slow wave (stage 3 and stage 4) sleep and accounts for the excessive daytime sleepiness.

It is important to identify patients with sleep apnoea because effective treatment of the condition is available in the form of nasal CPAP ventilation[1]. Drivers who are involved in road traffic accidents should be asked whether they suffer from sleep apnoea. A seriously impaired driver with sleep apnoea is putting himself and others at risk. Once identified, such a patient must be warned about the risks of driving and should be advised to stop driving until they are successfully treated.

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Concordant Childhood Acute Lymphoblastic Leukemia in Monozygotic Twins

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Summary

Two 4-year-old monozygotic Chinese, female twins developed concordant childhood acute lymphoblastic leukemia (ALL) within an interval of about 2 weeks. Based on morphology and cytochemistry findings of the bone marrow blast cells, a diagnosis of ALL, L1 was made. Immunophenotyping showed the blast cells of both twins expressed similar antigens, i.e. HLA-DR, CD10, CD13, CD19, CD22 and CD34.

Identical blood group, same HLA (human leucocyte antigen) genotype, sex and similar appearance suggest that the twins are monozygotic. Since the bone marrow leukemic cells of both twins were identical in morphology and expressed the same antigens with almost similar percentages of positivity, it is likely that the blast cells were derived from the same single clone. Based on the single clone hypothesis, the leukemogenic event must have arisen in utero in one twin and the cells from the abnormal clone then spread to the other twin via shared placental anastomoses.

Key Words: Concordant, Acute lymphoblastic leukemia, Twins, Monozygotic

Introduction

The incidence of childhood leukemia in children below 15 years of age is estimated to be about 3.5 per 100,000 person per year. If one monozygotic twin develops acute leukemia in childhood, the chances of acute leukemia in the other twin may be as high as 20% to 25%. One possible hypothesis for the high degree of concordance could be due to genetic susceptibility to leukemia since monozygotic twins have similar complement of genes. Another possibility is that the leukemogenic transformation might have arisen in utero as a single clone in one twin. Haemopoietic stem cells are exchanged between monozygotic twins via shared placental anastomoses. This could result in the bone marrow tissue of one twin being colonised by the stem cells of the abnormal clone from the other twin[1].

We report the clinical presentation, morphology, cytochemistry and immunophenotyping findings in two monozygotic twins who developed acute lymphoblastic leukemia (ALL) at about the same time.

Case Report

Twin I, a 4-year-old Chinese girl presented with low grade fever and pallor for 2 weeks, bone and joint pain for 1 week. On examination the liver was 4 cm below the costal margin and lymph nodes were palpable in the cervical, both inguinal and axillary regions. No bruises were noted. A full blood count showed: Hb, 11.2 g/dL; platelets, 29 x 10^9/L and white cell count (WCC), 4.7 x 10^9/L (differential counts: neutrophils 25%, lymphocytes 60%, monocytes 5%).

Ten days later, her twin sister, Twin II, presented with low grade fever, pallor, bone and joint pain for 3 days. On examination the liver was 2 cm below the subcostal margin. There was no bruising and enlargement of the lymph nodes. A full blood count revealed: Hb, 7.6 g/dL; platelets, 120 x 10^9/L and WCC, 2.4 x 10^9/L (differential counts: neutrophils 24%, lymphocytes 70% and monocytes 1%). Phenotypically both twins look exactly alike.

Morphology, cytochemistry and immunophenotyping studies were almost similar in both twins. The bone marrow showed 90% and 85% blasts in Twin I and Twin II respectively, and were hypercellular with markedly reduced hemopoiesis. The blasts were small with scanty cytoplasm and inconspicuous nucleoli. The blast cells were unreactive for myeloperoxidase in both twins and periodic acid-Schiff-positive in Twin II only. A diagnosis of ALL, L1 was made in both twins.

Immunophenotyping was performed by direct flourescence labelling using flow cytometry. Monoclonal antibodies against the following antigens were used: HLA-DR; progenitor stem cells: CD34; B-lymphoid: CD10, CD19, CD20 and CD22; T-lymphoid: CD2, CD3 and CD7 and myeloid: CD13 and CD33. Both twins were positive for HLA-DR, CD10, CD13, CD19, CD22 and CD34 and negative for CD2, CD3, CD7, CD20 and CD33. Dual flourescence labelling showed 45% and 40% of the blasts of Twin I and Twin II respectively coexpressed CD10 and CD13. The phenotypes of the blast cells were designated as B-lineage ALL with a single myeloid-associated antigen expression.

Table I summarises the clinical, hematological, morphological, cytochemical and immunophenotyping findings in both twins at diagnosis. Both twins have the same blood group (group A, rhesus positive) and the same human leukocyte antigen, HLA genotype (A_{1}, A_{24}, B_{22}, B_{62}, Cw_{01}, Cw_{04}).

The twins were treated according to the modified Berlin-Frankfurt Munster (BFM) protocol for ALL. Remission was achieved in both twins using vincristine, prednisolone, L-aspariginase and daunorubicin. At the time of reporting in October 1995, that is about 14 months after diagnosis, the twins are in continuous complete remission.

Discussion

The identical blood group, HLA (A, B, and C) genotype, sex and phenotype of both twins suggest that the twins are monozygotic. However, molecular studies to evaluate polymorphic markers were not done on the twins and their parents to confirm this. It is not known whether the twins had a monochorionic or dichorionic placenta.

Table I
Presenting clinical and laboratory features

Clinical and laboratory features	Twin I	Twin II
Clinical presentation		
Low grade fever	Yes	Yes
Pallor	Yes	Yes
Bone & joint pain	Yes	Yes
Bruises	None	None
Liver (below costal margin)	4 cm	2 cm
Palpable lymph nodes	Yes	None
Hematological data		
White cell count (x 10^9/L)	4.7	2.4
Hemoglobin (g/dL)	11.2	7.6
Platelets (x 10^9/L)	29	120
Bone marrow data		
Bone marrow blasts	90%	85%
French-American-British Classification	ALL, L1	ALL, L1
Myeloperoxidase	negative	negative
Periodic acid-Schiff	negative	positive
Immunologic cell marker*		
HLA-DR	96%	85%
CD10	91%	80%
CD13	48%	48%
CD19	93%	81%
CD22	82%	84%
CD34	92%	66%

** Both twins were negative for CD2, CD3, CD7, CD20 and CD33 antigens*

The leukemic cells of both twins have identical morphology (ALL, L1) and also express the same antigens (HLA-DR, CD10, CD13, CD19, CD22 and CD34) in almost similar percentages. These findings suggest that the leukemic cells in the twins were derived from the same single clone. Recent studies on the clonal nonconstitutional *MLL* gene rearrangement in monozygotic infant twins who develop concordant ALL provide supportive evidence for the intrauterine single cell origin hypothesis with twin to twin transmission[2,3], but this tests are not done in our patients. According to the single clone hypothesis, the leukemogenic event must have arisen in utero in one twin and the cells from the abnormal clone then spread to the other twin via shared placental anastomoses. This would not be impeded by an immunological barrier since monozygotic twins are genetically identical and histocompatible. The hypothesis is based on epidemiologic findings that concordant leukemia in monozygotic twins decreases with age, being highest during the first year of life and the risk of leukemia occurring after six years of age is similar to that of siblings, and also that the interval of onset of concordant leukemia between twins may be simultaneous or short, usually within weeks or months[1].

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Spontaneous Rupture of Renal Angiomyolipoma Presenting as Acute Abdomen

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Summary

Five cases of renal angiomyolipoma which underwent spontaneous rupture are described. These patients presented as an "acute abdomen" for which the diagnosis was not initially apparent. A high index of suspicion is required to make the diagnosis even with modern imaging techniques. The treatment of these tumours is discussed.

Key Words: Renal angiomyolipoma, Spontaneous rupture

Introduction

Renal angiomyolipomas are uncommon hamartomas of the kidney which have a propensity for bleeding. These patients often present acutely as a result of the haemorrhage which may be life-threatening. We report five patients who presented with an acute abdomen and discuss the role of radiological imaging in reaching the diagnosis.

Case 1

The patient was a 48-year lady known to have tuberous sclerosis. She presented with the sudden onset of right sided abdominal pain and swelling. On examination she was pale and tachycardic with a blood pressure of 90/40mmHg. Abdominal examination revealed a large tender mass in her right flank. Her haemoglobin was 9g/dl on admission and she was treated conservatively with blood