Multiple chondromatous hamartomas of the lung

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Received 12 June 2002; received in revised form 21 August 2002; accepted 26 August 2002

Abstract

Multiple chondromatous hamartomas (MCH) of the lung are very rare: only 16 cases have been reported to date. In young women, the tumours may be a manifestation of Carney triad (gastric leiomyoblastoma, pulmonary hamartoma and extra-adrenal paraganglioma) or Cowden syndrome (mucocutaneous lesions, multiple benign tumors of internal organs and increased risk for breast, thyroid, urogenital and digestive tract cancer). We report the 17th case of MCH of the lung, diagnosed accidentally in a 66 year-old male patient, with suspected concomitant hamartomas and malignant tumours. © 2002 Elsevier Science B.V. All rights reserved.

Keywords: Multiple chondromatous hamartomas of the lung; Carney triad; Cowden syndrome

1. Introduction

Hamartoma is the most common benign tumor of the lung, accounting for 8% of pulmonary neoplasms. On histologic section they are composed mainly of cartilage and gland-like formations and may include a significant amount of fat [1].

Hamartomas present a peak incidence in the sixth decade of life and are two to four times more common in males than in females [2]. They typically present as single, well circumscribed round nodules. Sixteen cases of multiple chondromatous hamartomas (MCH) have been described in the world since the report by Bateson in 1967 [2–4].

We describe the 17th case in the world of MCH of the lung, localized in the middle and lower right lung (Fig. 1).

2. Case report

A 66-year-old male heavy smoker was admitted to our department for multiple lung nodules in a COPD patient with high blood pressure, hypothyroidism and prostatic hypertrophy. A DDDR pace maker had been applied 2 months before to treat a Luciani Wenckebach periodic block 5:4.

CT scan disclosed a nodule 3 cm in diameter, with ‘pop corn’ calcification, compatible with hamartoma, in the posterobasal segment of the right lower lung lobe. Two other lesions were detected by CT scan in the lower and middle right lobe: they were 1 cm in diameter with a dubious aspect.

Brain and abdominal CT scan and whole body positron emission tomography were negative. Bronchoscopy and broncoalveolar lavage were negative. Suspecting pulmonary metastases, gastroscopy and colonoscopy were also performed and both were negative. Transthoracic fine needle aspiration biopsy did not establish diagnosis so we decided to operate. Respiratory frequency was 16 a.p.m.; arterial hemogasanalysis showed: Pa O$_2$ = 79 mmHg; Pa CO$_2$ = 39 mmHg; pH = 7.40, Sat O$_2$ = 96%; spirometry revealed an obstructive syndrome (FEV$_1$ = 2.07 l 57% – FVC = 4.02 l 85% – FEV$_1$/FVC = 51).

Thoracoscopy was performed but no nodule was identified so we practised thoracotomy: manual palpation revealed three nodules, resected by wedge resections. Histologic examination was suggestive for three chondromatous hamartomas (Fig. 2).

The patient was discharged 10 days after surgery in good general conditions.

3. Discussion

Pulmonary chondromatous hamartoma, originally considered a developmental malformation, is now classified as a benign neoplasm derived from the peribronchial mesenchyme [5]. Pulmonary hamartomas can be divided into parenchymal (90%) and endobronchial (10%) types. Usually hamartomas are located in a peripherical parenchymal location and are predominantly composed of cartilage.
Other components include fibromyxoid connective tissue, fat, bone and smooth muscle [5].

In biphasic lesions composed of a mixture of both stromal and epithelial cells, such as pulmonary chondromatous hamartomas, the mesenchymal component is the site of the High Mobility Group proteins (HMG) genetic alterations [6], involving the chromosomal region 12q14 through q15 [7]. Slow growth is the norm for these tumours; malignancy is rare, if it occurs at all, and only a few cases have been reported.

In general simple excision of a benign lesion is curative; often excision may be done without the need for an anatomic resection, although the type of resection is obviously dependent on the location of the lesion. Recent advances in minimally invasive techniques for the removal of these lesions makes it less important to look for ways to avoid removing a lesion that may be benign. Wedge excision through a thoracoscopic approach provides adequate therapy without the attendant morbidity of a thoracotomy; patients leave the hospital sooner and return to normal activities in a shorter period compared with that following thoracotomy. Benign pulmonary lesions presenting in an endobronchial location often may be definitively treated with rigid bronchoscopic excision [8].

Multiple pulmonary chondromatous hamartomas are a rare entity and only 16 cases have been described in the world. They often represent a manifestation of Cowden syndrome or Carney triad, but our patient presented none of the concomitant features of these diseases.

Cowden disease is a multiple hamartoma syndrome characterized by mucocutaneous lesions, multiple benign tumors of internal organs and increased risk for breast, thyroid, urogenital and digestive tract cancers. Carney triad is characterized by the association of gastric leiomyoblastoma, pulmonary hamartoma and extra-adrenal paraganglioma [9,10]. In our case none of these diseases were observed.

Radiographically, chondromatous hamartoma presents as a single peripheral nodular lesion, often with calcifications. The presence of fat density in a well demarcated lesion is suggestive of a benign tumor. In the past, needle aspiration biopsies were frequently done in suspect chondromatous hamartoma. However, positive findings are seldom obtained from an aspiration biopsy of hamartoma [1]. The gold standard to diagnose hamartoma remains surgical resection.

References

Fig. 1. CT scan showing multiple chondromatous hamartomas in the right lung.

Fig. 2. Histologic section.
lobe. Medical history of this patient was negative for malignancy, and its calcified CT scan detected a pulmonary nodule, compatible with hamartoma for sive cardiopaty, hypothyroidism and prostatic hypertrophy) in which thoracic HRCT scan detected within the lesion.

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I thank Dr. Filosso for his message and interesting information about the diagnostic assessment and management of solitary pulmonary nodules; however I would stress at least three points:

1. The two smaller lesions were both 1 cm in diameter; according to Lee et al. (Chest 2001; 120:1791-1797) PET false – negative cases frequently occur when the lesion is 1.2–1.5 cm in size.
2. We performed VATS to exclude pleural carcinosis before thoracotomy; we do not perform CT guided anchorage in case of multiple pulmonary nodules because this would require manual palpation of lung parenchyma.
3. Surgical resection is still the best way to histological diagnosis, especially when comparing to CT guided FNAB. We agree on the diagnostic algorithm proposed by Dr. Filosso, when radiological diagnosis is strongly suggestive for hamartomas.