Case Report

Kaposi sarcoma in a paediatric renal transplant recipient

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Introduction

The increased incidence of certain neoplasms is one of the major complications of immunosuppression introduced after renal transplantation. Among these Kaposi's sarcoma (KS) accounts for about 5.6% of the neoplasms in organ transplant recipients [1,2]. We herein report a rare case of paediatric KS developing 8 months after renal transplantation.

Case Report

A 15-year-old girl was transplanted from a living-unrelated donor in April 1994. Her primary disease was reflux nephropathy and she received the renal graft in a transplantation centre in India. When the patient was first admitted 2 weeks later, she was treated with pulse methylprednisolone for an acute rejection episode. Triple therapy with cyclosporin A (CsA) 5 mg/kg, azathioprine 2 mg/kg, and oral corticosteroid was continued. After the rejection episode she developed pericardial effusion and fever. Cytomegalovirus (CMV) was diagnosed from the recent increase of CMV IgM titre and she received ganciclovir. Her renal and liver function tests were normal and the pericardial effusion had disappeared when she was discharged 2 weeks later.

In September 1994 the patient’s azathioprine was decreased to 1 mg/kg because of a slight elevation of liver function tests. Serum CsA level was normal and this drug was continued at the same dosage. The markers for hepatitis viruses and Epstein–Barr virus were negative.

In early December the patient presented with violaceous papules and nodules. Dermatological examination revealed a red violaceous papule on the left forearm, 0.5 cm in diameter, which faded under pressure, and a similar papule of the same size which did not fade on pressure on the medial aspect of the right ankle (Figure 1). She also had two desquamated, violaceous macular lesions on the distal one-third of the leg (Figure 1). Skin adnexial structure and mucosae were normal. She did not have any lymphadenopathy. Her physical examination was otherwise normal.

KS was suspected and was confirmed by biopsy. The patient’s HIV antibody, repeated twice, was negative. Renal function tests, and CsA level were within normal ranges. Abdominal and thoracic ultrasonographies were normal.

Fig. 1. Two Kaposi sarcoma lesions on the right leg of the child.
A diagnosis of purely cutaneous Kaposi was made; azathioprine and CsA were stopped. Oral corticosteroid was continued and increased to 1 mg/kg per day. Two months later there had been no marked regression in her lesions. Cyclophosphamide at a dose of 2 mg/kg per day was given for 3 months. At follow-up the macular lesions had increased in number to 17, in different sizes, with again no apparent organ involvement. The patient was thus put on systemic vinblastine therapy. Two months later the new macular lesions had completely disappeared and there was marked regression in the initial three lesions. The graft was functioning well and there was no systemic involvement. A month later she is fine with further regression of lesions.

Discussion

With the use of more potent immunosuppressives, malignancies have arisen as an important focus for morbidity and mortality in transplant recipients. CsA has been the most frequently implicated drug among the immunosuppressives employed for renal transplantation. In adult patients Kaposi sarcoma arises on average 16.5 months after transplantation [3], but although there are only a few paediatric cases, the time interval among childhood patients seems to be much shorter: Fournet et al. [4] reported a case in 7 months, similar to ours, and Al Sulaiman et al. [5] reported three cases who were diagnosed in 3–5 months. Thus in the few cases reported Kaposi seems to develop earlier in childhood cases. Another epidemiological aspect of this malignancy is its predilection for certain ethnic groups. It is noteworthy that this was the first and only malignancy observed among the 25 transplanted paediatric cases from our centre in Turkey.

Herpesvirus-like particles have recently been suggested to have a pathogenic role in Kaposi sarcoma [6]. These particles have been reported to be highly homologous to Epstein–Barr virus. We have not been able to look for herpesvirus-like sequences in our patient; however, serology for these two viruses were negative. On the other hand our patient had had a recent CMV infection. Whether this was a coincidence or whether this virus is also effective in the pathogenetic process awaits further clarification.

Immunosuppressive treatment must be stopped in cases with visceral involvement; various treatments are employed afterwards [3,7]. Interferon has been advocated in some although there is much concern about graft rejection. Vincristine and bleomycin, or vincristine and vinblastine have been used as treatment [1]. Prognosis in childhood cases with visceral involvement has been quite poor [4,5]. On the other hand our patient did not have any visceral involvement. In purely cutaneous cases decrement of immunosuppressives, especially cyclosporin, are suggested in an attempt to preserve the graft [7]. A trial of cyclophosphamide in our patient was unsuccessful. Since there was an increase in the lesions vinblastine was started, with apparent success. We suggest that in purely cutaneous cases reduction of immunosuppressives may not be sufficient and that vinblastine is an effective form of treatment in childhood cases. In purely cutaneous cases conserving the graft should be attempted.

References


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