Infective endocarditis complicated by rupture of cerebral mycotic aneurysm in a child with obstructive HCM A case report with review of the literature

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Abstract

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A case report with review of the literature

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Abstract

Left ventricular (LV) hypertrophy is defined by a thickness $>$ 15 mm in the adult index case or $>$ 13 mm in the adult related case [1]

In children, it is defined according to a nomogram based on age and body surface area. Hypertrophy is usually asymmetrical predominating over the interventricular septum. [1]

The literature [2,3] , infective endocarditis in patients with hypertrophic cardiomyopathy (HCM) is virtually limited to isolated reported cases or groups of cases, therefore the risk of infective endocarditis during HCM remains poorly defined and thought to be low because HCM is a disorder of the myocardium and not the endocardium.[4]

Spirito et al[1] assessed the occurrence of IE in HCM was 4.3% at 10 years and discerned an HCM subgroup with both obstruction and atrial dilatation prone to develop IE [5]

It is considered low and mostly limited to patients with a subaortic obstruction at rest and or with an intrinsic mitral defect. [2]

The site of the vegetation is usually the anterior leaflet of the mitral valve but vegetations have been reported to occur at the septal level (area of contact with the mitral) and or on the valve [2].

We present a case of infective endocarditis on culture-negative native mitral valve in a previously asymptomatic 10-year-old child with HCM.

This case is not only a rare pediatric HCM with IE, but it also demonstrates the invaluable role of imaging in the diagnosis of HCM and its complications, especially in the case of IE.

Key words :

Obstructive, hypertrophic cardiomyopathy, mitral endocarditis, cerebral mycotic aneurysm, complicated

INTRODUCTION

Infective endocarditis complicating HCM is a rare entity with a <5% 10-year risk of disease progression [1].

The incidence of IE among HCM patients has been described to be 18 to 28 times higher than in the general population and left ventricular outflow tract obstruction (LVOTO) and enlarged left atria have been reported as factors

that increase the risk of IE in HCM [6]

Prognosis is worse in patients with IE associated with HOCM than in patients with IE alone or associated with congenital heart disease [6]

This report describes a pediatric case of HOCM with IE and its cerebral complication

Case presentation

A 10-year-old child with no particular history was admitted to the infectious diseases department for prolonged fever and dyspnea.

On examination he had a temperature of 38.5°C and a systolic murmur at the mitral focus

EKG: sinus tachycardia at 150bpm, systolic left ventricular hypertrophy (figure 1)

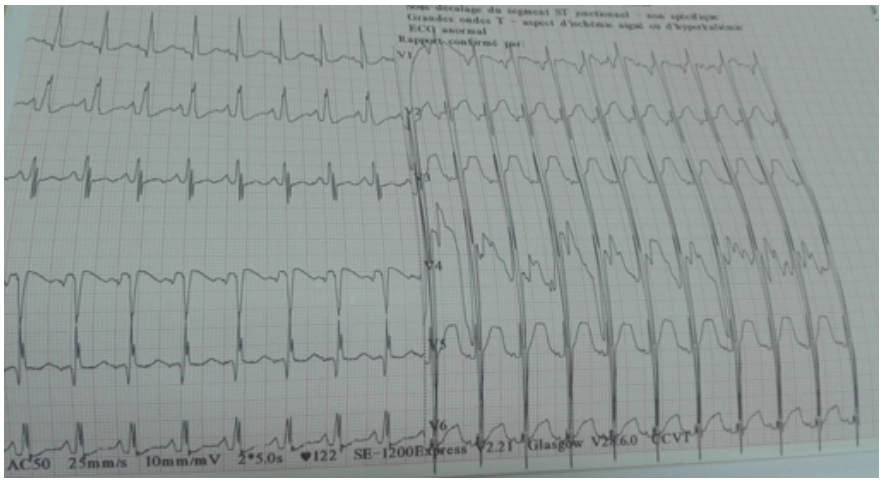

Figure 1 : left ventricular hypertrophy with repolarization disorder and biphasic T wave

On biology: he had a biological inflammatory syndrome with white blood cells at 22000 and CRP 80 with negative blood cultures.

A chest X-ray showed cardiomegaly

A trans thoracic echography showed a left ventricle of low compliance with a parietal hypertrophy located at the level of the basal and medial septum mesuring 21mm (figure 2 and 3)

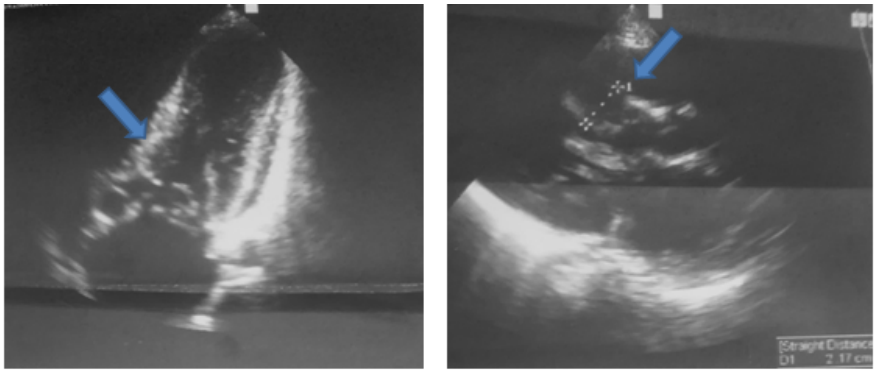

Figure 2 : transthoracic ultrasound centered on the left ventricule (longitudinal view) showing asymmetrical thickening of the left ventricular wall, especially the septum (arrow)

LVEF at 75%.

Voluminous mobile vegetations mutilating the anterior mitral valve measuring a total of 40mm long axis prolapsing into the OG which is dilated with significant mitral insufficiency. (figure 3 and 4)

On the aortic pathway: a septal hypertrophy obstruction measuring 27mm at this level.

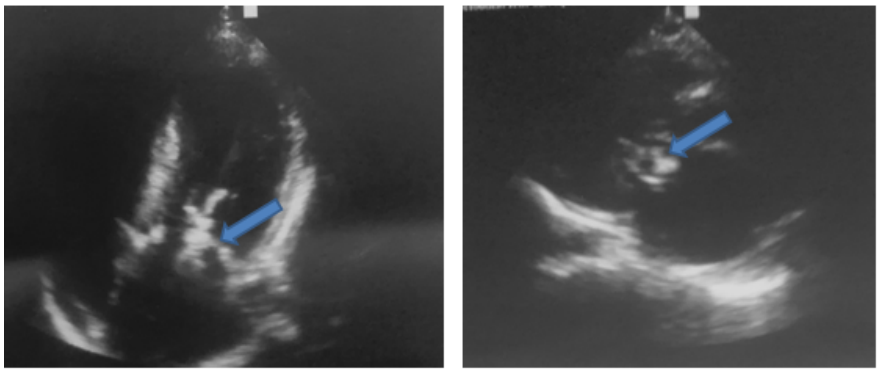

Figure 3 : transthoracic ultrasound centered on the left ventricule (longitudinal a and axial b view in the mitral level) showing vegetation (arrow) on the anterior leaflet of mitral valve

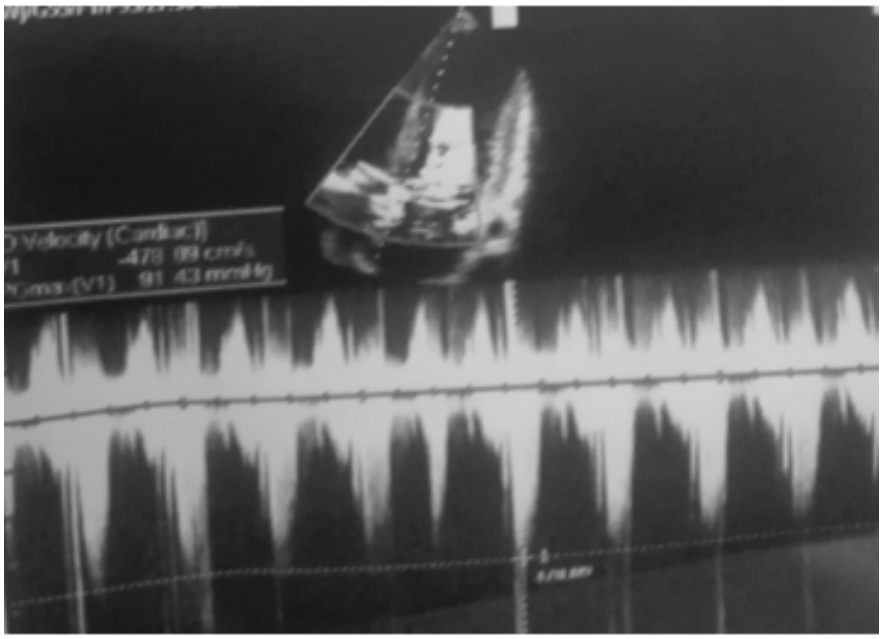

Figure 4 : Pulsed doppler shows significant regurgitation

The patient was treated with ampicillin, oxacillin and gentamicin.

A thoraco abdomino pelvic and cerebral CT scan was performed to assess the extent of the infection.

No notable complication or secondary infectious location was found.

Below are the sections centered on the heart. (figure 5)

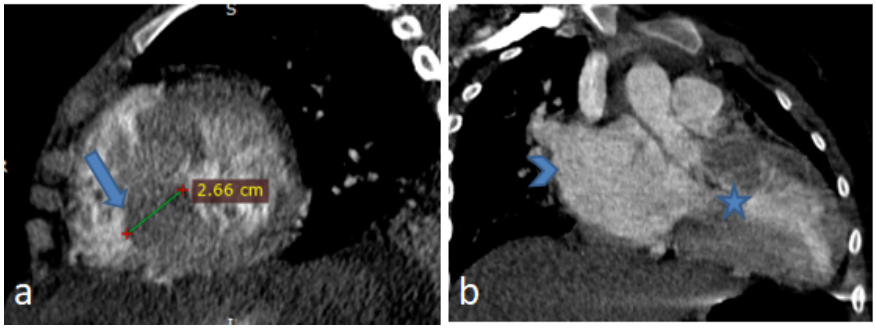

Figure 5 : Reconstructions of the heart : short axis (a) and sagittal oblique showing the hunting chamber (b)

We note the important thickness of the septal wall at 26mm, (arrow) the reduction of caliber of the LV and the hunting chamber (star) with dilatation of left atrium (arrow head)

The patient was operated on day 3 of his admission with mitral valve replacement + septal myomectomy

Then he was put on an intravenous antibiotic treatment (imipenem, teicoplanin and gentamicin)

The evolution was marked by an improvement with apyrexia, regression of the biological inflammatory syndrome and disappearance of the sub-aortic gradient and mitral insufficiency.

During the follow-up :

7 days after the operation, the patient presented a convulsive seizures.

A cerebral CT scan was performed, it showed a cerebral hematoma with quadri ventricular flooding, sub arachnoid hemorrhage and a diffuse cerebral edema responsible for cerebral herniation; sub falcoriel, central, temporal (figure 6).

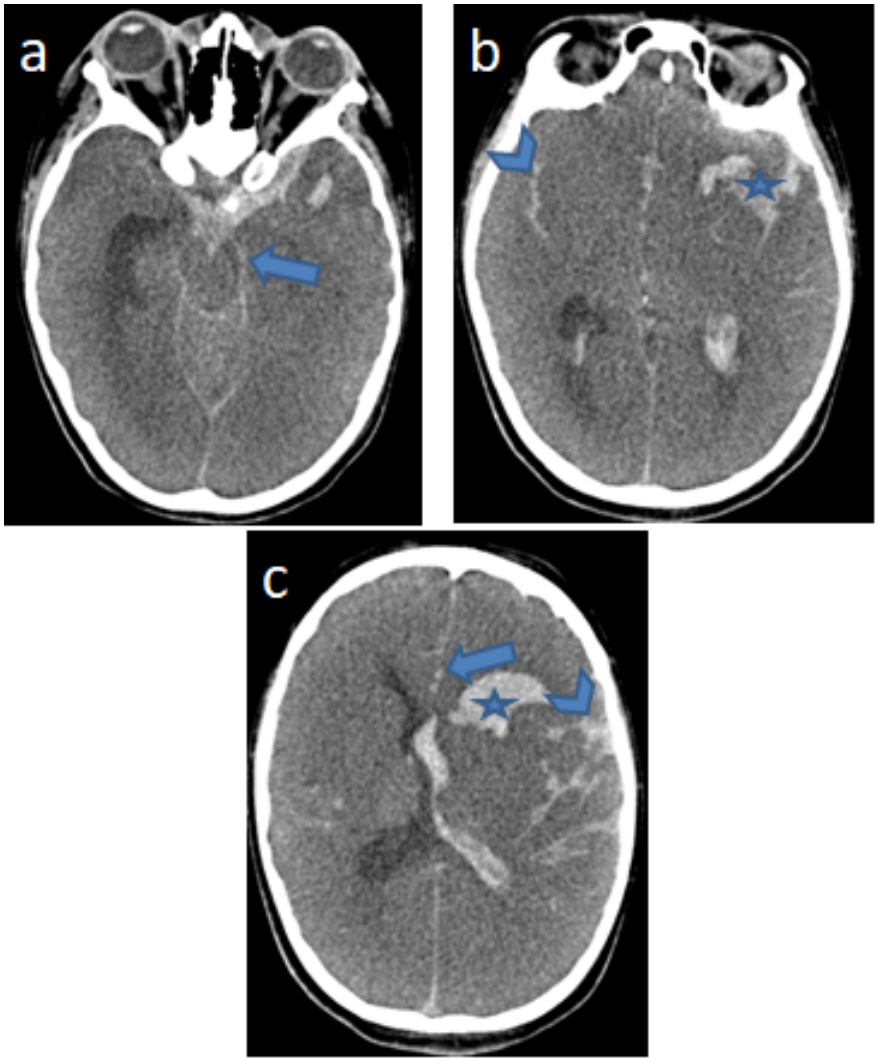

Figure 6 : cerebral CT scan showing : left frontotemporal hematoma (star) with quadri ventricular and sub arachnoidal hemorrhage (arrow head, b,c) with subfalcine (arrow, c), homolateral uncal transtentorial (arrow, a) cerebral herniations

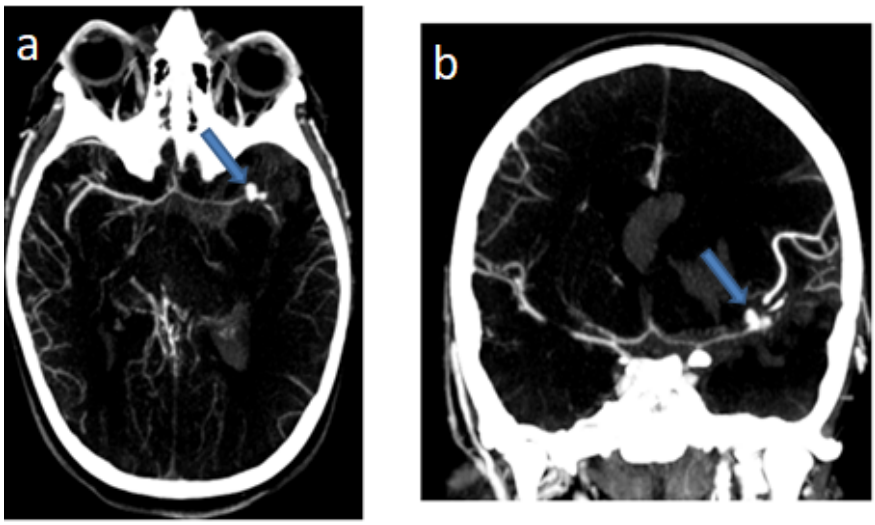

Figure 7 : cerebral CT arteriography , axial plane (a) and reconstruction in the frontal plane with MINMIP (b) showing a saccular aneurysm of the left sylvian artery (arrow)

Arterial phase of CT arteriography was done and showed a ruptured saccular aneurysm of the left middle cerebral artery M2 .(Figure 7)

Discussion

Infective endocarditis in hypertrophic cardiomyopathy is a serious complication associated with high morbidity and mortality.[1]

Although it is not common, it has been reported in the literature, but the data are limited to single cases or groups of cases.

The most cited study was that of Spirito and colleagues who followed the evolution of 810 patients with HCM and the incidence was reported to be 1.4 cases/person/year and the main risk factors were outlet obstruction and OG dilatation [2].

Another chinese study of Wang et al [7] showed that the proportion of HCM patients with IE was 0.19%,with the estimated incidence of 0.15/1 000 person-years in HCM patients.

The mitral valve was the most affected in studies conducted by Spirito et al in 1999 and Fernando et al in 2013 with a percentage of 70 and 71% respectively [2]

An obstructive HCM was present in all cases of infective endocarditis [10] with a statistically significant association between obstruction and infective endocarditis in the study of Spirito et al [2] but the multicenter, prospective, cohort study of Dominguez et al [3] and Sims et al [8] found a similar incidence of IE with or without left ventricular outflow tract obstruction (LVOTO). Nevertheless, the small sample size could be just a statistical outlier, limiting its universality . In addition, the latter study found no clinical difference between infective endocarditis in HCM patients with and without obstruction..

Because HCM is a disorder of the myocardium and not the endocardium, it has been considered to have low risk of IE. In fact the occurrence of infective endocarditis in HCM patients can be explained by endocardial lesions secondary to turbulent flow during ejection and contact between the anterior leaflet of the mitral valve and the septum during systole which is called « Venturi effect » , thus direct trauma resulting from septal-anterior mitral leaflet contact may predispose to infection and vegetation formation [2,4]

We note that the vegetation in our case was indeed located on the anterior leaflet of mitral valve.

The most common germs found in the literature were staphylococcus aureus and streptococcus , with this latest being more found among native-valve HCM patients [2, 3].

The germ in our case was not identified with blood culture remaining negtive.

Mitral valve replacement surgery and septal myomectomy are the two accepted therapeutic methods for symptomatic obstructive HCM refractory to medical treatment but the larger series on the surgical treatment of HCM suggests that the two operations are rarely combined [5,6].

We recall that our patient had both operations at the same time.

The occurrence of a complication such as rupture of an intracerebral mycotic aneurysm with hematoma, as was the case in our patient, is very rare and limited to a few isolated cases reported in the literature [7, 8], in a recent study of Jason R Sims et al [8], symptomatic embolic complications occurred in 33% of cases.

Conclusion

Infective endocarditis is an uncommon but serious complication in HCM that can occur in patients with or without LVOTO. Septic embolization and valve lesions are common in these patients. IE patients with HCM might have a poor prognosis compared to those without HCM and should receive cardiac surgery as early as possible.

Mortality is high but similar to that found in patients with or without indication for endocarditis prophylaxis. However, predisposing factors and Streptococci infections are more frequent among native-valve HCM patients.

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