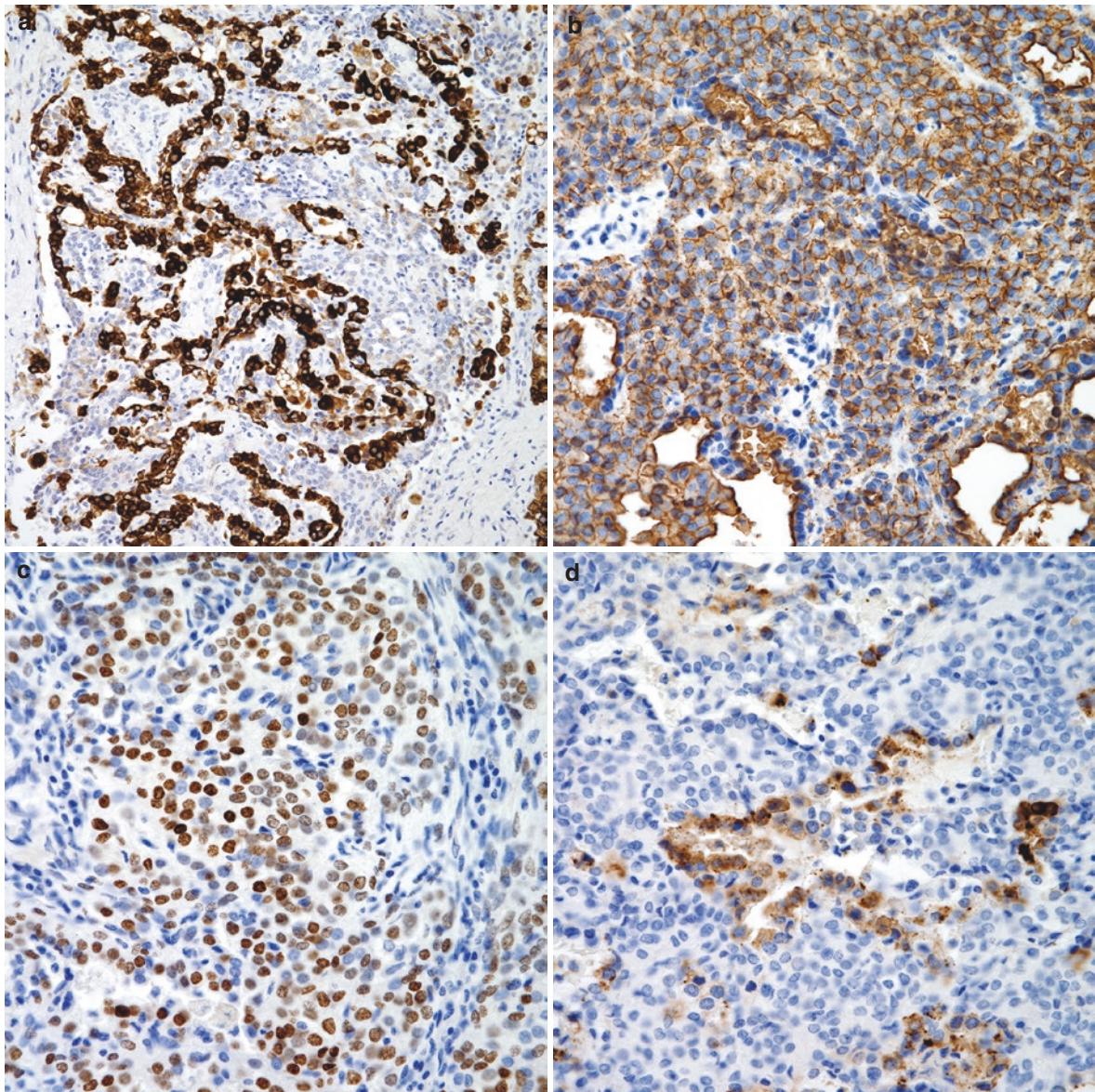


Fig. 16.9 (a) Keratin CAM 5.2-positive predominantly in cuboidal cells, (b) epithelial membrane antigen (EMA)-positive predominantly in stromal cells, (c) TTF-1 positive, (d) surfactant apoprotein positive mainly in cuboidal cells

Granular Cell Tumor

This tumor has ubiquitous distribution and the lung is among the most unusual locations as a primary neoplasm. It is considered a benign tumor and although there is a malignant counterpart, the benign tumor is the one that is much more common in the lung. Originally, the tumor was considered to be of muscle origin and was reported as myoblastoma, while other authors have considered it to be of Schwann, fibroblastic, or histiocytic origin.

Since the majority of these tumors have a central location, patients often have symptoms of cough and dyspnea. However, there are no specific symptoms [34–37]. More often, the tumor is identified during routine chest films for other reasons. The tumor in the lung more often is a single lesion; however, in other locations multiple tumors have been identified. By diagnostic imaging, this tumor cannot be distinguished from other more common bronchial neoplasms. Surgical resection is the treatment of choice for these tumors and appears to be curative.

Pathological Features

Grossly, the central tumor may present as a polypoid lesion obstructing airway or be in endobronchial location. Generally, they are less than 3 cm in largest diameter but tumors up to 5 cm have been described. In general, the tumor shows a whitish color and firm consistency and does not show hemorrhage or necrosis.

Histologically, the tumors show sheets of medium-size cells with ample granular eosinophilic cytoplasm, round small nuclei, and inconspicuous nucleoli. The tumor does not display cellular atypia or mitotic activity (Fig. 16.10a–e). The presence of cellular atypia or mitotic activity coupled with areas of necrosis should alert to the possibility of a malignant granular cell tumor. However, in some cases, despite the benign appearance of the tumor, the tumoral cells may involve adjacent peribronchial lymph nodes, which should not be construed a sign of malignancy.

Immunohistochemical stains may aid in the diagnosis, as these tumors are positive for S-100 protein. Other immune markers that have been reported positive include actin,

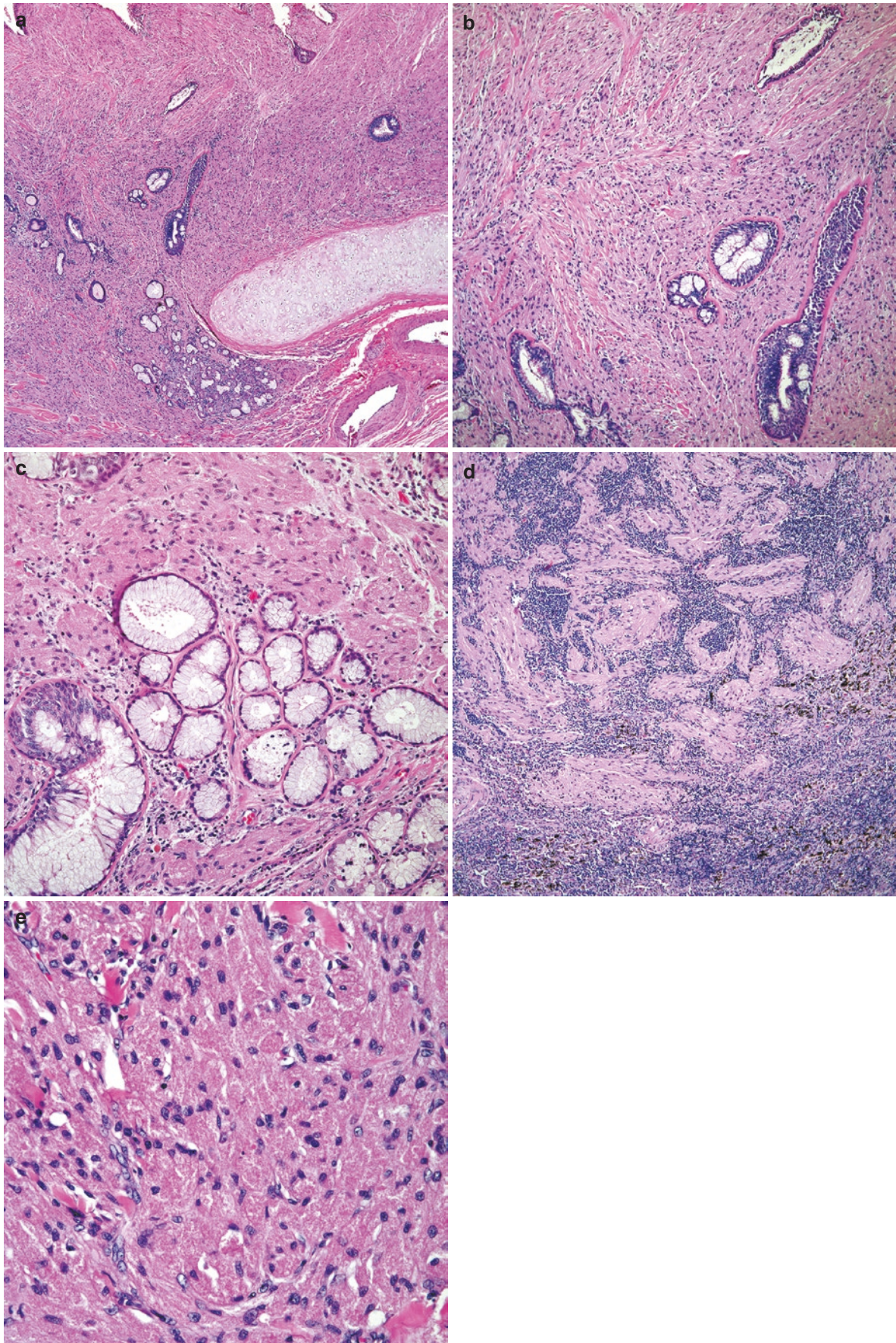


Fig. 16.10 (a) Low power view of a centrally located granular cell tumor, (b) tumor in endobronchial location adjacent to endobronchial glands, (c) tumor present but not involving mucous endobronchial

glands, (d) tumor extends to adjacent peribronchial lymph node, (e) cellular proliferation without atypia or mitotic activity

neuron-specific enolase, vimentin, myelin basic protein, cathepsin B, Leu-7, and anti-chymotrypsin. In general, granular cell tumors are negative for desmin, EMA, CEA, keratin, and TTF-1.

Hemangioblastoma-like Clear Cell Stromal Tumor

This is relatively a new entity described in adult patients. In the series by Lindholm [38], the patients were four women and one man with an average age of 45 years. The patients presented with non-specific symptoms including cough, dyspnea, and chest pain.

Diagnostic imaging in those tumors was described as an intrapulmonary mass without any specific features and, likely due to the rarity of the neoplasm, there are no specific radiological details. All the patients were treated by surgical resection and no documented recurrences or metastasis were reported.

Pathological Features

Grossly, the tumors appear to be well circumscribed but not encapsulated with focal areas of hemorrhage. None of the tumors described was above 3 cm in greatest diameter.

Histologically, the tumor is characterized by the presence of a neoplastic cellular proliferation in either solid or nested growth pattern alternating with dilated vascular spaces. In some cases, focal areas of hemorrhage are also present. The cells are of medium-size with clear to light eosinophilic cytoplasm, ovoid nuclei, and inconspicuous nucleoli (Fig. 16.11a–e). The tumor characteristically does not show marked cellular atypia or mitotic activity.

The immunohistochemical profile of these tumors is rather non-specific as the tumor shows negative staining for epithelial markers (keratin and EMA) as well as neuroendocrine, vascular, neural, muscle, and melanocytic markers. The only marker that is consistently positive is vimentin.

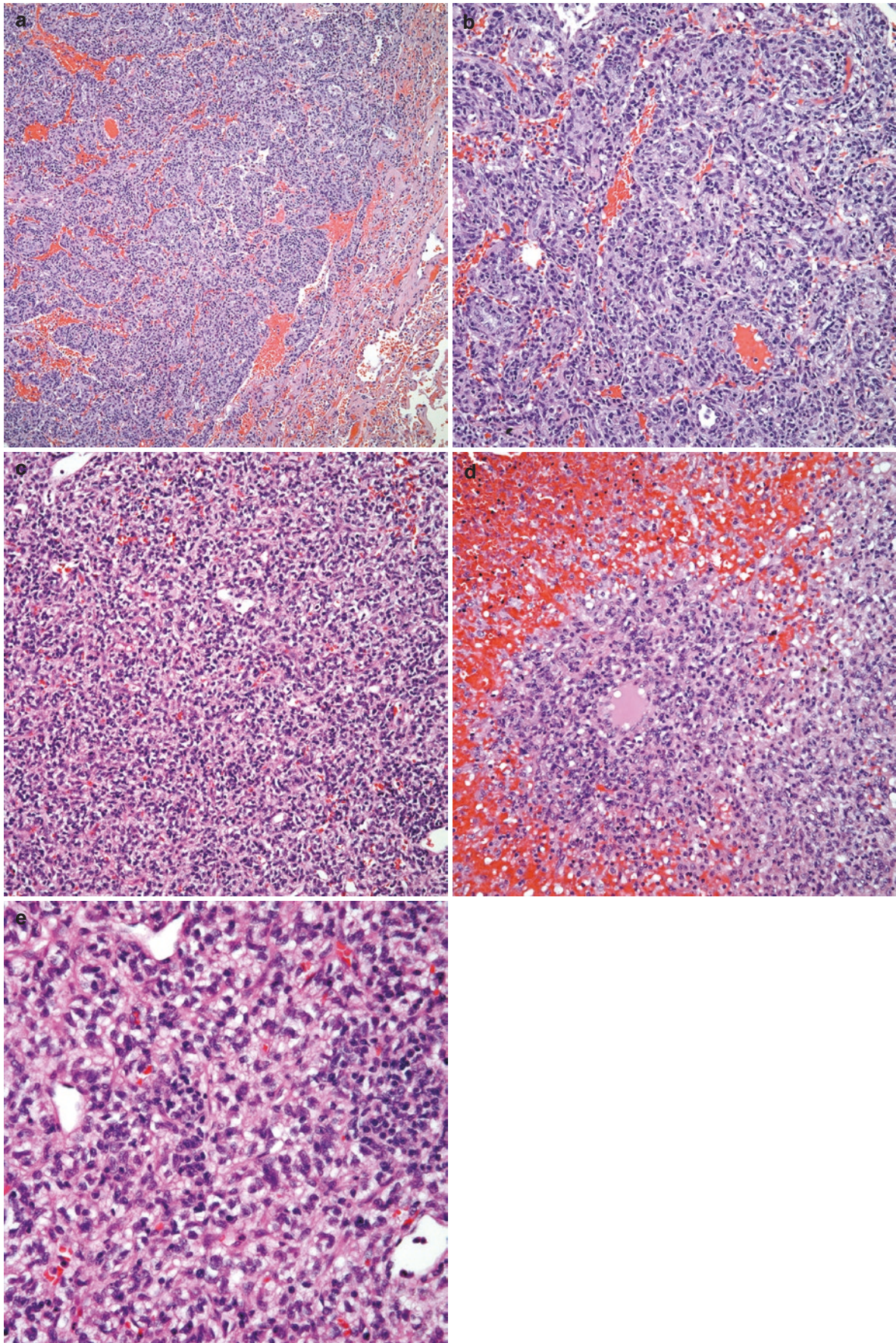


Fig. 16.11 (a) Low power view of a hemangioblastoma-like clear cell stromal tumor of the lung, (b) subtle nested growth pattern, (c) solid growth pattern with cells with clear cytoplasm and dilated vascular

spaces, (d) focal areas of hemorrhage may be seen in some cases, (e) higher magnification showing medium size cells with clear cytoplasm and ovoid nuclei, note the absence of cellular atypia and mitotic activity

Summary

This group of tumors, although unusual, poses a significant problem in diagnosis mainly when dealing with small biopsies. In the majority of cases, the diagnosis is established after surgical resection. Although diagnostic imaging plays an important role in determining the location of the tumor, there are no radiological details that correlate to a specific diagnosis. Even immunohistochemical analysis needs to be correlated with the morphological features of the tumor, and in some cases, large panels of immunohistochemical stains provide little support in the diagnosis except by excluding other more common neoplasms.

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