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Left atrial myxoma complicated with a myocardial infarction in a 10-year-old boy: A case report and review of the literature

Ahmed Sghaier, Nejeh Ben Halima, Ghada Hamila, Houssem Thabet, Marwa Ben Abdallah, Yossra Messaoudi

Cardiology Department of Ibn El Jazzar University Hospital, Kairouan, Tunisia

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SOUHRN

U desetiletého dítěte došlo k rozvoji námahové synkopy, jíž předcházely bolest na hrudi a palpitace. Fyzikální vyšetření nepřineslo žádné zvláštní nálezy až na diastolický roll v oblasti mitrální chlopně. Iniciální elektrokardiogram prokázal komorovou tachykardii úspěšně zrušenou kardioverzí. Kontrola úpravy elektrické aktivity srdce prokázala rozsáhlý přední infarkt myokardu. Emergentní dopplerovské echokardiografické vyšetření prokázalo dilataci levé komory, akinetické apikální a střední segmenty anterolaterální stěny a srdečního hrotu se sníženou ejekční frakcí na 35 %. Hmota vyplňující téměř celou levou síň připomínala myxom. Angiografické vyšetření hrudníku a koronárních tepen diagnózu potvrdilo; rovněž prokázalo ischemii přední stěny a nepostižené koronární tepny. Konečná diagnóza zněla myxom levé síně komplikovaný koronární embolií. Byla provedena chirurgická resekce; následné histopatologické vyšetření diagnózu potvrdilo. Rešerše literatury o myxomech levé síně komplikovaných koronární embolií u dětí, kterou jsme provedli pro účely tohoto článku, prokázala, že uvedená kombinace se sice vyskytuje vzácně, je však závažná a vyžaduje značné zkušenosti, aby bylo možno stanovit správnou diagnózu a léčbu neodkládat.

Keywords:
Child
Coronary embolism
Myocardial infarction
Myxoma

ABSTRACT

A 10-year-old child experienced syncope on exertion, preceded by chest pain and palpitations. The physical exam had no particularities except for a diastolic roll at the mitral area. The initial electrocardiogram showed a ventricular tachycardia successfully wiped out by cardioversion. The electrical control showed an extensive anterior myocardial infarction. Emergency Doppler echocardiography showed a dilated LV, akinesia of the apical and middle segments of the anterolateral wall and of the apex with a reduced LVEF of 35%. A mass covering almost all of the left atrium was noted evoking a myxoma. A thoracic angiography scan and a coronary-scan confirmed the diagnosis, showed anterior cardiac ischemia and normal coronary arteries. The final diagnosis was a left atrial myxoma complicated by a coronary artery embolism. The child underwent surgical resection. Histopathological examination confirmed the diagnosis.
In this paper, a review of the literature concerning left atrial myxomas complicated with coronary embolisms in children showed the rarity but severity of such association, thus requiring a high degree of clinical awareness to obtain a timely diagnosis and not to delay the treatment.

Address: Ahmed Sghaier, Cardiology Department of Ibn El Jazzar University Hospital, Kairouan, Tunisia, e-mail: Sghahmed991@gmail.com
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Introduction

Myxomas are frequent in children. They are more common in the left atrium. Systemic embolization is a complication that changes the prognosis. Embolization in the coronary arteries is quite rare but may cause myocardial infarction.

Observation

A previously healthy 10-year-old child experienced a syncope occurring during exercise associated with palpitations and an acute anterior chest pain radiating to the upper left limb. History noted no previous complaints including no on exertion angina and no other episodes of syncope.

The physical exam found a pale child. Heart rate was at 130 bpm, respiratory rate was at 25 cpm and blood pressure at 90/60 mmHg.

On the initial electrocardiogram (ECG), there was a ventricular tachycardia wiped out by external electric shock (Fig. 1). The electrical control noted an anterior ST segment elevation (Fig. 2). The chest radiograph showed no cardiomegaly. Biology was also normal except for high troponins. Based on the second ECG, the first diagnosis evoked was abnormal coronary artery birth.

The patient was placed on anticoagulant and anti-platelet aggregation therapy. An urgent Doppler echocardiography showed a dilated LV with an altered LVEF estimated at 35% in relation to an akinesia of the apex and of the apical and middle anterolateral wall segments. We noted the presence of a 3 cm hyperechoic mass covering almost all of the left atrium, partially obstructing the mitral orifice without any regurgitation of the latter. The tumor was mobile, and had a base implantation at the level of the septal atrial wall (Fig. 3).

A thoracic CT-scan confirmed the presence of a tissular mass in the left atrium, with a heterogeneous faint contrast enhancement evoking a myxoma. The coronary angiography eliminated a coronary artery birth anomaly and no intra-arterial obstruction was noted (Fig. 4).

The most probable diagnosis was a left atrial myxoma complicated with arterial coronary embolism.

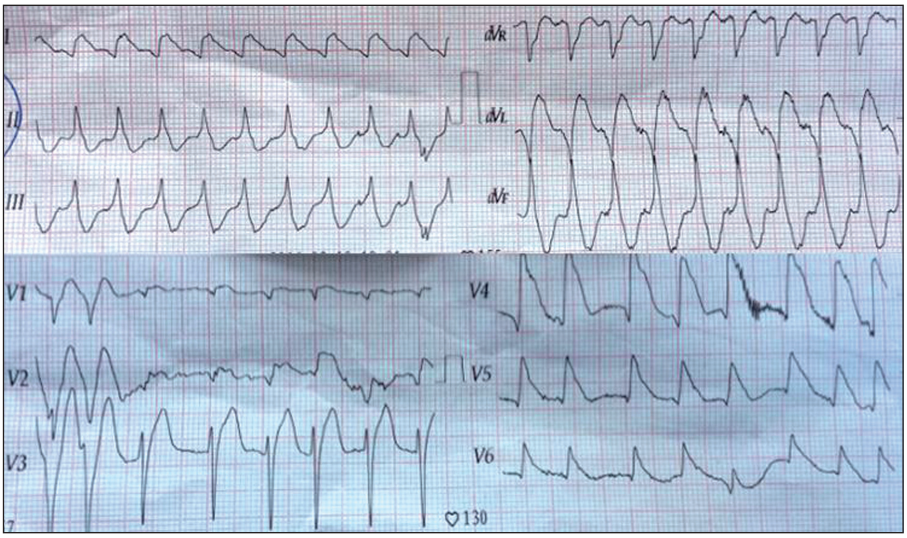

Fig. 1 – An ECG showing a ventricular tachycardia of 130 bpm.

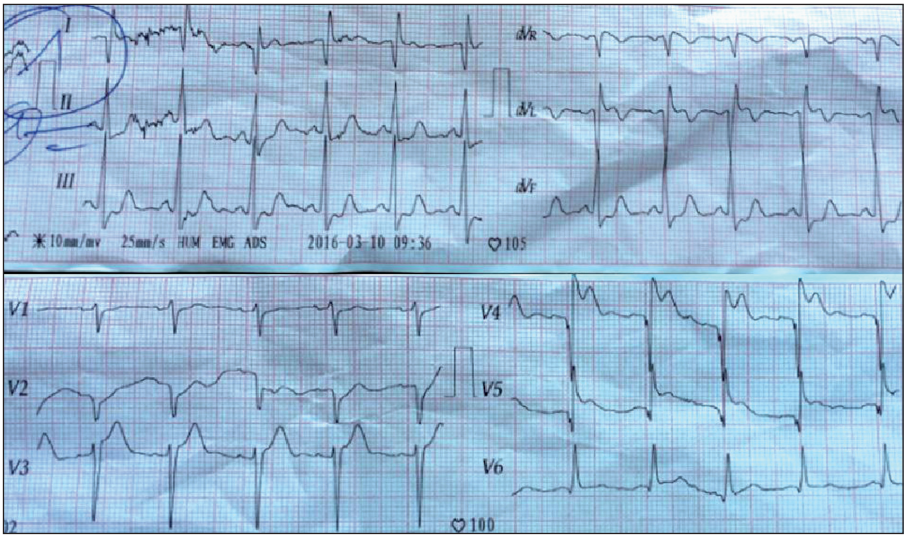

Fig. 2 – 2nd ECG: (after electrical choc) showing an anterior ST elevation myocardial infarction.

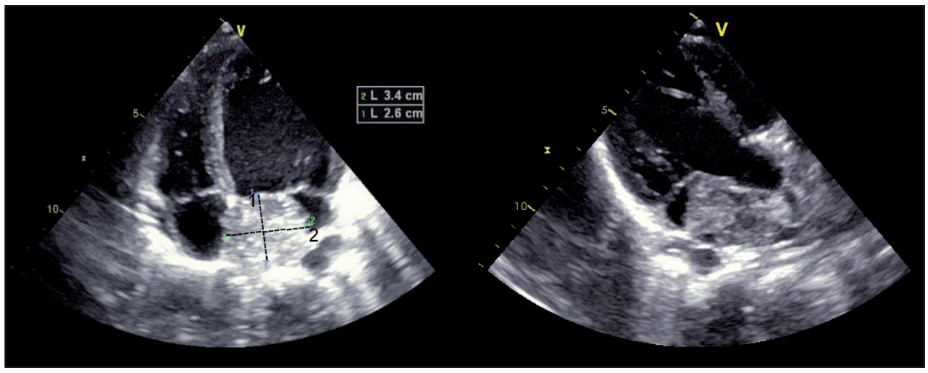

Fig. 3 – Transthoracic echocardiography showing a left atrial hyperechoic mass of 3.4 × 2.6 cm.

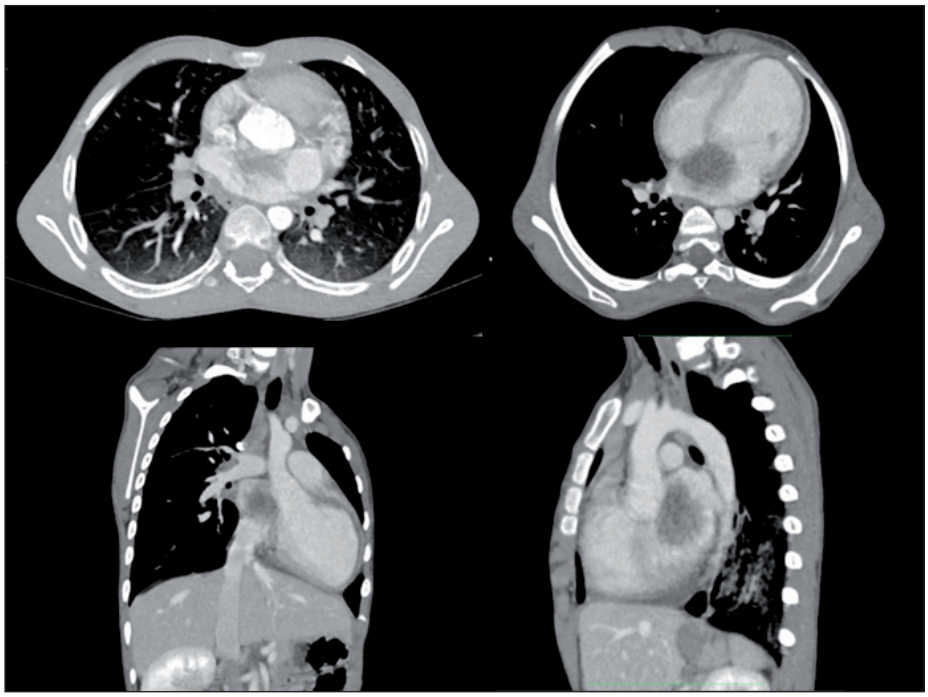

Fig. 4 – Different acquisitions on thoracic CT scan showing an intracardiac mass.

Given the presence of a large tumor with serious embolic event, the decision was to operate the child to prevent further episodes. Complete surgical resection was performed on day 3 of admission showing a gelatinous and friable left atrial mass. Per-operative coronary exploration had found a free left coronary artery. The surgical follow-up was simple and the anatomopathological exam confirmed the diagnosis of myxoma.

After a 1-year follow-up the patient showed no chest pain or palpitations. A stress-test was negative. Holter ECG lacked rhythm disorders but on TTE, we noted a persistent anterolateral myocardial infarction sequela with a LVEF at 42%.

Discussion

In adults, myxomas are the most common primary cardiac tumors. On the other hand, in children aged 1–16 years, rhabdomyomas are by far the most frequent, accounting for nearly 80% of the tumors in the Toronto series,[1] and then at about 15% each, comes fibroma, myxoma and teratoma and more rarely, hemangioma, mesothelioma and hamartoma.[2]

More common in the left atrium, myxomas originate from the mesenchymal cells typically located in the oval

Table 1 – Reported cases of myocardial infarction due to left atrial myxoma in the English literature in patients aged under 18 years old during the period of 1978–2016.

Case number	Year	First author	MI territory	Site of coronary obstruction	Age	Sex
1	1978	Tanabe J[15]	Inferior	Retroventricular	11	M
2	2003	Harikrishnan S[16]	Infero-lateral	No obstruction	9	F
3	2004	van Gelder HM[17]	Anterior	Cironflex	15	M
4	2017	Actual case	Anterior	No obstruction	10	M

pit.[3] Atrial myxomas occur predominantly sporadically but as an autosomal dominant inheritance familial form in less than 10% of the cases.[4]

Cardiac myxomas are usually diagnosed on echocardiography which often shows a heterogeneous hyperechogenic mass moving through the mitral valve.[5] Computed tomography and MRI can help determine the location and the extent of the tumor.[6]

Although the clinical presentation may be polymorphic, the majority of patients show symptoms related to Goodwin's triad including embolic phenomena, hemodynamic disorders due to flow obstruction, and vague constitutional symptoms or complaints.[7]

The syncope in our patient can probably be explained by a sudden and rapid obstruction of the mitral orifice. This was advanced by Gleason in his paper published in 1951.[8]

Systemic embolization may occur in 30–50% of patients with left atrial myxomas.[9] Embolization can occur in almost all organs, but in about half the cases, it is the central nervous system that is involved.[10] Occlusion of the coronary arteries is rare, but may cause acute myocardial infarction (AMI).[11,12]

Embolic fragments generally consist of myxomatous tissue and may also be linked to a thrombus dislodged from the surface of the myxoma.[13]

This case illustrates a very rare occurrence of an acute myocardial infarction in a pediatric patient. Management of this condition which is commonly encountered in adults but seldom seen in children was challenging. There are no controlled trials or specific recommendations to guide early treatment in this age group and the use of thrombolytics in children is commonly met with anxiety and an appropriate concern for bleeding complications, limiting the number of children who may benefit from early therapy. However, one recent paper showed that, when diagnosed in time, intravenous coronary thrombolysis must be the therapeutic strategy for AMI in children and severe complications are uncommon and do not require special facilities.[14]

A review of the English literature from 1978 to 2016 concerning left atrial myxomas embolized in coronary arteries in individuals under the age of 18 years old, showed the presence of only 3 cases. The characteristics of each of them and those of our patient are summarized in Table 1.

A clear male predominance is noticed. All patients were surgically treated with complete resection of the tumor. Indeed, once the diagnosis of left atrial myxoma is established in a patient with previous embolic events, rapid surgery is indicated to prevent further episodes.[18]

Recurrence of left atrial myxomas is rare.[19] Incomplete excision, however, may result in such phenomenon. Despite total excision, serial echocardiographic follow-up is recommended for patients with this disease.

Conclusion

Embolic events are one of the most frequent complications of a myxoma. This diagnosis should be evoked in front of an acute coronary syndrome in young patients. A high degree of clinical awareness is necessary to obtain a timely diagnosis and to not delay the treatment. Surgical resection could prevent potential embolic episodes.

Conflict of interest
None declared.

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Ethical statement
Authors state that the research was conducted according to ethical standards.

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