Case report

Incidental endobronchial hamartoma in a patient with enchondroma

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SUMMARY

Hamartomas are the most common type of benign tumours of the lung, constituting a small portion of all lung neoplasms. Hamartomas are rare benign tumours composed of multiple mesenchymal cell lines. Two clinical types have been defined according to the location: intraparenchymal and endobronchial, more frequently the former. We present a case of endobronchial hamartoma causing significant blockage of the right middle lobe. The finding was incidental on a CT scan of the chest done for staging purposes for a large mixed lytic and sclerotic lesion that was found within the proximal-mid portion of the tibial diaphysis. The endobronchial lesion was removed by hot electrocautery snare during bronchoscopy and identified as a hamartoma. Argon plasma coagulation was applied to the lesion's base afterwards and the patient was to follow-up in 3 months for a repeat CT scan.

BACKGROUND

Pulmonary hamartomas are third among possible causes of solitary nodules, accounting for around 8% of them.^{1,2} They are mostly found to be intraparenchymal, with only a small percentage being endobronchial. This case presents a rare occurrence of endobronchial hamartoma and the unique presentation incidentally found in the setting of enchondroma. It is always important to consider the importance of treating endobronchial hamartomas although they are benign tumours. This article serves as a reminder in the workup and treatment of pulmonary hamartomas. Careful and systemic methods of diagnosis are required despite low malignant potential given their propensity to cause symptoms and mimic other malignant lesions.

CASE PRESENTATION

The patient is a 59-year-old African American woman who was evaluated for her chronic left lower extremity pain. Per the patient, she was having chronic pain in her left leg, but recently developed swelling in the area that was concerning. An x-ray of the tibia was performed that revealed a large mixed lytic and sclerotic lesion within the proximal-middle portion of the tibial diaphysis. There was endosteal scalloping suggesting a chondroid matrix.

The appearance was concerning for chondrosarcoma. She subsequently had an MRI that showed a heterogeneous intramedullary lesion in the left tibia

with concerning features of endosteal scalloping, pinning, along with aggressive features of an irregular, lobulated expansion with marked cortical thinning and near disruption anteriorly (figure 1). There were sclerotic changes suggesting a potential osteoid matrix. She underwent a bone biopsy of the lesion and resection that showed cartilage with focal degenerative changes without significant cytologic atypia concerning for chondrosarcoma versus enchondroma and was later radiologically correlated with an enchondroma. At the time, the patient denied respiratory symptoms including shortness of breath and haemoptysis but reported a dry cough without other signs of infection.

INVESTIGATIONS

Due to the initial concern for chondrosarcoma, the patient had a CT chest that demonstrated a right middle lobe bronchus filling defect suggestive of an endobronchial lesion (figure 2). Based on the CT chest findings, the patient underwent a diagnostic bronchoscopy for airway evaluation that showed a polypoid endobronchial lesion from the lateral wall of the right middle lobe bronchus. The lesion was obstructing 80%–90% of the bronchus (figure 3A and B). Polypectomy of the lesion was performed and pathology of the lesion showed findings consistent with hyaline cartilage, calcification, smooth

Figure 1 Coronal cut of MRI of tibia showing a heterogeneous intramedullary lesion (blue arrow) in the left tibia.

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Figure 2 Transverse cut of a CT scan demonstrating a rounded lesion on the main right middle lobe bronchus (black arrow). The lesion is obstructing most of the right middle lobe bronchus.

muscle, fibrous tissues and fat, which were suggestive of an endobronchial hamartoma (figure 4).

DIFFERENTIAL DIAGNOSIS

Prior to the return of pathology, differentials included pulmonary chondroma, hamartoma and malignant lung cancers such as small cell lung carcinoma and non-small cell lung carcinoma.

TREATMENT

Hot electrocautery snare was used to perform a polypectomy of the lesion with full excision achieved. Argon plasma coagulation (APC) was then applied to the base of the lesion (figure 3C).

OUTCOME AND FOLLOW-UP

The patient tolerated the procedure well without complications. Afterwards, the decision was made to perform active surveillance with a repeat CT scan of the chest in 3 months, which showed no evidence of tumour, so a follow-up bronchoscopy was not performed.

DISCUSSION

Pulmonary hamartomas are well known to be the most common benign lung neoplasm with an incidence in the general population estimated to be 0.25% and the male to female ratio of 2:1 to 4:1.³ Studies have shown that hamartomas are true neoplasms

Figure 4 Microscopic view of a right middle lobe lesion (x20) demonstrating hyaline cartilage, calcification, smooth muscle, fibrous tissue and fat.

rather than developmental anomalies.⁴ They usually originate from the proximal part of the bronchial tree. According to Bateson, 80.5% of pulmonary hamartomas were intraparenchymal and 19.5% were endobronchial, although some authors reported an even lower percentage of endobronchial hamartomas.^{3 5} The incidence in the right side of the lung is slightly more than the left side (54.5% vs 45.5%).⁶

Pulmonary hamartomas contain mesenchymal and epithelial elements that differentiate into chondroid, adipose and smooth muscle cells. The feature that distinguishes hamartomas from other benign tumours is that they contain at least two mesenchymal components. The association of these benign tumours with pulmonary and extrapulmonary neoplasms has been mentioned. Carney's syndrome, also known as multiple hamartoma syndrome, is one such instance where chondromatous hamartomas of the lung are associated with gastric and adrenal tumours.⁷ The association of Cowden syndrome with pulmonary hamartoma has not yet been clearly established.⁸

Patients with intraparenchymal hamartomas are usually free of symptoms while endobronchial lesions cause a variety of symptoms due to bronchial irritation and obstruction. Patients may present with cough, fever, wheezing, dyspnoea or haemoptysis. Although rare, the diagnosis of pulmonary hamartoma should be considered as a possibility in cases of bronchial obstruction.⁹ Despite having a very low malignant potential, systemic workup

Figure 3 Bronchoscopic images of: (A) Bronchus intermedius demonstrating a right middle lobe endobronchial lesion (black arrow); (B) right middle lobe bronchus with an obstructing polypoid lesion; (C) right middle lobe bronchus postpolypectomy and application of Argon plasma coagulation.

Figure 5 An algorithm illustrating the treatment of pulmonary hamartomas.

and diagnosis are necessary, as bronchogenic carcinoma must be ruled out.¹⁰ Studies have demonstrated that hamartomas have an association with malignant lung tumours and are reported to have presented with synchronous and metachronous lung cancers.¹¹

Although non-specific, intraparenchymal hamartomas usually present in the form of solitary round pulmonary nodules that may contain popcorn calcifications and vary in size from a few millimetres to 30 cm in diameter. Chest CT typically shows collections of fat alternating with foci of calcification. In fact, CT densitometry is especially sensitive when it comes to diagnosing fat collections or popcorn calcifications.¹² MRI seems useful when neither fat nor calcification is present, which is the case in around 50% of hamartomas.^{12,13} Bronchoscopy may also be helpful for diagnosing and debulking of an endobronchial hamartoma. The histological examination provides a definitive diagnosis. The predominant component is cartilage, followed by fibromyxoid connective tissue, fat, bone and smooth muscle. Slit-like spaces lined by entrapped, benign pulmonary epithelium is a similar feature of both endobronchial and intraparenchymal hamartomas. On the contrary, pulmonary chondromas lack these slit-like spaces.¹⁴

Elsayed *et al* recommended that asymptomatic patients with an intraparenchymal hamartoma mandate close surveillance over surgical resection and that resection should be considered only in patients who are symptomatic or when a diagnosis is questionable.¹⁵ Depending on the size and location of the lesion, procedures can vary from wedge resection to pneumonectomy.¹⁶ Endobronchial hamartomas, unlike intraparenchymal hamartomas, are more likely to cause symptoms. The conventional treatment for endobronchial hamartomas is usually bronchoscopic polypectomy due to their propensity to mimic malignant endobronchial lesions and to prevent chronic post-obstructive lung injury.¹⁶ Laser treatment through rigid bronchoscopy or electrocautery with APC through flexible bronchoscopy are the most common endoscopic methods that are used to treat.^{17,18} In patients whose lung collapse has occurred for almost 4–6 weeks, the chances of getting complications like fibrosis or bronchiectasis are high and these patients would benefit from surgical resection.¹⁹ In patients with recurrent pneumonia, lobectomy

Learning points

- ▶ Endobronchial hamartomas are rare benign neoplasms of the tracheobronchial tree that may present as endobronchial lesions.
- ▶ Careful and systemic method of diagnosis is required despite the low malignant potential, including CT, MRI and bronchoscopy.
- ▶ The primary treatment approach for endobronchial hamartomas is bronchoscopic airway evaluation with further intervention including polypectomy with laser, electrocautery, Argon plasma coagulation and more extensive procedures including surgical wedge resection, lobectomy or pneumonectomy according to the lesion location and extent of postobstructive damage.
- ▶ Follow-up imaging is highly recommended to exclude the recurrence of pulmonary hamartomas.

may be indicated if bronchoscopic intervention is unfeasible.²⁰ Most solitary sub-pleural hamartomas may be removed by enucleation. A wedge resection is indicated if the lesion has become inflamed and involves nearby structures. In cases of irreversible damage of the lung distal to the obstruction, lobar resection or even pneumonectomy may be indicated (figure 5).¹⁸ Aggressive approaches like lobectomy are also considered if the lesion is a central intraparenchymal hamartoma with adherence to the hilum of the lung, or with multiple or large tumours that make wedge resection difficult. Due to the low potential for malignancy, usually a wedge resection is sufficient with recommended follow-up.²¹ Follow-up bronchoscopic examinations are necessary to exclude residual tumour or recurrence.²¹

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