



Teratoma That Making Aganglionic Colon Loop in an Infant: A Case Report

Süt Çocuğunda Aganglionic Kolon Anısı Yapan Teratom: Olgu Sunumu

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ÖZ

Over ve sakrokoksigeal bölgede nadiren görülen matür teratom literatür gözden geçirilerek sunuldu. Reteoperitoneal teratomlar çocuklarda ender olarak görülür. Teratomlar, ortaya çıktıkları organa veya bölgeye yabancı birçok dokudan oluşur. Barsakların organoid gelişimini gösteren teratomlar çok nadirdir. Retroperitoneal bölgede aganglionic kolonik loplara sahip teratoması olan üç aylık hastanın makroskopik ve histopatolojik bulgularını tanımladık.

Anahtar Kelimeler: Teratoma, Kolon parçası, Retroperitoneal neoplazmlar, Kız, Tanı

ABSTRACT

A case of an unusual occurrence of mature teratoma in ovary and sacrococcygeal region is reported with a review of the literature. Retroperitoneal teratomas are rare in children. Teratomas are composed of multiple tissues foreign to the organ or site in which they arise. Teratomas showing organoid development of intestine are extremely rare. We described the macroscopic and histopathology findings of an unusual case of teratoma with aganglionic colonic loops in a three months old child in retroperitoneal region.

Keywords: Teratoma, colon segments, Retroperitoneal Neoplasms, Female, Diagnosis.

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Introduction

The overall incidence of germ cell tumors has been estimated at about 0.9/100,000 population and of these, teratoma has been reported as the leading neonatal and infant neoplasm (1). Teratoma is a tumor composed of parenchymal cell types representative of more than one germ layer, usually all three (2). Extragonadal teratomas arise from totipotent cells in sequestered primitive cell rests. Only a few cases of rectal teratoma have previously been reported in the English literature (3). Histologically they show varying proportions of elements derived from ectoderm, mesoderm and endoderm, including neural tissue, teeth, cartilage, respiratory and gastrointestinal tract epithelium etc(4).

However, it is extremely rare to see complete segments of gastrointestinal tract including its total intestinal walls. We report a hitherto undescribed case of a mature teratoma harboring aganglionic loop of colon in retroperitoneal location.

Case report

A three months old female child presented with chronic constipation, abdominal swelling and restlessness since birth. On examination, it was a left abdominal mass measuring 20 cm in diameter was found. No congenital anomaly was detected. The laboratory findings, including haematological, biochemical and urine examinations, were within normal limits. Serum alpha-fetoproteins, measured by radioimmunometric assay, was 10 ng/ml (normal < 15 ng/ml). Ultrasonography of the abdomen revealed a round hypoechoic lesion measuring 20x15 cm in the retroperitoneum, located just below and close to the left kidney, along with moderate hydronephrosis of the left kidney. Contrast-enhanced computed tomography (CT) imaging showed a large cystic mass measuring 20x15x15 cm in close approximation and anterior to the lower half of the left kidney, with small areas of solid components present toward the inner peripheral portion of the mass. Radiologically, the possibility of a retroperitoneal teratoma compressing the left ureter leading to hydronephrosis of the left kidney was suggested.

Operation was made with supraumbilical transverse incision and revealed retroperitoneal

area. Per operatively, the retroperitoneal mass was found to be adherent to the outer. The surgically resected specimen revealed a thin-walled globular cystic mass that measured 20x15x15cm. , with smooth, mildly transparent capsule showing engorged prominent blood vessels and colon loop on its outer surface (Figure1).



Figure 1. The mass revealed smooth, mildly transparent capsule showing engorged prominent blood vessels and colon loop on its outer surface (Arrow)

Histopathological finding: The mass was composed of mature adipose tissue, cartilage, neural tissue and blood vessels; and full-layer of colonic wall. No ganglion cell in the area of between muscular layers. Histopathological diagnosis of mature teratoma with segments of aganglionic colon was given.

We followed up 4 years. Patient has no problem, and not given chemotherapy.

Discussion

Germ cell tumors are a varied group of benign and malignant neoplasms occurring in the perinatal period. They are found in various sites, both gonadal and extragonadal, the latter in midline locations such as the sacrococcygeal area, retroperitoneum, mediastinum, neck, and intracranial region (1). Teratomas are relatively uncommon tumors, representing only 3% of childhood malignancies. They arise in the midline or paraaxial regions such as at the

sacrococcygeal area 57%, gonads 29%, mediastinum 7%, retroperitoneum 4%, and cervix 3% (5). Retroperitoneal teratomas comprise 1% to 11% of primary retroperitoneal neoplasms and are the third most common retroperitoneal tumor in children after neuroblastoma and Wilms' tumor (6). These tumors consist of maturely differentiated derivatives of all three germ cell layers, the ectodermal layer being most prominent.

In mature teratomas, skin with dermal appendages, bronchial structures with bronchial glands and cartilage, neuroglial tissue, and teeth are commonly present and regarded as evidence of organogenesis (7). Organogenesis is extremely rare to find highly organized structures. The tumors consisting of many organized tissues such as small bowel, limbs or beating heart are called fetiform teratomas (homunculus).

The clinical and histologic features of a benign cystic teratoma with histologic evidence of almost full gastrointestinal tract development are presented in adult. This case is the first report of almost complete development of the gastrointestinal tract in a benign cystic teratoma. In this teratoma, the entire gastrointestinal tract from esophagus to colon is represented histologically (8). The literature has previously described only bowel epithelium, segments of bowel, appendix, and esophagus separately(4). In 2007 Agarwal et al. describe a benign cystic teratoma in a three month old child harboring multiple colonic loops in an unusual location. This is the first case report of a teratoma showing organoid development of a part of intestine in retroperitoneal location (9).

Treatment of pediatric GCT is based on a multimodal strategy that includes surgery, chemo- and radiotherapy. In these tumors the complete surgery represents the mainstay of treatment. Tumor resection is considered complete, if it is performed as en bloc resection of the tumor including the adjacent organ of origin (10). The paraaortic and other lymph nodes were checked, and evaluated normally.

We have presented a case of a teratoma with aganglionic colon loop that, even as a rare entity, presented atypically. The presence of this entity, with both an retroperitoneal location, aganglionic colon loop and in a 3 month old boy, is unusual.

Acknowledgments and Disclosure of Interest:

None

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