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A lung nodule unveiling a rare primary pulmonary leiomyosarcoma

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Abstract

Leiomyosarcomas are a group of rare heterogeneous malignant tumours, arising from smooth muscle mesenchyme. The tumour may arise at any site, however the uterus, gastrointestinal tract and soft tissues are the most frequently affected. Within the lung, the disease most commonly occurs as a consequence of distant metastasis, however may originate within the lung as a discrete disease entity which is exceptionally rare. We present the case of a 66-year-old gentleman who presented with dyspnoea and was found to have an incidental pulmonary nodule in which a biopsy subsequently confirmed an extremely rare primary pulmonary leiomyosarcoma.

Keywords

Primary pulmonary leiomyosarcoma; pulmonary nodule; lung cancer; emphysema

Introduction

Leiomyosarcomas are a group of heterogeneous tumours which arise from the mesenchyme of smooth muscle. The most common sites of origin include the uterus, gastrointestinal tract and soft tissues. In the lung, the disease most frequently arises as a consequence of distant metastasis from a primary lesion elsewhere, however it may arise as a distinct disease entity within the lung which is extremely rare. Primary pulmonary leiomyosarcoma was first described by Davidsohn *et al* in 1907, and accounts for less than 0.5% of all primary lung malignancies [1,2].

Whilst patients may present with similar symptoms to other more commonly encountered bronchogenic tumours, many patients remain asymptomatic and are identified incidentally. Although radiological investigation plays a pivotal role in establishing the diagnosis, the findings are often non-specific with pulmonary leiomyosarcomas presenting as solitary well-defined nodules, as was the case with our patient [3]. As such a definitive diagnosis can only be established by careful pathological examination of a tumour specimen combined with immunohistochemical staining to ascertain a likely site of origin.

Due to the high prevalence of pulmonary nodules often detected incidentally following routine chest radiography or computerised tomography, this case therefore highlights the importance of considering a primary pulmonary leiomyosarcoma in any patient presenting with a pulmonary nodule, particularly in those patients who enter a nodule surveillance pathway as early detection and resection

has demonstrated more favourable outcomes and improved rate of survival.

Case Report

A 66-year-old male insurance broker was referred to the general respiratory outpatient clinic with a several month history of exertional dyspnoea and a chronic cough productive of clear sputum. He was recently reviewed in primary care and a diagnosis of presumed chronic obstructive airway disease was suspected on the basis of a mild obstructive defect on hand held spirometry, and as such he was commenced on Salbutamol and Tiotropium inhalers.

Apart from dyspnoea and a chronic cough, he denied any other significant symptoms including anorexia, weight loss, haemoptysis or wheeze.

His past medical history included a recent Non-ST-elevation myocardial infarction, which required percutaneous intervention with stenting of the circumflex artery.

His current drug history included Aspirin 75mg once daily, Ramipril 10mg once daily, Bisoprolol 2.5mg once daily, Ticagrelor 90mg twice daily, Tiotropium 18mcg once daily and a salbutamol inhaler used on a *pro re nata* (PRN) basis. In terms of social history he was married and lived with his wife and was fully independent. He denied any exposure to asbestos and was an ex-smoker with an 80-pack-year history.

On clinical examination, he was apyrexial with a blood pressure of 120/85mmHg, pulse rate 85 beats-per-minute, respiratory rate of 16 breaths-per-minute and oxygen saturations of 98% on room air. He was comfortable at rest with no evidence of lymphadenopathy or finger clubbing. Full cardiovascular, respiratory and gastroenterological examinations were unremarkable.

His symptoms improved significantly, after commencing inhalers however, as part of a diagnostic work-up his general practitioner had requested a routine chest radiograph which demonstrated changes consistent with emphysema and a stable peripheral pulmonary nodule in the right lower zone. This had been noted on a previous chest radiograph taken six months earlier and was thought to represent a nipple shadow (Figure 1).

Given his significant smoking history, an urgent computerised tomography (CT) scan of the thorax was requested and demonstrated a 15mm well-defined lobulated lesion within the middle lobe with the presence of surrounding blood vessels. To exclude an arteriovenous malformation a further CT thorax was performed with contrast in the arterial phase and demonstrated no visible enhancement (Figure 2).

PET-CT revealed focused uptake of 12-fluorodeoxyglucose and accumulation in the right middle lobe lesion with a standardised uptake value (SUV) of 3.6 (Figure 3). There was no tracer uptake noted elsewhere and no lymphadenopathy. As such this was thought to represent a primary bronchogenic carcinoma, staged radiologically as T1a, N0, M0.

His case was discussed in the lung cancer multidisciplinary team meeting and a CT-guided biopsy was performed. Histology confirmed an infiltrated core of lung tissue with malignant spindle cells and immunohistochemistry positivity for actin, BCL2, desmin, vimentin with negativity for MNF116, CD34, CK5, CK6, CK7 and TTF-1 (Figures 4-5).

He subsequently underwent a video-assisted thoracoscopic lobectomy of the right middle lobe

and the final histology was consistent with a primary pulmonary leiomyosarcoma which was of bronchial arterial wall origin. He went on to make a full recovery and remains in remission twelve months later.

Case Discussion

Leiomyosarcomas are a group of heterogeneous malignant tumours, which originate in the smooth muscle mesenchyme and most often arise within the uterus, soft tissues and gastrointestinal tract. Primary pulmonary leiomyosarcoma (PPL) of the lung is extremely rare and accounts for less than 0.5% of all malignant lung neoplasms [1].

Within the lung, PPL may originate from the smooth muscle cells of the parenchyma, bronchial walls or pulmonary arteries and is classified according to the site of origin [4]. The intrapulmonary type is the most common type [5].

The disease typically affects middle-aged adults, with a 2.5:1 male to female predominance [6]. Several risk factors have been linked to the development of PPL and include radiotherapy, chemotherapeutic agents comprising of cyclophosphamide, melphalan and nitrosoureas as well as exposure to many occupational and environmental agents including arsenic and herbicides [6].

The presenting symptoms of PPL are similar to many other commonly encountered bronchogenic neoplasms and include persistent cough, dyspnoea, wheeze and haemoptysis alongside constitutional symptoms such as weight loss, anorexia and malaise. Alternatively, patients can remain asymptomatic or present as an incidental finding upon chest radiography, as was the case with our patient.

Radiologically, the features of a PPL are often non-specific, and such lesions typically occur in the lung peripheries as smooth, solitary well-defined nodules or masses [3]. As such, in order to differentiate a PPL from a primary bronchogenic tumour a biopsy is required with careful histological examination and immunohistological staining. Following an excisional biopsy, a diagnosis of PPL should only be considered when there is no evidence of a primary lesion elsewhere. In women, it is essential to exclude a concurrent uterine leiomyosarcoma [2].

Pulmonary leiomyosarcomas demonstrate a wide spectrum of differentiation and histologically are grossly characterised with a firm grey or white surface [4]. Microscopically, malignant spindle cells are observed with slender cigar-shaped nuclei arranged in interweaving fascicles [8]. Low grade tumours are characterised by low mitotic rates and absence of cellular atypia, necrosis and haemorrhage, whereas high-grade tumours have marked cellular atypia, high mitotic rates with significant haemorrhage and necrosis present [7]. Immunohistochemistry plays an essential role in differentiating PPL from primary bronchogenic tumours alongside identifying the source of smooth muscle and usually express positivity for smooth muscle actin, actin, vimentin, desmin and stain negative for carcinoembryonic antigens, cytokeratin, neuroendocrine filaments, S100 proteins and leukocyte common antigens [4].

Due to the rarity of the disease, the treatment of PPL has not yet been standardised. Current treatment modalities include surgical resection, chemotherapy and radiotherapy. For small solitary well-differentiated tumours, if amenable, complete surgical resection is considered curative. For larger tumours, pre-operative radiotherapy may be indicated to decrease tumour size prior to attempting

surgical resection [6]. In those, not amenable to surgical resection, then palliative chemotherapy andradiotherapy remains the modality of choice. Despite this, the prognosis remains poor with 5-year survival approaching only 35% [9].

Figures

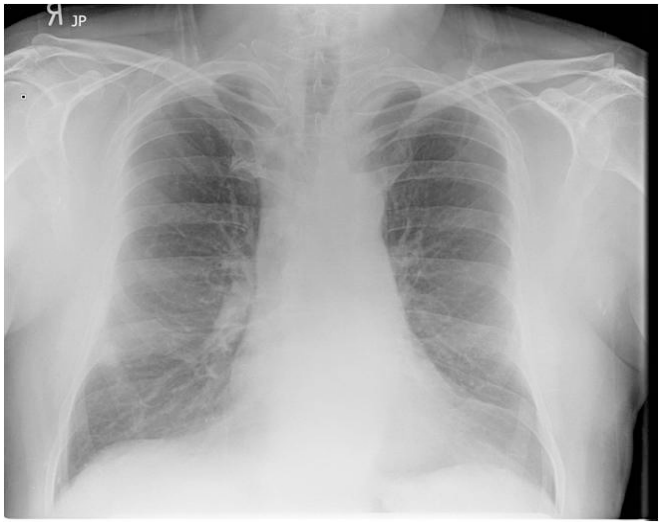

Figure 1: Chest radiograph demonstrating a well-defined right lower zone nodule.

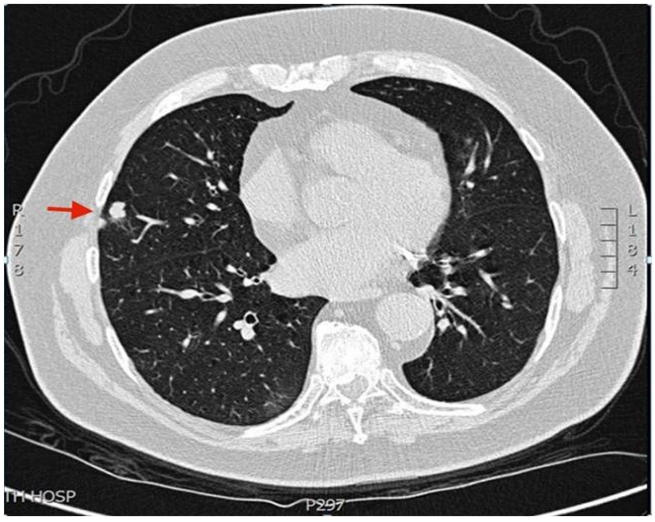

Figure 2: CT Thorax demonstrating a 15mm lobulated nodule in the right middle lobe.

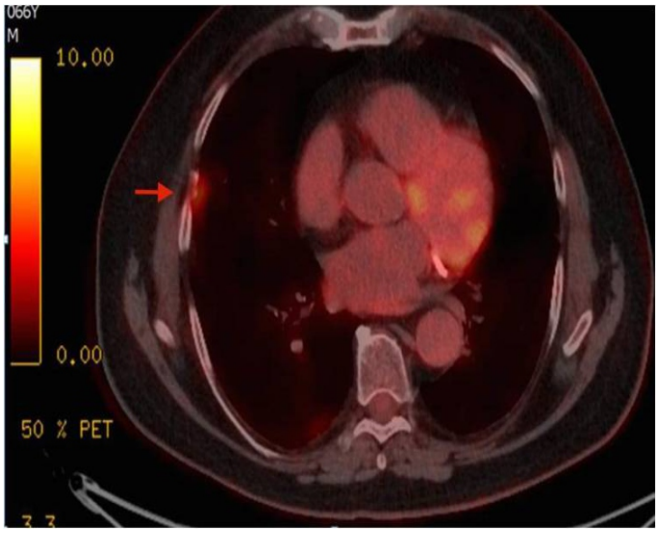

Figure 3: PET-CT revealing avidity of the right middle lobe nodule with an SUV of 3.6.

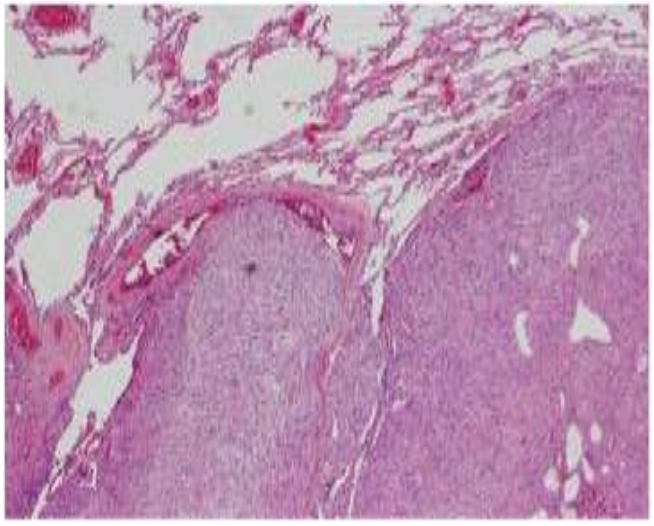

Figure 4: Standard haematoxylin and eosin stain showing a well differentiated grade 1 Leiomyosarcoma.

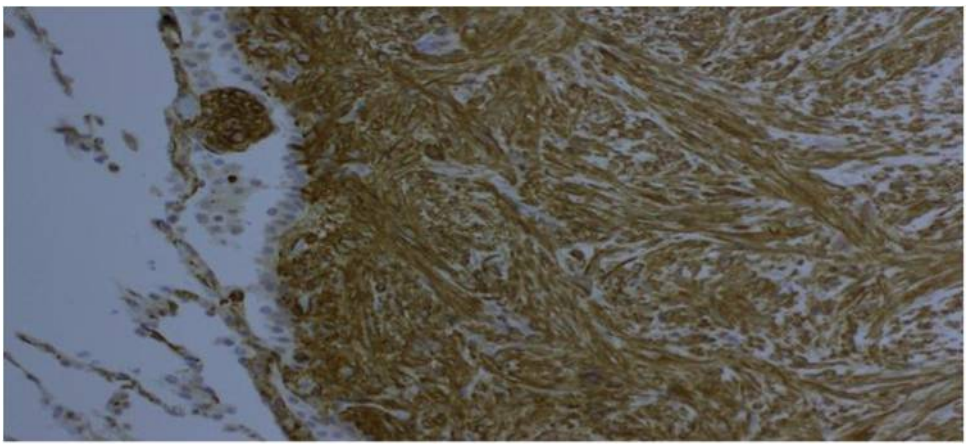

Figure 5: Positive immunostaining for smooth muscle actin.

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