

## Case Report

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# Cardiac Surgery in an Adult Patient with Congenital Asplenia and Thrombocytopenia

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### Abstract

Isolated congenital asplenia is a rare form of immunodeficiency often presenting with overwhelming and potentially fatal infection in early childhood. We report a case of an adult patient with congenital asplenia and severe chronic thrombocytopenia suffering from unstable angina pectoris. The patient underwent coronary artery bypass grafting with cardiopulmonary bypass. Consistent prophylaxis of infection, adequate surgical technique and control of thrombocytopenia prevented losing the patient from sepsis and uncontrollable bleeding.

**Keywords:** Cardiac surgery; Congenital asplenia; Thrombocytopenia

### Introduction

Isolated Congenital Asplenia (ICA) is a rare form of immunodeficiency, which characterizes by the absence of a spleen at birth in individuals with no other developmental defects. The patients are prone to overwhelming and potentially fatal infection. In these patients, high mortality rate in early childhood (up to 45%) was reported [1]. Patients who reach adult age are still at high risk of sepsis and meningitis [2]. We report a case of an adult patient with ICA in whom an uncommon form of thrombocytopenia was diagnosed and treated with corticosteroid therapy. The patient underwent coronary artery bypass grafting with an exceptional perioperative management in order to prevent severe infections and uncontrollable bleeding.

### Case Report

#### Patient's history

A 61-year-old woman with unstable angina pectoris and New York Heart association class II-III was referred to our hospital for Coronary Artery Bypass Grafting (CABG). Preoperative angiographic investigation demonstrated a 95% stenosis of Left Anterior Descending Artery (LAD) a 80% stenosis of dominant circumflex artery and a minor right coronary artery without coronary disease. Patient's history revealed repeated severe infections related to ICA and immune thrombocytopenic purpura, on daily corticosteroid treatment, with platelet counts above  $50 \times 10^9 /L$ . Patient was vaccinated with Pneumococcal vaccine.

#### Preoperative management

Preoperative platelet count was  $83 \times 10^9/L$ , but the platelet function tests and all the coagulation tests were normal. Biochemical markers of inflammation (WBC count, C-reactive protein and procalcitonin) were normal. In order to prevent a polysaccharide encapsulated bacterial

infection, a single dose of amoxicillin-clavulanic acid and polyclonal antibodies were administrated preoperatively. Since the patient had already been vaccinated with Pneumococcal vaccine, the response to immunization was checked. Measurement of pneumococcal antibodies revealed an adequate immunologic response to immunization. Routine prophylaxis of surgical site infection with cephuroxime was applied.

#### Anesthesia and operative course

Premedication consisted of atropine 0.5mg, midazolam 0.1 mg/kg and pethidine 50 mg intramuscularly. Anesthesia was induced with midazolam 0.2 mg/kg, fentanyl 8  $\mu\text{g}/\text{kg}$  and rocuronium 1 mg/kg and maintained by sevoflurane (1-1.5 MAC) with continuous infusion of fentanyl (1-5  $\mu\text{g}/\text{kg}/\text{h}$ ). CABG was performed with Cardiopulmonary Bypass (CPB) in moderate hypothermia ( $32^\circ\text{C}$ ). Membrane oxygenation (Medtronic Affinity.nt) and centrifugal pump (Jostra HL20) were used. Cardioplegic arrest was achieved by antegrade and retrograde administration of cold blood cardioplegia ( $4^\circ\text{C}$ ). LAD was grafted with left internal thoracic artery and a saphenous vein graft was targeted to obtuse marginal branch of circumflex artery. Intra-operative blood salvage, hemofiltration during CPB and meticulous surgical haemostasis were performed. Blood transfusion was not administrated during surgery. After CPB, platelets count fell on  $27 \times 10^9 /L$  and platelet transfusion was administrated.

#### Postoperative course

Complete postoperative blood loss through chest tubes was 350 ml. Patient was extubated 9 h after operation and released from ICU after 24 h. A single dose of amoxicillin-clavulanic acid was administrated daily. Inflammatory markers were monitored daily. Blood samples were collected for culture on the 1<sup>st</sup>, 3<sup>rd</sup> and 5<sup>th</sup> day after operation. No

	Day 1	Day 2	Day 3	Day 4	Day 5	Day 6
WBC ( $\times 10^9/L$ )	12.8	14.3	11.6	9.3	9.4	8.1
CRP (mg/L)	28	52	56	31	25	18
PCT (ng/mL)	0.37	0.59	0.34	0.24	0.19	0.12
PLT ( $\times 10^9/L$ )	48	56	61	54	67	62

WBC=White Blood Cell Count, CRP=C-reactive protein, PCT=Procalcitonin, PLT=Platelet Count.

**Table 1:** Markers of inflammation and blood cell count were monitored daily during postoperative hospitalization.

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infective or other significant complication occurred in postoperative course. Patient was released on 6<sup>th</sup> day after operation (Table 1).

## Discussion

Asplenia is an uncommon condition that may be acquired (following surgery), functional or congenital. It is associated with an increased incidence of life-threatening sepsis caused mainly by encapsulated bacteria. As is well known, there are two types of congenital asplenia, namely the Ivemark syndrome, which is usually associated with major cardiac malformations, and ICA. ICA is a very rare condition that occurs sporadically or has a familiar association [1,3]. In the review by Mahlaoui et al. they found 50 reported cases of ICA in the literature that included 24 sporadic and 26 familial cases: of these cases, 33 were children and 13 were adults (the age was not available for four cases) [1,4]. Severe infections and associated anomalies result in high mortality in early childhood. Most of these presented in the early years of life and rarely were diagnosed in the adult. Survival rate does not exceed 20% at one year [5]. It is evident that there is a significant difference in immune competence between patients with acquired and congenital asplenia. Contrary to acquired asplenia, only a few cases of adult ICA were reported, underlining the rareness of this condition in adults [2,6].

Immunization with pneumococcal vaccine and prophylactic antibiotics have become accepted as an integral part of the preventive strategy against serious infection in the asplenic population [7,8]. At least 87% infective episodes in asplenic patients were caused by *S. pneumonia* [6]. However, difficulties with antibody response in asplenic patients were found, especially in relation to the pneumococcal polysaccharide vaccine [8]. Therefore, we checked the response to immunisation by measuring pneumococcal antibodies in our patient prior to surgery.

In patients with severe thrombocytopenia (platelet count lower than  $100 \times 10^9/L$ ), the risk of bleeding is high. Furthermore, it is enhanced in intensive care patients when additional factors which interfere with normal haemostatic mechanisms (e.g. platelet function defects, hyperfibrinolysis, invasive procedures, CPB) are present.

Successful cardiac surgery in thrombocytopenia patients was reported, although transfusion of excessive amounts of erythrocyte suspensions and platelets was needed. Mathew et al. reported that 20 of 23 (87%) patients with thrombocytopenia required platelets replacement [9]. In patients with immune thrombocytopenia, immunosuppressive agents, corticosteroids and high dose immunoglobuline have been used in the preoperative period. Gamma globulin instead of immunosuppressives and corticosteroids was recommended in order to avoid any postoperative infectious complications [10].

Prior to surgery, we identified two major risk factors: asplenia and thrombocytopenia. Previous reports emphasized the significance of each of the factors. Due to a rare combination of congenital asplenia and thrombocytopenia, our patient was considered to have extremely high risk for cardiac surgery where significant bleeding can be expected. Preoperative prophylaxis of severe infections included dual large spectrum antibiotic prophylaxis (one targeting polysaccharide encapsulated bacteria) and polyclonal antibodies transfusion. Postoperative systemic inflammatory response was monitored daily and bacteria growth in blood cultures was controlled. Antibiotic prophylaxis of polysaccharide encapsulated bacterial infections was continued during the complete postoperative hospitalization.

Our patient underwent open heart surgery with severely reduced

platelet count but normally functioning platelets. Current options as the intra-operative blood salvage and hemofiltration during CPB reduce the necessity for massive postoperative platelets transfusion. Nevertheless, a single platelet transfusion had to be administered.

To the best of our knowledge, this is the first case report concerning the association between congenital asplenia in an adult, thrombocytopenia and cardiac surgery. Once in a blue moon, an adult patient with ICA is admitted to a major surgical intervention. Special care is needed to prevent losing the patient from postoperative infection. When risks are assumed properly and all possible measures are undertaken, postoperative course is most likely to be without complications.

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