VIA MEDICA

CASE REPORT

Cardiology Journal
2010, Vol. 17, No.2, pp. 189–191

ISSN 1897–5593

Myocardial infarction in a low risk patient with hereditary hemorrhagic telangiectasia

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Abstract

We describe the case of a 57 year-old woman with NSTE ACS, a history of recurrent and prolonged epistaxis, and low prior cardiovascular risk. Additional findings revealed anemia and an aneurysm in her central nervous system. During her hospital stay, hereditary hemorrhagic telangiectasia (HHT) was diagnosed. After application of two antiplatelet drugs, the patient was scheduled for coronarography, followed by coronary artery bypass grafting. During her hospital stay, only a minor episode of epistaxis was observed. We conclude that anemization due to HTT may significantly accelerate the progress of ischemic heart disease, resulting in acute coronary syndrome. Moreover, coronarography preceded by routine application of two antiplatelet drugs seems not to increase the risk of hemorrhage in HHT patients complicated with myocardial infarction. (Cardiol J 2010; 17, 2: 189–191)

Key words: acute coronary syndrome, hereditary hemorrhagic telangiectasia, anemia

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant systemic fibrovascular dysplasia in which telangiectases, arteriovenous malformations, and aneurysms may be widely distributed throughout the body vasculature [1]. It affects approximately 1 in 8000 individuals worldwide [2], and cuts life expectancy by approaching seven years [3]. Major clinical manifestations include recurrent bleeding from mucosal telangiectases and arteriovenous malformations [1]. Epistaxis is the most frequent first clinical symptom and usually develops before the age of 20 [4]. However, age of onset and severity are highly variable. Patients may not be diagnosed until a life-threatening complication is presented.

We are presenting a case of myocardial infarction (MI) in a patient with HHT.

A 57 year-old woman with negative past medical and family history was admitted for chest pain and suspicion of Non-ST Elevation Acute Coronary Syndrome (NSTE ACS). She was a non-obese non-smoker who had never received substitutive hormonal therapy or any other medical treatment. However, she reported a one week history of progressive chest pain (initially only at exertion, then also at rest) and a two month history of recurrent and prolonged epistaxis.

Electrocardiography on admission revealed ST depression in the inferior leads and transient ST elevation in the leads V1–V3. Over the following days, negative T waves in all chest leads developed.

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Received: 30.12.2008 Accepted: 3.05.2009

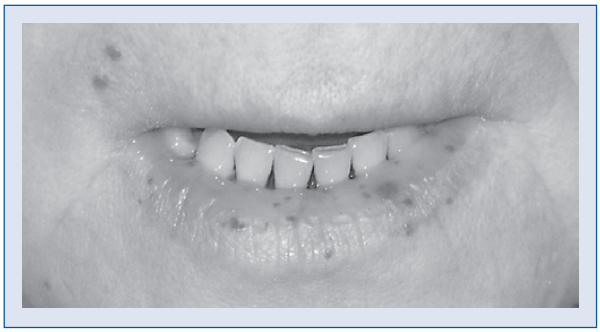

Figure 1. Angiomas in lower lip.

Laboratory findings displayed serum troponin elevation only in the third sample (the prior two samples were within normal limits), microcytic anaemia — Hgb 8 g%, Htc 26%, MCV 62 FL, Fe 16 μg/dL, UIBC 324 μg/dL.

According to risk stratification, an early conservative strategy was applied. Therefore, two units of packed red blood cells were transfused, achieving satisfactory improvement. Physical examination revealed multiple angiomas in the epithelium of nasopharynx (Fig. 1) which enabled diagnosis of HHT.

Applied treatment involved: clopidogrel, acetilsalicilic acid (ASA), metoprolol, perindopril, simvastatin, oral folic acid and iron supplements.

Magnetic resonance imaging, performed to assess potential malformation in the central nervous system, displayed a 2 mm aneurysm of the left posterior communicating artery (Fig. 2). Initially, echocardiography revealed hypokinesis of the anterior wall and interventricular septum, accompanied by slight mitral and tricuspid insufficiency with left ventricular ejection fraction of 55%. However, all symptoms of contractility disorders withdrew before discharge.

Coronary angiography revealed multilevel critical stenoses in the medial and distal portion of the left anterior descending (LAD) artery with the ostium of the second diagonal branch involvement (Fig. 3). The patient underwent left internal mammary artery (LIMA)-LAD off pump coronary artery bypass grafting followed by coronary artery bypass grafting to diagonal artery. The patient, with good early post-operative results, was transferred to another unit for further rehabilitation.

Acute coronary syndrome (ACS) risk model according to GRACE at admission revealed a 1% probability of death during hospital stay, and 6% within six months. The risk of in-hospital death or death/MI reached 14% and 21% in a six month horizon. The TIMI risk score was three (moderate).

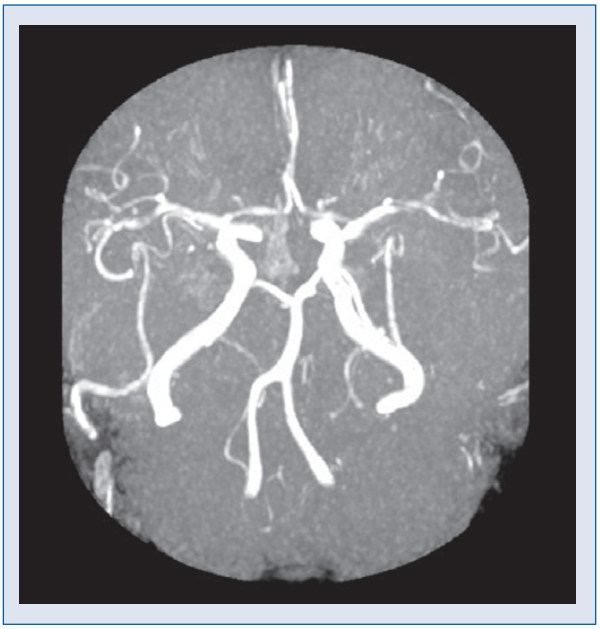

Figure 2. Aneurysm of left posterior communicating artery.

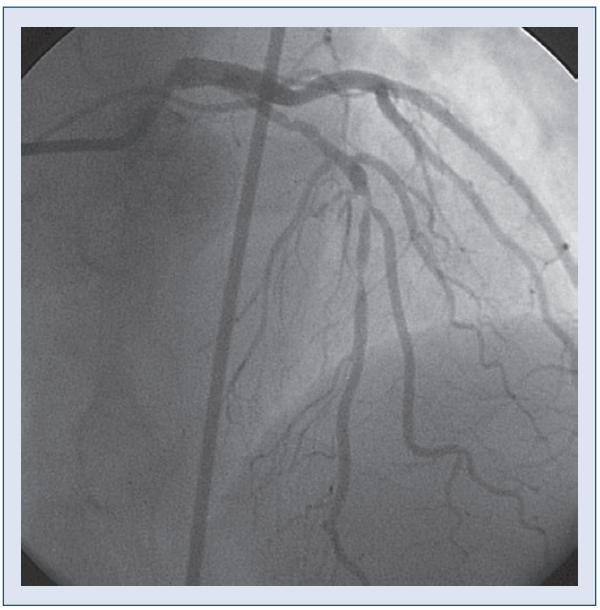

Figure 3. Narrowings in left anterior descending.

At discharge, the six month risk dropped to 1% for death and 2% for death or MI, which we considered satisfactory.

Myocardial infarction frequently (in 50–70% of cases) appears as the first manifestation of ischemic heart disease [5, 6], especially in normotensive [6] young males and often is associated with impairment of a single vessel [7, 8]. Our patient seemed

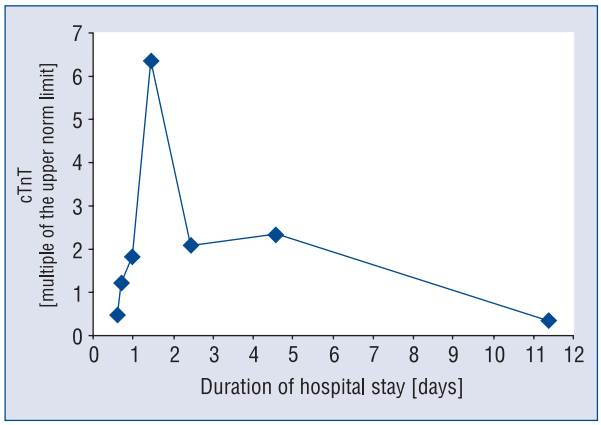

Figure 4. Changes in cardiac troponin levels.

to have experienced progression from stable angina to NSTE ACS within one week. Nevertheless, apart from anemisation, which also is a significant and independent predictor of major adverse cardiovascular events in ACS patients [9], she had no other risk factors.

Baseline hemoglobin level of 8–9 g/dL at admission in NSTE patients is associated with a 2.45 (95% confidence interval 1.80–3.33) fold increase in 30-day risk of cardiovascular death, MI or recurrent ischemia [9]. Therefore, we decided to transfuse 2 units of packed red blood cells. However, it has also been proved that in such patients blood transfusion may contribute to in-hospital death or MI with estimated odds ratio 1.44 (95% confidence interval 1.30–1.60) [10].

The application of two antiplatelet drugs is a routine management of patients scheduled for coronarography. Due to the increased risk of bleeding in HTT patients, particularly with intracranial aneurysm, we were concerned about applying the second antiplatelet drug. Acute complications considering heart and ischemic stroke were evaluated respectively at 4.3% and 7.5% [11]. Literature provided us with one interesting, but only partially relevant, case report, which was not enough to support a clear clinical decision. The reported patient was a middle-aged woman with NSTE and previously diagnosed HHT treated only with acetyl salicylic acid [12]. Nevertheless, we decided to combine ASA with clopidogrel and during the hospital stay we observed only one episode of minor epistaxis.

We also observed an interesting evolution of cTnT levels (Fig. 4). The increase was relatively late and very transient, which in association with echocardiographic findings may be considered a result of minor subendocardial ischemia. Moreover, some studies indicate that stored blood may promote vasoconstriction and trigger ischemic events [13].

It is probable that accelerated progression of angina resulting in ACS was caused by decompensation due to recurrent blood loss (first clinical manifestation of HHT) in combination with preexisting multilevel narrowings in LAD. Human hereditary telangiectasia may sometimes be revelatory for ischemic heart disease but does not seem to require major changes in the standard management of ACS.

Acknowledgements

The authors do not report any conflict of interest regarding this work.

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