[Downloaded free from http://www.ijrc.in on Tuesday, November 15, 2022, IP: 122.180.177.97]

Case report

Chronic thromboembolic pulmonary hypertension in an adult with spina bifida and a ventriculo-atrial cerebrospinal fluid shunt *in situ*

Rajendram R*, Joseph A

Email: rajkumarrajendram@doctors.org.uk

Abstract

A 31 year old man with spina bifida and a ventriculo-atrial (VA) cerebrospinal fluid (CSF) shunt *in situ* presented with sudden onset dyspnoea and pleuritic chest pain. He reported a nine month history of increasing breathlessness. Clinical signs of right heart strain were present. Echocardiography estimated the pulmonary artery pressure to be 86 mm Hg. Computed tomography pulmonary angiogram confirmed acute pulmonary embolism (PE) and chronic pulmonary thromboembolic disease. He was anticoagulated but unfortunately had a cardiac arrest and succumbed. VA CSF shunts were used for the treatment of hydrocephalus between the 1950s and 1980s. Although most VA shunts sited for hydrocephalus in childhood have been removed, some may remain *in situ* in adults. These patients are at risk of PE and development of chronic thromboembolic pulmonary hypertension (CTEPH). Removal of the VA shunt should be considered when patients are shown to be shunt independent. We advise the regular screening of patients with VA CSF shunts for pulmonary hypertension with pulse oximetry, electrocardiography, chest radiography and echocardiography as it is preventable, detectable and treatable.

Keywords: Hydrocephalus, pulmonary thromboembolism, ventriculoatrial shunt.

A 31 year old man with spina bifida presented with acute dyspnoea and left-sided pleuritic chest pain. He also reported a 9 month history of increasing breathlessness on exertion. Normally able to transfer independently and use a wheelchair, he was finding this increasingly difficult.

Past medical history included correction of the meningomyelocele in 1974 at 7 months of age. Progressive hydrocephalus was treated at 8 months with a ventriculo-atrial (VA) cerebrospinal fluid (CSF) shunt. This was subsequently revised at 3 and 10 years of age.

R Rajendram, AKC, BSc (Hons), MBBS (Dist), MRCP, FRCA, FFICM, EDIC
Locum Consultant in Anaesthesia, Intensive Care and Perioperative Medicine, Royal Free London NHS Trust, Barnet and Chase Farm Hospitals, Wellhouse Lane, Barnet, UK

Alex Joseph, MD, FRCA
Consultant, Dept of Anaesthesia and Surgical Intensive Care, Changi General Hospital, Singapore

On admission, he was afebrile, tachypnoeic, hypotensive and tachycardic. Oxygen saturation was 91% on air but improved to 98% with oxygen. Examination suggested right heart strain and pulmonary hypertension with a raised jugular venous pulsation, a left parasternal heave and a loud pulmonary second heart sound. There were no signs of deep vein thrombosis.

The Wells score[1] for pulmonary embolism (PE) was 4.5 [moderate risk; tachycardia (1.5); PE was the most likely diagnosis (3.0)]. Low molecular weight heparin was started empirically for presumed PE. The patient's condition improved and so thrombolysis was not administered.

Full blood count, renal, liver and bone profile were unremarkable. C-reactive protein was elevated (64 mg/L) but D-dimer was massively raised (3234 µg/L). Arterial blood gas (ABG) analysis on air

How to cite this article: Rajendram R, Joseph A. Chronic thromboembolic pulmonary hypertension in an adult with spina bifida and a ventriculo-atrial cerebrospinal fluid shunt *in situ*. Ind J Resp Care 2015; 4:578-80.

[Downloaded free from http://www.ijrc.in on Tuesday, November 15, 2022, IP: 122.180.177.97]

revealed PaO_2 6.7 kPa, pH 7.55, $PaCO_2$ 2.7 kPa, HCO_3^- 18 mmol/L, base excess +10 mmol/L. Electrocardiograph (ECG) showed sinus tachycardia, P pulmonale, right axis deviation, right bundle branch block and right ventricular hypertrophy with strain. Chest radiography demonstrated cardiomegaly, dilated proximal pulmonary arteries and peripheral pruning.

Echocardiography revealed right heart dilatation with right ventricular hypertrophy. Pulmonary artery pressure was estimated at 86 mm Hg. Valvular heart disease and intracardiac shunt were excluded. Computed tomographic pulmonary angiogram (CTPA) confirmed acute pulmonary emboli in the lobar arteries of the left lung and also revealed a chronic saddle embolus in the pulmonary trunk. A 'mosaic' pattern of lung perfusion was also reported. This is highly suggestive of pulmonary hypertension secondary to recurrent pulmonary emboli.[2]

Acute PE and chronic thromboembolic pulmonary hypertension (CTEPH) were diagnosed. The patient was referred for pulmonary thromboendarterectomy, but unfortunately deteriorated suddenly 2 days later. Attempted cardiopulmonary resuscitation was unsuccessful.

Discussion

VA CSF shunts were an established intervention for hydrocephalus from the 1950s until the early 1980s. The mortality of hydrocephalus was substantially reduced. However, VA shunts have many complications including malfunction, infection and thromboembolism.

CTEPH is characterised by organised thromboemboli that obstruct the pulmonary vascular bed. CTEPH may be a complication of acute symptomatic PE but 4% of cases do not present with signs, symptoms or classical risk factors for venous thromboembolism.[4]

In patients with a VA shunt, PE and CTEPH are clinically recognised in only 0.3% and 0.4% respectively.[3] However, at autopsy, the incidence of these complications in children were 60% and 6% respectively.[3] The incidence in adults is unknown.

The increased incidence of PE and CTEPH is not entirely explained by the physical presence of the VA CSF shunt within the right atrium. CSF may alter the pulmonary vascular endothelium resulting in *in-situ* thrombosis, promoting pulmonary hypertension.[3]

In a case series of children treated with CSF shunts for nontumour hydrocephalus, shunt independence could be demonstrated in 3.2% (27 cases).[5] Of these 27 cases, six had hydrocephalus associated with meningomyelocele. Most VA shunts sited for hydrocephalus in childhood will have been removed, but some, as in this case may remain *in situ* in adults. These patients remain at risk of developing CTEPH. Removal of VA CSF shunts should be considered for those who are shunt independent.

Conclusion

The use of ventriculoperitoneal (VP) CSF shunts has reduced the risk of PE and CTEPH in patients treated for hydrocephalus. However, VA shunts remain an alternative treatment for hydrocephalus if a VP shunt is not feasible. This raises issues of screening and treatment for CTEPH. In the absence of any guidelines, we recommend active identification of patients with VA CSF shunts from hospital and general practice records. Removal of the VA shunt should be considered for shunt independent patients. Regular clinical review of patients with VA shunts *in situ* with screening for pulmonary hypertension (pulse oximetry, chest radiography, ECG and TTE) must be done. If pulmonary hypertension has developed, CTPA should be performed. Lung function tests and TTE may be used to exclude other causes of pulmonary hypertension. Patients with CTEPH should be anticoagulated and considered for pulmonary thromboendarterectomy.

References

1. Wells PS, Anderson DR, Rogers M *et al.* Excluding pulmonary embolism at the bedside without diagnostic imaging: management of patients with suspected pulmonary embolism presenting to the emergency department by using a simple clinical model and d-dimer. *Ann Intern Med* 2001; **135**: 98-107

[Downloaded free from http://www.ijrc.in on Tuesday, November 15, 2022, IP: 122.180.177.97]

2. Stern EJ, Swensen SJ, Hartman TE, Frank MS. CT Mosaic pattern of lung attenuation: Distinguishing Different causes. *Am J Roentgenol* 1995; **165**:813-6.
3. Pascual JM, Prakash UB. Development of pulmonary hypertension after placement of a ventriculoatrial shunt. *Mayo Clinic Proc* 1993; **68**:1172-82.
4. Bonderman D, Jakowitsch J, Adlbrecht C *et al.* Medical conditions increasing the risk of chronic thromboembolic pulmonary hypertension. *Thromb Haemost* 2005; **93**:512-6.
5. Iannelli A, Rea G, Di Rocco C. CSF shunt removal in children with hydrocephalus. *Acta Neurochir (Wien)* 2005; **147**:503-7.