

Primary chondrosarcoma of the lung with cutaneous and skeletal metastases

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ABSTRACT

Extraosseous chondrosarcomas are rare tumours. Primary chondrosarcoma of the lung is very rare, and is considered to be a slow-growing, well-circumscribed tumour, with rare incidence of extra-thoracic metastasis. We report a 60-year-old man who had chondrosarcoma of the lung with two local recurrences, namely: recurrent cutaneous metastases and a skeletal metastasis. Cutaneous metastases were treated by excision with adequate margins each time and they did not recur at the same site. Metastasis in the C5 vertebrae was treated by corpectomy and bone grafting. The patient is well six years after diagnosis.

Keywords: chondrosarcoma, cutaneous metastases, lung chondrosarcoma, lung neoplasm, skeletal metastasis

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INTRODUCTION

Extraskeletal chondrosarcoma is a rare tumour.⁽¹⁾ Pulmonary metastasis of skeletal chondrosarcoma is common, but primary chondrosarcoma of the lung is very rare.^(1,2) We report a case of primary chondrosarcoma of the lung with multiple cutaneous and skeletal metastases.

CASE REPORT

A 60-year-old man presented with complaints of dry cough for a period of 3–4 months. He was a non-smoker and there was no past history of pulmonary tuberculosis. Examination of the respiratory system revealed decreased air entry in the left lower zone. His chest radiograph showed opacity in the lower lobe of left lung. Computed tomography (CT) showed a low-attenuation lesion in the paracardiac region of the left lower lobe, measuring 77 mm × 72 mm × 140 mm, with consolidation in the surrounding lung fields (Fig. 1). Needle biopsy confirmed a chondroid tumour. Thoracotomy was done and the tumour was

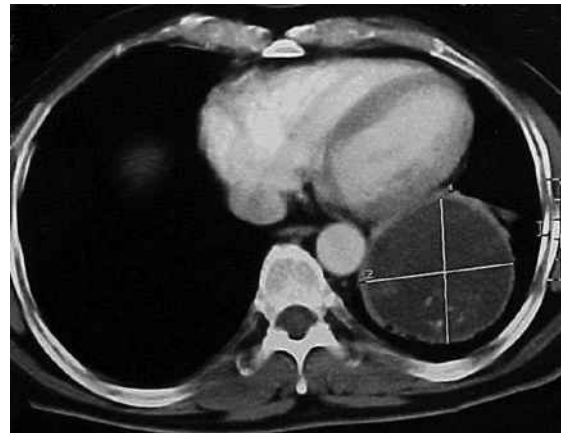


Fig. 1 Axial CT image of the primary tumour in the left lung shows a large, well-defined low-attenuation lesion with small scattered specks of calcification within the periphery, situated in the left posterior paracardiac region.

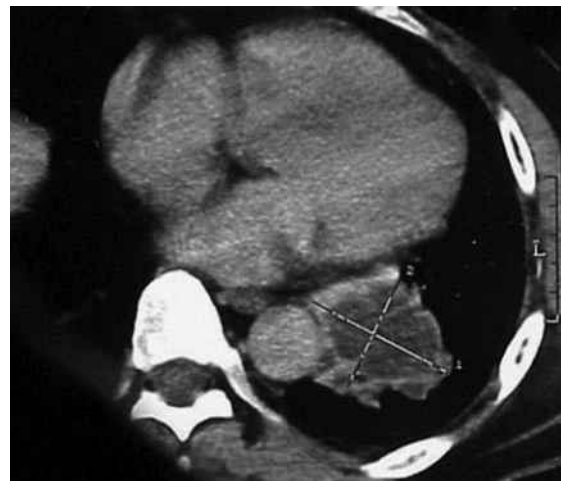


Fig. 2 Axial CT image of the first recurrence of the tumour shows the lesion is located in the vicinity of the posterior cardiac border and aorta.

excised with adequate margins. Histopathological examination revealed well-differentiated chondrocytes with enlarged nuclei in a cartilaginous matrix and was consistent with a low grade chondrosarcoma. The patient recovered uneventfully.

He was symptom-free for one year. He then had an episode of haemoptysis. CT scan was repeated, showing recurrence of the lesion in the posterior basal

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Fig. 3 Axial CT image of the second local recurrence shows a well-margined mass lesion with an intrinsic necrotic component, situated in the left lung posteriorly at the level of the arch of aorta, adherent to the descending thoracic aorta and adjacent pleural reflection. The mass has also extended to the left posterior chest wall.



Fig. 4 Sagittal T1-W MR image of the cervical spine section taken through the region of the left neural foramina shows a low-signal intensity lesion involving the C5 vertebra with an extension anteriorly into the retropharyngeal space and posteriorly to the neural foramina, with the involvement of adjacent disc space.

segment of the left lower lobe, measuring 54 mm × 42 mm × 70 mm (Fig. 2). The lesion was in the vicinity of the aorta and posterior cardiac border. This time, a left lower lobectomy was done. Histopathological

examination confirmed low-grade chondrosarcoma of the lung. At one month after the second surgery, the patient presented with a soft swelling in a periumbilical region. The excision biopsy of this lesion showed low-grade chondrosarcoma similar to the one found in the lung. Bone scintiscan and bronchoscopy were performed, and skeletal or pulmonary lesions were ruled out.

At eight months after surgery, the patient had a massive haemoptysis again. This time showed a 43 mm × 54 mm well-margined mass in the left thoracic paraspinal region, adherent to the arch and descending thoracic aorta, and extending to the left posterior chest wall (Fig. 3). Needle biopsy confirmed the recurrence. After considering the location of the tumour, the patient was subjected to radiotherapy and chemotherapy. The patient received 63 Gy in 35 fractions, by intensity modulated radiotherapy technique, to the left thoracic paraspinal region. After completion of radiotherapy, he received six cycles of iphosphamide (2 g/m², Days 1–5, two-hour infusion) with adriamycin (25 mg/m², Days 1–3, continuous infusion) every three weeks. Postchemotherapy CT showed regression of the tumour (25 mm × 42 mm). The tumour was then excised with safe margins. Histopathology confirmed chondrosarcoma adherent to the chest wall. After that, the patient developed lesions in the anterior abdominal wall, left paraspinal region and left thigh at an interval of six months. Each time, it was excised and confirmed to be of similar histopathology as the primary tumour.

Four and a half years after the detection of first tumour, he complained of pain radiating down to the left upper limb, with tingling sensations and numbness in the left hand. There was no restriction of neck movements. Neurological examination revealed hypoaesthesia in the distribution of the C5 and C6 nerve roots. There was no motor deficit. Radiograph of cervical spine did not reveal any abnormality. Magnetic resonance imaging of the cervical spine showed a metastatic lesion involving the C5 vertebrae affecting the pedicle, transverse process with compression of the C5 and C6 nerve roots, and indentation over the spinal cord (Fig. 4). The patient was operated, with C5 corpectomy, autologous bone grafting and anterior plate fixation. At one year after surgery, he had full neurological recovery, and there was no recurrence of any skin lesion.

DISCUSSION

Morgan and Salama described the criteria for diagnosing primary chondrosarcoma of the lung in 1972, which were then followed by other reviewers.⁽¹⁾

According to them, a tumour is not a primary chondrosarcoma if: (1) there is total absence of clinical history; (2) possible origin of the tumour is from the thoracic cage; (3) presence of skeletal chondrosarcoma discovered immediately after thoracotomy; (4) amputation of a limb or excision of tissue potentially containing cartilage before the pulmonary lesion became apparent; (5) evidence of pre-existing pulmonary hamartoma; and (6) histology that is vague, equivocal or confined wholly to a small biopsy. Morgenroth et al described 18 cases of primary pulmonary chondrosarcoma of the lung in their review of the literature in 1989.⁽³⁾ Four of them originated in the trachea, nine in segmental, or more peripheral bronchi, and five in a major bronchus. We found a few other cases reported in the literature since then.⁽⁴⁻⁷⁾

The natural history of primary chondrosarcoma of the lung is similar to that of the skeletal chondrosarcoma. It grows slowly in the initial phase and the patient may remain symptom-free for a long period. Following that, a symptomatic phase with a more rapid course is usual. The most common symptoms include a non-productive cough due to bronchial obstruction, followed by chest pain, dyspnoea and haemoptysis in the late stages of the disease.⁽⁸⁾ The average age of presentation is 55.3 years and both gender are equally affected.⁽⁷⁾

The growth pattern is reported to be expanding and significant invasion to the pulmonary arteries has been reported.⁽²⁾ Infiltration of the pleura, pericardium and mediastinal lymph nodes is also reported.⁽¹⁾ Local recurrence is reported, even after excision with adequate margins,⁽⁸⁾ as in our case. Chondrosarcomas are exceedingly transplantable. Jazy et al reported recurrences at the site of the previous thoracotomy tube.⁽⁸⁾ In our case, the subsequent intrathoracic and cutaneous lesions could be the result of the initial thoracotomy incision approach. It has been suggested that tumours arising in the tracheobronchial tree have a better prognosis than those originating in the more peripheral part of the lung. Ten of 13 cases with peripheral neoplasm reviewed by Daniels et al had a fatal outcome within a year of diagnosis.⁽⁹⁾

Extrathoracic metastasis of the pulmonary chondrosarcoma is infrequent. Metastases to the cervical lymph node, skull and kidney have been reported.⁽⁷⁾ Jazy et al reported the chondrosarcoma extending from the apex of lung into the epidural space, causing symptoms of Horner's syndrome and paraparesis.⁽⁸⁾ We found six cases of skeletal chondrosarcomas with bony metastases; all had metastases to the vertebral column.⁽¹⁰⁾ Four of the six patients had tumours containing myxoid elements, and this type of tumour is believed to have a higher

incidence of metastases. Skeletal metastasis is even rarer for extraskeletal myxoid chondrosarcoma, with only four known cases reported to date.^(11,12)

Cutaneous metastases of the sarcoma are also quite rare. Brownstein and Helwig reported the largest series (724 patients) of malignancies with cutaneous metastases.⁽¹³⁾ A review of the published cases of chondrosarcomas metastatic to the skin yielded a total of 14 cases, out of which 12 primary tumours were in the bone, one in the heart and one in the soft tissue of the forearm.^(13,14) These metastases can be single or multiple, with a slightly higher occurrence in the head and neck region. The time interval between the diagnosis of chondrosarcoma and the appearance of cutaneous metastases varies from two weeks to 115 months, with a mean of 46 months. Most of the patients die within a mean period of six months since the appearance of cutaneous metastases.

Surgery is the optimal treatment for lung chondrosarcoma. Jazy et al reported a significant response to radiation in one of their two cases, and a partial remission was achieved with a combination chemotherapy of adriamycin and cytoxan in their second case.⁽⁸⁾ Adriamycin is the single, most effective chemotherapeutic agent for chondrosarcoma.⁽¹⁵⁾ The synergistic effect of iphosphamide with adriamycin is well documented, as is the superiority of the combination chemotherapy over a single drug treatment in most malignancies.⁽¹⁶⁾ In our case, a combination of preoperative radiotherapy and chemotherapy at the time of the second recurrence gave excellent results. It reduced the size of the tumour, making resection with adequate margins possible. Although surgery remains the best treatment, preoperative radiotherapy and chemotherapy should always be considered for tumours which appear difficult to resect with adequate margins.

This case is unique in more ways than one. It is the first case of chondrosarcoma of the lung with multiple cutaneous metastases. It is also the first reported case involving cervical vertebrae by metastases from lung chondrosarcoma. The cutaneous metastases first appeared 14 months after diagnosis of a primary tumour. These recurred periodically and were resected with adequate margins. The metastases did not recur at the same site. The patient is now living a healthy active life, six years after the diagnosis of the primary tumour.

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