INTRODUCTION

Beta thalassemia major is a hereditary anemia secondary to deficient production of the beta-globins in structurally normal hemoglobin, with a consequent precocious destruction of red globules present in the circulation. Coagulation and cardiac complications due to the accumulation of iron are the main causes of death in these patients.

CLINICAL CASE

The authors present a case of cardiac iron overload complicated by fatal multi-organ embolism due to a left ventricular thrombus in a beta-thalassemia major patient.

DISCUSSION

The presence of chronic hypercoagulability related to pro-thrombotic anomalies in beta-thalassemia major patients may be complicated by thrombotic embolism events. On the basis of our experience, we believe that to prevent rapid deterioration of the patient to fatal multi-organ embolism secondary to thrombolytic detachment it’s advisable to perform systemic thrombolysis rather than surgery for a single occlusion which precludes the use of thrombolytic agents.

KEY WORDS: beta-thalassemia; echocardiography; thrombophilia, multi-organ failure, ferrochelate treatment.
tion presented revealed reduced compliance to fer-
rochelate treatment. The patient was conscious in
the emergency room, and well-oriented in space
and time (GCS 15). A chest exam revealed a
reduced vesicular murmur in all pulmonary fields
during breathing, and SpO2 97% in ambient air. The
ECG revealed sinus rhythm with widespread anom-
alies of the ventricular repolarization. The ECG
also revealed normal dimensions of the left ven-
tricle, mild systolic dysfunction (injection fraction
47%), with conserved diastolic function, normal
arterial pulmonary pressure (27mmHg), and an
absence of pericardial effusion. Myocardial necro-
sis enzymes were normal, as were inflammation
indices.

In contrast, serum ferritin was particularly
high (10,065ng/mg). A standard chest x-ray demon-
strated a normal cardio-thoracic ratio. Hence, a
diagnosis of moderate heart failure secondary to left
ventricular systolic dysfunction was proposed, due
to cardiac iron overload in a thalassemia patient.
The patient was then admitted to the Cardiology
department of our hospital, and on the second day
the patient complained of a sudden and intense pain
in the iliac fossa and the left side. An ultrasound
and CT of the abdomen were performed, and
revealed hepatic iron overload and hydro-abdomen.
The chest CT also revealed the presence of an apic-
al thrombus of the left ventricle that was not pre-
sent on the admission ECG, and was confirmed by
a new ECG immediately after the CT scan, which
showed the presence of a pedunculated and mobile
thrombus adhering to the apex of the left ventricle.

Anticoagulant therapy was then started by
administering low molecular weight Heparin 5000,
combined with Warfarin 1 cp/day, until reaching an
INR value over 2, followed by oral anticoagulant
alone(46). About 24 hours after the thrombus episode
in the left ventricle, a generalized abdominal pain
suddenly appeared in the lower limbs (which were
cold to the touch, and peripheral pulse was absent),
and associated signs of shock. An urgent cardiac
U.S. revealed a disappearance of the apical throm-
bus previously observed. An angio CT revealed an
occlusion of sub-renal abdominal aorta with a bilat-
eral extension at the common iliac arteries. So the
patient underwent urgent percutaneous endovascu-
ar treatment with placement of an endoprosthesis.
Immediately after the operation multi-organ failure
occurred due to systemic embolism until the death
of the patient.

Discussion

Transfusion therapy improved the patient's prog-
nosis significantly, reducing both the hepatic-
splenomegaly from extra-marrow erythropoiesis and
the bone deformations due to abnormal hematopoi-
esis marrow. The iron overload due to the
continued transfusions and increased intestinal
absorption could not be completely eliminated and
was the main cause of morbidity and mortality in this
patient(4). Iron deposits develop in many organs caus-
ing tissue damage and reduced capacity for catalysis
of free radicals(10). The serum ferritin level is the most
used indicator to evaluate iron deposits; however, that
lab value is only approximate and becomes unreliable
in the presence of hepatic insufficiency(4). Cardiac events secondary to iron overload are
the main cause of death in thalassemia patients(2,3).
Heart failure due to iron overload usually develops in
patients with less than optimal ferrochelate therapy
and multiple endocrine pathology. The clinical course
of heart failure is extremely variable, from sudden
death in patients with insufficient chelating therapy to
sporadic cases in which optimization of chelating
therapy causes a regression of cardiomyopathy(7).

In addition, the existence of pro-thrombotic
anomalies of hemostasis in these patients (low levels
of C and S protein, endothelial activation and
platelets, high levels of thrombin-antithrombin III),
have led to recognition of the existence of a chronic
state of hypercoagulability, more serious in
splenectomized subjects. This condition corre-
lates with a higher than normal incidence of throm-
embolic events, that principally consist of cerebral
ischemia, deep vein thrombosis, and pulmonary
embolism(45). In our patient anti-coagulant prophylax-
is before the operation and after anti-thrombolytic
therapy as indicated in guidelines(8), was not sufficient
to prevent systemic embolism. Hence, we believe that
in beta-thalassemia patients it's advisable to perform
short-term systemic thrombolytic therapy with a cuta-
neous plasminogen activator rather than surgery for a
single occlusion that can preclude the use of throm-
bolytic agents.
References


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