Case Report

Kaposi sarcoma and gonadoblastoma dysgerminoma with gonadal dysgenesis following cadaveric renal transplantation

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Introduction

It is known that secondary malignancies are the most important long-term complications of transplantation. Among these cases malignancy incidence is 100 times higher than that of an age-matched general population [1–3]. Lymphomas are the most frequent post-transplantation tumours [2,3]. Our case, a female phenotype with 46XY gonadal dysgenesis, already carries a risk for malignancy, especially for gonadoblastoma and dysgerminoma [4]. The patient was pre-pubertal and with no genital abnormality before the transplantation. When she was 15 years old, 16 months after transplantation, the tumour started to appear. Immunosuppressive treatment appeared to give rise to early tumour development. Although there was no recurrence of ovarian tumour, Kaposi sarcoma (KS) was diagnosed 2 years later. This case is interesting since it presents primary and secondary malignancies together.

Case report

A 14-year-old female phenotype patient with end-stage renal disease who did not have any previous family history was transplanted from a cadaveric donor on 27 May 1992. Her condition began as nephrotic syndrome when she was 3.5 years old. At the age of 9 years, she experienced convulsions and started to feel fatigue. By this time her renal function had failed and she was treated with regular haemodialysis until the transplantation. Following the transplantation a triple immunosuppressive regimen was performed with anti-thymocyte globulin (ATG), azathioprine (Aza), and prednisolone (PRD).

As soon as the patient’s renal function returned to normal ATG was replaced by cyclosporin (CsA). During the post-transplantation period she experienced 10 days of anuria and underwent five haemodialysis sessions. Twenty-five days later the serum creatinine declined to 1.5 mg/dl. Antirejection steroid pulse treatments were performed in the 6th and 22nd months.

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After 16 months of transplantation she presented to the hospital with fever and headache, but our attempts to find an origin of infection were unsuccessful.

By this time ultrasonographic examination had revealed a possible dermoid cyst in the right ovary and pelvicalyceal ectasia in the transplanted kidney. Gynaecological examination also showed a palpable semimobile mass in the pouch of Douglas. Later, β-hCG started to increase (150 mIU/ml; normal, 5 mIU/ml) and ultrasonography revealed an 11.2 × 6.2 × 7.4-cm round mass in the right inguinal region next to the bladder, showing both cystic and solid structures. There was also ascites in the peritoneal cavity, and the patient’s uterus was compatible with her age.

These findings prompted the decision to perform exploratory laparotomy, and a 300-g ovarian tumour was removed measuring 11.5 × 9 × 6 cm. This mass showed lobulation and calcification, and contained necrotic and haemorrhagic areas. Histopathological examination revealed malignant germ-cell dysgerminoma and gonadoblastoma. (Figures 1 and 2). After the operation azathioprine was stopped and the patient received chemotherapy with etoposide and carboplatin for 6 months. Chromosomal analysis showed 46XY gonadal dysgenesis.

For 2 years the tumour showed no recurrence, but at the end of this time a right supraclavicular mass with a histopathological diagnosis of KS was eventually diagnosed. (Figure 3).

Discussion

Gonadal dysgenesis are very rare conditions, appearing approximately once in every 2000 female births [5].

The expectancy of gonadal tumour in such cases appear to be about 20–30% [5–7]. Our case showed normal female phenotype. Her pretransplant examination revealed no genital abnormality and her genital development was compatible with her age. Gonadoblastoma dysgerminomas are potentially low-grade malignant tumours. Our case did not show any distant metastases or local invasion. It is difficult to say whether there is any effect of immunosuppressive therapy in our case to enhance the development of tumour. A similar case was observed previously [5]. It is reasonable to comment that immunosuppressive agents might cause early development of tumour. Secondary malignancy incidences are 5–6% after renal transplantation, which means that among these patients the risk of secondary malignancy has increased 100 times. Most of these tumours are derived from the reticuloendothelial system; 8% are gynaecological.
tumours, mostly squamous cell carcinomas of the cervix [5]. Other than this a metastatic dysgerminoma case at the age of 18 and an endometrial carcinoma at the age of 28 were also reported [5]. Kaposi sarcoma is also a frequently seen secondary malignancy after renal transplantation and AIDS cases. In the Netherlands the incidence is reported as five cases in 5606 allograft recipients [8]. Hereditary predisposition has been discussed as a risk for KS. Jewish, Mediterranean, black, or Arabic ancestries have been reported by the Cincinnati transplant tumour registry to carry higher risk for KS. Also in Saudi Arabia KS is reported as the most frequent malignancy after renal transplantation. It has been reported that HLA-A2 has an increased incidence among these patients [9]. It has also been noted that HLA-DR5 is increased in KS patients with AIDS [10]. The type of immuno-suppressive regimen is also important for the development of malignancy. Patients treated with CsA and ATG have higher risk of KS. Our case had all the risk factors mentioned above; first azathioprine and later CsA had to be discontinued, and our patient is now under only minimal corticosteroid therapy, and surprisingly, her renal function still under control.

References

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