History
16 month old male with irritability.

Diagnosis
Presacral Neuroblastoma

Additional Clinical
Initial US to evaluate for intussusception revealed a pelvic mass.

Discussion
Neuroblastoma originates from neural crest cells along the sympathetic chain, as well as in the adrenal medulla. Primordial neural crest cells may undergo differentiation into mature ganglion or Schwann cells or may remain undifferentiated and immature neuroblastic cells. Neuroblastoma accounts for about 10% of all pediatric cancers and is the most common malignancy in the 1st year of life. Neuroblastoma is, overall, the fourth most common malignancy in childhood, after leukemia, lymphoma, and central nervous system tumors. The median age at diagnosis is 22 months; nearly all are diagnosed by the end of the first decade. Approximately 70% of neuroblastomas and ganglioneuroblastomas are found in the abdomen (adrenal gland and retroperitoneal area), 20% in the posterior mediastinum, 5% in the neck region, and only 2%–3% in the pelvis.

The clinical manifestations of neuroblastoma are nonspecific and may include constitutional symptoms such as fever, weight loss, malaise, and failure to thrive. Symptoms secondary to mass effect also are common and may include pain, urinary retention or frequency of evacuation, and constipation. Other symptoms are related to the effects of hormonal secretion, such as diarrhea due to vasoactive intestinal peptides, or opsomyoclonus, a nonmetastatic paraneoplastic condition. In patients with a presacral tumor, the pelvic mass also may be detected during physical examination.

Radiographic findings are nonspecific and include calcification, which has been observed in 30% of cases. Presacral tumors may extend into the spinal space through the neural foramina and cause erosion and remodeling of bone. In addition, indirect indicators of the presence of a presacral tumor, such as displacement of the bowel or urinary bladder, can be seen radiographically. Neuroblastomas may include areas of hemorrhage or necrosis, but they do not contain fat. On sonography, neuroblastomas have a heterogeneous appearance secondary to hemorrhage, necrosis and calcification. Doppler US may help determine the relation of the mass to regional vessels. CT demonstrates the location of the tumor, its boundaries, and any extension, including involvement of neural foramina. Calcifications are seen in more than 80% of neuroblastomas at CT. Neuroblastoma has heterogeneous signal intensity at MR imaging, with low signal intensity and variable contrast enhancement on T1-weighted images and with high signal intensity on T2-weighted images again related to hemorrhage, necrosis and calcification. MR has the additional benefit of detecting marrow and intraspinal involvement.

Findings
Fusion US-Utilizing navigation technology, the CT image is reconstructed from the previously obtained CT DICOM data set in the same plane as the real time ultrasound image. A large heterogeneous presacral soft tissue mass with intralesional calcification is present. MR-Multilobulated presacral mass demonstrating isointensity on T1 and hyperintensity on T2 and postgadolinium sequences and intraspinal extension. NM-Anterior and posterior 24 hour delayed I-123 MIBG images demonstrate marked avidity of the pelvic mass.

Reference
24 HR I-123 MIBG SCAN
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