CASE-BASED SESSION: SEE THIS CASE AT LEAST ONCE

Saturday 6 December 2014, 10:00–11:00
Location: Agora

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Evaluation of mucopolysaccharidosis cardiovascular disease: About four clinical cases
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Background: Mucopolysaccharidosis (MPS) is an autosomal recessive lysosomal storage disorder. Purpose: To describe cardiovascular disease in four patients with MPS
Methods: Case 1: A 14-year-old boy, was referred to our echocardiographic laboratory for the follow-up of valvulopathy in the context of Hurler–Scheie MPS I. Echocardiography revealed thickening of the mitral valve with increase of the mitral stenosis tricuspid regurgitation and pulmonary hypertension since 2012. Case 2: A 9-year-old boy (Hurler, MPS I) had history of respiratory failure, coarse facial features, otitis, Umbilical and scrotal hernia. we observe an excessive tissue of the mitral valve with mitral and tricuspid regurgitation and pulmonary hypertension. Case 3: A 10-year-old girl (Hurler, MPS I) had history of glaucoma, Corneal opacities, articular steepness, umbilical hernia and carpal tunnel syndrome. Auscultation revealed mid systolic murmur in the apex and we observe an increase in thickening of the mitral valve with regurgitation and stenosis, and pulmonary hypertension. Case 4: A 4-year-old boy with Hunter syndrome (MPS II) an history of articular steepness and umbilical hernia. Echocardiography shows an aspect of left ventricular dilatation with globally reduced contraction, central mitral regurgitation and increased distance between Emitral and septum.
Conclusion: They are common MPS features including cardiac valve disease, coarse facial features, hepatosplenomegaly, joint restriction and corneal clouding. Hurler, MPS I and Hurler–Scheie have common features regarding cardiovascular disease as thickening of the mitral valve with regurgitation and mitral stenosis who differ from the Hunter syndrome, MPS type II.

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Cardiac angiosarcoma presenting as an acute pericarditis in a young boy: the fantom of the pericardium
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A 23 years old young men was referred from a local district hospital to our center for fever and chest pain, exacerbated by inspiration and left lateral decubitus, one week after a gastroenteritis episode. Cardiomegaly was detected at chest X-Ray; ECG showed sinus rhythm and aspecific ST -T abnormalities. Echocardiography demonstrated large pericardial effusion, with no emodinamic instability; regional wall motion and global left ventricular ejection fraction were preserved. Laboratory tests showed an increase in inflammatory markers, while Troponin T was normal. Patient was treated with non steroid anti-inflammatory drugs because of the suspicious of a viral pericarditis. Echocardiography repeated before discharge showed only a slight pericardial effusion behind the right atrium.

One month later the patient underwent a new cardiologic evaluation: he was asymptomatic for chest pain and dyspnoea; no significant changes from discharge were detected at the ECG. Conversely, echocardiography showed an undetected large inhomogeneous mass within the pericardium, compressing and infiltrating the right atrium lateral wall. Subsequent thoracic CT and a cardiac RMN were performed: the presence of a large mass within the pericardium compressing the right atrium was confirmed, associated with increased pre-tracheal lymphonode. Transthoracic biopsy of the pre-tracheal lymphonode was performed and the histological work-up revealed a grade 3 malignant angiosarcoma. Metastasis were excluded and the patient was treated with a double cycle of Epirubicin plus Ifosfamide, with a notable reduction of the mass at the current 6 months follow up.

Cardiac angiosarcoma is a rare clinical entity, with an higher prevalence in men aged 20-50 years old, characterized by a very poor prognosis; survival time from diagnosis is about 6 month. Among patients with pericarditis an undiagnosed malignancy was detected in 4-7% at post-mortem examination. We report the case of a young man with malignant angiosarcoma located in the pericardium that onset as a typical acute pericarditis. We would like to emphazise on a rare but hard etiology of pericardial effusion in young patients, stressing on the importance of repeated echocardiograms, in order to detect a possible late growing (up to 2 months from symptoms) of a malignant cause.
Multi trouble cardiomiopathy - an unusual case of VSD, PDA, DCRV, BAV & CoA coexisting in a young women

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We present a unique case of a young female patient with Eisenmenger’s Syndrome (patent ductus arteriosus - PDA and ventricular septal defect - VSD) concomitant with double-chambered right ventricle (DCRV), severe aortic stenosis due to congenital bicuspid valve and also coarctation of the aorta. It is probably the first reported case of coexistence of these defects in a single patient.

The echocardiogram (Picture A) revealed right heart enlargement (38 mm), and thickened myocardium within the left (16 mm) and right (12 mm) ventricle. The defect was revealed in membranous portion of the intraventricular septum (Picture B), causing the left-right leakage (pressure gradient approx. 180 mmHg). The bicuspid aortic valve was revealed, with the limited leaflet mobility, causing aortic stenosis (APG max = 71 mmHg) and mild incompetence. The tricuspid incompetence was assessed as moderate, but the tricuspid pressure gradient was 113 mmHg.

Cardiac MRI confirmed the left-to-right jet within the membranous portion of the interventricular septum (Picture C), the presence of a 19 mm thick muscular fold within the right ventricular outflow tract (RVOT), which caused the RV lumen narrowing (Picture D). The PDA (9-10 mm in diameter) diagnosis with the flow from the extended pulmonary trunk to the aorta was confirmed (Picture E), also the impaired function of the aortic valve and coarctation of the aorta (Picture F) was shown.

During the right heart catheterization the mean pulmonary artery pressure was 93 mmHg, whereas the pulmonary vascular resistance was 8.56 W.U.

The conservative therapy with Bosentan (0.5-1, b.i.d.) was started. The patient is currently evaluated as NYHA II (6MWT 452m, NT-proBNP 1675.0 pg/ml). In the future she could be considered as the recipient of heart and lung transplant.

Left Ventricular mural endocarditis associated with Hereditary Haemorrhagic Telangiectasia

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Introduction: Heredity haemorrhagic telangiectasia (HHT) is the most common cause of pulmonary arteriovenous malformations (PAVMs), which due to paradoxical embolization may cause cerebral abscess. We report a patient with HHT who developed left ventricular mural endocarditis presenting as an intraventricular mass.

Case Report: A 55-year-old man with HHT was admitted for brain abscess. In spite of appropriate antibiotic therapy, he had persistent fever. Echocardiogram revealed a mobile mass attached to the posterior papillary muscle (PPM) with no valvular involvement, mimicking an intracardiac tumor (Panel A). Surprisingly, the mass grew from 9 mm to 20 mm in five days (Panel B). Because of the high risk of embolization, the patient underwent surgical resection (Panel C) and mitral valve replacement. The wall of mass was very friable, therefore, attempt of excision led to the expression of purulent fluid. Histopathologic examination of PPM revealed the aggregation of leukocytes (Panel D). Blood cultures grew methicillin-resistant Staphylococcus aureus. A diagnosis of infective endocarditis with mural vegetation was made. Giant PAVM causing abscesses was treated by coil embolization (Panel E, F). The clinical course was uneventful after operation.

Discussion: Mural endocarditis secondary to PAVM is extremely rare. Moreover, it is noteworthy that the mass grew at a fast pace. In view of the gangrenous partial necrosis of PPM (Panel C) and internal content filled with purulent fluid, it seems likely that the contraction of surrounding myocardium squeezed out the abscess to the left ventricular cavity.

Conclusion: This case highlights that PAVM provides cause of mural endocarditis in HHT patients.
Angio-CT application in an unusually acute Giant Cell Arteritis case

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Purpose: Giant Cell Arteritis (GCA) is a systemic large vessel vasculitis, with an extracranial arterial involvement described in 10-15% of cases, usually affecting the aorta and their branches. Patients with GCA are more likely to develop aortic aneurysms, but they are rarely present at the moment of the diagnosis.

Methods: We report a case of an 80 year-old caucasian woman, who described proximal muscle pain in the arms with morning stiffness of the shoulders for 8 months. In the 2 previous months, she developed progressively worse bilateral arm claudication, severe pain, cold extremities and digital necrosis. She had no palpable radial pulses and a non-measurable blood pressure.

Results: The patient had a normochromic anemia, an ESR 120mm/h, and a negative infectious and autoimmune workup. The Angio-CT revealed concentric wall thickening of the aorta extended to the aortic arch branches, mainly subclavian and axillary arteries, which were seriously stenotic, with areas of bilateral occlusion; an aneurysmatic formation of the ascending aorta and no involvement of the carotid branches. Despite the corticosteroid therapy there were no resolution of the acute critical ischemia. Subsequently, she was successfully submitted to a surgical revascularization procedure using a bilateral carotid-humeral bypass.

Conclusions: GCA, usually a chronic benign vasculitis, was exceptionally presented in this case as an acute critical upper limb ischemia, resulting from a massive inflammatory process of the subclavian and axillary arteries, treated with a surgical rescue revascularization procedure. Moreover, the simultaneously presence of an aneurysmatic dilation of the ascending aorta is also an unusual feature. This case heightens Angio-CT’s major relevance in this condition diagnosis and treatment.

Abstract 1199 Figure.