26-MONTH-OLD GIRL was evaluated for stunted growth (weight, height, and head circumference less than the 5th percentile for age) and a 2-month history of altered bowel patterns with perianal pain. The decelerating growth pattern was first noted between the ages of 6 and 12 months, when her weight and length for age-growth percentiles fell from the 25th and 50th, to 5th and 25th, respectively. They later fell to less than the 5th percentile at the time of consultation. The 3-day diet record obtained at consultation revealed an average caloric intake of 121 kcal/kg per day (protein, 12%; carbohydrate, 57%; fat, 31%). Loose stools alternating with constipation characterized the altered bowel pattern. There was no history of mucus in stools, melena, or hematochezia. The perianal pain was most prominent when she sat in her car seat and immediately prior to bowel movements. She was born after a full-term pregnancy and had normal gross motor milestones, fine motor skills, and language development. Apart from small stature and minimal subcutaneous fat tissue, the rest of her physical examination results were within normal ranges. There was no anal fissure or other external perianal lesion. Laboratory tests included a normal hemogram, hepatic panel, sweat chloride, serum vitamin E level, and antigliadin/antiendomysial antibodies negative for celiac disease. Upper gastrointestinal endoscopy showed normal small-bowel mucosa and histology. The digital rectal examination performed prior to flexible sigmoidoscopy (normal) revealed a firm presacral mass that was further delineated by axial computed tomography (Figure 1). The serum α-fetoprotein level subsequently obtained was 1980 ng/mL (normal range, 0.6-11.1 ng). Histologic sections from the tumor mass resected at laparotomy are seen in Figure 2.
Denouement and Discussion

Type IV Sacrococcygeal Teratoma With Yolk Sac (Endodermal Sinus) Tumor Presenting With Failure to Thrive and Perianal Pain

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acrococcygeal teratomas account for 40% of all germ cell tumors and up to 78% of extragonadal germ cell tumors.1 The incidence of germ cell tumors in children in the United States is 2.4 cases per 1 million children, with a biphasic age distribution. The first peak occurs at 2 years of age and a second peak at 15 to 20 years of age.3 There are 3 distinct clinical presentations of germ cell tumors: (1) tumors of the adolescent testis and ovary; (2) extragonadal germ cell tumors of older children; and (3) tumors of infants and young children. The origin of extragonadal germ cell tumors is presumed to be either aberrantly migrated germ cells, or alternatively, totipotent embryonic cells. The most common locations for extragonadal germ cell tumors are sacrococcygeal, mediastinal (including the pericardium, heart, and lung), intracranial, and retroperitoneal, respectively.

The clinical presentation of sacrococcygeal tumors is classified into 4 types. Type 1 tumors (47%) are predominantly externalized, with limited extension into the pelvic region; type 2 (34%) have similar external components and intrapelvic extension; type 3 (9%) have a minimal external component, with significant pelvic and intra-abdominal extension; and type 4 (10%) tumors are internalized presacral tumors with no external evidence of disease.3 The single most important histologic distinction to be made with sacrococcygeal germ cell tumors is whether or not they also contain yolk sac tumor or embryonal carcinoma, which are the malignant components. The incidence of malignancy is highest in type 4 tumors.5 Benign sacrococcygeal lesions are commonly seen in newborns, whereas malignant tumors most commonly present in children older than 1 year of age.

The histologic features of yolk sac (endodermal sinus) tumors are characterized by several patterns with varying cytology, making histological identification sometimes difficult. The majority of yolk sac tumors show more than 1 histological type, though none are believed to have specific prognostic significance. Generally, however, yolk sac tumors are characterized by primitive tubular or “microscopic” structures lined by somewhat flattened primitive cuboidal epithelial cells. One of the most common histological variants is the “papillary type,” in which the characteristic Schiller-Dural bodies are seen, as in the present case. Periodic acid–Schiff (PAS)–positive hyaline droplets can be frequently recognized in the cytoplasm of many of these cells. Immunohistochemically, these tumors characteristically stain for α-fetoprotein. Yolk sac tumor is the most common malignant germ cell tumor in prepubertal children and may be histologically differentiated from embryonal carcinoma on the basis of virtual nonexistence of the latter in young children. Yolk sac tumor is commonly, but not always, associated with elevated serum α-fetoprotein levels.

Preoperative diagnosis may be accomplished by one or more imaging modality techniques. Prenatal ultrasound can identify the tumor when it is sufficiently large. Postnatal computed tomography and/or magnetic resonance imaging can show the mass and its location. Identification of fluid, tissue fat, and calcium densities within the mass is characteristic of teratoma, but it is not determinate for malignant potential.

Treatment is complete surgical resection, including excision of the coccyx. In the absence of malignancy, the cure rate is 95%.1,2 Presence of malignancy necessitates addition of chemotherapy with platinum-containing regimens, and this is associated with inferior prognosis. Level 1 or 2 fetoprotein may be used to monitor for recurrence and metastases. It is necessary to differentiate high serum α-fetoprotein levels from the normal infancy-related elevation,3 spurious increase secondary to chemotherapy-induced tumor lysis,4 hepatic disorders, cholestasis secondary to anesthesia, and medications such as phenytoin or methotrexate.1

Impaired growth and malnutrition are not uniquely related to malignancy. Nonetheless, disturbances related to tumor cell turnover and by-products may contribute to altered nutrient metabolism and energy imbalance in vulnerable hosts.2,3 Other than malignancy, there was no other satisfactory clinical explanation for the poor growth pattern observed in the subject of this report. The perianal pain and altered bowel patterns were presumed to be secondary to pressure effects from the tumor.

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REFERENCES