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Ein Symptom von thrombotic thrombocytopenic purpura ist die Erhöhung des Herzinfarkts im ST-Segment.

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ABSTRACT

In emergency rooms all over the world is a myocardial infarction with ST segment elevation (STE) on electrocardiography (ECG) a common presentation. In patients with thrombotic thrombocytopenic purpura (TTP) are myocardial injury and necrosis rarely the initial presentation. A 48-year-old woman was presented for primary percutaneous coronary intervention with STE myocardial infarction from outside hospital. Her klinische Darstellung war jedoch unkonsistent. Durch eine schnelle Untersuchung wurden Anzeichen von microangiopathic hemolytic anemia, thrombocytopenia, einer akuten Kidneyverletzung mit Waxing und einem abnehmenden psychischen Zustand festgestellt. TTP mit niedriger ADAMST-13 Aktivität wurde diagnostiziert. In conjunction with intravenous steroid therapy, plasmapheresis was initiated. Nach vollständiger Genesung der left ventricular ejection fraction und normalen myocardial perfusionsuntersuchungen ging der Patient nach Hause. Eine schnelle Bewertung ist erforderlich. to determine uncommon STE myocardial infarction causes. In order to ensure quality control in the emergency room and catheterization laboratories, it is equally important to integrate all facets of the patient's clinical and objectiv data.

Key words: Acute coronary syndrome, electrocardiography, microangiopathy, plasmapheresis, ST segment elevation myocardial infarction, thrombotic thrombocytopenic purpura

INTRODUCTION

Patienten mit Brustschmerzen benötigen eine schnelle Triage, Untersuchung und Behandlung. In emergency rooms all over the world is a myocardial infarction with ST segment elevation (STE) on electrocardiography (ECG) a common presentation. Es ist erforderlich, eine plötzliche Herzcatheterisierung und eine mögliche percutaneous coronary intervention (PCI) zu berücksichtigen, wenn ein ACS auf dem Elektrokardiogramm durch STE gekennzeichnet ist. In fact, core measures and quality metrics exist in

place that grade a hospital's efficiency at caring for such patients.[1] However, the time pressure to optimize such quality metrics may lead to an unintended rush to treatmentprior to adequate evaluation.[1]

In this report, we present a rare case where myocardial infarction was seen as a presenting feature of an underlying hematologic disease, thrombotic thrombocytopenic purpura (TTP). This case highlights the importance of a thorough, yet efficient, clinical evaluation in which the history, physical exam, ECG and laboratory data were needed to make the appropriate triage decision and not miss an unusual diagnosis.

CASE REPORT

A 48-year-old woman with no known coronary risk factors was transferred to our hospital's cardiac catheterization laboratory from an outside facility with

the diagnosis of STE myocardial infarction for primary PCI. On arrival to our catheterization laboratory, the ECG from the referring hospital showed sinus tachycardia with normal axis and intervals. There was STE in leads I, II, aVL, V4–6 and reciprocal ST segment depression in lead III [Figure 1]. Laboratory data were not yet available. However, the patient's history of present illness was significant for malaise, fever, chills and lethargy that began 3 days prior to hospitalization. Further questioning established that she had mild generalized abdominal pain and one episode of non-bloody diarrhea. The family also noted that she had been intermittently confused and was talking gibberish. On the morning of admission, she had severe chest pain associated with nausea, vomiting and dyspnea on exertion, which led her to seek medical care. Her medical history was notable for a transient ischemic attack 7 years prior. An extensive thrombophilia work-up at that time was negative. She also had a history of two miscarriages in the past.

On examination, she appeared toxic and in respiratory distress. Vital signs revealed a blood pressure of 126/70 mmHg with a heart rate of 121 beats per minute. Her temperature at admission was 34.4°C. The respiratory rate was 30 breaths per minute. Oxygen saturation was 100% on a non-re-breather mask. She was pale, cold and clammy with delayed capillary refill. She had cyanosis in all fingers with mild cyanosis of her tongue and lips. In addition, mottling of her skin and livedo reticularis over the thighs was noted. There were a few purpuric skin lesions observed in her antecubital fossa and upper arms. Her jugular venous pressure was elevated up to the angle of the jaw. Cardiac exam revealed a normal first and second heart sound along with a fourth heart sound. There were no murmurs. Peripheral pulses were not palpable in the feet and were only faintly palpable in the arms. The lungs were clear to auscultation. Abdominal exam was unremarkable. There was no peripheral edema. Neurologically, she was somewhat confused, but the sensory and motor exam was essentially normal. Given that the patient was not having

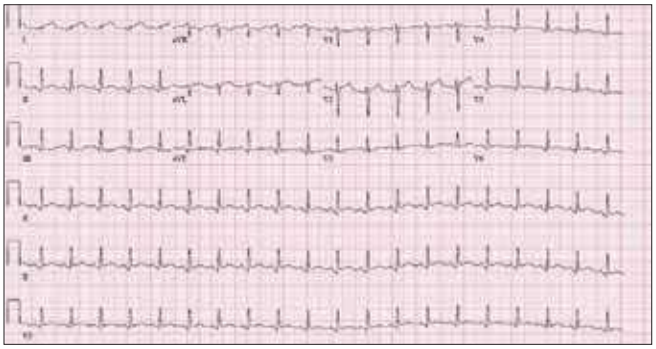

Figure 1: Sinus tachycardia at 121 beats per minute with ST segment elevation in Lead I, II, aVL, V4–6

active chest pain, the history was inconsistent with ACS and she appeared more toxic than expected for a lateral wall myocardial infarction; cardiac catheterization was deferred and emergent laboratory studies were obtained.

Initial laboratory studies revealed a white blood cell count of $13.5 \times 10^3/mm^3$; hematocrit of 24%; mean corpuscular volume of 88.4 fL and platelet count of $6 \times 10^9/L$. Her lactate dehydrogenase was elevated at 2820 units/L and haptoglobin was low at less than 10 mg/dL. Coagulation profile showed international normalized ratio of 1.2, prothrombin time of 12.6 seconds, fibrinogen 199 mg/L and D-dimer 1.27 feu mg/L. Electrolytes were within normal limits; acute kidney injury was noted with blood urea nitrogen 51 mg/dL and creatinine 1.9 mg/dL. Total bilirubin was markedly elevated at 32 mg% with an indirect bilirubin of 2.1 mg%. Cardiac biomarkers were elevated with creatinine kinase of 487 units/L and MB fraction of 28.8 ng/mL. Troponin-T was 0.86 ng/mL. Urinalysis showed pH of 6.0, 3+ albumin, 3+ hemoglobin, eight WBCs and greater than two RBCs with some amorphous crystals. Peripheral smear showed moderate schistocytes, few spherocytes and low platelet count. Chest radiograph showed no cardiopulmonary abnormalities. Echocardiography demonstrated an ejection fraction of 40–45% with severe hypokinesis of the inferior and basal anteroseptal wall. No significant valvular lesions were noted.

In view of the acute onset of symptoms associated with microangiopathic hemolytic anemia, thrombocytopenia, acute kidney injury and waxing and waning mental status, the presumptive diagnosis of TTP was made. Further coagulopathy testing was negative. A Disintegrin and Metalloproteinase with Thrombospondin Motifs (ADAMST-13) activity was found to be low with presence of ADAMST-13 inhibitors in the plasma.

Plasmapheresis was started immediately. Intravenous steroid therapy was also initiated. She improved clinicallywith this treatment and normalized her platelets and lactatedehydrogenase, and STE in her ECG resolved [Figure 2].

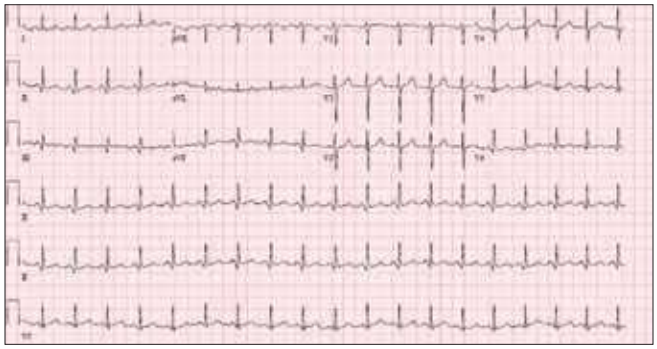

Figure 2: Sinus tachycardia at 113 beats per minute with resolved ST-segment changes

She was discharged home after a full recovery. Outpatient cardiovascular follow-up demonstrated normalization of the left ventricular ejection fraction by echocardiography and a completely normal nuclear myocardial perfusion stress study. Further testing with invasive catheterization was not pursued as the entire episode was considered to be transient, secondary to metabolic derangement.

DISCUSSION

TTP ist eine schwere, thrombotische Microangiopathy, die hauptsächlich durch systemische Platelet-von Willebrand-Faktor-Aggregation, organische Ischämie, schwere Thrombozytopenie und Erythrocytenfragmentation gekennzeichnet ist.Die intravaskuläre Blutgerinnsel wird nicht als bedeutendes Merkmal des Zustands betrachtet. Oftmals tritt eine ausgedehnte Organdysfunktion klinisch auf. Die Pankreatitis, die Adrenalen, das Herz, der Gehirn und die Nieren können pathologische Bereiche von Nekrose und Blutungen aufweisen.[4-5] Viele Patienten mit TTP zeigen myocardial injury and necrosis, aber die erste Symptome sind selten [6]. Es wird wahrscheinlich von microthrombi aus massiver platelet aggregation statt einer plaque rupture-thrombosis cascade verursacht.[7] In TTP varies the incidence of myocardial infarction from 15 to 41 percent.[6-9]

Early recognition of myocardial injury in a case of TTP is crucial as it identifies higher risk. However, invasive therapy in the form of cardiac catheterization and PCI may be fraught with complications and is precluded by acute kidney injury and low platelet count.[8] Thrombocytopeniaalso prevents the use of usual medical management in ACS such as antiplatelet and anticoagulant therapy.[9] Angiotensin converting enzyme inhibitors are also not used because of concomitant renal injury. Beta blockersand HMG CoA reductase inhibitors may be used althoughtheir role is questionable.[9] In acute bouts of TTP, suchas this case, the treatment of choice is rapid initiation ofplasmapheresis. In addition, immunosuppressive therapy including steroid therapy is helpful, especially in the setting of auto-antibodies against ADAMST-13 factor.[13,14] Relapsing cases of TTP have been treated with rituximab, a monoclonal antibody against CD20 on memory B cells with good effect. However, large clinical trials are lackingfor this.[13]

Finally, STE myocardial infarction is very common in the emergency rooms. Eine schnelle Untersuchung ist erforderlich, um ungewöhnliche Ursachen für STE-Herzinfarkte zu finden. Cardiac involvement is a common occurrence in TTP, but as an index event it can be misleading. In order to ensure quality control in the emergency room and catheterization laboratories, it is equally important to integrate all facets of the patient's clinical and objectiv data. Once TTP is identified, a multidisciplinary approach is needed to treat it quickly.

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