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#### RESEARCH ARTICLE

## PULMONARY MUCOSA-ASSOCIATED LYMPHOID TISSUE LYMPHOMA: TWO CASES REPORT

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## Manuscript Info

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#### Key messages:

Primary pulmonary lymphoma is rare Lung MALT lymphoma are the most common lung lymphomas The diagnosis is based on immunohistochemical analyses

## Abstract

Primary pulmonary lymphoma (PCL) is rare and represents only 0.5-1% of the malignant lung tumour. Pulmonary MALT lymphoma is the most common histological aspect. The pathophysiological mechanism remains poorly known, unlike other locations of MALT lymphoma, no pathogen has so far been associated with the lung localization of the disease. It is characterized by slow evolution and atypical clinical presentation which delays diagnosis. The form «pneumonic» on the imaging allows to evoke the diagnosis while the confirmation is based on the immunohistochemical study which detects the presence of positive CD20 cell. Therapeutic options include surveillance of indolent types of LPP, surgery in localized tumors, chemotherapy in diffuse forms. We report two observations of diagnosed pulmonary MALT lymphoma on a scanno-guided bronchial and pulmonary biopsy.

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## Introduction:-

Primary pulmonary lymphoma (PPL) is rare, representing 0.5-1% of primary malignant lung tumours<sup>1</sup>. They can be primitive or secondary to lymphomas of extra-pulmonary origin <sup>2</sup>.

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PPL are defined histologically by a clonal lymphocyte pulmonary proliferation without extra-pulmonary involvement within three months of diagnosis. The most common histological aspects are: Mucosa-Associated Lymphoid Tissue lymphoma [MALT]; large cell B lymphomas; lymphomatoidgranulomatosis<sup>2</sup>.

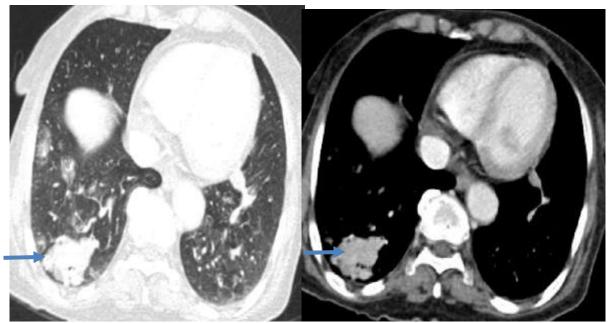
Lung MALT lymphoma are indolent phenotype B lymphomas and are the most common lung lymphomas. They are often asymptomatic in appearance radiological little specific and their diagnosis is mainly based on immunohistochemical analyses <sup>2</sup>.

We report two observations illustrating the clinical, radiological and pathological aspect of this lymphoma.

#### Case 1:

An 80-year-old woman, hypertensive undercalcium channel blockers, who had stage II dyspnea of mMRC associated with chest pain and epigastralgia, all evolving in a context of conservation of general condition. The clinical examination remains without abnormalities. The thoracic scanner (Figure 1) objected to the presence of lower right lobar pulmonary parenchymal condensation associated with peripheral and central bilateral pulmonary parenchymal frosted glass lesions. The bronchoscopy showed a diffuse thickening of the spurs at the right bronchial shaft . Histological study of bronchial biopsies was negative. The anatomopathological study of the scanno-guided trans parietal pulmonary biopsies showed lung localization of non-Hodgkin's type B malignant lymphoma of the

marginal zone (Malt)(figure 2). The assessment of extension in search of another location was without particularity (thoraco-abdominal-pelvic scanner, medullary biopsy and oeso-gastroduodenalfibroscopy). the patient was put on polychimiotherapy, the clinical course was favorable after sixth cure.



**Figure 1:-** A thoracic CT scan showing lower right lobar pulmonary parenchymal condensation associated with peripheral and central bilateral pulmonary parenchymal frosted glass lesions.

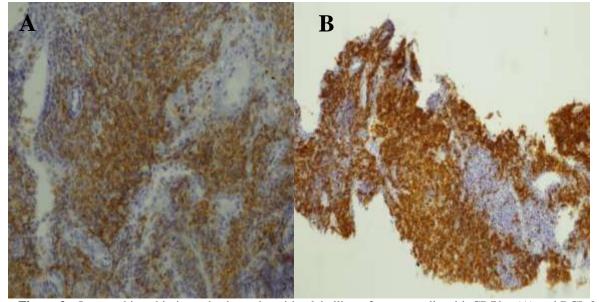


Figure 2:- Immunohistochimie study showed positive labelling of tumour cells with CD79 a (A) and BCL 2 (B).

### Case 2:

A 62-year-old woman, followed for tetracyclic depression, consults for a dry, associated stage II dyspnea of the mMRC evolving for 3 months without any other associated respiratory or extra-respiratory signs, All evolving in a context of alteration of the general state. The clinical examination found a patient in good general condition, pleuropulmonary examination and lymph nodes were peculiar. The chest CT scan (Figure 2) involved a left pulmonary hilo process. Bronchoscopy showed a budding formation (Figure 3) in the left stem bronchus. The histological and immuno-histochemical study (Figure 4) of bronchial biopsies was suggestive of marginal-zone B

lymphoma. The cerebral CT and abdominal pelvic bone marrow biopsy performed as part of the extension workup were normal. The therapeutic conduct was to put the patient on polychimiotherapy. The evolution was favorable after 6 cures. Simple monitoring was decided with a 2-year setback.

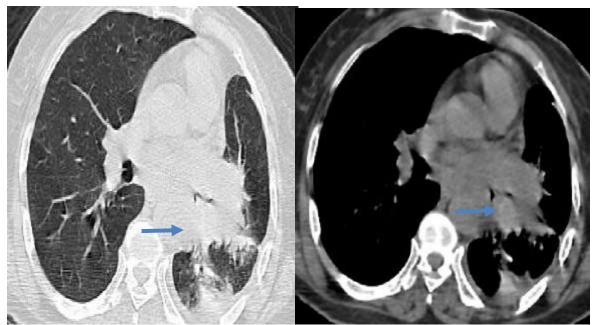


Figure 3:- The chest CT scan showing a left pulmonary hilo process.



**Figure 4:-** A Bronchoscopy showing a budding formation in the left stem bronchus.

## Discussion:-

Pulmonary MALT lymphoma belongs to the marginal-zone lymphoma group representing about 1% of primary malignant lung tumours and 3% to 4% of non-Hodgkin's extra-lymph node malignant lymphomas (4)It usually affects women between the ages of 50 and 60 and smoking is not an obvious risk factor <sup>2-5</sup>. Both of our patients are middle-aged women at 71.

MALT is a lymphocyte tissue specialized in the defense of the mucous membranes, the digestive tract is most likely affected in connection with chronic stimulations of infectious origin such as helicobacter pylori <sup>6</sup>. In the lung, certain autoimmune diseases (lupus, Hashimoto's thyroiditis and, Sjogren's syndrome) are recognized as factors favouring the occurrence of MALT lymphoma <sup>5</sup>.

Many cytogenetic abnormalities have been described in MALT lymphomas. The most common translocation is t(11;18)(q21;q21) (API2-MALT1) found in 42% of lung lesions <sup>2</sup>.

Half of the patients are asymptomatic or may have aspecific signs such as cough, dyspnea, chest pain. Pulmonary auscultation found crackling rails in 20% of cases. The presence of general signs points to aggressive lymphoma<sup>2-7</sup>. In our cases the clinical signs were dyspnea, chest pain and dry cough

The most evocative radiological presentation of pulmonary MALT lymphoma is the medium lobar alveolar condensation, often multifocal, an aerated alveolar-bronchogram. The other forms, less frequent and less characteristic, include: the «pseudo-tumoral» presentation, In less than 10% of cases, the scanner shows reticulonodular opacities, atelectasias, pleural effusion or medial lymphadenopathy <sup>2</sup>. One of our patients had pleural effusion.

Many pathologies result in chronic alveolar opacities, the real diagnostic trap is that of sarcoidosis which is more common and may have the same radiological presentation as lymphoma<sup>7</sup>.

Bronchoscopy is an essential examination for diagnosis, it can show different aspects ranging from a normal endoscopic aspect to a bronchial stenosis <sup>4</sup>. In our cases, bronchial endoscopy showed a budding formation in one patient, and showed only thickening of the spurs in the other patient. The diagnosis of pulmonary MALT lymphoma can be made on a bronchial biopsy, scanno-guided trans parietal biopsy or surgical lung biopsy <sup>8</sup>. Histologically, when the sample is small, it poses a differential diagnostic problem with diffuse lymphoid hyperplasia or lymphoid interstitial pneumonia,thus,a bronchoalveolar lavage is indicated <sup>5-8</sup>. The diagnostic confirmation of our patients was obtained on a scanno-guided bronchial and trans-parietal lung biopsy.

The immunohistochemical analysis confirms the diagnosis and eliminates other differential diagnoses of LPP type Malt. It makes it possible to specify the phenotype B CD20 positive with co-expression of CD43 <sup>9</sup>.

An extension assessment is necessary as soon as the diagnosis is confirmed, the chest-abdominal-pelvic scanner helps eliminate a secondary lung localization of a nodal lymphoma. Mucous involvement may present at the time of diagnosis including stomach and ENT mucous membranes<sup>2</sup>. Our two patients benefited from a thoraco-abdominal-pelvic CT and a bone marrow biopsy and only one patient benefited from a gastroduodenalfibroscopy . In our patients the results came back negative.

The therapeutic attitude of pulmonary MALT lymphoma is based on a simple surveillance initially because of their indolent character. When the disease is localized but symptomatic, radiation therapy may be offered in resectable but non-operable lymphomas. When the disease is bilateral or non-operable, the standard treatment is chemotherapy <sup>9-10</sup>, as the case of our patients. This lymphoma has good prognosis with a five-year survival greater than 80% <sup>10</sup>.

### **Conclusion:-**

Pulmonary MALT lymphoma is a rare pathology and difficult to diagnose. The clinical-radiological aspects are very specific. The diagnosis is based on immunohistochemical analyses of bronchial or lung biopsies. Their prognosis is excellent and the modalities of treatment will be discussed.

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