

Jaundice and Rash 24 Days after Cardiac Surgery
Case study by Jim Perkins, M.D. and Gerald Hoeltge, M.D. (©2009)

A sixty four year-old man was admitted to a hospital for repair of a descending thoracic aortic aneurysm without rupture or dissection. The patient had a history of hypertension, adult-onset diabetes mellitus, smoking and chronic obstructive lung disease, a CVA four years earlier, renal insufficiency, and peptic ulcer disease. He had previously undergone coronary bypass grafting. His work-up for the operation revealed multiple cardiac abnormalities in addition to the 9 cm long saccular aneurysm just distal to the left subclavian artery.

After uneventful repair of the aneurysm the patient's course was complicated by late post-operative extubation due to fluid overload and poor respiratory toilet. He was transferred out of the ICU on the second post-operative day and discharged on the ninth day. During the admission he received seven units of RBCs without immediate complications. On discharge his creatinine level was 2.1.

He was well for the first week after discharge, but over the following week developed progressive weakness and jaundice, and was admitted to another hospital for confusion. Neutropenia and fever were noted and on the 24th postoperative day the patient was transferred to the hospital at which he had his operation. The patient was hypotensive with severe jaundice and a deep red macular rash over his entire body. He had worsening renal function (creatinine ~5), neutropenia, and thrombocytopenia. He was drowsy but had no localizing neurologic signs.

Skin biopsy demonstrated a scant perivascular chronic inflammatory infiltrate in the upper dermis as well as epidermal hyperplasia, acanthosis, and apoptosis with a lymphocytic infiltrate consistent with an erythema multiforme-like drug eruption. A CSF culture grew *Candida albicans* the day following admission and 4 days later a blood culture yielded the same organism. Multiple antibiotics were given, including amphotericin, and he received high dose steroids. Respiratory distress prompted ventilation.

The patient's jaundice, pancytopenia, and renal failure progressed and on the 31st post-operative day he developed acidosis, unresponsive to bicarbonate infusion and ischemic extremities. He died on the 32nd post-operative day.

HLA typing was performed on the patient and 5 of the 7 donors of blood received during the first hospitalization.

	HLA-A	HLA-B	HLA-Cw	HLA-DRB1	HLA-DQB1	HLA-DPB1
Patient	1,24	7,8	7,-	03011,15011	02,0602/11	0401,1901
Donor 1	2,24	18,60	3,5	0401,1302	0302,0604	0401,-
Donor 2	2,24	35,63	4,7	1302,1402	0301,0604	0402,1001
Donor 3	1,-	8,-	7,-	0301,-	02,-	0401,0402 (2301,5101)
Donor 4	3,26	37,60	3,±6	15011,1302	0602/11,0604/08	0401,0301
Donor 5	2,11	35,60	3,4	03011,15011	02,0602/11	0401,0201 (0402,3301)

At autopsy the patient was found to have a hypoplastic marrow, an inflammatory reaction in the liver and skin, disseminated candidiasis with fungal enteritis at all levels of the GI tract and abscesses in the brain, liver, kidneys, lungs, spleen and thyroid, recent atheroemboi in the spleen, pancreas and heart, and extensive atherosclerosis.

Questions:

1. What transfusion reaction is present in this case? What clinical features allow you to make that diagnosis? How would you classify this reaction within the various classification systems used for transfusion reactions?
2. This type of transfusion reaction was originally described in patients who were immunosuppressed. Do you think that this patient was immunosuppressed? What is the pathogenesis of the reaction in this case? Which donor do you think was responsible for this reaction? Why?
3. Why did the patient have a hypoplastic marrow? How does this reaction differ from the more common presentation of this problem in the clinical situation in which it is typically seen? How does this explain his most prominent cause of death?
4. How might this reaction have been prevented? What immunosuppressed patients should receive this form of prophylaxis? What type of donor would increase the likelihood of this reaction?