UNUSUAL BEHAVIOUR OF A LUNG INFLAMMATORY MYOFIBROBLASTIC TUMOUR

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Abstract

Inflammatory myofibroblastic tumours (IMTs) are rare lesions. We report a case of a 55 year-old male, admitted with a pneumonia. Further investigation revealed a left lower lobe mass and enlarged mediastinal lymph nodes. Cytology of the bronchoalveolar lavage suggested a squamous cell carcinoma. He received four cycles of chemotherapy followed by a left lower lobectomy. Pathological analysis was compatible with IMT. Three months after surgery, a new IMT nodule located in the lingula was excised. Four months later,endobronchial involvement and the presence of liver nodules were detected.Ten months after the first surgery a CT revealed a sacrum lesion. Histology was compatible with undifferentiated sarcoma and a sarcomatous transformation was assumed.

INTRODUCTION

IMTs, which were first described by *Brunn in* 1939, represent approximately 0.02-1.2% of all lung tumours.[1,2] There is a lack of information about the natural history, clinical presentation and effective treatments.[1]

The prevalence is not sex dependent,[1,2,3] and may present at any age.[1,2,4] Preoperative diagnosis is difficult to obtain, which may delay appropriate treatment.[1,4,5]

CASE REPORT

55 year-old male, with history of heavy smoking and chronic obstructive pulmonary disease (COPD), was admitted to the hospital due to a left pneumonia. A fiberoptic bronchoscopy with bronchoalveolar lavage suggested squamous cell carcinoma. The CT and PET-Scan revealed a left lower lobe mass with 92x89 mm (SUVmax 49). There was an increased uptake of FDG on stations 4R, 5,7 and 10L. EBUS of stations 4R, 4L and 7 without evidence of malignancy. The diagnosis of a stage IIIA (T3N2M0) squamous cell carcinoma was assumed.

The case was discussed in a multidisciplinary meeting and neoadjuvant therapy (four cycles of cysplatine/gemcitabine) was administered based on station 5 with suspected metastatic disease.

Regardless of a slight reduction of the lymph nodes, the main lesion remained stable. After a negative cervical mediastinoscopy, a left lower lobectomy was performed.The histology was compatible with IMT, measuring 115x87x35 mm. Immunohistochemistry was positive for ALK and Vimentin. ALK gene rearrangements were not found. No mediastinal lymph node involvement was reported. The bronchial stump was 5 mm from the tumour but revealed a squamous metaplasia in all its length. The patient was re-staged as IIB (ypT3N0M0).

Three months after surgery, a CT-scan showed a new nodule in the lingula, which was removed. Pathology confirmed an IMT.

Four months later, a CT-scan revealed several lung and liver nodules. An occlusion of the left main bronchus, causing left lung atelectasis, required debulking of an endobronchial IMT mass. Finally, palliative radiotherapy to the left main bronchus was decided (total dose of 20Gy in 4Gy daily fractions).

Ten months after the first surgery, after the onset of back pain, a new CT-scan revealed a sacrum lesion, whose needle biopsy was suspicious for multiple myeloma.

Histological reassessment of all samples was performed. Sacrum lesion was compatible with undifferentiated sarcoma and a sarcomatous transformation was admitted

Shortly after, he developed neurological symptoms on lower limbs and urgent spinal decompression took place.

The patient died of disease progression 18 months after the initial diagnosis.

DISCUSSION

IMTs are considered benign or low-grade malignant tumours,[1,3] because of possible malignant invasion, recurrence and, exceptionally, malignant transformation.[3,5,6]

The exact etiology is unknown, although several reports suggest that might be related to recurrent respiratory infections or autoimmune mechanisms (such as increased IgG4).[2,3,7,8]

The size of the tumour(<3 cm) and free surgical margins resection are the major determinants for avoiding recurrence and increasing survival.[3,5,7] In our patient, the lesion was larger than 3 cm but the resection was complete. Endobronchial involvement is considered uncommon, although it is reported in up to 12% of cases.[6]

In most series, patients area symptomatic and the tumour is accidentally discovered.[1,3,4,5] Symptoms are usually related to location and are nonspecific.[2,4,5]

Diagnosis before surgery is difficult. Usually a needle biopsy shows a nonspecific mixture of inflammatory cells.[1,4,5] In our patient, the preoperative diagnosis suggesting a squamous carcinoma may be related to the squamous metaplasia later found in the bronchial stump.

In most cases, nodules are solitary and regular,[2,3] being PET-Scan uptake similar to malignant lesions.[1] Although rare, there are some reports of distant metastasis and sarcomatous transformation.[1,5,6,7,9]

ALK gene rearrangements can be present in 50% of IMTs.[7,8] Based on that, drugs like Crizotinib may have a role.[8] Corticosteroids are usually not useful in adults,[2,3,5] except for those with increased IgG4,[7] butgood results have been reported in children.[2,3,5] Chemotherapy is an option in cases of multifocal or invasive lesions or in cases of local recurrence.[2,3]

CONCLUSION

Despite the past assumption of benign behaviour of IMTs, this report as well as others found in the literature, show us that this is not always true. The lack of valid therapeutic options is still an issue for those who present with disseminated disease.

Reviewer's note:

Initial decision for induction chemotherapy lacks support after a negative EBUS!

Fernado Barata

Author's reply: We thank the reviewer for his comment. Induction chemotherapy was decided upon in a multidisciplinary meeting, based on a large (92mm) squamous cell carcinoma with a possible single station N2 disease in station 5 (stage IIIA) supported by the NCCN clinical practise guidelines.

Daniel Cabral

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