Scalp Metastasis of Paraspinal Alveolar Rhabdomyosarcoma

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We describe a 10-year-old girl with paraspinal alveolar rhabdomyosarcoma (RMS) who presented with an asymptomatic soft tissue mass on the left aspect of the parietal scalp of 2 weeks' duration. Biopsy demonstrated metastasis of her RMS. A full-body positron emission tomography (PET) scan revealed multiple areas of increased uptake consistent with extensive metastases. We also discuss the literature on RMS classification and metastasis to the skin.

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Rhabdomyosarcoma (RMS) is one of the most common tumors found in children, accounting for up to 5% of all tumors of childhood[1] and about half of the sarcomas in patients younger than 15 years. There are approximately 250 cases diagnosed each year.[2] These tumors are thought to come from primitive mesenchymal cells in the skeletal muscle that develop into distinct heterogeneous tumor cells.[3] The World Health Organization recognizes 3 major classifications of RMS based largely on the work performed by the Intergroup Rhabdomyosarcoma Study Group (IRSG).[4] Embryonal RMS is the most common type, encompassing 70% to 80% of all RMS cases; it includes the botryoid, spindle cell, embryonal (not otherwise specified), and anaplastic variants. Alveolar RMS comprises approximately 20% of RMS cases. The rare pleomorphic classification occurs almost exclusively in adults and often involves the extremities.[4,5] We describe an unusual occurrence of paraspinal alveolar RMS that metastasized to the scalp in a 10-year-old girl.

Case Report

A 10-year-old girl with previously diagnosed L5-S1 paraspinal alveolar RMS presented to our clinic with asymptomatic soft tissue swelling on the left aspect of the parietal scalp of 2 weeks' duration. Physical examination revealed a nondescript, 4-cm, somewhat firm, nontender, soft tissue swelling without surface changes; a skull radiograph corroborated soft tissue findings (Figure 1). A biopsy of the deep fibrous tissue attached to the calvarium demonstrated a slight bluish tinge (Figure 2). Immunohistological staining patterns with actin, desmin, and myogenin (Figure 3) were consistent with RMS.

Due to this finding of a distant metastasis, a full-body positron emission tomography (PET) scan was performed, which revealed multiple areas of uptake in the bilateral knees, bilateral iliac crests, and left axillary lymphatic chain, as well as the biopsy site on the scalp. Subsequent bone marrow biopsy of the right and left iliac crests showed 50% of the bone marrow spaces filled with a metastatic tumor. Chemotherapy and radiation treatments were reinstituted; however, after 3 months, the patient died.

Comment

Rhabdomyosarcoma is the most common malignant soft tissue tumor in children[6] and thus should be considered in the differential diagnosis of any abnormal soft tissue growth in patients younger than 18 years. In children, the most common site of RMS is the head and neck; there is an increased predilection for the nasal cavities and paranasal sinuses. There also is a high incidence of genitourinary system involvement. Occasionally RMS is detected on the distal extremities and many suggest a worse prognosis with hand or foot involvement.[5] Less common presentations are parameningeal tumors in the cranial vault or in the

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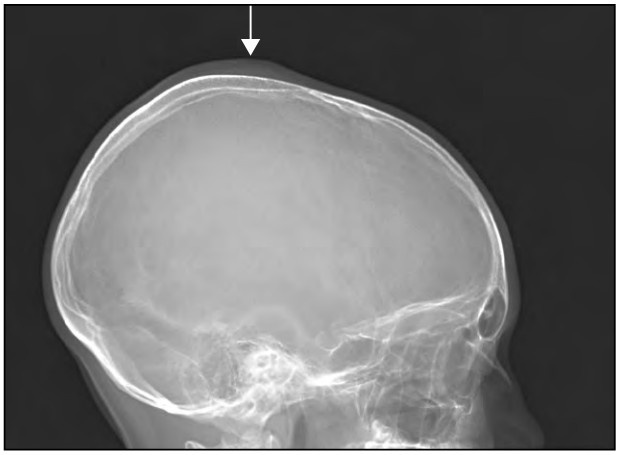

Figure 1. A lateral view of the skull showed soft tissue swelling (arrow).

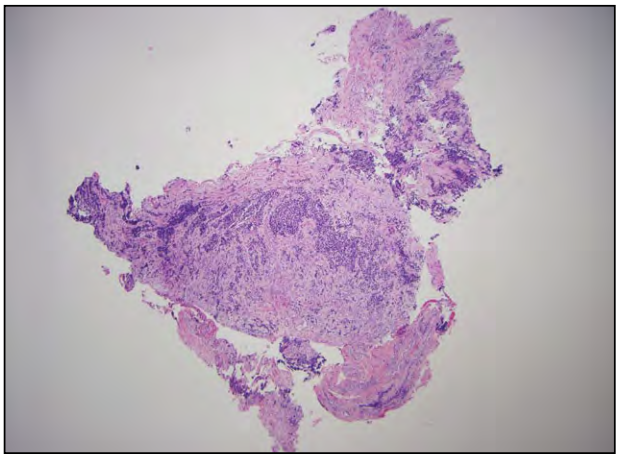

Figure 2. A deep section of a biopsy specimen demonstrated apparent basophilic cells (H&E, original magnification ×4).

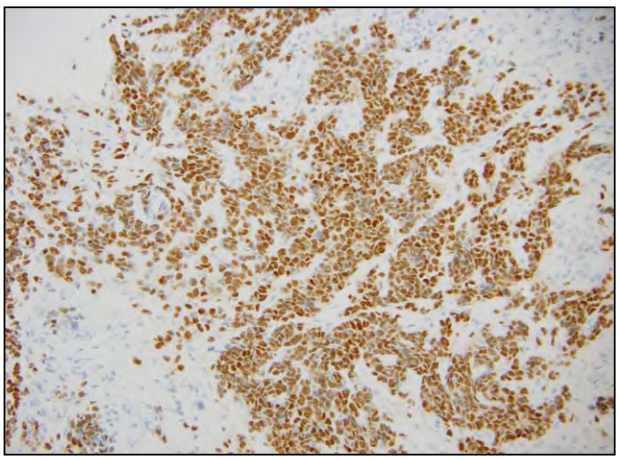

Figure 3. Positive myogenin stain (original magnification ×20).

vertebral canal. Overall, approximately one-fourth of patients present with metastases, most commonly to the lung as well as the lymph nodes, bone, and bone marrow, followed by distant metastasis.[7]

Rhabdomyosarcoma lesions typically are fleshy in appearance but may appear as an erythematous mass with overlying telangiectatic vessels or as a soft tissue swelling. The differential diagnosis to be considered when encountering RMS may be hemangioma, vascular malformation, fibrosarcoma, cyst, infection, and neoplastic or nonneoplastic tumors such as lipoma or angiosarcoma. Workup for RMS typically includes plain radiographs, computed tomography, magnetic resonance imaging, and PET.

The definitive treatment of RMS is complex, usually requiring multiple treatment modalities that are largely based on the size and extent of the tumor as well as the size and extent of metastases. Surgical excision or debulking can improve outcomes; however, certain extensive organ involvement only allows for tissue sampling for determination of subtype and to help appropriately align therapy. Chemotherapy can reduce the size of a tumor to a more manageable size for surgical excision. Typical protocols have shown vincristine, dactinomycin, and cyclophosphamide to have favorable results. Radiotherapy most commonly begins at or around week 20 and has beneficial effects on the microscopic and gross tumor burden. Established treatment guidelines are organized by the RMS subtype, TNM staging system, and IRSG protocols of the surgical-pathologic group system derived after evaluating 4292 RMS patients.[2,8] A review of studies produced by the IRSG consistently reveals that the following factors indicate a good prognosis for RMS: older age of detection, embryonal subtype, female sex, lack of meningeal or lymph node involvement, and no metastasis on diagnosis.[8]

Rhabdomyosarcoma may locally extend and/or metastasize to the skin; metastases usually present as asymptomatic lesions but can be painful on occasion and first present as a mimic of primary skin tumors.[9] Most cutaneous metastases are found when the primary tumor is on an extremity or when the disease has spread to multiple sites, which portends a poor prognosis. The rare congenital (infantile) extramedullary hematopoiesis usually will present with multiple cutaneous lesions and is one of the known causes of the "blueberry muffin baby" appearance.[10] The number of cases of cutaneous metastasis of RMS is difficult to quantify, as the literature often reports metastasis to a particular body region (ie, shoulder, hand) without delineating if it was found on the skin or in deeper tissues.

Few cases of RMS involving scalp lesions have been reported in the literature; a few cases of primary

scalp RMS have been reported. Foxx et al[11] reported a case of primary scalp RMS that then spread locally into the cranial vault. Actual metastases of RMS to the scalp from distant sites are exceedingly rare. Wesche et al[12] reported scalp metastases from a tumor originating in the retroperitoneum. Potenza and Winslow[13] reported multiple metastases from a primary lesion originating on the right thenar eminence. Chaudhuri et al[14] described a patient with lesions that metastasized from the calf to the orbit and scalp.

Our patient presented with paraspinal alveolar RMS that metastasized to the scalp, which adds to the small number of existing cases in the literature. Scalp RMS is likely a sign of distant and notable tumor metastasis, and further testing with PET scanning is likely indicated to determine the extent of tumor burden.

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