Thrombus on the eustachian valve leading to recurrent pulmonary embolism: A rare problem requiring aggressive management

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The eustachian valve is an embryological remnant of the inferior vena cava valve that is absent or inconspicuous in the adult. Even when prominent, it is considered to be a benign finding. The present report describes a patient with deep venous thrombosis who had recurrent pulmonary embolism despite thrombolysis and anticoagulation. He was found to have an adherent thrombus on the eustachian valve and his symptoms resolved completely following surgical thrombectomy. The present report highlights that the eustachian valve can, on rare occasions, harbour pathology and can adversely impact the outcomes of coexisting medical problems such as deep venous thrombosis. Infective endocarditis, pulmonary embolism and systemic embolism via a patent foramen ovale are the major complications of eustachian valve pathology. Transesophageal echocardiography appears to be superior to transthoracic echocardiography in identifying eustachian valve pathology and should be considered in all patients with thromboembolism without a known source.

Key Words: *Eustachian valve; Eustachian valve thrombosis; Valve of inferior vena cava; Venous thromboembolism*

The eustachian valve is an embryological remnant of the inferior vena cava (IVC) valve. It is usually absent or inconspicuous and has no known function in the normal adult. However, on rare occasions, a persistent eustachian valve may be associated with thrombus, infective endocarditis or tumour. Herein, we report a case of recurrent pulmonary embolism from an adherent thrombus on a prominent eustachian valve.

CASE PRESENTATION

A 52-year-old man presented to a peripheral hospital with acute dyspnea. Computed tomography scanning revealed pulmonary embolism with a nonocclusive clot in the main pulmonary artery. A transthoracic echocardiogram (TTE) also demonstrated the thrombus in the main pulmonary artery but revealed no evidence of right ventricular dysfunction. Doppler ultrasonography revealed acute thrombosis of the left femoral vein extending into the left iliac vein and distal IVC, which was confirmed by radionuclide venography. The patient was hemodynamically stable but had mild hypoxia that was easily corrected with minimal supplemental oxygen. Despite the absence of hypotension, respiratory failure or right ventricular dysfunction, the patient was thrombolysed with streptokinase, presumably due to the high clot burden. He received a bolus of 250,000 units followed by 100,000 units/h infusion for 24 h and was subsequently started on

Thrombus dans la valvule d'Eustache à l'origine d'une embolie pulmonaire récurrente : Tableau rare nécessitant une prise en charge énergique

La valvule d'Eustache est un reliquat embryologique de la valvule de la veine cave inférieure qui est absente ou imperceptible chez l'adulte. Même quand elle est proéminente, elle est considérée bénigne. Le présent rapport décrit le cas d'un patient souffrant de thrombose veineuse profonde qui a subi une embolie pulmonaire récurrente malgré la thrombolyse et l'anticoagulation. Ce patient s'est révélé porteur d'un thrombus adhérent au niveau de la valvule d'Eustache et ses symptômes sont entièrement rentrés dans l'ordre après une thrombectomie chirurgicale. Le présent rapport rappelle que la valvule d'Eustache peut, quoique rarement, héberger une pathologie et exercer un impact négatif sur l'issue des problèmes de santé coexistants, comme la thrombose veineuse profonde. L'endocardite infectieuse, l'embolie pulmonaire et l'embolie systémique rendues possibles par un foramen ovale perméable sont les principales complications d'une pathologie de la valvule d'Eustache. L'échocardiographie transœsophagienne semble supérieure à l'échocardiographie transthoracique pour l'identification d'une pathologie de la valvule d'Eustache et elle est à envisager chez tous les patients souffrant de thromboembolie d'étiologie inconnue.

heparin and warfarin. The patient had marked improvement with the above therapy and, within one week, was ambulating comfortably without supplemental oxygen. Complete blood count and the metabolic panel were normal, and evaluation for inherited and acquired thrombophilic states (proteins C and S, antithrombin III, factor V Leiden, and antiphospholipid antibody) was unremarkable. Approximately 10 days after onset of his initial symptoms, the patient developed recurrent shortness of breath and progressively became hypoxic. A repeat TTE showed complete resolution of thrombus in the main pulmonary artery, mild pulmonary arterial hypertension and normal right ventricular function. The patient was then transferred to Nizam's Institute of Medical Sciences (Hyderabad, India) for further workup and management.

Evaluation at Nizam's Institute of Medical Sciences revealed an unchanged chest x-ray and absence of new infiltrates on a computed tomography scan. Repeat Doppler examination revealed resolution of the left femoral vein, left iliac vein and IVC thrombi. This was confirmed by conventional venogram (Figure 1). A subsequent transesophageal echocardiogram (TEE) revealed a small freely mobile mass at the IVC-right atrium (IVC-RA) junction (Figure 2 and Video). The primary diagnostic consideration at this point was a residual adherent thrombus on the eustachian valve. Therapeutic alternatives included repeat systemic thrombolysis, local

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Figure 1 Conventional venogram showing no residual left iliac or inferior vena cava thrombus

catheter-directed thrombolysis, surgical removal or continuation of anticoagulation. Given the risks of repeat thrombolysis, it was preferred to manage the patient conservatively with anticoagulation alone. However, the patient continued to have progressive worsening of dyspnea and hypoxia along with distinct episodes of pleuritic chest pain presumed to be due to recurrent pulmonary embolism. This prompted the decision to proceed with surgery after an in-depth discussion with the patient, his family and the cardiac surgeon. A preoperative coronary angiogram was normal. The patient duly underwent open heart surgery on cardiopulmonary bypass and a reddish-white mass (1 cm × 1.5 cm) attached to the IVC-RA junction was excised. There was no atrial septal defect or patent foramen ovale. Histopathology was consistent with layered thrombus attached to valve tissue (dense plate of collagenous fibrous tissue covered by endocardium). Postoperatively, the patient improved rapidly and returned to baseline functional status within two weeks. At six months follow-up, he remains asymptomatic and continues to be on warfarin.

DISCUSSION

The eustachian valve is named after the famous Italian anatomist Bartolomeo Eustachi, who first described it in the mid-16th century. In the majority of adults, the eustachian valve is either inconspicuous or appears as a thin crescentic endocardial fold originating at the orifice of the IVC. However, there is a large variability in size, shape, thickness, texture, length and the extent of the persistent eustachian valve. Irrespective of its size, a persistent eustachian valve is usually non-pathologic and requires no treatment in the absence of associated cardiac anomalies. Infrequently, a large eustachian valve can cause obstruction at the IVC-RA junction or augment right to left shunting in infants with an atrial septal defect. Other abnormalities of the eustachian valve include myxoma, fibroelastoma and endocarditis (especially in intravenous drug users) (1,2).

Thrombosis involving the eustachian valve is exceedingly rare but has previously been reported. The thrombus may originate in situ or may result from adhesion of embolic material from a distant source (2,3). In either case, there is significant risk of embolization. This is particularly concerning in light of the evidence demonstrating a causal link between a large eustachian valve and patent foramen ovale (4). It is, however, not known whether the risk of clot development or embolism is related to the size, shape or length of the eustachian valve. Adding to the confusion, there has been at least one report of a prominent eustachian valve that proved beneficial by trapping a clot in transit and preventing pulmonary embolism (5). Prompt diagnosis and emergent treatment is therefore necessary to prevent pulmonary or systemic embolization.

Figure 2 Preoperative transesophageal echocardiogram showing the thrombus at the inferior vena cava (IVC)-right (Rt) atrium junction

Available evidence suggests that TEE is superior to TTE in the detection of eustachian valve endocarditis and thrombus (1).

Successful therapy with thrombolytics has been reported (2,3). Our patient was determined to have a eustachian valve thrombus after he received thrombolytic therapy. However, we could not determine whether the thrombus had been present before thrombolysis or had developed later. To our knowledge, there has been no previous report of surgery primarily for eustachian valve thrombus removal in patients without a coexistent cardiac anomaly.

CONCLUSIONS

A persistent eustachian valve is mainly a benign entity but can occasionally be associated with significant pathology such as thrombus, endocarditis or tumours. A eustachian valve thrombus carries grave implications because of the potential for pulmonary and systemic embolism. The possibility of intracardiac thrombus, specifically eustachian valve thrombus, should be considered in all cases of venous thrombosis with recurrent embolic manifestations despite adequate therapy. TEE seems to be superior to TTE in detecting eustachian valve pathology and should be considered in all such patients. Thrombolytic therapy is usually adequate but surgery might be necessary in some patients.

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