Case report 612

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Radiological studies and legends

Clinical information

This 21-month-old white female presented to an outside hospital with a 1-week history of fever, intermittent abdominal pain, vomiting, and diarrhea. A firm, nontender mass was palpable to the right of the patient’s 11th thoracic vertebra. Abdominal radiographs revealed a posterior soft tissue mass at the T11–12 level. Sonography (Fig. 1 A) demonstrated a 3 × 5 cm retroperitoneal mass superior to the right kidney, with an irregular, stellate, cystic-appearing center. A large mass, displacing the right kidney without involvement of the collecting system, was noted by intravenous pyelography. (Fig. 1 B, C) The patient was transferred to The Johns Hopkins Hospital for further evaluation of a suspected neuroblastoma.

Computed tomographic (CT) scans (Fig. 2 A, B) were obtained and demonstrated a 5 cm solid mass with areas of necrosis in the right suprarenal zone displacing the right kidney downward. The mass extended medially into the spinal canal. A presumptive diagnosis of adrenal neuroblastoma was made.

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Diagnosis: Rhabdomyosarcoma of the right psoas muscle

At surgery a tumor arising within the psoas muscle, with a normal right adrenal gland, was found. Pathological review of the mass was consistent with an embryonal rhabdomyosarcoma. (Fig. 3A, B) [5].

Discussion

Rhabdomyosarcoma is the most common soft tissue sarcoma in children, comprising 10% of malignant soft tissue neoplasms and exceeded in frequency only by brain neoplasms, neuroblastoma and Wilms' tumor [5, 14, 19, 22]. The peak incidence of rhabdomyosarcoma in children is between 2 and 6 years [12]. One-third of patients present in the third decade, however, suggesting a bimodal distribution [12]. The incidence is generally greater in males.

The tumor is thought to develop from primitive rhabdomyoblasts or undifferentiated mesoderm [11]. Histologically, rhabdomyosarcoma is divided into three basic subtypes: embryonal, alveolar, and pleomorphic (undifferentiated) [5, 6, 16]. In children the embryonal subtype is the most common and originates most frequently in the head and neck or genitourinary tract [5, 16]. The tumor is characterized by its bulky nature, propensity for local invasion, and rapid hematogenous and lymphatic dissemination [1].

Prior to CT, most patients with rhabdomyosarcoma were evaluated using standard plain films, intravenous urography, barium studies, and infusion venacavagrams [6, 11, 16]. With the introduction of CT, the primary tumor site, extent of local and distant spread and potential resectability, can be accurately determined [1]. CT has been reported to be the optimal method for defining the extent of rhabdomyosarcomas of the prostate and bladder by identifying their relationship to pelvic organs and musculature [1].

The majority of neuroblastomas are diagnosed in children less than 5 years of age, and over one-half manifest clinically in patients less than 2 years of age. The incidence of neuroblastoma, like rhabdomyosarcoma, is slightly higher among males than females [21]. Neuroblastoma can occur in a variety of paraspinal locations and in parenchymal organs. Common sites include the adrenal medulla, extra-adrenal retroperitoneal abdominal areas, thorax, and, less frequently, cervical ganglia and other areas, (e.g., thorax, other areas of head and neck) [2]. Paraspinal neuroblastomas may extend into the spinal column through the intervertebral foramina [17].

In the past, it was attendant upon the pathologist to provide differentiation between rhabdomyosarcomas, neuroblastoma, and other neoplasms of the small, blue-cell tumor group. The differential diagnosis of these tumors can be difficult by conventional morphology [18]. A comprehensive body of literature on the ultrastructural features of these neoplasms has become available in the past several years [3, 7–10, 13, 15, 20]. Ultrastructural features used in differentiation of neuroblastoma from other blue-cell tumors include the presence or frequency of dense core granules, glycogen, various filaments, and cell attachments [21].

As demonstrated by the case presented here, the correct classification of a "small blue-cell tumor" can of-

Pathological material

Fig. 3. A Histopathology of the tumor reveal small, round-to-oval cells with scanty cytoplasm, admixed with small spindle-shaped cells with tails of eosinophilic cytoplasm. Cytoplasmic cross-striations are identified in several of the tumor cells. A loose myxoid stroma is present. B Immunoperoxidase studies show the tumor cells to stain positively for muscle-specific actin and desmin (BioGenex, San Ramon, Calif.), while staining for myoglobin was negative. Staining for desmin highlighted the presence of cross striations in several of the tumor cells. These findings are diagnostic of an embryonal type rhabdomyosarcoma.