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Case Report

Gastric Ewing Sarcoma identified on a Meckel's scan

Lavi Nissim, MD, Gerald Mandell, MD*

Department of Radiology, Phoenix Children's Hospital, 1919 E Thomas Rd. Phoenix, AZ 85016 USA

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ABSTRACT

Ewing Sarcoma is a malignant small round blue cell tumor most commonly found in bones and soft tissues of the axial skeleton and extremities. The Ewing family of tumors, including peripheral neuroectodermal tumor, represent the second most common malignancy in the pediatric population and second most common primary bone tumor after osteosarcoma. In a majority of Ewing Sarcoma cases, there is a translocation between chromosomes 11 and 22. Extraskeletal Ewing Sarcoma of the stomach is exceptionally rare, with only a handful of case reports. Here we report a case of primary Ewing Sarcoma of the stomach found initially as a filling defect in the stomach on technetium-99m pertechnetate scintigraphy to evaluate for gastrointestinal bleeding.

Case Report

An otherwise healthy 17-year-old female presented to the Emergency Department with worsening abdominal pain and fatigue for 1 month. Upon further review, she reported melena of uncertain duration. Abdominal radiograph showed moderate stool burden, but was otherwise unremarkable. On presentation, the patient was noted to be anemic with serum hemoglobin of 5.2. Her stool was positive for occult blood, but she did not have bright red blood per rectum. The patient was administered 2 units of packed red blood cells with appropriate correction. A nuclear medicine technetium-99m pertechnetate scan ("Meckel's scan") was performed to assess for a Meckel's diverticulum (Fig. 1). Based on the findings on the scintigraphy,a contrast-enhanced computed tomography (CT) of the abdomen and pelvis was ordered, revealing a large mass (Fig. 2). The patient then underwent upper endoscopy with tissue sampling (Fig. 3). Biopsy showed malignant infiltrate in the lamina propria with staining characteristics of Ewing Sarcoma. Genetic testing showed the tissue to be positive for abnormality of the EWSR1 (22q12) locus, confirming diagnosis.

Discussion

Primitive neuroectodermal tumors (PNET)are high-grade small round blue cell tumors of neuroectodermal origin [1]. This family of tumors includes Ewing Sarcoma, Askin tumor (thoracopulmonary PNET) and extraskeletal PNET(also known as Extraskeletal Ewing Sarcoma or EES) [2,3]. These tumors are found to have a translocation between chromosomes 11 and

* Corresponding author.
E-mail address: lnissim@phoenixchildrens.com (L. Nissim).
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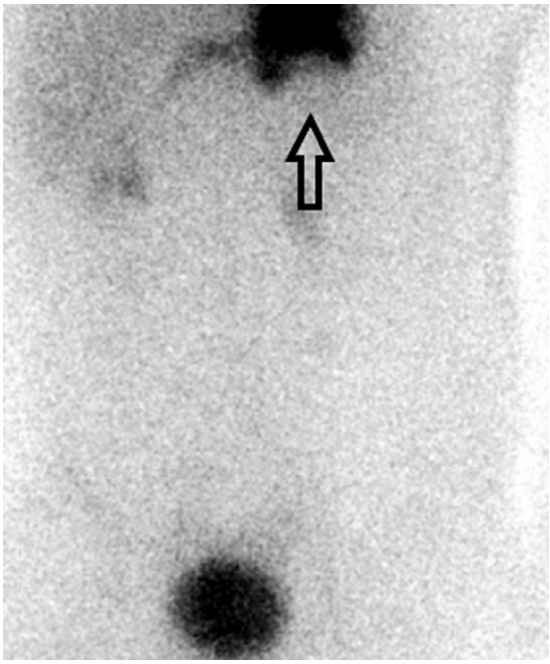

Fig. 1 – Anterior 1 hour delayed image of the abdomen under gamma camera after intravenous administration of 4.9 mCi of Technitium-99m pertechnetate. A filling defect is identified within the stomach body along the greater curvature (arrow). No ectopic gastric mucosa identified.

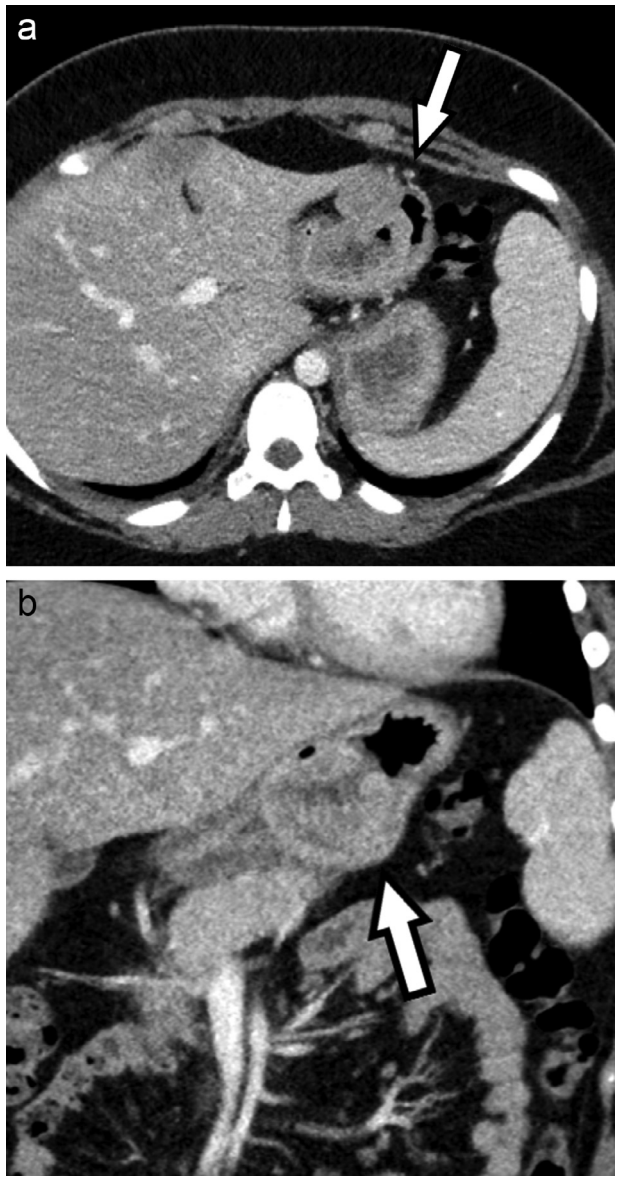

Fig. 2 – (a) Axial contrast-enhanced CT image shows a large ulcerated mass within the body of the stomach, measuring 6.7 x 5.2 x 4.6 cm (arrow).(b) Coronal reconstructed CT image demonstrating the large ulcerated mass within the stomach body (arrow), at the site of abnormality seen on scintigraphy.

22 involving the Ewing Sarcoma breakpoint gene 1 (ESWR1), resulting in abnormal interaction with regulators of cell division [3–6]. Neoplasms in this family are thought to arise from the same precursor cell [1,5]. The majority of PNET tumors arise in the skeleton, with approximately 400 patients presenting annually in the United States, though any part of the body may be affected [5,6]. They tend to occur in childhood or young adulthood and there is slight male predominance [2,7]. Skeletal Ewing Sarcoma may be distinguished by specific imaging features in children or young adults such as cortical destruction at the metadiaphysis of long bones [3]. Treatment of PNET tumors typically involves combined modality, with neoadjuvant chemotherapy followed by surgery and/or radiation therapy [3,6].

EES account for only 15%-20% of the family of tumors and may occur in a slightly older patient population, with average age of 20 years, and may not have male predominance [2]. Of the EES, the most prevalent is Askin tumor (thoracopulmonary) [5]. Other locations include the small bowel, esophagus, pancreas, or genitourinary system [6,8]. Because of slow growth, these tumors may be insidious and present as massive lesions up to 20 cm in size, encasing other organs. Unfortunately, increased tumor volume conveys a worse prognosis. Systemic symptoms such as malaise, fever, or elevated markers of inflammation may occur in metastatic disease [5].

Neuroectodermal tumors of the gastrointestinal tract (GNET) are exceptionally rare, with less than 100 described in the literature; these were recently redesignated from previous classification of clear cell sarcoma-like tumor of the gastrointestinal tract (GI) [4,7]. Typical symptoms on presentation include abdominal pain, weight loss, or intestinal obstruction [2,7]. Pathologists unaware of the diagnosis may errantly misdiagnose the entity as the more common GI stromal tumor or malignant nerve sheath tumor [7,9]. The tumors are avid on fluorine-18 fluoro-deoxyglucose positron emission tomography/CT and typically show an enhancing mass with possible central necrosis on CT or magnetic resonance imaging 3. In one study of 19 patients with GNET, most common site of origin was the small intestines (57.9%) followed by stomach (15.8%), colon (10.5%), ileocecal junction (5.3%), esophagus (5.3%), and anal canal (5.3%); of 15 patients followed at 29.7

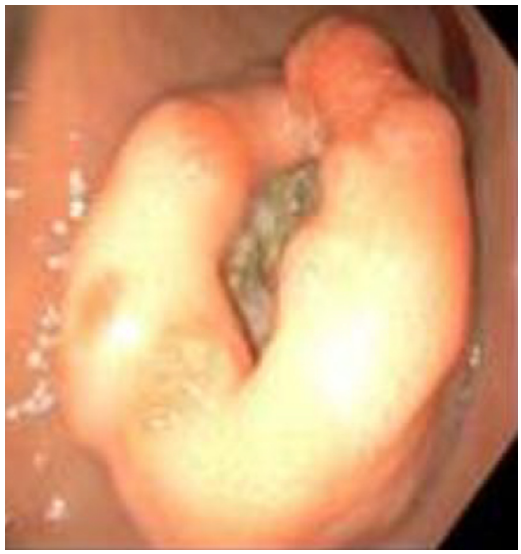

Fig. 3 – Endoscopy shows large ulcerating mass in the stomach body.

months, 8 had no evidence of disease, 5 patients were alive with disease, and 2 succumbed to their disease [4].

Differential considerations for mass or mass-like lesion associated with lower GI bleeding in a pediatric patient include GI stromal tumor, metastatic implants to the GI tract, Meckel's diverticulum, and intussusception [7]. EES tumors in the GI tract may present with vague abdominal symptoms of chronic standing leading to chronic anemia, but acute-onset bleeding with painful abdomen is possible [5]. Elevation of serum markers such as CA-125 may be minimal [8]. GI tract perforation as a result of EES has been reported in 2 cases [1].

In summary, this case represents a unique presentation of rare GNET identified initially on nuclear medicine imaging to rule-out a Meckel's diverticulum. The findings presented in this case should alert the clinician to the possibility of a gastric tumor incidentally identified on the pertechnetate scintigraphy study, as well as the possibility of a neuroectodermal tumor as etiology of GI tract mass with occult bleeding, albeit rare.

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