A 7-year-old boy presented with a history of intermittent abdominal pain and vomiting for 1 month. Findings of the abdominal examination were unremarkable. A computed tomography scan revealed a 6.9 × 7.3-cm heterogeneous mass in the right side of pelvis extending from below the level of aortic bifurcation and abutting the superior surface of the urinary bladder. A positron emission tomography scan showed a positron emission tomography-avid mass in the right side of pelvis without any other uptake elsewhere (Figure 1, A; available at www.jpeds.com). A core biopsy revealed histopathologic features of spindle cell/sclerosing rhabdomyosarcoma, which on immunohistochemical analysis strongly expressed desmin and was negative for myogenin and smooth muscle actin. MyoD1 showed an equivocal staining pattern (Figure 2; available at www.jpeds.com). The patient received 5 weeks of chemotherapy based on the Intergroup Rhabdomyosarcoma Study IV protocol for rhabdomyosarcoma, following which a computed tomography scan was performed. The mass remained unchanged in size; however, it was now located on the left side of the pelvis, pushing the dome of the urinary bladder (Figure 1, B). On exploration, the tumor was found to be arising from the Meckel diverticulum (Figure 3). The tumor was excised along with the adjacent small intestine, and an ileoileal anastomosis was performed. Findings of the histopathologic examination revealed prominent chemotherapy-induced changes, in the form of hyalinization and chronic inflammation, with scanty residual tumor. The resection margins were negative for the presence of tumor. The patient received maintenance chemotherapy based on the Intergroup Rhabdomyosarcoma Study IV protocol and whole abdomen radiotherapy (41.4 Gy). The patient is disease free at 8 months of follow-up.

Rhabdomyosarcoma is the most common soft-tissue malignancy in children. Intra-abdominal rhabdomyosarcoma account for 10%-12% of all rhabdomyosarcomas and arise from the intestinal tract, the retroperitoneum, and pelvic sites excluding the genitourinary organs. The Meckel diverticulum is a remnant of the omphalomesenteric duct and is the most common congenital anomaly of the gastrointestinal tract, with an estimated prevalence of 2%. Bleeding and obstruction are the most common complications associated with the Meckel diverticulum, whereas malignancies account for only 0.5%-3.2% of the complications. The Surveillance, Epidemiology, and End Results database review between 1973 and 2006 identified 163 cases of Meckel diverticulum cancers. The mean age at diagnosis was 60.6 years, and no cases were recorded in patients younger than 10 years of age in this review. The most common cancers associated with the Meckel diverticulum include carcinoids, adenocarcinoma, stromal and smooth muscle tumors (gastrointestinal stromal tumor/leiomyosarcoma), sarcomas, and lymphoma. We previously have reported desmoplastic small round cell tumor of the Meckel diverticulum in a 6-year-old boy. A thorough review of indexed medical literature did not reveal any cases of rhabdomyosarcoma involving the Meckel diverticulum or their presentation as a wandering pelvic mass.

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References


Figure 1. A, Maximum intensity projection of positron emission tomography scan performed for staging shows a mass (long arrow) with heterogeneous fluorodeoxyglucose uptake in the right side of the pelvic cavity. B, A coronal reformatted image of the computed tomography scan done for response assessment shows the residual mass (short arrow) in the left side of the pelvic cavity.

Figure 2. A, Cellular tumor composed of cells arranged in cords, clusters, and singly in a hyalinized/sclerotic stroma. Hematoxylin and eosin ×200. B, Greater magnification showing oval- to spindle-shaped cells arranged in cords and clusters in a sclerotic stroma. Hematoxylin and eosin ×400. C, Diffuse, strong desmin positivity in tumor cells. Diaminobenzidine ×400.