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Case Report

The positive therapeutic effect in a patient of Evans syndrome combined with acute myocardial infarction

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Abstract: Evans syndrome (ES) is a rare combination of autoimmune hemolytic anemia and thrombocytopenia. This case report details an old male ES patient with acute myocardial infarction. He was successfully treated by primary percutaneous coronary intervention in the case of low hemoglobin level (60 g/L). Considering ES recurrence after surgery, he was given human immunoglobulin, methyl prednisolone and TPO treatment. On the basis of his platelet count, the patient was required to take only one anti-platelet drug or stop all anti-platelet drugs. To the best of our knowledge, this is the first report of ES with AMI. This case suggests that primary PCI can be a useful therapeutic strategy even if patient has low hemoglobin level, but careful balance between anti-platelet therapy and efforts to raise platelet count are needed after surgery.

Keywords: Evans syndrome (ES), acute myocardial infarction (AMI), PCI, anti-platelet therapy

Introduction

Evans syndrome (ES) is the rare simultaneous or subsequent development of autoimmune hemolytic anemia (AIHA) and idiopathic thrombocytopenic purpura (ITP). AIHA is an infrequent group of diseases defined by autoantibody mediated red blood cell destruction. Most patients with AIHA require therapy. In warm antibody AIHA, standard first line therapy is glucocorticosteroids with or without high dose immunoglobulins, whereas splenectomy is considered second-line therapy [1]. ITP is an autoimmune disorder with a low platelet count characterized by premature platelet destruction mediated by autoantibodies and often causes mucocutaneous bleeding. The decision to treat ITP is based on the platelet count, the degree of bleeding, and the patient's lifestyle [2-4].

No case of ES with acute myocardial infarction (AMI) has been reported and there are no precise recommendations about treatment. When invasive treatment such as primary percutaneous coronary intervention (PCI) is chosen, it is necessary to pay special attention to antiplatelet therapy and patients' platelet count [5-9]. We herein presented an old male patient of Evans syndrome with AMI.

Case report

An 80-year-old male patient who diagnosed with Evans syndrome twenty years ago felt chest pain on June 30, 2013. ST-segment elevation was on his electrocardiogram and cTnI was 0.860 ng/ml. He was diagnosed with recurrent myocardial infarction and admitted in cardiovascular department because he was once diagnosed with acute myocardial infarction and a drug-eluting stent was placed in his left anterior descending coronary artery (LAD) in 2009. His peripheral blood platelet count was 187 × 10^9/L, but hemoglobin was only 60 g/L and red blood cell cross matching difficulties happened. In the case of such low hemoglobin level, the patient was still successfully given primary percutaneous coronary intervention (PCI) on July 23. Coronary angiography showed in-stent restenosis occurred and another drug-eluting

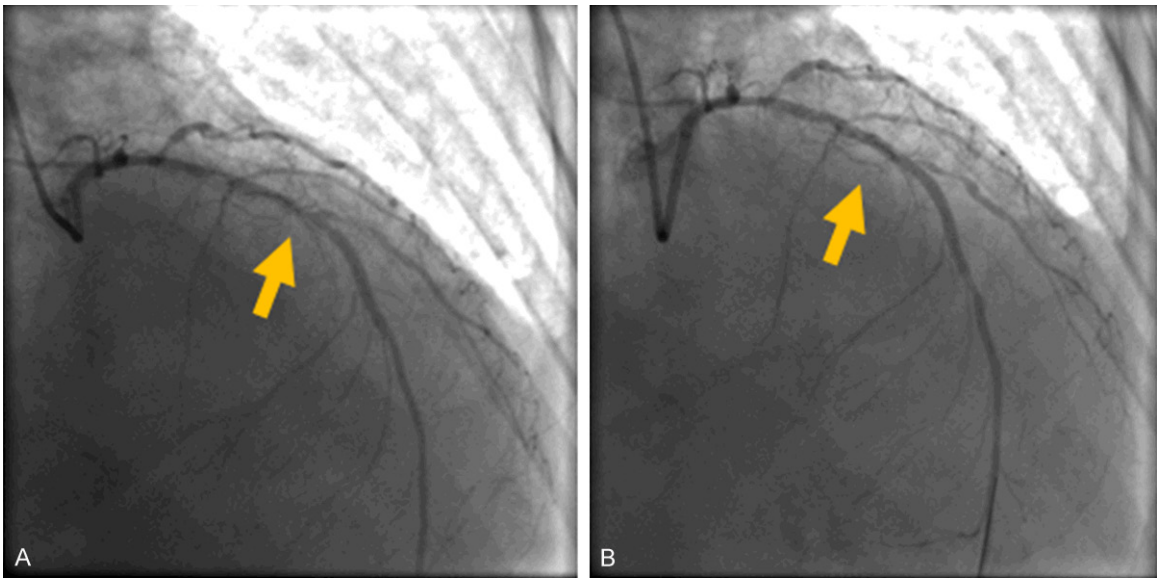

Figure 1. Coronary angiography results. A. Coronary angiography showed that 90% in-stent restenosis (the pointed) occurred in this patient's LAD; B. Another drug-eluting stent (2.5 × 29 mm) was placed in his LAD (the pointed).

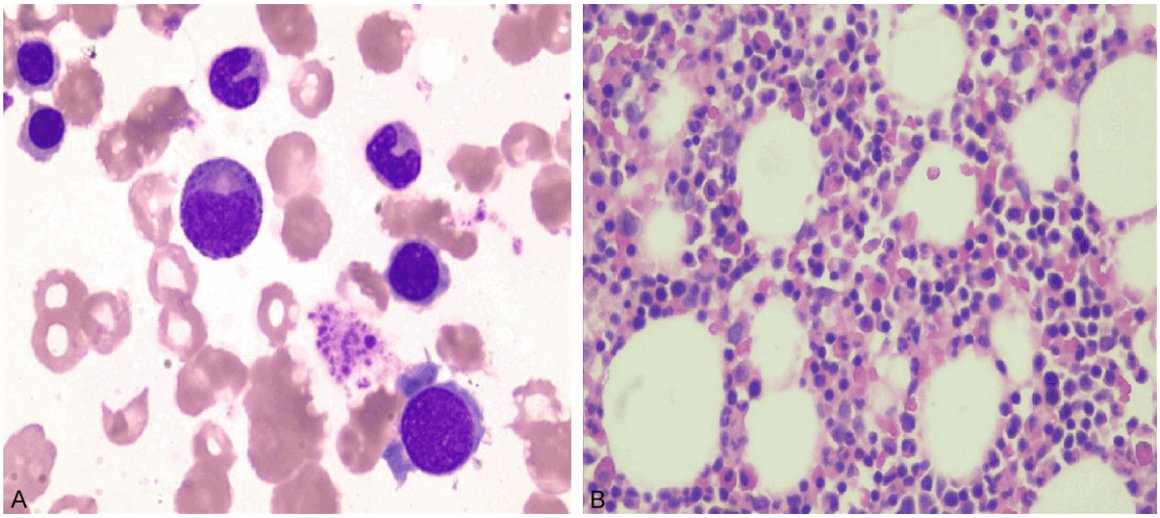

Figure 2. Bone marrow examination results. A. Bone marrow smear (1000 ×). B. Bone marrow histology (400 ×). Both A and B showed erythroid hyperplasia of bone marrow.

stent (2.5 × 29 mm) was placed in his LAD with 90% stenosis (**Figure 1**).

After surgery, he felt comfortable and accepted anti-platelet therapy (bayaspirin 100 mg, qd, po and clopidogrel 75 mg, qd, po). However, on the 13th day after PCI surgery, the patient felt chest distress. ST-segment had no dynamic change on his electrocardiogram, but blood test showed his hemoglobin was only 72.4 g/L. Considering low hemoglobin level inducing chest distress, the patient accepted some examinations such as Coombs test, bone marrow aspiration and biopsy. His direct Coombs test was positive. Both bone marrow cytology and histology results showed marrow erythroid hyperplasia (**Figure 2**). Because of the recurrence of ES, he was transferred to hematology department to continue treatment. He still accepted the previous anti-platelet therapy. From the 21st to 23rd day after PCI surgery, he was given intravenous infusion of human immunoglobulin 25 g/d and his hemoglobin increased to 89 g/L. He clearly felt chest dis-

tress relieved and asked to stop human immunoglobulin on the 24th day after PCI surgery.

On the 22nd day after PCI surgery, his platelet count began to decrease to 64 × 10^9/L. On the 27th day after surgery, his platelet count was found only 26 × 10^9/L. Then he was given human immunoglobulin (25 g/d ivgtt, from the 27th to 29th day), methyl prednisolone (40 mg p.o. on the 27th day, 20 mg on the 28th day, 8 mg on the 29th day) and thrombopoietin (15 KU/d, hypo). Fortunately, on the 30th day after surgery, his platelet count was up to 140 × 10^9/L and his hemoglobin level was up to 99 g/L.

However, the patient began to complain insomnia and dreaminess, so he asked to stop methyl prednisolone on the 30th day after surgery. On the 32nd day after surgery, the patient got cough and expectoration, his platelet count reduced to 18 × 10^9/L again and hemoglobin was 94 g/L. Chest CT showed his right lower lung infection. So he was given intravenous infusion of antibiotics and methyl prednisolone 40 mg every day. The next day his platelet was up to 102 × 10^9/L and hemoglobin was 87 g/L. Methyl prednisolone was reduced to 20 mg/d on the 38th day, and maintained 8 mg/d after the 40th day. In this condition, the patient's platelet count kept normal and hemoglobin ranged from 90 g/L to 100 g/L. Besides, his symptoms of cough and expectoration disappeared gradually, so antibiotics were stopped.

It was worth mentioning that after PCI surgery, the patient's platelet count was continuously monitored and anti-platelet treatment strategy was adjusted according to his platelet level. When his platelet count was less than 50 × 10^9/L, one kind of anti-platelet drug (bayaspirin) was discontinued. When his platelet count was less than 30 × 10^9/L, all anti-platelet drugs were stopped taking. Platelet transfusion was necessary when his platelet count was less than 20 × 10^9/L.

The patient was discharged from hospital after his hemogram almost returned to normal and discontinued corticosteroid on the 43rd day after PCI surgery. Follow-up so far, the patient's heart works well in the case of taking two previous anti-platelet drugs. And his hemoglobin level is close to the normal and platelet count is totally normal.

Discussion

This case report detailed an 80-year-old male patient who diagnosed with ES some years ago combining with acute myocardial infarction (AMI). Although the patient's hemoglobin level was low (60 g/L) and red blood cell cross matching difficulties happened, he was still successfully treated by primary PCI. It was proved that such disposal was very necessary, because the patient had very serious left coronary artery lesions confirmed by coronary angiography. If he had not been given emergency PCI, he could die of AMI.

In fact, AIHA can cause ABO type identification and blood cross matching difficulties are mentioned in some literatures [10-12]. High titer of autoantibodies can cause blood type identification difficulty and difficult blood matching. For these patients, if they required transfusion, their autoantibodies should be absorbed by their own cells firstly. Then, they are admitted to match blood and washing RBC is priority selection. It has been not well-known that what level of hemoglobin should be maintained and what the lowest hemoglobin level is when one patients should be treated by primary PCI. The present case suggests that primary PCI can be still one of the therapeutic strategies for AMI patients with severe anemia. This patient, at the low level of hemoglobin (60 g/L), was successfully treated by primary PCI, therefore it can be used as a reference to deal with such cases in the future.

It was worth mentioning that this ES patients' hemogram were continuously monitored and paid close attention to. Although both anemia and thrombocytopenia were caused by abnormal autoimmune reaction, their changes were found not parallel. Namely, when his anemia occurred, his platelet count was normal and when his platelet count declined to the lowest, his hemoglobin didn't decrease obviously. So it is amazing that AIHA and ITP can occur simultaneously or in succession in ES patients.

After PCI surgery, he developed thrombocytopenia (minimum platelet count only 18 × 10^9/L). Some measures were taken such as intravenous infusion of human immunoglobulin, glucocorticoid, TPO hypodermic injection and platelet transfusion to enhance PLT level and reduce bleeding risk. Considering pulmonary infection

could make his thrombocytopenia heavier, antibiotics were timely adopted. Patients who will be treated by PCI need accept anti-platelet therapy (aspirin and clopidogrel) not only in pre-operation but also in post-operation [13-15]. However, long-term anti-platelet therapy may increase the risk of bleeding for ES patients. Careful balance between usual anti-platelet therapy and efforts to raise platelet count was important. On one hand it was needed to enhance his PLT level to prevent bleeding, on the other hand, the patient needed anti-platelet drugs to prevent in-stent re-stenosis after PCI surgery. It was a contradiction in the treatment. So far, there is no clear treatment strategy reported in literatures or guidelines that can be followed. In this case, treatment strategy was appropriately adjusted according to platelet count of this patient. When his platelet count was less than 50×10^9/L, one kind of anti-platelet drug (bayaspirin) was discontinued. When his platelet count was less than 30×10^9/L, all anti-platelet drugs were stopped taking. Platelet transfusion was necessary when his platelet count was less than 20×10^9/L.

Follow-up so far, the patient's heart works well. His hemoglobin level is close to the normal and platelet count is totally normal. To the best of our knowledge, this is the first report of ES with AMI. This case suggests that primary PCI can be a useful therapeutic strategy even if the patient has low hemoglobin level, but careful balance between anti-platelet therapy and efforts to raise platelet count is needed after surgery. Treatment should be individualized, but it is still a challenge to avoid reinfarction and bleeding complication.

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Disclosure of conflict of interest

None.

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