Rare tumours of the chest

E. Muñoz, S. Cedrés, E. Felip

Vall d'Hebron Hospital, Barcelona, Spain

Malignant thoracic tumours include non-small cell lung cancer (NSCLC) – adenocarcinoma, squamous cell carcinoma, large cell carcinoma, undifferentiated carcinoma – and small cell lung cancer (SCLC) which account for more than 95% of all primary neoplasms. However, a variety of non-bronchiogenic malignant tumours may occasionally affect the lung and represent 3–5% of all primary chest tumours. The most frequently encountered malignant primary thoracic tumours are thymoma, sarcomas, malignant melanoma and carcinoid tumours [1–3].

Thymomas and thymic carcinomas constitute 30% of anterior mediastinal masses in adults and arise from thymic epithelial cells [4]. Thymomas commonly affect people aged between 40 and 60 years. Nearly one half are asymptomatic but when patients do have symptoms, they are due to local invasion in 65% of cases (chest pain, dyspnea, haemoptysis, stridor, cardiac arrythmias and Horner's syndrome) and paraneoplasic syndromes. They rarely metastasise outside of the thorax. Surgery with complete resection is the mainstay of therapy for patients with completely encapsulated tumours and it plays a key role in locally invasive disease with increase in survival even if invasion is found to be present [5]. The role of subtotal resection in advanced disease remains controversial.

Adjuvant radiotherapy can decrease recurrences in invasive disease but its benefit in encapsulated tumours has not been proved. In subtotal resections, radiotherapy maximises the chance of local control. Retrospective reviews show that 20–30% of patients may be candidates for systemic therapy including corticosteroid, single-agent or combination chemotherapy (platinum, doxorubicin, cyclophosphamide, taxanes), chemoradiotherapy or cytokine therapy, although the optimal regimen has not yet been determined [2,3,6].

Primary pulmonary sarcomas (PPS) account for less than 0.2% of all malignant pulmonary tumours. These tumours may occur at any age and there is no evidence of a gender predisposition. Leiomyosarcoma represents the most frequent subgroup followed by fibrosarcoma and haemangiopericytoma. Other types of PPS include solitary fibrous tumour, synovial sarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma and angiosarcoma. PPS arise from the stromal elements of the bronchial tree, sanguineous vessel and lung parenchyma. Diagnosis frequently reveals local invasion, patients presenting with dyspnea, a cough, chest pain or haemoptysis. Pulmonary thromboembolism, pulmonary hypertension or right heart failure occasionally occur in angiosarcomas, and paraneoplasic syndromes may occur in patients with haemangiopericytoma. Occasionally, PPS metastasises to distant organs including contralateral lung, liver, brain, bone or soft tissues. Surgical treatment with complete resection (lobectomy or neumonectomy) is the mainstay treatment and the most important prognostic factor. Other prognostic factors are histologic grade and tumour size (poor prognosis tumours >5 cm) [3]. The addition of chemotherapy and radiotherapy to surgical treatment (neo- or adjuvant) can increase the results but no definitive evidence of benefit has been shown. Chemotherapy, either as a single agent or in combination regimens, may have palliative benefit in unresectable disease. Experience for PPS with chemotherapy is predominantly gathered from patients with traditional soft tissue sarcomas and is extrapolated to pulmonary sarcomas. The most commonly used agents include doxorubicin (20% response rate), ifosfamide and dacarbazine [2].

Although malignant melanomas principally affect the skin and ocular area, there is well documented information about primary melanomas in the lungs [2, 3,7]. Primary lung melanomas are defined by minimal criteria including no history of a previous melanoma (cutaneous or ocular), no demonstrable melanoma in any other organ at the time of surgery, a solitary tumour in the surgical specimen from the lung, tumour morphology compatible with a primary tumour, and no evidence at autopsy of a primary melanoma elsewhere. Lung melanomas are unifocal and are commonly presented as a polypoid obstruction lesion within the tracheobronchial tree or as a mass in the lung parenchyma. Similar to NSCLC, lung melanomas metastasise to regional lymph nodes in hilar and

mediastinal regions and can metastasise to the brain, liver, pleura, pericardium or heart. The treatment of choice for primary lung melanoma is radical surgical excision with regional lymph node dissection. In local recurrence, if surgery is not feasible, systemic chemotherapy or radiotherapy may be considered in symptomatic cases.

Finally, pulmonary carcinoid tumours make up 1–2% of all primary lung cancers [2,8]. There is equal gender distribution, they can occur at any age in a person's lifetime, and they are more frequently located centrally (2/3) in the lung. As with NSCLC, the TNM staging system is used to classify pulmonary carcinoids into prognostic categories. At the moment of diagnosis, patients are symptomatic, presenting with coughs, haemoptysis, recurrent chest infections, chest discomfort, pain and shortness of breath. The 'carcinoid syndrome' is rare in patients with pulmonary carcinoids with an incidence of around 2%. Surgical treatment (thoracotomies or laser resections) remains the treatment of choice and offers the only chance of cure with 5-year survival rates of 70–100%. Local recurrence is rare although atypical carcinoids have a higher risk of relapse [3]. The percentage of patients who develop metastatic disease is variable and its treatment includes somatostatin analogues, interferons, radionucleotides and local treatments. Chemotherapy for metastatic carcinoid tumours has been disappointing.

Although the clinical and pathological features of rare malignant chest tumours are initially similar to those of common pulmonary carcinomas, the treatment approach is uncertain.

Conflict of interest statement

None declared.

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