115 年度奇美醫學院胸腔內科臨床病例討論會

1 時間：115 年 09 月 22 日 PM: 4:00-5:00
2 課程活動題目: Pulmonary lymphoma in CXR
3 主持人：柯獻欽
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6 摘要：

Lymphoma is a type of cancer that arises in the lymph nodes in the body. In the chest, the involved lymph nodes are usually located in the mediastinum, near the trachea and large vessels flowing to and from the heart and lungs. Patients with relatively mild symptoms can frequently present with striking chest x-ray abnormalities. CT is used to confirm the cause of the mediastinal abnormality on CXR, delineate the full extent of tumor, and evaluate complications. CT of the abdomen and pelvis are often performed to stage the tumor and evaluate for involved lymph nodes below the diaphragm.

Low-grade pulmonary B-cell lymphoma is the most frequent form of primary pulmonary clonal lymphoid proliferation. It arises from mucosa-associated lymphoid tissue. It is usually indolent and appears in the form of a chronic alveolar opacity. The prognosis is excellent, but treatment is controversial (simple monitoring, surgery or single-agent chemotherapy). High-grade pulmonary B-cell lymphoma is far rarer and usually occurs in individuals with an underlying disorder (*e.g.* immunodeficiency). The prognosis is poor and therapeutic options depend on the underlying disorder.

Clinical and radiological signs

Nearly half these patients are asymptomatic at diagnosis and are identified fortuitously on the basis of a radiological pulmonary anomaly. When present, symptoms, such as cough, mild dyspnoea, chest pain and occasionally haemoptysis, are nonspecific. Pulmonary auscultation reveals crackles in <20% of cases. By definition, extra-pulmonary manifestations are restricted to general signs (fever and weight loss) and occur in less than one quarter of patients. The usual radiological aspect (50–90% of cases) is a localised alveolar opacity, with a diameter of <5 cm and blurred or well-defined contours (according to the series); it is associated in nearly 50% of cases with an air bronchogram. Computed tomography (CT), which is more sensitive than standard radiography, has demonstrated that the lesions are usually bilateral (60–70%) and multiple (70–77%). Nearly all these lesions contain clear areas corresponding to an intact bronchial lumen. The presence of distended bronchi within the lesions is a good diagnostic sign, although the underlying mechanism is unexplained. Less than 10% of patients have bilateral diffuse reticulonodular opacities, atelectasia or pleural effusion. CT can reveal hilar and mediastinal adenopathies. The interval between the first clinical or radiological manifestations and diagnosis ranges from 5 months to 8 yrs.

Course, prognosis and treatment

Course and prognostic factors

The outcome of MALT-type PPL is generally favourable in most series, with a 5-yr survival rate of >80% and a median survival time of >10 yrs. The overall survival of these patients is longer than that of patients with lymphoma of the nodal or splenic marginal zone. In contrast, it has not yet been demonstrated that the survival of patients with MALT-type PPL is equivalent to that of the general population . The median survival of patients with gastrointestinal MALT-type PPL does not differ from that of patients with other localisations, but progression-free survival appears to be shorter in the latter, especially in patients with pulmonary forms. Long-term surveillance is necessary, owing to the frequency of late local or extrathoracic relapse after surgical resection (almost 50% of patients after >2 yrs). No clear prognostic factors have been identified in MALT lymphoma. In univariate analysis, some all-comer series of MALT lymphoma have shown a pejorative influence of age over 60 yrs, elevated ß$_2$ microglobulin and failure to enter a complete response during first-line treatment . The only prognosticator in multivariate analysis is elevated ß$_2$ microglobulin. According to other authors, intratumoural amyloid deposition is a factor of poor prognosis, while lymphoepithelial lesions are associated with a good prognosis. A recent series of 48 patients with PPL identified no prognostic factors among the following: presentation, bilaterality, tumour/nodes/metastasis (TNM) stage, surgical resection, adjuvant chemotherapy and several histological criteria.

Some authors have suggested that low-grade PPL can transform into high-grade proliferation, based on the existence of mixed forms or transitional forms identified by serial biopsy. This conflicts with recent studies showing differences between the cytogenetic abnormalities of low-grade and high-grade PPL. For example, the t(11;18) translocation is only present in low-grade PPL. For this reason, the latest revision of the WHO classification recommends the use of the term "large B-cell lymphoma" rather than "high-grade MALT lymphoma".

Principles of treatment

There is no consensus on treatment. The lack of an identified culprit antigen in the lung, contrary to the stomach (*H. pylori*), means that antibiotics effective on low-grade localised gastric lymphoma are inappropriate. Current treatment options are surgery, chemotherapy and radiotherapy–. The respective efficacy of these treatments cannot be analysed, however, owing to a lack of comparative series, and some authors even propose simple clinical monitoring. Nevertheless, surgical resection is commonly preferred for localised tumours–. Exclusive chemotherapy is generally used for patients with bilateral or extrapulmonary involvement, relapse or progression. Combination regimens, such as cyclophosphamide, adriamycin, oncovin and prednisone (CHOP), have not proven more effective than single-agent regimens with chloraminophene, cyclophosphamide, azathioprine or steroids. Radiotherapy is rarely used.