Scientific letters

Management of Primary Immune Thrombocytopenia With Ertrombopag in a Patient With Recent Acute Coronary Syndrome

Tratamiento de trombocitopenia inmune primaria con ertrombopag en un paciente con síndrome coronario agudo reciente

To the Editor,

To date, no consensus has been reached on the treatment of patients with primary immune thrombocytopenia (ITP) who require antiplatelet therapy after experiencing acute coronary syndrome (ACS).

We report the case of a 55-year-old male smoker with hypertension who, as a child, was diagnosed with primary ITP, which responded to steroids. The patient had an asymptomatic relapse as an adult, without medical follow-up. He was admitted to the emergency room with non-ST-segment elevation ACS and a platelet count of $13 \times 10^9$ cells/L, confirmed by a peripheral blood smear. After treatment with 40 mg of intravenous dexamethasone and intravenous immunoglobulins (IVlg) at a dose of 1 g/kg body weight, his platelet count increased to $82 \times 10^9$ cells/L in 24 hours. However, this increase was accompanied by severe precordial pain and ST elevation in anterior leads. Cardiac catheterization confirmed severe coronary artery disease in the proximal and mid anterior descending artery. The patient underwent primary percutaneous transluminal coronary angioplasty with implantation of 2 XIENCE V drug-eluting stents (DES). Anticoagulation therapy consisted of enoxaparin and antiplatelet therapy consisting of clopidogrel and aspirin. After 15 days of treatment with prednisone (1 mg/kg body weight), an attempt to reduce the dose led to progressive thrombocytopenia, with generalized ecchymosis that developed at a platelet count of $65 \times 10^9$ cells/L. Six months after stent implantation, iatrogenic damage due to chronic corticosteroid therapy was evident, and we had to select the best therapeutic option for a patient with symptomatic corticosteroid-dependent ITP, with a platelet count $< 65 \times 10^9$ cells/L, but whose bleeding was attributed not so much to the thrombocytopenia as to the dual antiplatelet therapy (which, given the patient’s cardiovascular risk and the double stent placement, would need to be maintained for 6 more months to complete the 1-year treatment regimen).

In emergency situations, injection of IVlg may be appropriate, but not as regular treatment: several cases of ACS associated with IVlg therapy have been reported in patients with primary ITP, and in our patient, ST elevation coincided with its administration.

There were 2 long-term options: to administer aspirin alone for antiplatelet therapy—which would probably permit safe steroid reduction, but would increase thrombotic risk—or maintain dual antiplatelet therapy and attempt a second therapeutic line for primary ITP.

Splenectomy is the second-line treatment most widely recommended by current therapeutic consensus guidelines but was not an option for our patient at this stage of his disease for 2 reasons: first, it would require clopidogrel discontinuation, which would increase the thrombotic risk 6 months after ACS with double DES and, second, the change would be definitive (after the first year, he would foreseeably be taking a single antiplatelet agent; if the patient, although thrombocytopenic, maintained a safe platelet count without treatment, splenectomy might prove to have been unnecessary).

The thrombopoietin receptor agonists, ertrombopag and romiplostim, are recommended as second-line therapy when splenectomy is not indicated. However, they are discouraged in patients with high thrombotic risk. Although no cases of ertrombopag use have been published in patients with recent ACS, acute stent thrombosis has been reported in a patient receiving romiplostim. Other treatments for primary ITP are available for compassionate use, but there is no experience in ACS.

After considering the different options, we decided to maintain the dual antiplatelet therapy and administer ertrombopag to achieve a count of 60 to $80 \times 10^9$ platelets/L. After 1 week with 50 mg/d of ertrombopag, it was possible to discontinue prednisone and, with a dose of 25 mg/d, to maintain counts of 49 to $91 \times 10^9$ platelets/L until 1 year had elapsed since the double stent implantation, and there were no further bleeding episodes or thrombotic complications. Subsequent withdrawal of clopidogrel allowed discontinuation of ertrombopag and maintenance of antiplatelet therapy with aspirin alone. Three months later, the patient remained asymptomatic with a platelet count of around $30 \times 10^9$ cells/L.

A review of the limited literature on the topic shows that some authors propose dual antiplatelet therapy in ACS patients with platelet counts $> 30 \times 10^9$ cells/L without bleeding, and implantation of bare metal stents to reduce the period of clopidogrel therapy. In a case of chronic asymptomatic ITP (platelets $> 100 \times 10^9$ cells/L), DES were implanted and the patient received dual antiplatelet therapy with no bleeding complications. However, we have found no reports describing the treatment of patients with DES implanted during an episode of asymptomatic ITP, when the risk of stent thrombosis is very high if dual antiplatelet therapy is discontinued, even when the platelet count is low, but the bleeding risk is also high due to the concomitance of thrombocytopenia and antiplatelet therapy. Given the lack of high-quality scientific evidence regarding the management of these patients, recommendations cannot be made concerning their treatment, which should be individualized to minimize both risks. Special attention should be paid to the choice of the stent, as it will influence the duration of dual antiplatelet therapy. In our experience, with dual antiplatelet therapy, ertrombopag can be a safe and effective alternative after failed corticosteroid therapy, when splenectomy is not an option.

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REFERENCES


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Acute Clinical Presentation of Pseudoaneurysm and Aortopulmonary Fistula as a Very Late Complication of Aortic Coarctation Patch Repair

Presentación clínica aguda de seudoaneurisma y fistula aortopulmonar como complicación tardía de la reparación quirúrgica de la coartación de aorta

To the Editor,

A 57-year-old woman was admitted to the emergency room of our hospital for sudden lipotimia and copious hemoptysis. Twenty-seven years previously she had undergone Dacron patch aortoplasty for aortic coarctation with the Cooley technique. On admission, the patient was hypotensive, dyspnecic and anemic (Hb 6.3 mg/dl). A computed tomography scan showed a 36-mm pseudoaneurysm in the proximity of the aortic isthmus, perforated through a 6-mm fistula into the posterolateral aspect of the left lung (Figure 1A-D).

Thoracic endovascular aortic repair was not indicated due to unsuitable vascular access and lack of appropriate landing zones for endograft placement. The patient underwent emergency surgical reoperation.

The chest was entered through a fourth intercostal space left thoracotomy. The proximal thoracic aorta immediately distal to the left subclavian artery and the descending aorta at the seventh intercostal space level were isolated for subsequent cross clamping. At the level of the previously coarctated segment, a pseudoaneurysmatic dilatation of the aorta was observed, tenaciously adherent to the left upper pulmonary lobe. On moderate (32 °C) hypothermic cardiopulmonary bypass, by using left femoral vein drainage and dual femoral artery and distal aortic arch arterial return, the descending aorta was proximally and distally cross clamped. The pseudoaneurysm was partially dissected free and was then excised, leaving the anteromedial wall attached to the lung, thus penetrating into the pulmonary parenchyma. The Dacron patch showed a 1-cm disruption of the suture line (Figure 2A).

The continuity of the descending thoracic aorta was reconstructed with a 24-mm Dacron tube graft (Figure 2B). The patient was hemodynamically stable after the intervention, with no evidence of hemorrhage, but unfortunately died 2 months later from sepsis.

Aortic coarctation represents approximately 5% of all congenital heart malformations, with an incidence of 0.2–0.6 per 1000 live births. Surgical correction in young patients is indicated and many

Figure 1. Computed tomography angiography scan shows the 36-mm pseudoaneurysm (arrows), located 13 mm distal to the origin of the left subclavian artery, perforated through a 6-mm fistula with the bronchial tree. A: 2D computed tomography angiography scan, axial view. B: 2D coronal view. C: 3D sagittal plane reconstruction. D: 3D left posterior reconstruction. Arrows show the 36 mm pseudoaneurysm located 13 mm distal to the origin of the left subclavian artery, perforated through a 6 mm fistula with the bronchial tree.