CT assessment of unusual primary lung tumors and pathologic correlation

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Learning objectives

Exhibiting some notions about epidemiology, radiological semiology and pathological correlation of some unusual primary lung tumors.

Background

There is a number of unusual primary lung lesions, both neoplastic and pseudotumoral that are part of the differential diagnosis of a pulmonary nodule or mass. The study of these lesions by CT allows definition of its radiological characteristics. Although there are no specific images of each one, some typical findings can be found such as the presence of fat in exogenous lipoid pneumonia.

We performed a retrospective study, collecting the cases seen in our hospital since 1996. We got a large iconography and we correlated radiological semiology and histological findings.

We collected 31 cases classified in 5 groups: tumors of the lymphoreticular system (5 low-grade small B-cell Lymphoma of BALT origin, 1 angiotropic lymphoma), neuroendocrine tumors (6 typical carcinoid, 2 atypical carcinoid, 5 large cell neuroendocrine carcinoma, 1 squamous carcinoma combined -large cell and 1 mixed large and small cell carcinoma), epithelial tumors (2 sarcomatoid carcinomas, 1 adenosquamous carcinoma), mesenchymal tumors (3 myofibroblastic inflammatory tumors, 1 pulmonary blastoma, 1 fibrosarcoma) and pseudo (1 primary pulmonary amyloidosis, 1 lipoid pneumonia).

Imaging findings OR Procedure details

1.-TUMORS OF THE LYMPHORETICULAR SYSTEM.

1.1-Low-Grade Small B-Cell Lymphoma of BALT Origin (Pseudolymphoma)

It’s a slow-growing neoplasm (before called benign pseudolymphoma). It’s most common primary lymphoma. It affects both sexes equally, between 25-85 years of age. About 50% of cases are asymptomatic at the time of diagnosis (incidental finding). Prognosis is
usually good, and depends on predominant cell type and stage of disease. The treatment is local excision with a recurrence of 10-15%.

**Imaging findings.**

They appear as nodules or nodular consolidation (Fig.1 and Fig.2), or, more commonly, as multiple nodules (Fig.4). The diffuse involvement of the axial and interlobular interstitium, and lobar infiltrates are other possible presentations. These opacities are usually centered around a bronchus, and CT image shows presence of air bronchogram in them.

**Histologic findings.**

Whitish masses without necrosis, with infiltrative and non-destructive growth. The normal architecture of the lung is preserved.

Microscopic examination usually shows a pattern typical lymphoepithelial: epithelial cells infiltrated by lymphocytes (Fig.3).

1.2- Intravascular large B-cell lymphoma.

Also referred to as malignant angioendoteliosis or angiotropic lymphoma. It is usually a B-cell lymphoma, characterized by massive proliferation of neoplastic mononuclear cells within the capillaries, arterioles and venules. The obstruction of small blood vessels by lymphoma cells is responsible for the clinical multiorgan ischemic manifestations. The prognosis is very poor.

**Imaging findings.**

Semiology is very varied. The most common findings are the presence of areas of "ground glass" consolidations with air bronchograms, centrilobular ill-defined nodules etc (Fig.5).

**Histologic findings.**

Microscopic examination of the sampled tissue revealed an intravascular capillary and small vessel accumulation of large atypical pleomorphic mononuclear cells, morphologically lymphoid in origin (Fig.6).

2.-NEUROENDOCRINE TUMORS.
Neuroendocrine tumors represent 25% of all lung neoplasms. Small cell tumors are the most frequent.

2.1.- PULMONARY CARCINOID TUMORS.

Pulmonary or bronchial carcinoid tumors account for over 25% of all carcinoid tumors and for 1%-2% of all pulmonary neoplasms. About 80%-90% of pulmonary carcinoids are typical carcinoid, and 10-20% are atypical carcinoids.

2.1.1.- Typical carcinoid.

It affects males and females equally with a mean age of 40-49 years. Most of them are central tumors, causing early obstructive symptoms (a high percentage are diagnosed at stage I). These tumors are not associated with cigarette smoking. The overall 5-year survival rate for typical carcinoids is 87%.

Imaging findings.

Well-defined round or ovoid nodule or mass with lobulated margins (Fig. 7). On CT images calcification appear in up to 30% of tumors as a punctate or diffuse pattern. Although uncommon, these lesions may cavitate especially the larger ones. Typical carcinoids may cause narrowing, deformity or bronchial obstruction (Fig. 9 and fig.10). Occasionally, these tumors manifest as a small nodule within the bronchial lumen. They are usually vascular tumors with a large enhancement (more than 30 UH).

Histologic findings.

Irregular lesion, whitish-gray, with homogeneous appearance. The microscopic study show small nests or interconnected trabeculae of uniform cells with round nuclei and scant cytoplasm, separated by a prominent vascular stroma with numerous thin-walled blood vessels (Fig.8). These tumors do not show necrosis, atypia or significant mitotic activity (<2 mitosis/10 high-power fields).

2.1.2.- Atypical carcinoid.

It affects men more frequently than women. The mean age is 50-59 years. They tend to be peripheral lesions and cause symptoms later than typical carcinoids (approximately 50% of atypical carcinoids are stage I). Most of the patients have a history of cigarette smoking. The overall 5-year survival rate for atypical carcinoids is 56%.
Imaging findings.

Both typical and atypical carcinoids have similar radiological features.

Atypical carcinoids are larger than typical carcinoids and they have a peripheral location more often than typical carcinoids. These tumors may be associated with hilar and mediastinal lymphadenopathy secondary to recurrent pneumonias, or neoplastic involvement (Fig. 11 and Fig. 12).

Histologic findings.

Irregular, grayish brown with areas of hemorrhage. Compared with typical carcinoid, atypical carcinoid has a greater cytologic pleomorphism, more areas of tumor necrosis, increased cellularity and architectural irregularities, and higher mitotic activity (2-10 mitosis/10 high-power field) (Fig. 13).

2.2.- LARGE CELL NEUROENDOCRINE CARCINOMA (LCNEC)

Men are more frequently affected than women. The mean age is 60-69 years. The overall 5-year survival rate for atypical carcinoids are 13-45%.

Imaging findings.

The radiologic features are a spectrum between the atypical carcinoid and small cell tumor, with nonspecific findings. LCNEC tend to be central tumors (Fig. 14): well-defined and, lobulated masses or nodules with air bronchogram, cavity, necrosis or bubble lucency. Calcifications are seen in 9% of cases. 24% of LCNEC are presented with pleural effusion. Mediastinal involvement is common.

Histologic findings.

Heterogeneous and brown mass, with necrotic areas. These tumors are composed of large polygonal cells with granular eosinophilic that exhibit organoid nesting, palisading, trabecular, and rosette-like growth patterns with a high mitotic rate (> 10/10 high power fields) and a conspicuous tendency for necrosis (Fig. 15).

2.3.- MIXED TUMORS.

These neuroendocrine lung tumors are unusual lesions. Histologic findings show different cell types that make up the tumor (Fig. 16). Imaging findings tend to be nonspecific, because these tumors have common features of several tumors. As an example of them,
we present a case of a neuroendocrine mixed small cell/large cell tumor (Fig. 17 and Fig.18).

3. EPITHELIAL TUMORS.

3.1. Sarcomatoid carcinoma.

A very uncommon malignant lung tumor. It is a biphasic neoplasm with a mixed epithelial/mesenchymal growth pattern. It is more common in men than in women. These tumors are strongly associated with smoking. It may manifest as peripheral lung mass without symptoms (incidental finding) or as an endobronchial mass with obstructive symptoms (early diagnosis). The 2-year survival rate is 2-5%.

Imaging findings.

These tumors can be found as a peripheral solid mass of irregular margins without calcifications, or as an endobronchial lesion (Fig. 19 and Fig.20).

Histologic findings.

Heterogeneous irregular and grayish mass. Histologic findings show connective tissue infiltrated by solid individual cells or groups of cells with dense, misshapen and bulky cores. The epithelial component is well defined while the mixed component has a marked pleomorphism, findings consistent with an anaplastic tumor (Fig.21).

3.2. Adenosquamous carcinoma.

Adenosquamous carcinoma is a type of tumor that contains two types of cells: squamous and gland-like cells. These tumors usually show an endobronchial growth with obstructive symptoms.

Imaging findings.

Hilar mass that may have a polypoid growth into the bronchial lumen and invasion of cartilage. The central necrosis and cavitation are common (Fig.22).

Histologic findings.

There are a mixed component, keratinized squamous cells and mucous glandular cells (Fig.23).
4. - MESENCHYMAL TUMORS.

4.1. - Inflammatory myofibroblastic tumor.

Inflammatory myofibroblastic tumor of the lung (also known as plasma cell granuloma, inflammatory pseudotumor, fibrous histiocytoma, fibroxanthoma, and xanthogranuloma) includes a spectrum of pulmonary lesions. It is currently unclear whether these lesions represent a primary inflammatory process versus a low-grade malignancy with a prominent inflammatory response. Similar lesions can also develop in the gastrointestinal tract, salivary glands, mediastinum, trachea and bronchial wall.

Both sexes and any age group appear to be equally affected.

Surgical resection is recommended as diagnosis and treatment.

Imaging findings.

CT findings are variable and nonspecific. Unique mass with lobulated or spiculated margins, variable size and slow growth, that most often affects the lower lobes (Fig. 24 and Fig.25). Sometimes it is detected postobstructive pneumonitis or atelectasis. Calcification is rare, more common in children.

Histologic findings.

Whitish-yellow, well circumscribed and not encapsulated mass. Microscopic examination shows a polyclonal infiltration of normal plasma cell with other inflammatory cells such as lymphocytes, histiocytes, myofibroblasts and neutrophils (Fig.26).

4.2. - Pulmonary blastoma.

Pulmonary blastoma is a rare type of non small cell lung cancer. Less than one in a hundred (1%) of all non small cell lung cancers are pulmonary blastomas. It shouldn't be confused with pleuropulmonary blastoma, a different condition that mainly affects children. Pulmonary blastoma tends to affect people at a younger age than those who develop more common types of lung cancer. The average age at diagnosis is between 40 and 60 years of age, but it is also diagnosed in older people. It also seems to affect women more often than men. It generally has a poor prognosis because most of them have distant metastases.

Imaging findings.
It usually appears as a unique and well-defined pulmonary mass, with peripheral location, but sometimes appears as multiple masses. These lesions may cavitate and are sometimes accompanied by pleural effusion (Fig.27).

**Histologic findings.**

Whitish mass with embryonic bronchial structures identified in a background of immature sarcoma.

It is characterized histologically by a primitive, variably mixed blastematous and sarcomatous appearance (Fig.28).

**4.3.- Pulmonary sarcoma.**

Primary pulmonary sarcomas are extremely rare tumors. Most of them are metastasis from extrathoracic tumours. It is necessary to exclude pulmonary metastatic disease before the diagnosis.

Primary pulmonary sarcomas are usually fibrosarcomas, leiomyosarcomas or sarcomas of the pulmonary artery.

**Imaging findings.**

Solitary pulmonary mass or nodule or endobronchial lesion that usually produces atelectasis such as bronchogenic carcinoma. Angiosarcomas are intravascular lesions.

**Histologic findings.**

We show the histology of metastatic fibrosarcoma, with spindle-shaped and with elongated nuclei without marked pleomorphism. Mitotic index is not very high, and there is minimal stroma invasion with plasma cells (Fig.29).

**5.- PSEUDOTUMORS.**

**5.1.- Pulmonary nodular amyloidosis.**

Amyloidosis is characterized by abnormal deposition of material consisting of light chain proteins of a monoclonal inmunoglobulin.

Pulmonary amyloid deposition not associated with systemic disease occurs in three anatomic distributions: tracheobronchial, parenchymal, and senile (or diffuse).
Nodular pulmonary amyloidosis is a limited form of amyloidosis. It is characterized by single or multiple intrapulmonary nodules or masses and is also known as amyloidoma. These may be mistaken as a tumour.

It usually affects patients around 60 years. Patients more often are asymptomatic, so the diagnosis is an incidental finding. Surgical resection is diagnostic and curative because pulmonary nodular amyloidosis it is a localized form of amyloidosis.

**Imaging findings.**

Imaging of nodular amyloidosis shows solitary or multiple nodules with a smooth or lobular contour. Nodules are often seen in a subpleural or peripheral location (Fig.30 y Fig.31). Calcification may occur in up to 50% of nodules seen at CT scan. Amyloidoma usually occur in the lower lobes.

**Histologic findings.**

Amyloid nodules are either yellow, gray, or pale tan. The nodules range in size from 0.6 to 15 cm. Larger lesions may cavitate or exhibit hemorrhage, necrosis, fibrosis, or calcification.

Microscopic features: Amyloid appears as an amorphous sheet of eosinophilic extracellular material that surrounds vessels.

The protein takes up Congo red stain and exhibits apple-green birefringence at polarized microscopic analysis.

**5.2. - Exogenous lipoid pneumonia.**

Exogenous lipoid pneumonia, which results from aspiration of oil (animal, vegetal or mineral oil), presents a difficult diagnostic problem for both the clinician and the radiologist, because it may mimic many diseases, particularly lung tumors. Most of the patients are asymptomatic. Others present with symptoms of dyspnea, cough and fever. Acute aspiration of large amounts of oil, is very rare, but causes a marked respiratory distress.

**Imaging findings.**

CT imaging shows consolidations or pulmonary nodules / masses, usually located in the LLII (fig. 32), and "crazy-paving" pattern in HRCT.

Puede condicionar la presencia de consolidaciones o de nódulos/masas, habitualmente localizados en los LLII. También se ha descrito el "patrón en empedrado" en los exámenes de TCAR.
CT imaging show fat deposits (with negative attenuation) within the pulmonary opacities.

**Histologic findings.**

Histology demonstrates lipid laden macrophages in the alveolar walls and interstitium, with association of lipid material, inflammatory cells and variable fibrosis.

**Images for this section:**

*Fig. 1:* Low-Grade Small B-Cell Lymphoma of BALT Origin. Chest radiograph: large consolidation in middle and right upper lobes such as pulmonary mass. Smaller consolidations are seen in right lower lobe and left upper lobe.
**Fig. 2:** Low-Grade Small B-Cell Lymphoma of BALT Origin. Axial and sagittal CT scan: bilateral consolidations as a pulmonary mass, and air bronchograms.
**Fig. 3:** Low-grade NHL TYPE B (BALT). 400x HE lymphoepithelial lesion.
Fig. 4: Low-Grade Small B-Cell Lymphoma of BALT Origin. Axial CT scan with lung and mediastinal windows shows multiple, bilateral and ill-defined pulmonary nodules. Mediastinal lymphadenopathy.
70-years-old man with evening fever, excessive sweating and general decline.

**Fig. 5:** Lymphoma Intravascular large B-cell lymphoma. Coronal and axial CT scan show diffuse and bilateral thickening of interlobular septa, predominantly in upper lobes. Millimetric interstitial well-defined nodules. Mosaic perfusion pattern.
Fig. 6: Lymphoma Intravascular large B-cell lymphoma. Alveolar septa with intravascular infiltration by large neoplastic lymphocytes CD20 positive (CD20 immunohistochemistry, x100).
70-years-old woman with cough and dyspnoea

**Fig. 7:** Typical carcinoid tumor. Axial chest CT scan: 2 cm nodule in left upper lobe, with lobulated margins and pleural tail. Lymphadenopathy are not seen.
**Fig. 8:** Typical carcinoid tumor: solid nests and cords of small cells with uniform nuclei without mitotic activity and vascular hyaline stroma (HE, x100).
Fig. 9: Typical carcinoid tumor. Chest radiograph shows a middle lobe atelectasis.
**Fig. 10:** Typical carcinoid tumor. CT scan showed that the middle lobe atelectasis was secondary to a central tumor.
Fig. 11: Atypical carcinoid tumors. Chest CT scan, mediastinal window: pulmonary lesion in left lower lobe that causes narrowing of the left lower bronchus.
Fig. 12: Atypical carcinoid tumors. Chest CT scan with mediastinal window: left pneumonectomy. Recurrence at the left bronchial stump as a heterogeneous mass. Collection produced by pleural pneumonectomy, and pleural implants. Necrotic lymphadenopathy.
Fig. 13: Atypical carcinoid tumors: solid nests rosette-like with atypia and mitotic activity (HE, 400x).
Fig. 14: Large cell neuroendocrine tumor. Chest CT scan with lung and mediastinal window: pulmonary mass in the right lung that which contacts with the pleura. Hilar lymphadenopathy and pleural right effusion are seen.
**Fig. 15:** Large cell neuroendocrine carcinoma: CD56 immunohistochemistry, 400x.
**Fig. 16:** Combined small cell carcinoma-cell carcinoma: small cell undifferentiated carcinoma with foci of keratinizing squamous differentiation (HE, 200x).
**Fig. 17:** Mixed large and small cell carcinoma. Chest radiograph shows right pleural effusion that occupies half of the hemithorax. Nodular-like opacity in left lower lobe that probably corresponds to an incipient infiltration.
Fig. 18: Mixed large and small cell carcinoma. Chest and abdominal CT with mediastinal window: 4 cm right hilar mass that close the right interlobar bronchus producing complete atelectasis of middle and right lower lobes. Necrotic mediastinal lymphadenopathy. Multiple liver metastases.
Fig. 19: Sarcomatoid carcinoma. Chest radiograph: left hilar mass and left pleural effusion. Cavitary lesion in left upper lobe.
**Fig. 20:** Sarcomatoid carcinoma. Chest CT scan with lung and mediastinal windows: heterogeneus left hilar mass, with foci of necrosis, which causes narrowing of left pulmonary artery almost completely. Necrotic mediastinal lymph nodes. In addition there is a 2 cm cavitary lesion with ground glass opacities adjacent parenchyma, probably of infectious etiology.
**Fig. 21:** Sarcomatoid carcinoma: proliferation of spindle or pleomorphic tumor cells with sarcomatoid appearance (HE 100x) and immunohistochemical expression of cytokeratin (detail)
Fig. 22: Adenosquamous carcinomas. Chest CT scan with lung window shows a right parahilar 5 cm mass that contacts with the posterior wall of the right interlobar bronchus. Right pleural effusion. Mediastinal lymphadenopathy.
Fig. 23: Adenosquamous carcinoma: keratinizing squamous cell component (left) and mucosecretory cells with vacuoles positive diastase PAS (right), (400x).
Fig. 24: Inflammatory myofibroblastic tumor. Chest radiograph: cavited pulmonary nodule in the apical segment of left lower lobe.
**Fig. 25:** Inflammatory myofibroblastic tumor. Axial and sagittal chest CT scan: pulmonary mass in the apical segment of the left lower lobe, with irregular and spiculated margins and central air-fluid level, which pulls the fissure and contact with the pleura. There are no lymphadenopathy.
**Fig. 26:** Inflammatory myofibroblastic tumor: polymorphous inflammatory infiltrate with proliferation of spindle and stellate cells (myofibroblast), (actin immunohistochemistry, x100).
Fig. 27: Pulmonary blastoma. Chest radiograph: massive right pleural effusion with contralateral mediastinal shift. Chest CT, mediastinal window, right lung mass, necrotic, with pleural effusion, which produces a significant contralateral mediastinal shift.
Fig. 28: Biphasic tumor with neoplastic glands with subnuclear vacuolization, spindle cells and myxoid stroma. Pulmonary blastoma. (Hematoxylin-eosin 400x)
Fig. 29: Fibrosarcoma, monomorphic proliferation of atypical fibroblasts arranged in intersecting fascicles(HE, 400x).
80-years-old woman with generalized weakness and pain in both legs

Fig. 30: Pulmonary nodular amyloidosis. Chest radiograph: Cardiomegaly. Nodule-like opacities in left lung apex. Smaller and less well-defined opacities in the right lung.
Fig. 31: Pulmonary nodular amyloidosis. Chest CT, axial sections: multiple pulmonary nodules of variable size and well-defined poly-lobed margins, more numerous in the upper lobes. There is a cavitate nodule.
**Fig. 32:** Lipoid pneumonia exogenous. Presence of an infiltrate in the left lower lobe in a patient with aspirations of Paraffin wax poured into a drink container. NOTE: because of technical problems we can not show CT with mediastinal window where you can see the characteristic fat density within the infiltrate.
**Conclusion**

We want to highlight a group of an unusual primary lung tumors that sometimes we should manage in our clinical practice.

Although most lung tumors do not show typical radiological features that allow diagnosis only with imaging, CT is a useful tool to define the location and extent of these tumors and the choice of treatment. It also narrows the differential diagnosis of these lesions.

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