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Whole-Body MRI Showing Unusual Pancreatic Metastasis from a Forearm Rhabdomyosarcoma without Lung Involvement

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Rhabdomyosarcoma (RMS) is the most common soft tissue malignancy in children and constitutes 4.5% of all pediatric malignancies.[1] Histologically, RMS is classified into embryonal, alveolar, and pleomorphic subtypes.[2,3] Embryonal RMS is commonly seen in young children in the head and neck region followed by the genitourinary system, whereas alveolar RMS is commonly seen in older children in the extremities. Common sites of metastases are lungs, bone, bone marrow, lymph nodes, and brain for both types.[4] Metastasis to the pancreas is extremely rare, several from an alveolar subtype primary[3] and only one from an embryonal subtype in a patient who presented with acute pancreatitis.[5]

An 11-year-old boy was referred to our clinic with a lump on his right forearm. On physical examination, an approximately 10 × 5 cm subcutaneous mass was evident at the ulnar side of the forearm. The patient's magnetic resonance imaging (MRI) from an outside center revealed a solid neoplastic mass sized 9.5 cm longitudinally and 5.1 × 3.8 cm transversely. A follow-up MRI one month later showed a two-fold increase in the mass (Figure 1). Histological examination of the excisional biopsy showed round cells with an oval nucleus and increased mitotic activity. The immunohistochemical stains of the tumor demonstrated positivity for desmin diffusely, and myogenin, MyoD1, and CD99 focally. S100, TdT, leukocyte common antigen (LCA), Pan-Keratin, and epithelial membrane antigen (EMA) were negative (Figure 2). Based on histopathological examination, the diagnosis of alveolar RMS was confirmed. Positron emission tomography-computed tomography (PET/CT), thorax CT and whole-body MRI were performed for staging. PET/CT revealed an avid fluorodeoxyglucose (FDG) uptake in the primary mass, the right axillary, cervical, paraaortic, and bilateral inguinal lymph nodes. Increased FDG uptake was also evident at multiple vertebrae, right ischium, left pubic ramus, bilateral distal femora, left clavicle, bilateral scapulae, and left humeral head (Figure 3A). In the abdomen, there was increased FDG uptake in a soft tissue mass at the level of the T11 vertebra, indistinct from the left adrenal gland and pancreatic tail (Figure 3C). Thorax CT revealed no lung metastasis. Whole-body MRI displayed the metastatic pancreas lesion, aforementioned musculoskeletal metastases, and additional lesions in the lumbar vertebrae and ribs (Figure 3B and D). Findings were consistent with stage 4 RMS. Radiotherapy was not recommended, chemotherapy with ifosfamide, vincristine, and actinomycin regimen was started and the patient has been still following. Written informed consent for publication was provided by the patient's legal guardian.

Although pancreas is a common metastatic site for alveolar RMS (67% in the autopsy series), this entity is not well-known.[3] In a study by Jha et al.[3] 8 patients had pancreas metastases out of 71 cases. Two metastatic lesions of pancreas were missed on CT and diagnosed with PET-CT. In that series, whole-body MRI was not performed to detect metastatic lesions. Breneman et al[6] evaluated 1116 patients with RMS, divided to metastatic (n = 127) and non-metastatic (n = 889) groups. Among them, 663 (59.4%) cases had embryonal histology and 49 (7.3%) of those had metastatic disease. Sites of metastatic involvement were only given for overall patients, with the lung being the most common site; pancreatic involvement was not seen in any subtype.[6]

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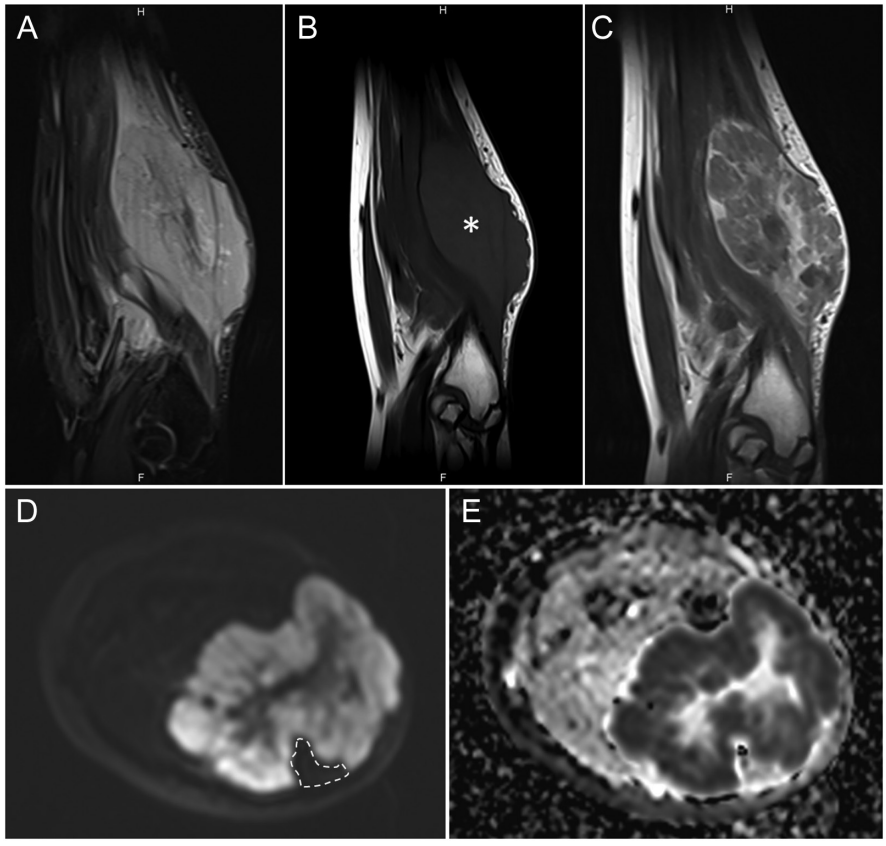

Figure 1. Right forearm magnetic resonance imaging (MRI) of an 11-year-old boy. Long axis fat suppressed T2-weighted image demonstrates that the mass has heterogeneous hyperintense signal intensity. Precontrast (B) and postcontrast (C) T1-weighted images reveal that the mass has low T1 intensity relative to the muscle and enhances heterogeneously. Axial diffusion weighted image (DWI) (D) and apparent diffusion coefficient (ADC) (E) map of the mass reveals restricted diffusion of the mass peripherally. Flexor carpi radialis muscle is partially surrounded and compressed upon by the mass centrally (D, dashed outline).

In children, metastatic disease to the pancreas is less often than primary pediatric pancreatic masses. Differentiation of pediatric pancreatic masses can be challenging due to their overlapping imaging features. Pancreatoblastoma generally occurs in younger children and is suggested by an elevated serum α-fetoprotein level, while solid pseudopapillary neoplasm of the pancreas is the most common tumor in older children, particularly girls. Neuroendocrine tumor is typically seen in the setting of von Hippel–Lindau syndrome or tuberous sclerosis. If a small, round, solid mass is revealed incidentally in the tail of the pancreas, the most probable diagnosis is intrapancreatic accessory spleen. Metastatic involvement of the pancreas may manifest as a single mass, multiple masses, or diffuse pancreatic enlargement.[7]

In our case, we performed whole-body MRI lasting 19 minutes, and PET/CT to find out metastatic lesions. Whole-body MRI is an emerging imaging technique with wide field-of-view imaging and high resolution to provide robust soft-tissue characterization. It is particularly advantageous in children owing to the absence of ionizing radiation which is a major concern for PET/CT regarding cumulative radiation exposure on follow-up. Especially radiation-free techniques should be preferred in patients with cancer predisposition syndromes that comprise up to 10% of pediatric cancer cases. Whole-body MRI is at least equivalent to FDG PET/CT for skeletal metastases, which are the primary concern in many small round cell tumors.[8] In the study of Siegel et al.[9] whole-body MRI for staging distant metastases in 66 pediatric cancer patients was not less successful than conventional imaging techniques (CT, scintigraphy studies, and/or PET). In our patient, whole-body MRI revealed more metastatic bone lesions than PET/CT, better localized the pancreatic mass and demonstrated scalp lesions more conspicuously.

On the other hand, several diagnostic limitations of whole-body MRI are worthy of mention. Magnetic resonance imaging evaluation is primarily anatomic, lacks specificity for certain types of findings, such as post-treatment changes, necrosis, and scar tissue, which can lead to diagnostic uncertainty, especially in lymphoma. Diffusion weighted image can provide functional data, however reliable apparent diffusion coefficient thresholds for distinguishing benign from malignant tissue have yet to be widely validated.[8] Although good concordance ($\kappa = 0.91$-0.94) was reported between whole-body MRI and PET/CT for initial staging in pediatric lymphoma by Littoij et al.[10] pure anatomic staging of MRI is insufficient to supersede the PET for functional evaluation by comparing SUV values.[8]

In conclusion, unusual metastatic lesions can be seen in RMS even in the absence of metastases to the more frequent sites. We suggest whole-body MRI especially in the pediatric population for staging regarding concerns for ionizing radiation and ability to detect metastases as well as PET-CT.

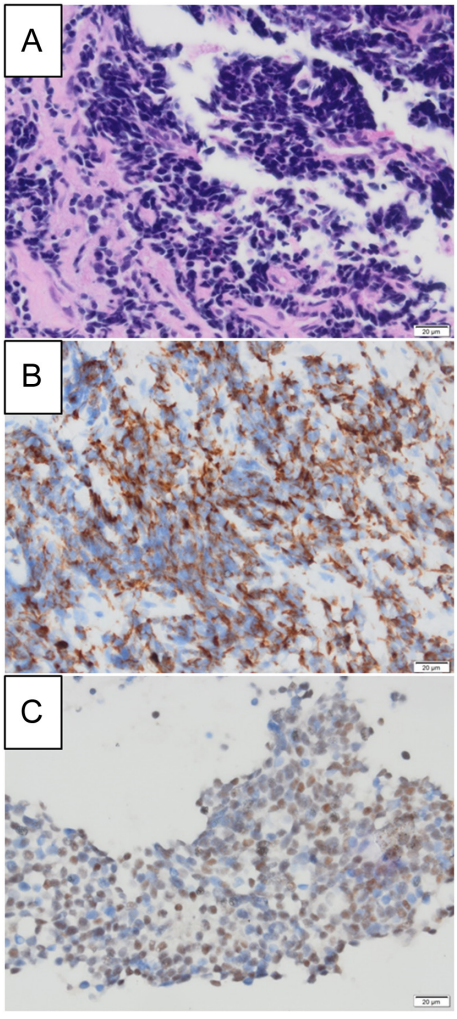

Figure 2. (A) Tissue array section composed predominantly of primitive round to ovoid cells with marked mitotic activity (H&E stain; 460×) (B) Desmin immupositivity is seen with immunoperoxidase stain. 460×. (C) Strong nuclear staining with the myogenin antibody is revealed. 460×.

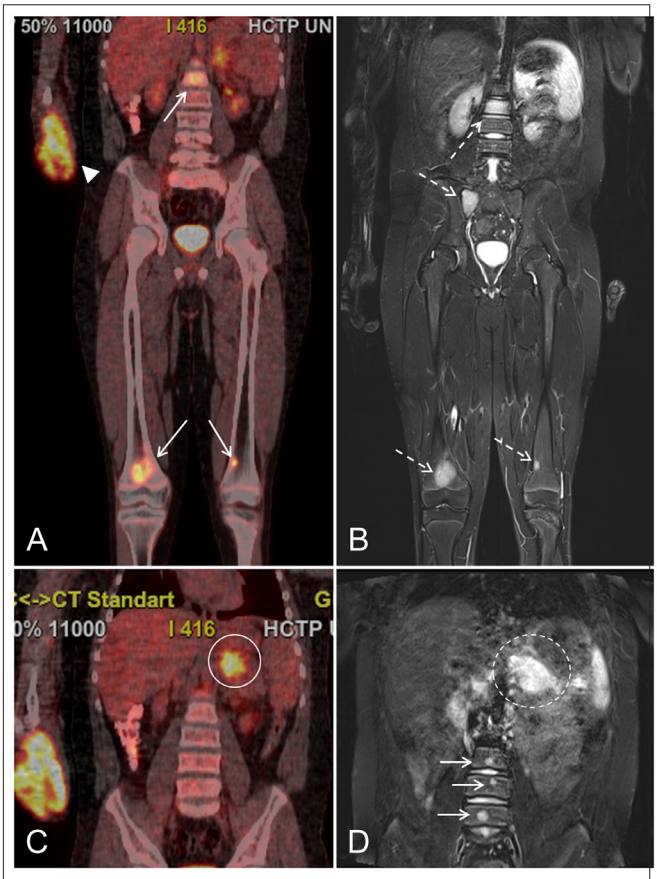

Figure 3. (A) Coronal plane positron emission tomography/computed tomography (PET/CT) demonstrates primary mass (arrowhead) and multiple bone metastases (arrows) with avid fluorodeoxyglucose (FDG) uptake. (B) Equivalent coronal plane fat-suppressed T2-weighted magnetic resonance imaging (MRI) reveals multiple bone metastases (dashed arrows) as well as PET/CT. (C) Another section from the coronal plane PET/CT shows the metastatic mass at the left upper quadrant with intense FDG uptake (white circle). (D) A whole-body MRI from the similar coronal section reveals the T2 hyperintense mass as well (dashed circle). Moreover, there are round nodular metastatic lesions (arrows) in the body of L3, L4, and L5 vertebra which are not identified on PET/CT.

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