A 64-YEAR-OLD MAN WAS REFERRED TO OUR institution after he was found to have a retrorectal mass on a computed tomographic scan he underwent for persistent rectal pain and discomfort. Family and medical history were unremarkable. No laboratory abnormalities or specific findings were noted on physical examination. At digital rectal examination, the mass was not palpable and the rectal mucosa was intact. The computed tomographic scan showed a 40-mm mass in the ischiorectal fossa adherent to the levator ani muscle and rectum, and while it was suspicious for invasive rectal carcinoma, the exophytic appearance was suggestive of a soft tissue neoplasm. The patient also underwent magnetic resonance imaging, which revealed an enhancing, 35-mm soft tissue mass in the right ischiorectal fossa adherent to the wall of the rectum with questionable involvement, extending through the right levator ani muscle, suspicious for a rectal carcinoma vs a soft tissue mass (Figure 1). Transrectal ultrasonography was performed and a 30-mm solid hypoechoic mass with a 4-mm calcification in the left lateral perirectal space 9 to 11 cm from the anal verge was noted without signs of rectal wall invasion. No lesions suspicious for locoregional invasion were identified. Fine-needle (22-gauge) aspiration was performed and the cytological analysis revealed the presence of neoplastic elements positive for AE1/AE3 and synaptophysin but not for chromogranin and p63.

Based on the radiological and cytological findings, which did not rule out a potential malignancy, the patient underwent surgical excision of the mass. An arcuate incision was performed, extending from the posterior midline of the anal margin, with preservation of the sphincter complex to the right of the coccyx. The coccyx was excised to achieve adequate exposure, the anococcygeal ligament was divided, and the retrorectal space was entered. A 5-cm calcified mass involving the posterior aspect of the retrorectal space adjacent to the tip of the coccyx extending toward the right was identified and excised to normal healthy tissue, confirmed by multiple frozen sections. At gross examination, the specimen consisted of a well-circumscribed mass of rubbery consistency with a cystic component and a 1-cm calcified area (Figure 2).

The postoperative course was uneventful and the patient was discharged on postoperative day 1.

What Is the Diagnosis?

A. Metastatic mesorectal lymph node
B. Neuroendocrine carcinoma of a tailgut cyst
C. Dermoid cyst
D. Chordoma

Figure 1. Magnetic resonance imaging with gadolinium contrast showing a well-circumscribed 2.5 × 3.5 × 4-cm soft tissue mass with contrast enhancement in the right ischiorectal fossa, containing a 10-mm calcification (arrow), apparently in continuity with the rectal wall (arrowheads). The mass seems to invade the right levator muscle, with deformity/hypoplasia of the sacrum and coccyx.

Figure 2. Gross appearance of the specimen, with a dusky pink to gray-tan glistening mass with focal gray-white fibrotic areas. A cystic component (*) and a calcified area (arrow) are clearly visible.