

## LIPOBLASTOMA AND LIPOSARCOMA IN CHILDREN: AN ANALYSIS OF 9 CASES AND A REVIEW OF THE LITERATURE

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**OBJECTIVES:** To review the experience at a children's hospital of lipoblastoma and liposarcoma and to identify any factors that would differentiate one type of tumour from the other.

**DESIGN:** A retrospective case series.

**SETTING:** British Columbia's Children's Hospital a tertiary-care pediatric centre.

**PATIENTS:** All patients with a pathological diagnosis of lipoblastoma and liposarcoma recorded over 12 years.

**MAIN OUTCOME MEASURES:** The frequency of lipoblastoma and liposarcoma, identified from biopsy specimens of pediatric adipose tumours. The clinical, pathological and cytogenetic variables between lipoblastoma and liposarcoma.

**RESULTS:** One hundred and forty-nine adipose tumours were recorded. Seven (4.7%) were lipoblastomas and 2 (1.3%) were liposarcomas. All tumours presented as asymptomatic, slow-growing, soft-tissue masses. The children with lipoblastoma tended to be younger, but 29% were over 3 years of age. The liposarcoma patients were aged 9 and 14 years. One liposarcoma was of myxoid type and the other was a round cell variant. Karyotypes were reported for 1 lipoblastoma and 1 liposarcoma. The myxoid liposarcoma karyotype was 46,XY,t(12;16)(q13;p11), and the lipoblastoma was reported as 46,XY,der(8)?t(8q;?),+mar.

**CONCLUSIONS:** Lipoblastoma is an unusual childhood neoplasm and liposarcoma is very rare in children. Both tumours may present in a similar fashion, and differentiating them histologically can be difficult. Age cannot be relied upon to accurately predict their behaviour. The tumour karyotype is very helpful in differentiating these neoplasms.

**OBJECTIFS :** Étudier l'expérience, à un hôpital pour enfants, des lipoblastomes et des liposarcomes et définir les facteurs qui permettraient de distinguer un type de tumeur de l'autre.

**CONCEPTION :** Étude de cas rétrospective.

**CONTEXTE :** British Columbia's Children's Hospital, centre pédiatrique de soins tertiaires.

**PATIENTS :** Au cours d'une période de 12 ans, tous les patients chez lesquels on a diagnostiqué la présence pathologique d'un lipoblastome et d'un liposarcome.

**PRINCIPALES MESURES DE RÉSULTATS :** La fréquence du lipoblastome et du liposarcome, déterminée à partir de spécimens prélevés par biopsie pratiquée dans des lipomes chez des enfants. Les variables cliniques, pathologiques et cytogénétiques entre le lipoblastome et le liposarcome.

**RÉSULTATS :** On a consigné 149 lipomes, dont sept (4,7 %) lipoblastomes et deux (1,3 %) liposarcomes. Toutes les tumeurs se manifestaient sous la forme de masses de tissus mous asymptomatiques et à croissance lente. Les enfants atteints d'un lipoblastome avaient tendance à être plus jeunes, mais 29 % avaient plus de trois ans. Les patients atteints d'un liposarcome avaient 9 ans et 14 ans. Un liposarcome était de type myxoïde et l'autre était une variante à cellules rondes. On a signalé la présence de caryotypes dans un lipoblastome et un liposarcome. Le caryotype du liposarcome myxoïde était 46,XY,t(12;16)(q13;p11) et celui du lipoblastome, 46,XY,der(8)?t(8q;?),+mar.

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CONCLUSIONS : Le lipoblastome est un néoplasme inusité et le liposarcome est très rare chez les enfants. Les deux tumeurs peuvent se présenter de la même façon et il peut être difficile de les distinguer sur le plan histologique. On ne peut compter sur l'âge pour prédire avec exactitude leur comportement. Le caryotype tumoral est très utile lorsqu'il s'agit de distinguer ces néoplasmes.

The majority of soft-tissue tumours occurring in childhood are benign and of vascular or fibroblastic origin. Adipose tumours are relatively unusual, making up about 6% of soft-tissue neoplasms reported in the first 2 decades of life.<sup>1</sup> Approximately two-thirds of these are simple lipomas, up to 30% are lipoblastomas and liposarcomas are very rare. Although benign in nature, lipoblastoma bears a striking histologic similarity to myxoid liposarcoma, a low-grade malignant tumour that is rare in childhood.<sup>2,3</sup> Traditionally, pathologists have used the patient's age to differentiate these neoplasms. Recently, cytogenetic analysis of the tumour tissue has revealed specific markers that can be very helpful in differentiating the neoplasms.

We undertook a review of all cases of lipoblastoma and liposarcoma over a 12-year period at British Columbia's Children's Hospital (BCCH) and compared the clinical, pathological and cytogenetic features of these tumours.

## PATIENTS AND METHODS

We searched the records of the BCCH Department of Pathology from 1982 to 1994 for adipose tumours, using the following tissue codes: lipoma — not otherwise specified (NOS), lipomatosis — NOS, lipoblastomatosis, liposarcoma — NOS, liposarcoma — well differentiated and myxoid liposarcoma. The BCCH health records of children whose pathologic diagnosis was lipoblastoma or liposarcoma were reviewed for demographic features, clinical presentation, treatment, pathological features and outcome.

## FINDINGS

One hundred and forty-nine cases of adipose tumours were identified. There were 7 cases of lipoblastoma and 2 of liposarcoma. The remaining tumours were lipomas. Table I summarizes the clinical data of the 9 lipoblastoma and liposarcoma cases. The

median patient age at the time of excision for the lipoblastomas was 2.2 years (range from 0.1 to 9.6 years). The 2 children with liposarcoma were aged 9 and 14 years. All tumours presented as painless, progressively enlarging soft-tissue masses. The liposarcomas were both located on extremities, and their maximum dimensions were 2 and 10 cm respectively. Six of the 7 lipoblastomas were located on the trunk. The average maximum dimension of the lipoblastomas was 4.7 cm (range from 2.5 to 7.5 cm). All tumours were completely excised except 1 lipoblastoma (case 1, Table I). In 6 of the 9 cases there were no tumour recurrences reported an average of 1.6 years (range from 3 months to 2 years) after excision. Two children with lipoblastoma and 1 with liposarcoma were lost to follow-up.

Generally, the tumours had a similar gross appearance with partial or complete encapsulation and a multilobular configuration. On sectioning, they were of tan, brown or dull white

**Table I**

**Data on 9 Children With Lipoblastoma or Liposarcoma**

Case no.	Sex/age, yr	Tumour location	Tumour size, cm*	Pathological findings	Tumor karyotype
1	M/8.3	Abdominal wall	4.0	Lipoblastoma	NR
2	F/0.1	Buttock	3.5	Lipoblastoma	NR
3	M/9.6	Foot	2.5	Lipoblastoma	NR
4	F/2.2	Axilla	6.0	Lipoblastoma	NR
5	M/0.3	Thorax	NR	Lipoblastoma	NR
6	F/0.6	Axilla	NR	Lipoblastoma	NR
7	M/14	Leg	2.0	Liposarcoma, round cell	NR
8	M/9	Thigh	10.0	Liposarcoma, myxoid	46,XY,t(12;6)(q13;p11)
9	M/1.6	Buttock	7.5	Lipoblastoma	46,XY,der(8)?t(8q;?),+mar

\*Maximum tumour dimension  
NR = no record

appearance with soft, sometimes fluctuant myxoid or gelatinous-like areas. Histologically, the lipoblastomas were characterized by lobules of mature and immature fat cells, primitive mesenchymal cells, and monovacuolar and multivacuolar lipoblasts. The lobules were separated by fibrous trabeculae containing small blood vessels. Two of the lipoblastomas were diffuse in nature (lipoblastomatosis) and the remainder were encapsulated. The myxoid liposarcoma had a similar gross and microscopic appearance, although the lobules were not as large and occasional mitosis and nuclear atypia were noted. The round cell liposarcoma also had a similar appearance but with predominantly closely packed, undifferentiated, monomorphic round cells and occasional interspersed lipoblasts, and a branching trabecular network in the less cellular portions.

Cytogenetic analysis of the tumour tissue was performed in the 2 most recent cases. The karyotype of case 8 (Table I) revealed a reciprocal translocation of chromosomes 12 and 16 with breakpoints involving band 13 of the long arm of chromosome 12 and band 11 of the short arm of chromosome 16. The final full karyotype was 46,XY,t(12;16)(q13;p11). In case 9 (Table I) the karyotype demonstrated a structural rearrangement of chromosome 8 and a deletion of the long arm of chromosome 8, with or without additional unidentified chromosome material. A small marker chromosome with the morphology of a ring was also present. The full karyotype was 46,XY,der(8):t(8q:?),+mar.

## DISCUSSION

Lipoblastomas are reported to occur primarily in infants and children less than 3 years of age.<sup>4,5</sup> In this series 2 of the 7 children were over the age

of 3 years. Liposarcomas occur most commonly in the 3rd to 7th decades of life. They are extremely unusual in children younger than 10 years. Of 2500 cases of liposarcoma reviewed by the United States Armed Forces Institute of Pathology (AFIP), 17 occurred in children under 16 years of age, but only 2 children were younger than 11 years.<sup>3</sup> In this report the 2 children with liposarcoma were aged 9 and 14 years.

In children, liposarcomas usually occur on an extremity, as in both cases reported here. A truncal location is less common and retroperitoneal tumours are uncommon. In a large series of lipoblastomas, 70% were reported to occur on an extremity.<sup>5</sup> Others reported a predilection for the trunk.<sup>6,7</sup> We found only 1 occurring on an extremity.

There are 2 types of lipoblastoma. One is well encapsulated, superficial and may mimic a lipoma. The other is deeper with an infiltrative growth pattern. It is sometimes referred to as lipoblastomatosis. Two of our cases were poorly encapsulated.

One of our liposarcomas was a myxoid variant, which is the most common histologic type reported in children. The other was a round cell type, which is very unusual in children.

Lipoblastoma is definitively treated by complete resection. Recurrence rates are reported in the range of 13% to 20%.<sup>4,5,8,9</sup> In this series, all tumours but 1 were completely resected and there have been no recurrences. Of the 17 pediatric liposarcomas in the AFIP series there were 3 recurrences (18%) and 1 death (6%). One of our children with liposarcoma was lost to follow-up and the other had complete resection and was disease free at 6 months. In adults, liposarcoma recurs in 53% to 86% and the 5-year survival rate is 47% to 77%.<sup>10</sup> The use of adjuvant therapy for childhood liposar-

coma is not advocated for most completely resected tumours.<sup>3</sup>

The translocation anomaly of the tumour karyotype in case 8 is characteristic for myxoid liposarcoma.<sup>11,12</sup> This anomaly has not been reported in lipoblastoma. The tumour karyotype of a number of lipoblastomas has been reported and 6 of 7 (86%) reveal a breakpoint in chromosome 8 with variable rearrangements of the nuclear material.<sup>13-16</sup> We examined the tumour karyotype in our most recent case of lipoblastoma and found a similar breakpoint in chromosome 8. This anomaly has not been reported in liposarcoma.

The tumour karyotype plays a significant role in differentiating these tumours. Microscopically, lipoblastoma can be indistinguishable from myxoid liposarcoma.<sup>10</sup> Clinically, the tumours often behave in a similar fashion. Age has been traditionally used to substantiate the diagnosis. However, as we found, age cannot be relied upon.

In summary, adipose tumours other than lipoma, are rare in children. Both lipoblastoma and liposarcoma can present as nontender, slow and progressively growing soft-tissue masses. Lipoblastoma tends to occur on the trunk of children less than 3 years of age. Liposarcoma is much less common and tends to occur on an extremity of older children and adolescents. These clinical features, however, cannot be relied upon to accurately distinguish between lipoblastoma and liposarcoma, nor can the histologic findings. However, the tumour karyotype seems to be specific. Therefore, we recommend that for a child with an unusual tumour of adipose tissue (i.e., mucinous, invading fascia, not well localized) a fresh specimen of tumour tissue to be submitted for tissue culture and biologic evaluation.

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