EXTREMITIES AT RISK CASE STUDY: ALVEOLAR RHABDOMYOSARCOMA

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History:

A 12-month-old, otherwise healthy, Asian male presented to his primary care physician with a right thigh mass - he did not have other symptomatology nor did the mass appear to bother him. His birth history was unremarkable and he displayed normal growth and development, walking and starting to talk. Immunizations were up to date. Since he had recently begun to walk and may have incurred some minor trauma, the differential diagnosis included hematoma. His PCP followed him closely and ordered an MRI when the mass did not resolve rapidly. He was then referred to pediatric oncology for further evaluation.

Clinical Exam:

The patient appeared to be an active, healthy child with a right posterolateral thigh mass near his buttock which measured approximately 6 x 7 cm. The right thigh circumference was 28 cm and the left was 25 cm. He had no abnormal adenopathy and no palpable inguinal nodes.

Radiographic Evaluation:

MRI showed a well-defined, non-enhancing mass rising from the inferior aspect of the gluteus maximus muscle (Fig 1). CT of the chest was negative for any focal nodules and a CT of the abdomen/pelvis was notable for the primary mass but revealed no evidence of metastasis. A total body bone scan and skeletal survey were negative. Other staging studies included bilateral bone marrow aspirates and biopsies that were also negative for tumor spread.

Pathology:

A needle biopsy of the mass performed by an orthopedic oncologist was diagnostic for rhabdomyosarcoma. Cytogenetic studies and molecular translocation studies were consistent with the alveolar sub-type of rhabdomyosarcoma.

Treatment:

Following definitive resection with negative margins, he commenced adjuvant chemotherapy per the national treatment protocol for children with rhabdomyosarcoma. The protocol included cycles of vincristine, actinomycin-D and cyclophosphamide (VAC) administered for a total of 42 weeks.

Results:

This patient has now been off chemotherapy for 12 months and is without evidence of recurrent disease. He is approaching his third birthday and has returned to all normal activities without any physical sequelae. He is seen frequently by his pediatric and orthopedic oncologists.

Discussion:

Rhabdomyosarcoma (RMS) is a highly malignant small blue cell tumor. It is the most common soft tissue sarcoma in children under age 15. Of the 350 new cases reported in the U.S. each year, two-thirds are diagnosed in children under age seven. RMS is believed to arise from immature mesenchymal cells and diagnosis is made by histological pattern. Embryonal RMS is the most common histologic sub-type, accounting for 70-80% of all RMS. Alveolar RMS has a slightly worse prognosis than embryonal and characterizes 10-20% of RMS cases. Tumors can occur anywhere in the body but are predominantly in these areas: 1) head and neck, 2) genitourinary tract and retroperitoneum, and 3) upper and lower extremities. An adequate biopsy is critical in order to obtain sufficient tissue for accurate diagnosis and molecular characterization.

Prior to 1960, childhood RMS was an almost uniformly fatal neoplasm that recurred and metastatisized in a high percent of cases. Today, advances in combination therapy, including conservative surgery, multimodality chemotherapy and, in certain situations, radiotherapy, have maximized cure rates. The prognosis is based on location, extent of disease, and histologic subtype. With modern treatment, 75% of low to intermediate-risk patients remain free of disease at five years.

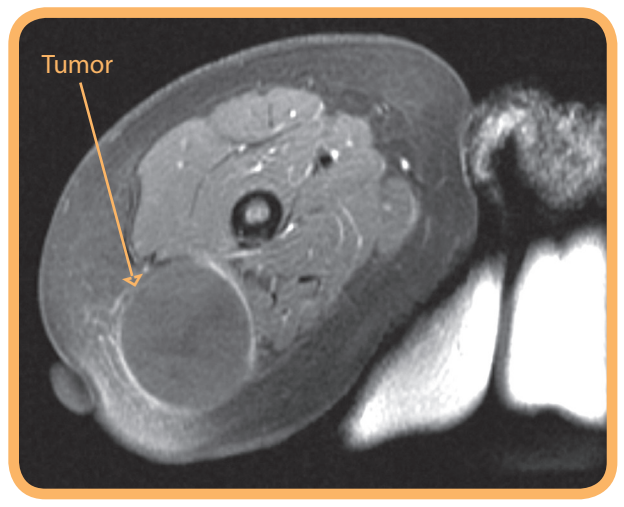

Fig. 1

Further Reading:

Breitfeld, PP, Meyer WH: Rhabdomyosarcoma: new windows of opportunity. Oncologist 2005;10:518-27.

Stevens, MCG, Rey A, Bouvet N, et al: Treatment of nonmetastatic rhabdomyosarcoma in childhood and adolescence: third study of the international society of paediatric oncology. J Clin Oncol 2005;23:2618-28.

Lanzkowsky P: Rhabdomyosarcoma and other soft tissue sarcomas. In Manual of Pediatric Hematology and Oncology. Pgs 561-573.4th edition. Elsevier Academic Press, 2005, p. 561-73.

It is our hope that this information will help you with difficult patient diagnoses you may encounter in your own practice. If you have a patient you would like to present at our conference, or request a consultation, please notify our staff, locally at 303-832-5462 or toll-free at 1-800-262-5462. Visit our website: www.TheDenverClinic.com.

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> Save the Date

Monday, December 6, 2006
7am – 1pm

Extremities at Risk Annual Conference:

Assessment & Treatment of Orthopaedic Complications

Keynote Speaker:
Howard Rosenthal, MD

"Use of CyberKnife/ Radiosurgery for Extremity & Spine Tumors"

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Extremities At Risk Case Study:
Alveolar Rhabdomyosarcoma

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