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Case Report

Neuroendocrine tumor arising from tailgut cyst with spinal cord tethering: case report and literature review

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Abstract

BACKGROUND CONTEXT: Neuroendocrine tumors (NETs) from tailgut cysts are rare; only 15 cases have been reported until now. A tailgut cyst with spinal cord tethering has not been previously reported, although both diseases are congenital anomalies in the early stage of gestation.

PURPOSE: To report a rare case of NET from tailgut cyst associated with spinal cord tethering and review the literature.

STUDY DESIGN: Case report and literature review.

METHODS: We describe the clinical course of a 53-year-old man, who presented with gluteal pain and bladder dysfunction. Magnetic resonance images showed that a tumor of the sacral spinal canal extended into the retrorectal space and connected to a thickened fatty filum terminale, which was tethering the spinal cord.

RESULTS: Because of tumor malignancy on a computed tomography-guided biopsy and the imaging data of involvement of presacral lymph nodes, we performed total removal of the tumor. Pathologic examination revealed NET (Grade 2) arising from a tailgut cyst. The patient received somatostatin analog therapy after surgery, followed by local radiation because of the further enlargement of the lymph nodes. Later, we started everolimus therapy for the metastases to the retroperitoneal lymph nodes. He presented with no local recurrence or further disease progression at 28 months after surgery. The review indicated that tumors in Grade 2 or 3 showed progressive clinical course after surgery and three of seven patients with biopsy were misdiagnosed.

CONCLUSIONS: The correct preoperative diagnosis of NETs from tailgut cysts is difficult, but extremely important because Grade 2 or 3 tumors show disease progression even after surgery. Presacral congenital tumors, such as tailgut cysts, have the potential of malignant transformation into neuroendcrine tumors or adenocarcinomas. Comorbidity of spinal cord tethering and tailgut cyst suggests some relationship to common developmental errors in embryogenesis.

Keywords: Embryologic failure; Malignant transformation; Neuroendocrine tumor; Somatostatine analogue; Spinal cord tethering; Tailgut cyst

Introduction

A tailgut cyst is a rare benign developmental lesion from the remnants of an embryonic hindgut [1]. It rarely undergoes malignant transformation, such as neuroendocrine tumor (NET), carcinoma, and adenocarcinoma [2]. Neuroendcrine tumors, defined as epithelial neoplasms with predominant neuroendocrine differentiation, are relatively rare

FDA device/drug status: Approved (Octreotid, Everolimus).

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tumors arising from most organs of the body [3]. Most of these tumors are more indolent than other epithelial malignancies; however, a subset of them behave aggressively and are resistant to treatment [4]. Neuroendcrine tumors from tailgut cysts are very rare and only 15 cases have been reported with clinical details in the medical literature [5–19]. We describe here a new case and summarize the clinicopathologic features of NETs from tailgut cysts with a literature review. In addition, a tailgut cyst with spinal cord tethering, to the best of our knowledge, has not been previously reported. We discuss this comorbidity in the context of embryogenesis.

Case presentation

History and examination

A 53-year-old man presented with 1 year history of left gluteal pain, impaired defecation urge, difficulty in starting urination, and sensory disturbance in the perianal and gluteal areas. The gluteal pain was related to prolonged upright position. He showed weakness of anal sphincter muscles and Valsalva voiding; however, he was not incontinent for urine and feces. No skin lesion was detected on his back and buttocks. He realized to have coccygeal deviation since childhood and previous lumbar radiographs had shown erosion of his coccyx (Fig. 1). He had no associated anomalies in the central nervous system and other organs. In addition, he had no family history of presacral tumors and did not present with sacral agenesis or anorectal anomalies. Magnetic resonance images revealed an extradural tumor in the sacral spinal canal (Fig. 2, A–E). It was mostly hypointense both on T1- and T2-weighted images and moderately enhanced with gadolinium. The tumor extended anteriocaudally to the retrorectal space with some cystic components and rostrally to the caudal end of the thecal sac. It connected to a thickened fatty filum terminale, which was tethering the spinal cord and showing partially high intensity both on T1- and T2-weighted images. Computed tomography (CT) scans showed enlargement of the sacral spinal canal with scalloped vertebral bodies. Positron emission tomography revealed mild accumulation of the radioisotope in the sacral tumor (standardized uptake value: 2.11–2.35) without any abnormal accumulation at other sites, and whole-body CT scans with contrast-enhancement detected no distal metastasis. Serum levels of key tumor markers were within normal limits. Bone erosion and tethered cord suggested a congenital malformation, whereas progressive neurologic symptoms and the mild tracer accumulation in positron emission tomography were features suggestive of malignancy. It was difficult to narrow the differential diagnosis of this tumor only with neuroradiologic findings. We performed a CT-guided biopsy to obtain a pathologic diagnosis, which was necessary to formulate a surgical strategy.

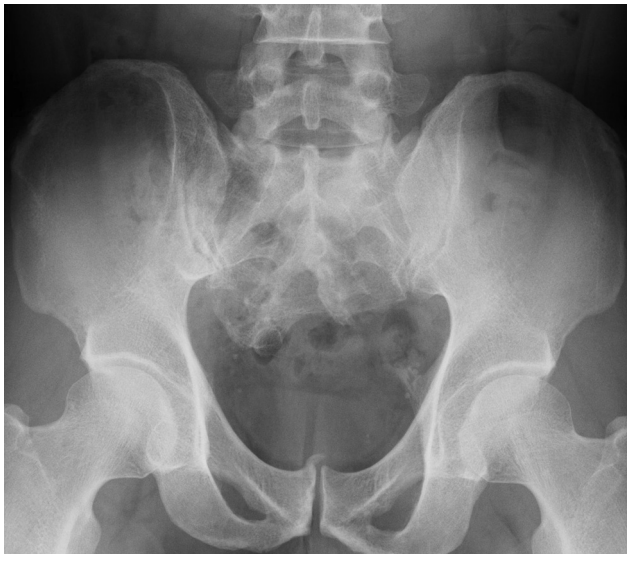

Fig. 1. Pelvic X-ray showing erosion of the coccyx.

Biopsy

Hematoxylin-eosin stained section of the biopsy specimen showed dense proliferation of round cells. The proliferation rate, as highlightened by the Ki-67 immunohistochemical stain, was 15%. The tissue was positive for vimentin and slightly positive for pan-cytokeratin. It was negative for epithelial membrane antigen, S-100 protein, and CD99 antigen. The biopsy did not lead to a definitive diagnosis, although some tumors such as schwannoma, chordoma, ependymoma, and Ewing sarcoma were excluded. Rhabdoid tumor remained as a possible pathologic diagnosis at biopsy; however, it was less likely because it usually develops in infants and young children. Tumor removal was necessary for a final pathologic diagnosis that will determine the treatment strategy. In view of the high Ki-67 index, a retrospective review of all neuroimagings revealed an enlargement of the presacral lymph nodes because of regional metastases (Fig. 2E). We planned maximum tumor removal because of its increased proliferative ability and metastatic potential to the regional lymph nodes.

Operation

We performed osteoplastic sacral laminectomy. The tumor occupied most of the epidural space and covered the caudal portion of the thecal sac. The extradural tumor invaded into the intradural space and continued to the thickened fatty filum terminale at its rostral end (Fig. 3, Left). At first, we divided the filum terminale to untether the spinal cord. Next, we dissected the intradural tumor from adjacent nerve roots and totally removed it. Then, we removed most of the extradural tumor with detachment of the extradural