Idiopathic Pulmonary Artery Aneurysm Detected with Multidetector Computed Tomography: a Rare but Potentially Lethal Vascular Abnormality

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A 39 year old woman presented with upper respiratory infection symptoms that did not respond to conventional treatment. Physical examination was unremarkable and there was no history of any systemic disease. A chest radiograph was obtained and revealed a left hilar enlargement and normal lungs [Figure 1A]. The patient then underwent transthoracic echocardiography to rule out structural cardiac abnormalities or pulmonary hypertension. The TTE was normal except for mild mitral regurgitation. For further evaluation, she was referred by her physician to our department for chest computed tomography examination. Pulmonary CT angiography was performed using 64-row multidetector computed tomography and revealed saccular aneurysmal dilatation of the left inferior lobar pulmonary artery, associated with small mural thrombus [Figure 1 B and C]. The maximum diameter of the aneurysm was 35 mm. No cardiac or pulmonary cause was found for the pulmonary artery aneurysm.

PAA is a vascular abnormality morphologically characterized by focal dilatation of the vessel involving all three layers of the vessel wall, with or without mural thrombi or wall calcifications. A pseudoaneurysm does not involve all layers of the arterial wall. Pulmonary aneurysms may occur in association with congenital cardiac defects, particularly patent ductus arteriosus. More common causes include vasculitides and alterations in connective tissue (e.g., Behçet disease, Takayasu disease, Marfan syndrome), and pulmonary hypertension [1]. Pulmonary pseudoaneurysms are usually caused by trauma (especially iatrogenic: during placement of a catheter, chest tube insertion, surgical procedures). A less common cause is penetrating chest trauma. Infection with tuberculosis, pyogenic bacteria, and fungi can also cause pulmonary pseudoaneurysm. Malignant lung tumors can cause erosion into the pulmonary arteries and result in pseudoaneurysm formation. Idiopathic PAA without any

Figure 1. Pulmonary artery aneurysm in a 39 year old asymptomatic woman. [A] Chest radiograph show left hilar enlargement (arrows) and normal lungs. Contrast-enhanced axial [B] and 3D volume rendering [C] CT images show saccular aneurysmal dilatation of the left inferior lobar pulmonary artery with small mural thrombi (arrows). PAA = pulmonary artery aneurysm, PT = pulmonary trunk.
associated diseases is a rare lesion and has infrequently been reported. Most patients with PAA are asymptomatic or have non-specific symptoms. Patients with PAA may present with hemoptysis. The mortality rate for patients with a ruptured PAA is very high; thus early and accurate diagnosis of PAA is essential. Owing to its high spatial resolution, contrast-enhanced MDCT is considered the primary technique for diagnosing PAA since it offers a unique opportunity to evaluate the presence, size, shape and exact location of the aneurysm, and concomitant cardiovascular abnormalities [2]. The aneurysms appear as saccular or fusiform areas of dilatation of various sizes, with homogeneous contrast material filling that occurs simultaneously with that in the pulmonary artery. Based on the imaging findings only, the differentiation between pulmonary aneurysms and pseudoaneurysms is very difficult.

In patients with idiopathic PAA, surgical treatment may be considered when the aneurysm is large and when it is associated with pulmonary regurgitation. However, the role of surgery in main pulmonary artery aneurysms is still undetermined. Pulmonary CT angiography could emerge as an ideal non-invasive technique for percutaneous intervention (embolization) or surgical treatment planning. Magnetic resonance imaging can also be used for establishing the diagnosis; furthermore, it may show the arterial wall thickening in connective tissue disease, and provide information regarding blood flow direction in cases of post-stenotic dilatation due to disease involving the pulmonary valve.

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References

Erratum
In the article “Treatment of Crohn’s Disease with Cannabis: An Observation Study” that appeared in the August issue [IMAJ 2011; 13 (8): 455-8], a mistake occurred in the spelling of the third author’s name. The correct spelling is D. Yablecovitch and not D. Yablekovitz as printed.

Capsule
Genetic risk and a primary role for cell-mediated immune mechanisms in multiple sclerosis

Multiple sclerosis is a common disease of the central nervous system in which the interplay between inflammatory and neurodegenerative processes typically results in intermittent neurological disturbance followed by progressive accumulation of disability. Epidemiological studies have shown that genetic factors are primarily responsible for the substantially increased frequency of the disease seen in the relatives of affected individuals, and systematic attempts to identify linkage in multiplex families have confirmed that variation within the major histocompatibility complex (MHC) exerts the greatest individual effect on risk. Modestly powered genome-wide association studies (GWAS) have enabled more than 20 additional risk loci to be identified and have shown that multiple variants exerting modest individual effects have a key role in disease susceptibility. Most of the genetic architecture underlying susceptibility to the disease remains to be defined and is anticipated to require the analysis of sample sizes that are beyond the numbers currently available to individual research groups. In a collaborative GWAS involving 9772 cases of European descent collected by 23 research groups working in 15 different countries, The International Multiple Sclerosis Genetics Consortium & The Wellcome Trust Case Control Consortium 2 have replicated almost all of the previously suggested associations and identified at least a further 29 novel susceptibility loci. Within the MHC they have refined the identity of the *HLA-DRB1* risk alleles and confirmed that variation in the *HLA-A* gene underlies the independent protective effect attributable to the class I region. Immunologically relevant genes are significantly overrepresented among those mapping close to the identified loci and particularly implicate T helper cell differentiation in the pathogenesis of multiple sclerosis.

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Eitan Israeli

“Where is the life we have lost in living? Where is the wisdom we have lost in knowledge? Where is the knowledge we have lost in information?”
T.S. Eliot (1888-1965), American-born British playwright, literary critic and Nobel Prize laureate

“Everybody wants to save the earth; nobody wants to help mom do the dishes”
P.J. O’ Rourke (born 1947), American writer and satirist