Thrombolytic treatment as first option in recurrent tricuspid prosthetic valve thrombosis and Ebstein's anomaly

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ABSTRACT

Ebstein's anomaly is the most frequent cause of congenital tricuspid regurgitation. The coexistence of a mechanical heart prosthesis in a low-pressure circuit and poor compliance in the anticoagulant therapy contributed decisively to the appearance of recurrent mechanical heart valve thrombosis in these patients. A 49 years old female patient is reported where thrombolytic therapy with recombinant Streptokinase (TT-rSK) was the first treatment choice in seven recurrent episodes of prosthetic valve thrombosis.

INTRODUCTION

An abnormal junction of the bicuspid valves produces a pathological descent of the tricuspid valve in the right ventricle characterizing Ebstein's Anomaly (EA). According to the degree of tricuspid regurgitation or the association of other malformations, the clinical picture of this disease ranges from the absence of symptoms to severe heart failure. In case surgical treatment is required, its success would depend upon the possibility of reconstruction (or replacement) of the tricuspid valve (TV) (1, 2).

A patient is reported with EA who has undergone three tricuspid valve replacements, and has suffered eight episodes of prosthetic heart valve thrombosis that has been treated with thrombolytic drugs in the course of 7 years.

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The main purpose of this paper is report a case where multiple courses of thrombolytic treatment were used for prosthetic heart valve thrombosis as first therapeutic option.

CLINIC CASE

We present a 49 year old female patient, diagnosed at age 31 with an EA, atrial septal defect (ASD) and severe tricuspid regurgitation. She was operated on to close the ASD and right atrial plicature and to replace the TV by a bioprosthesis. Four years later, the bioprosthesis degenerated and was replaced by a monodisc mechanical prosthesis. Warfarin was maintained as anticoagulant therapy. Four years later, the patient was admitted with right cardiac failure and reported absence of prosthetic sounds. The transthoracic echocardiogram (TTE) revealed a thrombosis of the tricuspid prosthesis (increase of echogenicity in the auricular side of the prosthesis and a marked decrease of the movements of the disc).



Figure 1: Trans-thoracic echocardiogram where a tricuspid prosthesis is observed with increase in its echogenicity that extends toward right atrium corresponding to a massive thrombosis.

With the consent of the patient, a thrombolytic treatment (TT) with recombinant streptokinase (rSK), baseline doses of 250,000 IU in 30 minutes followed by a continuous infusion of 100,000 IU/hour, was initiated lasting 48 hours. The total doses administered were 5,000,000 IU.

The symptoms disappeared and the TTE showed a total disappearance of the thrombus and a recovery of the disc movements. There were no complications due to the thrombolytic therapy.

After this episode, the patient further experienced seven similar crises of rethrombosis. They all responded to therapy. In the last episode, despite the disappearance of the symptoms,

hepatomegaly and ascitis remained. A nonobstructive residual image was observed in the heart prosthesis after thrombolysis, so the valve had to be replaced again. Abundant pannus was observed around the prosthesis. Presently, two years after the surgical procedure, the patient remains asymptomatic.

DISCUSSION

Ebtein's anomaly is the most frequent cause of congenital tricuspid failure. The magnitude of the tricuspid failure, together with the extension of the atrialized right ventricle, determines the degree of deterioration of the right ventricular function. The coexistence of a mechanical heart valve prosthesis in a low pressure circuit and irregularities in the anticoagulant therapy contributed decisively to the appearance of recurrent prosthetic thrombosis in this patient (3).

We reported a previous series of cases where tricuspid heart valve thrombosis was involved (4) and TT prescribed with good results.

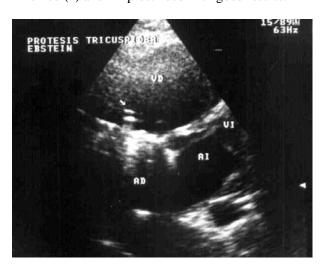


Figure 2: Trans-thoracic echocardiogram, in the same tricuspid prosthesis where you can see that thrombus has disappeared and normal functioning of the mechanical prosthesis 72 hours after thrombolysis.

There was agreement with others (5-7), in considering thrombolysis as the treatment of choice. In these patients with high thrombotic risk, the antithrombotic prophylaxis and the adequate follow-up of the anticoagulation are extremely important. In case any thrombotic complication occurred, there should be no doubt in continuing to use "thrombolytic drugs" as a first choice, even in recurring episodes (8).

The presence of periprosthetic pannus can contribute to a partial response to the thrombolytic drug therapy.

In our patient was noticed no allergic reactions.

A streptokinase obtained by means of recombinant-DNA (rSK) has been evaluated previously in two clinical trials in acute myocardial infarct (AMI) patients. In the first trial, 224 patients were randomized to receive rSK or natural streptokinase (nSK). Both groups behaved similarly regarding coronary patency at eight days after thrombolysis and the changes induced on fibrinogen degradations products, and thrombin time (9). They were also similar with respect to anti-SK antibodies titer and their anti-SK neutralizing activity (10,11).

Streptokinase (SK) is a 47 kDa protein produced by various strains of hemolytic streptococci and is a potent activator of the fibrinolytic enzyme system in humans. SK is in widespread clinical use to treat acute infarction because of its function as an activator of vascular fibrinolysis. Since streptococcal infections are common, normal individuals are immunized with SK and antibodies (Abs) to SK can be detected in most of them. This therapy generates significant T-cell responses to SK and the neutralizing capacity of the Abs rises significantly. Neutralizing Abs reduces the efficiency of thrombolytic therapy and may cause allergic reactions (12).

Incidence of rethrombosis is approximately 20% of the patients treated with thrombolysis (13). Torrado and co-workers recommend treatment of rethorombosis with further administration of thrombolytics (3). Lengyel have reported a 75% rate of success in recurrent prosthetic heart valves thrombosis treated with a second thrombolytics administration (14).

Shapira and collegues evaluated the effectiveness and safety of thrombolytic treatment in stuck bileaflet heart valves in the absence of risk thrombi. In 12 patients they had 83, 3 % of success with 25 % of rethrombosis. All recurrent episodes were treated successfully with thrombolytic therapy. They concluded thorombolytic therapy is highly successful and safe, both in the primary episode and in recurrence (15).

In our case with a right-sided mechanical prosthetic valve due to correction of an EA, where the incidence of thrombosis is high, we decided thrombolytic therapy as first choice of treatment because there were no contraindications and the previous knowledge of effectiveness and safety of thrombolysis, also because we share the concept

that it constitutes the first line of treatment in prosthetic heart valve thrombosis independently of functional class and size of thrombus when there are no contraindications. To trhe best of our knowledge this is the first such a case report in the literature.

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