

An unusual cause of chest pain in a young man: bronchogenic cysts and their cardiac manifestations

¹A Magro, ²J Micallef, ³S Falzon, ⁴M Agius, ⁵W Busutil, ⁶S Montefort

¹Basic Specialist Trainee; ²Resident Specialist; ^{3,4}House Officer, Department of Medicine; ⁵Consultant Cardiothoracic Surgeon, Department of Cardiothoracic Surgery; ⁶Professor of Medicine and Consultant Respiratory Physician, Department of Medicine, Mater Dei Hospital, Msida, Malta

ABSTRACT We report a case of a 24-year-old man who presented with chest pain and electrocardiographic evidence of myocardial ischaemia. An abnormal structure located behind the heart on the urgent transthoracic echocardiogram and a computed tomography scan of the mediastinum led to prompt surgery with eventual resection of the lesion. The histology revealed fragments of connective tissue covered by squamous epithelium and ciliated epithelium, consistent with a bronchogenic cyst. The case study is accompanied by a literature review of the pathogenesis, diagnosis and management of bronchogenic cysts and their association with cardiac symptoms.

KEYWORDS Bronchogenic cyst, chest pain

DECLARATION OF INTERESTS No conflict of interests declared.

Correspondence to A Magro,
Department of Medicine,
Mater Dei Hospital, Tal-Qroqq,
Msida MSD 2090, Malta

tel. +31 064 929 1062
e-mail lexmeli@yahoo.com

CASE REPORT

A 24-year-old previously healthy man was admitted to our casualty department with a two-day history of worsening central retrosternal chest pain, diaphoresis and dyspnoea. An electrocardiogram showed ischaemic changes (Figure 1) and a subsequent echocardiogram showed a mass, posterior to the right atrium. The mass, which was not arising from the heart, was impinging on the oesophagus (Figure 2) and right pulmonary arteries. A computed tomography (CT) scan of the thorax revealed a homogeneous, hypodense mass, about 6 cm in diameter, behind the right atrium, and excluded the possibility of an aortic aneurysm (Figure 3). The patient was treated symptomatically with opiate analgesia due to the severity of the pain. A causal connection between the symptoms and clinical findings was assumed and surgery was carried out a few days after admission. Histological examination of the mass showed fragments of connective tissue covered by pseudostratified and squamous epithelium. The presence of ciliated epithelium in some places made bronchogenic cyst the most likely diagnosis. The patient has remained symptom-free to the present day, one year after surgery.

The mechanisms underlying the pathophysiology of the ischaemic changes in our patient are not clear. Given that he has remained free of symptoms post-operatively, the most likely cause of his symptoms was impingement of the cyst on the coronary vessels producing the ischaemic changes. In our literature review we draw attention to the association between cardiac ischaemia and anginal symptoms and the presence of a bronchogenic cyst. Infection is thought to have played a part in the

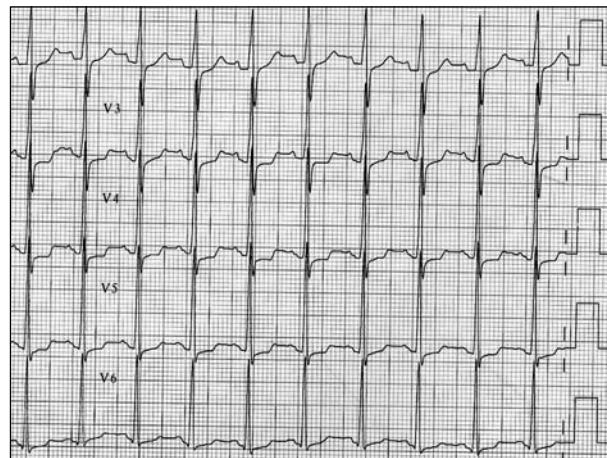


FIGURE 1 Electrocardiogram of a 24-year-old man, who was later shown to have a large retrocardiac bronchogenic cyst, during an episode of chest pain showing ST segment depression in the chest leads V3 to V5.

cyst becoming symptomatic, possibly due to an acute increase in fluid content, although the fluid cultures remained sterile.

Cysts of the mediastinum that are benign masses constitute a small but important diagnostic group, representing 12–18% of all primary mediastinal tumours.¹ The classification of mediastinal cysts is based on their aetiology, encompassing bronchogenic, oesophageal duplication cysts of foregut origin, mesothelial-derived pericardial/pleural cysts and thymic cysts. Bronchogenic cysts are classified as foregut cysts, together with oesophageal cysts, and occur more frequently than oesophageal cysts.¹ The first reported case of a bronchogenic cyst was by Meyer in 1859.²

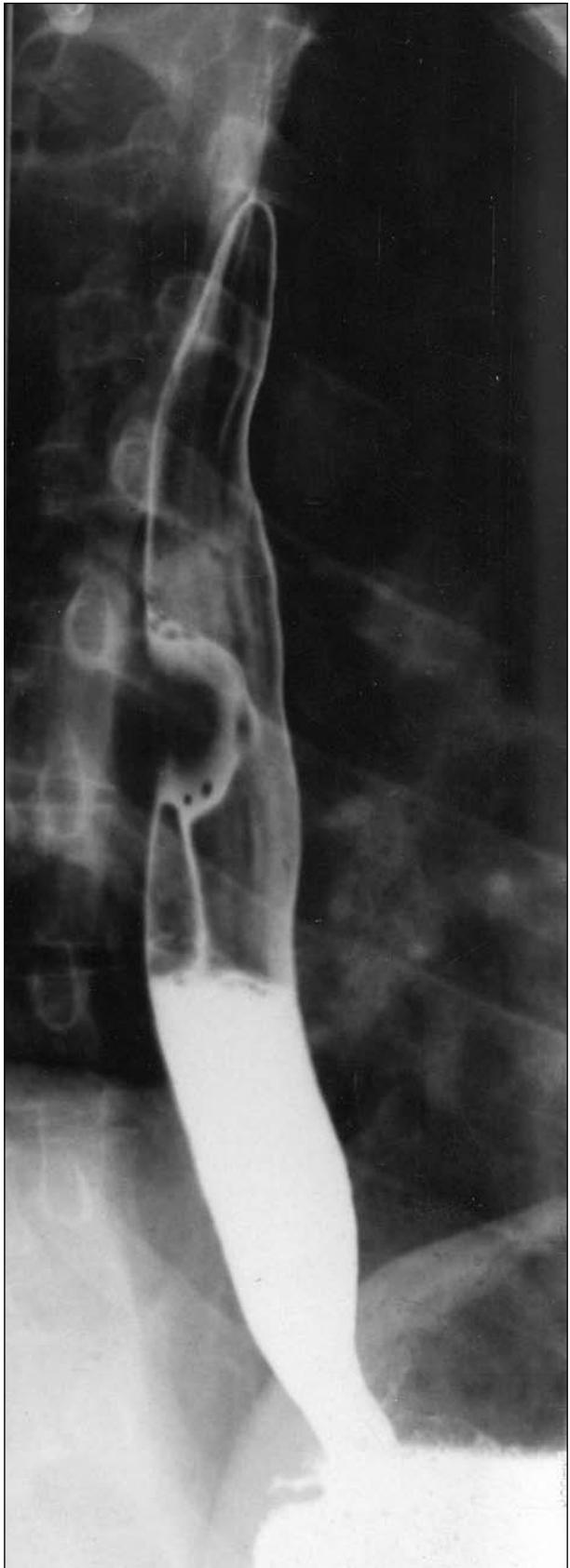


FIGURE 2 Barium swallow performed for associated dysphagia clearly showing a filling defect in the mid-oesophagus. The size of the filling defect underestimates the actual size of the bronchogenic cyst; this could be caused by fibrous adhesions later observed at surgery.

Bronchogenic cysts represent a spectrum of bronchopulmonary malformations that result from abnormal budding of the tracheobronchial tree. The tracheal bud develops from the primitive foregut as a ventral diverticulum around the fourth week of gestation and then undergoes further branching and differentiation.² The location of the cyst depends on the embryological stage of development at which the anomaly occurs. When this abnormal budding occurs early during development, the cyst tends to be located along the tracheobronchial tree. Cysts that arise later are more peripheral and may be located within the lung parenchyma.³ Most commonly, bronchogenic cysts are found along the tracheobronchial tree. However, they have been described in locations such as subcutaneous tissues, the neck, diaphragm, pericardium, spine, abdomen and skin.^{2,3}

CLINICAL PRESENTATION

Patients with a bronchogenic cyst can present with a variety of symptoms. Many patients are asymptomatic, and an incidental finding on chest radiographs is a common mode of presentation. The mean age in a study by Patel et al.² was 37.6 years, and this correlates with other studies. There is an age-dependent trend to the pathogenesis of symptomatic lesions. In the paediatric population, enlargement of cysts frequently leads to life-threatening situations due to impinging on adjacent structures. In adults, cysts tend to remain asymptomatic until there is enlargement secondary to secretion, infection or haemorrhage.⁴

Cough and chest pain are the most common symptoms. Other symptoms include purulent sputum, dyspnoea, fever, anorexia and weight loss, haemoptysis and dysphagia.^{2,3} Other serious reported complications include extrinsic pulmonary artery stenosis, superior vena caval obstruction, pericardial tamponade, arrhythmias, pulmonary oedema, pneumothorax, thrombosis and air embolism.² Carcinomatous and sarcomatous transformation have also been reported.

Acute presentations are very rare in adults, but when they occur they can be life-threatening. There are a small number of cases described in the literature in which cardiac ischaemia is a consequence of a bronchogenic cyst pressing on the coronaries. Patients may present with typical anginal symptoms: chest pain with left upper limb radiation, diaphoresis, nausea or dyspnoea which may even be exercise-induced.^{4,5} Cardiac infarction may be another presentation, and ventricular fibrillation secondary to ischaemia or infarction has also been described.^{4,6} Frequently, in such cases, the bronchogenic cysts were discovered during echocardiography. The age of the patient and the absence of risk factors act as a pointer to an alternative pathogenicity to atherosclerosis and thrombosis.

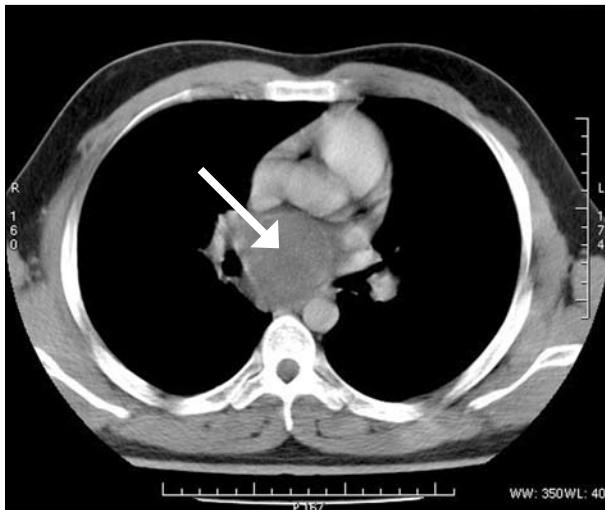


FIGURE 3 A computed tomography scan of the thorax showing the bronchogenic cyst (arrow) as a homogeneous, hypodense mass, about 6 cm in diameter, lying posterior to the right atrium and anterior to the oesophagus.

Bronchogenic cysts have been implicated as a cause of arrhythmias and other electrocardiogram changes in a number of cases. These include conduction abnormalities, atrial fibrillation and signs of altered haemodynamics. The mechanisms of these changes include external pressure from a cyst, internal growth into one of the heart chambers causing obstruction to blood flow, or growth into a septum of the heart causing arrhythmias.^{7–11}

DIAGNOSIS

Bronchogenic cysts must be differentiated from other causes of mediastinal masses, although this can be difficult. In all cases, the definitive diagnosis is through histological examination of a biopsy specimen or after surgery. The differential diagnosis includes other cysts of foregut origin, enteric-oesophageal and neuroenteric cysts, pericardial cysts and masses of thymic origin, thymomas and thymic cysts. Other benign possibilities are benign lymphadenopathy, extension of a cervical goitre into the mediastinum and vascular lesions. The percentage of mediastinal masses which are malignant is higher in children than in adults. These include lymphomas, thymic carcinomas, germ cell neoplasms and neurogenic neoplasms.

The chest X-ray is usually abnormal in patients with a bronchogenic cyst but is generally non-diagnostic, as in the case of our patient. The cyst might appear as a homogeneous mass. When the cyst communicates with the airways, it will appear as an air-filled mass or it might have an air-fluid level. Abnormalities in the surrounding lung parenchyma, atelectasis or consolidation, may occur and may make the diagnosis more difficult.

The CT scan is the investigation of choice as it can demonstrate the size, location and morphology of the

cyst. Generally, cysts have a homogeneous attenuation; with higher fluid levels, however, if they are infected, or if the protein content is high, they will appear much denser. A CT scan and three-dimensional CT reconstruction are useful in outlining the cyst in relation to other structures, and this helps in planning the most appropriate surgical approach. With the increasing availability of coronary CT angiography, the relation of the cyst to the coronaries and the cardiac structure can be evaluated and can shed light on the pathophysiology of the clinical presentation and help in the surgical planning. A CT scan on its own is usually sufficient for locating and characterising the cyst, but magnetic resonance imaging is better at delineating anatomic relations and in definition of the cyst. If the fluid within the cyst is serous, it shows low signal intensity in T1-weighted images and high signal intensity in T2-weighted images. However, if the cyst has a high protein content, it will have a high signal intensity on T1-weighted images.¹²

Fine-needle aspiration (FNA) performed transbronchially or percutaneously can be opted for prior to surgery and may be helpful in distinguishing a malignant from a benign lesion. This procedure may show bronchial epithelial cells, considered pathognomonic in certain quarters.² However, FNA in patients with certain mediastinal diseases unrelated to bronchogenic cysts also frequently reveals bronchial epithelial cells, so this remains controversial.

Other investigations might be needed for the diagnosis and in identification of complications. These include transthoracic and transoesophageal echocardiography, barium swallow, thyroid scans and digital subtraction angiography. Echocardiography is useful to distinguish between a vascular and a cystic structure by means of Doppler colour flow, and sonicated contrast helps in distinguishing a structure as extrinsic to the heart when it is found in such close proximity.⁸

Histopathological examination of the surgical specimen is required for a definitive diagnosis of bronchogenic cysts. The identification of columnar ciliated epithelium is diagnostic. Areas of cartilage, smooth muscle and seromucinous bronchial glands may also be found.¹²

TREATMENT

Transbronchial or transoesophageal needle aspiration is sometimes used as an alternative to surgery.¹³ However, this should be limited to urgent relief of pressure in the acutely compromised or in non-operable individuals. Although relief is effective, recurrence usually occurs.

Surgical excision remains the treatment of choice. This is usually done via a posterolateral thoracotomy or median sternotomy. Other alternatives are removal via media-

stinoscope, thoracoscopic resection and video-assisted thoracoscopic resection. In a series by Jiménez Merchán et al.¹⁴ eight patients with bronchogenic cysts were treated using video-assisted thoracoscopic resection with good results, minimal complications and fast post-operative recovery. Intrapulmonary cysts may require segmental or lobar resection. Complications mostly arise in the presence of adhesions, fistulae, infection and increasing cyst size. It was reported in several studies that greater intra-operative and post-operative complications and difficulty occur in those patients who were symptomatic before the operation, compared with asymptomatic patients.^{2,3} In the study by Patel et al.² and in other major studies, it is documented that it is

common for asymptomatic patients to develop symptoms. It is therefore recommended that even asymptomatic cysts are removed to limit operative morbidity.

CONCLUSION

In conclusion, bronchogenic cysts are rare but must be considered in the differential diagnosis in certain clinical scenarios, especially in young, healthy individuals. There is evidence that bronchogenic cysts can in the long run give rise to disabling symptoms and be lethal. With this in mind, bronchogenic cysts warrant surgical resection, whether symptomatic or not.

REFERENCES

- 1 Takeda S, Miyoshi S, Minami M et al. Clinical spectrum of mediastinal cysts. *Chest* 2003; 124:125–32. doi:10.1378/chest.124.1.125
- 2 Patel SR, Meeker DP, Biscotti CV et al. Presentation and management of bronchogenic cysts in the adult. *Chest* 1994; 106:79–85. doi:10.1378/chest.106.1.79
- 3 Aktoglu S, Yuncu G, Halilcolar H et al. Bronchogenic cysts: clinicopathological presentation and treatment. *Eur Respir J* 1996; 9:2017–21. doi:10.1183/09031936.96.09102017
- 4 Kennebeck GA, Wong AK, Berry WR et al. Mediastinal bronchogenic cyst manifesting as a catastrophic myocardial infarction. *Ann Thorac Surg* 1999; 67:1789–91. doi:10.1016/S0003-4975(99)00340-9
- 5 Hauber J, Rein J, Schaudig G et al. [High-grade coronary stenosis due to a bronchogenic cyst.] *Dtsch Med Wochenschr* 1995; 120:597–602. In German. doi:10.1055/s-2008-1055383
- 6 Azeem F, Finlay M, Rathwell C et al. A near fatal presentation of a bronchogenic cyst compressing the left main coronary artery. *J Thorac Cardiovasc Surg* 2008; 135:1395–6. doi:10.1016/j.jtcvs.2007.09.082
- 7 Mawatari T, Itoh T, Hachiro Y et al. Large bronchial cyst causing compression of the left atrium. *Ann Thorac Cardiovasc Surg* 2003; 9:261–3.
- 8 Kerut EK, Mills T, Helmcke F. Bronchogenic cyst with extrinsic pulmonary vein and left atrial compression presenting as exertional dyspnoea. *Echocardiography* 2007; 24:179–81. doi:10.1111/j.1540-8175.2007.00373.x
- 9 Prates PR, Lovato L, Homsi-Neto A et al. Right ventricular bronchogenic cyst. *Tex Heart Inst J* 2003; 30:71–3.
- 10 Martínez-Mateo V, Arias MA, Juárez-Tosina R et al. Permanent third degree atrioventricular block as clinical presentation of an intracardiac bronchogenic cyst. *Europace* 2008; 10:638–40. doi:10.1093/europace/eun056
- 11 Volpi A, Cavalli A, Maggioni AP et al. Left atrial compression by a mediastinal bronchogenic cyst presenting with paroxysmal atrial fibrillation. *Thorax* 1988; 43:216–7. doi:10.1136/thx.43.3.216
- 12 Limaïem F, Ayadi-Kaddour A, Djilani H et al. Pulmonary and mediastinal bronchogenic cysts: a clinicopathologic study of 33 cases. *Lung* 2008; 186:55–61. doi:10.1007/s00408-007-9056-4
- 13 Dhand S, Krimsky W. Bronchogenic cyst treated by endobronchial ultrasound drainage. *Thorax* 2008; 63:386. doi:10.1136/thx.2007.078071
- 14 Jiménez Merchán R, Congregado Loscertales M, Gallardo Valera G, et al. [Resection of 8 mediastinal bronchogenic cysts by video-assisted thoracoscopy.] *Arch Bronconeumol* 2008; 44:220–3. In Spanish. doi:10.1157/13119542

MYRE SIM GRANTS AND STUDENT BURSARIES

Grants

Applications are invited from Fellows and Members of the Royal College of Physicians of Edinburgh or medical graduates of Edinburgh University for grants to assist eligible individuals to further their professional competence and research activities. Any activity that assists a viable research project will be considered, including payments for secretarial, library or computer facilities or nursing, laboratory or student assistance. A grant may also be given to assist with travel expenses to attend scientific meetings, postgraduate courses, visits to special clinics and other educational activities. Awards will be up to a maximum of £2,000 and are not intended to be salary supporting. A short paper reporting on the research or activity undertaken will be required.

Student bursaries

Applications are invited from medical students of the University of Edinburgh for a limited number of bursaries of up to £750. The bursaries are intended to contribute towards the expenses of an eight-week elective period of study and research, either in the UK or overseas, during an attachment with a physician holding a non-academic post. A short paper reporting on the project undertaken will be required.

Further details and application forms are available from:

Mrs Roselin Combe
Secretary to the Myre Sim Committee
Royal College of Physicians of Edinburgh
9 Queen Street
Edinburgh EH2 1JQ
Email: r.combe@rcpe.ac.uk
Direct tel: 0131 247 3601