(n=2), thoracic wall and lung (n=1), cutaneous involvement (surrounding a central venous access) (n=1). All patients received high doses of amphotericin B and surgical debridement was performed in three. One patient is alive, three are dead (two died of mucormycosis and one of CMV pneumonia).

We conclude that mucormycosis is not a common infection in patients undergoing HT but a high morbidity and mortality follow it. Sustained neutropenia is the most important risk factor. The early diagnosis followed by prolonged treatment with amphotericin B and surgical debridement, when possible, can improve the survival of these patients.

Key words

Mucormycosis, transplantation, hemopoiesis

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Transfusion-related acute lung injury associated with an NA1-specific antigranulocyte antibody

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Transfusion-related acute lung injury (TRALI) is an infrequent complication of hemotherapy. Antigranulocyte antibodies, most of them present in donor's serum, have been implicated in its pathogenesis. We describe a case of TRALI, following red blood cell transfusion, associated with an antigranulocyte antibody with NA1 specificity in the patient's serum.

A 70-year-old female with a history of previous transfusions was admitted for an elective prosthetic hip implant. Following surgery a single unit of non-buffy-coat deprived packed red blood cells with saline, adenine, glucose, mannitol, (SAG-M) was transfused. Thirty minutes later the patient developed acute respiratory failure. A chest X-ray revealed bilateral alveolar infiltrates with a non-dilated heart, findings consistent with acute pulmonary edema. A Swan-Ganz catheter was placed, showing pulmonary and central venous pressures suggestive of TRALI. Systemic corticosteroids (prednisolone 2 mg per kg) were started, and the patient required mechanical ventilatory support. The clinical course was favorable with resolution within 48 hours. In order to establish a serologic diagnosis, antileukocyte antibodies were searched for in both the patient's and donor's serum. Anti-HLA antibodies were ruled out with a lymphocytotoxic test using the patient's serum and a lymphocyte panel (n=18) of known HLA phenotypes. The presence of specific antigranulocyte antibodies was studied with granuloagglutination and an indirect immunofluorescence (GIFT) test. Both tests showed the presence of an antigranulocyte antibody in the patient's serum, and when tested against granulocytes of known phenotype, the antibody was shown to be specific for NA1 (Table 1). The patient's and donor's granulocyte phenotypes were established by an immunofluorescence technique with flow cytometry (FACScan, Becton Dickinson, San José, CA, USA) using monoclonal antibodies specific for NA1, NA2 and CD16. The patient's phenotype was NA2/NA2, $CD16^+$, while the donor's was found to be NA1/NA2, $CD16^+$. Finally, once the antibody's specificity had been established, a confirmatory bidirectional crossmatch was perfomed with a positive reaction with the patient's serum and the donor's granulocytes (Table 1). The diagnosis was TRALI associated with an antigranulocyte antibody with NA1 specificity in the patient's serum.

TRALI is a relatively infrequent transfusion-related complication, although it ranks second in transfusion-related mortality.[1] TRALI has been described following the transfusion of the majority of blood components;[2-9] its incidence has been estimated as 1 in 5000 transfusions.[2] Clinically TRALI presents as an adult respiratory distress syndrome. Diagnosis requires a high index of suspicion and is made by exclusion. With appropriate supportive treatment, 80% of patients can be expected to recover fully, and mortality ranges from 5 to 10% in most studies.[2] The pathogenesis of TRALI is not fully understood,

Table 1.

Phenotype of tested granulocytes	*NA1NA1*	*NA2NA2*	*NA1NA2 (Donor)*	*NA1NA2*
Patient's serum	+++	–	++	++
Multispecific anti-HLA antiserum	+++	+++	+++	+++
AB serum	–	–	–	–

although there are two plausible hypotheses: an immune-mediated reaction or direct lung injury by biologically active lipids generated during the storage of the blood product.[10] Classically it has been attributed to the presence of antileukocye antibodies in the patient's or donor's serum, which are found in less than 50% of cases with specific tests.[12] In 90% of these cases the antibodies are, however, found in the donor's serum,[11] unlike our case. Previously described antigranulocyte antibodies have shown specificities for the NB and 5b antigens.[4,6,8] Other antibodies implicated have been anti-HLA class I antibodies,[11] anti-HLA A2[4] and B35-specific antibodies.[6] Currently the role of these antibodies in the pathogenesis of TRALI is controversial, since anti-HLA antibodies are found in 1-2% of the general population.[2] Recently, Silliman *et al.* suggested that TRALI may be mediated by biologically active lipids generated during the storage of blood products, particularly when transfused to patients with certain predisposing factors, leading to a *syndrome of neutrophil overactivation* with endogenous cytokine release, indiscriminate endothelial neutrophil adhesion and activation.[10] Although unproven, the concept of a multifactorial pathogenic mechanism, including the effect of specific antigranulocyte antibodies, active lipids and other as yet undefined factors, emerges as a more rational explanation of this complex syndrome.

Key words

Transfusion-related injuries, antigranulocyte antobodies

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Plasma and urinary endothelin-1 titers and plasma von Willebrand activity in *Pseudoxanthoma Elasticum*

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We found high endothelin-1 and von Willebrand factor plasma titers not only in two individuals (daughter and father) affected with *Pseudoxanthoma elasticum* but also in a young unaffected relative. These findings raise the possiblity that these molecules could be the first biochemical fingerprints of this, still not clinically evident, rare inherited disorder of elastic tissue.

Pseudoxanthoma elasticum (PXE) is a rare inherited disorder of elastic tissue characterized by progressive calcification of the elastic fibers in the skin, retina and cardiovascular system;[1] the estimated prevalence of this disease is 1 in 70,000-100,000. A more common autosomal recessive and a less common autosomal dominant pattern of inheritance, with high penetrance, have been described. Recently, an area on the long arm of chromosome 16 (16p13.1) was identified as the single gene that accounts for both the recessive and dominant forms of PXE.[2] The most characteristic clinical manifestations of PXE are yellowish grouped papules and plaques on the skin of flexure areas, angioid streaks in Bruch's membrane of the retina, calcified cardiovascular lesions, and severe hemorrhagic diatheses.[1] Diagnosis of PXE is based on clinical evaluation, histologic demonstration of abnormal, calcified elastic fibers in skin biopsy, and fundoscopic examination showing the presence of the typical angioid streaks.[1]

We recently cared for a 41-year-old woman affect-