

Tailgut Kisti Zemininde Gelişen Nöroendokrin Tümörün BT Bulguları

CT Imaging Characteristics of Neuroendocrine Tumor Arising From Tailgut Cyst

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ÖZ

Tailgut (postnatal gut), hintgutun en kaudal parçası olup anüsün gelecekteki distal kesimini oluşturur. Genelde embriyonik hayatın 8. haftasında involüsyona uğrar. Tailgut kisti involüsyona uğramayan kalıntılardan köken alır. Tailgut kistinden malignensi gelişimi çok nadir olup en sık adenokarsinom, nöroendokrin tümör ve sarkom gelişimi bildirilmiştir. Biz burada tailgut kisti zemininde gelişen nöroendokrin tümörün bilgisayarlı tomografi (BT) bulgularını sunmayı amaçladık. Otuzbeş yaşında endometriozis nedeniyle takipte olan kadın hasta, karın ağrısı şikayeti ile başvurdu. Hastaya ultrasonografi (US) ve kontrastlı abdominopelvik BT çekildi. BT'de presakral alanda büyüğü yaklaşık 6.5x6x6.5 cm çapta solid heterojen düzgün sınırlı multiple nodüler kiteller mevcuttu. Hastaya yapılan cerrahi operasyon sonrasında patoloji sonucu tailgut kisti zemininde gelişen nöroendokrin tümör olarak raporlandı. Tailgut kisti asemptomatiktir, en sık presakral bölgede rektum ile sakrum arasında izlenir. Kadınlarda daha sık rastlanır. Her yaş grubunda görülebilir de orta yaş grubunda (ortalama 35 yaş) daha sık tanı alır. Bizim hastamız da kadın ve 35 yaşındaydı. Tailgut kisti ile diğer presakral kistik lezyonların ayırıcı tanısı tailgut kistinin malign transformasyonu nedeniyle yapılmalıdır. Tailgut kisti nadiren malign transformasyon göstermekte olup; adenokarsinom, nöroendokrin tümör ve sarkom gelişimi bildirilmiştir. Hastamızda lezyonların eksizyonu sonucu patoloji nöroendokrin tümör olarak gelmiştir. Presakral bölgede saptanan kitelerde tailgut kisti malignite potansiyeli nedeniyle ayırıcı tanıda akılda bulundurulmalıdır.

Anahtar Kelimeler: Tailgut kisti; nöroendokrin tümör; presakral alan

ABSTRACT

The tailgut is the most caudal part of the hindgut, it forms the distal part of the anus. It normally involutes by the eighth week of gestational age. If a tailgut rest proceeds, it may give rise to a tailgut cyst. Malign transformation of the tailgut cyst is very rare. Adenocarcinoma, neuroendocrine carcinoma, and sarcoma developing within the tailgut cyst has been reported in the literature. We present a case of neuroendocrine tumor which is arisen from a tailgut cyst in a middle aged woman with its computed tomography (CT) imaging findings. A 35 year-old woman with a history of endometriosis admitted to our hospital with abdominal pain. An ultrasonography (US) examination and contrast medium enhanced tomography of the abdomen and pelvis was performed. CT showed multiple well-defined solid heterojen masses in presacral space. The largest dimensions of the masses were 6.5x6x6.5 cm. The patient underwent surgery. Pathology was reported as neuroendocrine tumor arising within tailgut cyst. It is usually detected as an asymptomatic mass in the presacral space between rectum and sacrum. Tailgut cyst is more common in female and usually presents in middle age (mean 35), but it can present at any age. Our patient was female and 35 years old. Discrimination of tailgut cyst from other presacral cysts is important because of the malignant potential of a tailgut cyst. Malign transformation is very rare. Adenocarcinoma, neuroendocrine carcinoma, and sarcoma developing within the tailgut cyst has been reported. In our case pathology was reported as neuroendocrine tumor arising within tailgut cyst. It should be kept in mind the differential diagnosis of presacral masses because of the malignant potential of a tailgut cyst.

Key words: Tailgut cyst; neuroendocrine tumor; presacral space

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Başvuru Tarihi: 16.04.2017

Kabul Tarihi: 21.11.2017

INTRODUCTION

The tailgut normally involutes by the eighth week of gestational age. If a tailgut rest proceeds, it may give rise to a tailgut cyst in the presacral space. Malignant transformation of the tailgut cyst is very rare. We report a case of neuroendocrine tumor which is arised from a tailgut cyst in a middle aged woman with computed tomography findings.

CASE REPORT

A 35 year-old woman with a history of endometriosis was admitted to our hospital for her routine control. Her physical examination and laboratory results were normal. Pelvic masses have been found on abdomen and pelvic ultrasonography. Contrast medium enhanced abdomen and pelvic CT (Aquilion 64 Model TSX-101A; Toshiba Medical Systems) was performed. CT showed multiple well-defined solid heterogenous masses in presacral space. The largest dimensions of the masses were 6.5x6x6.5 cm. There were no association between the tumor and the rectum radiologically. The patient underwent surgery. At histopathologic examination there were solid tumor in fibrous stroma. Tumor cells have uniform small and hyperchromatic nuclei with scant cytoplasm. Thin fibrous bands divided tumor cells in nests. Mitotic figures were scant. Initially neuroendocrine tumor was thought. Tumor cells were diffuse positive with chromogranin A (DAK-A3, 1/100; Dako) and synaptophysin (SY38, 1/100; Dako) and negative with inhibin α (R1, 1/100; Dako) in immunohistochemical evaluation. Ki-67 (MIB-1, 1/100; Dako) proliferation index was 2%. In resection material tumor was encapsulated and there were no colon segment. By these features tumor was diagnosed as well differentiated neuroendocrine carcinoma developed within retrorectal tail gut remnant.

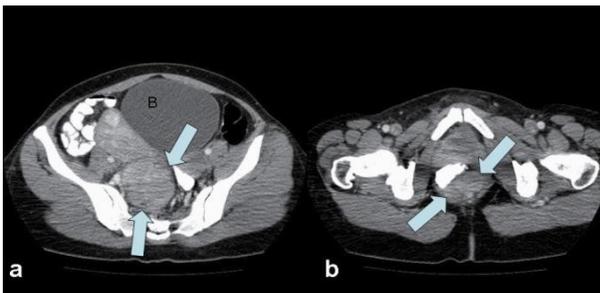


Figure 1: Axial contrast enhanced CT shows multiple well-defined solid heterojen masses in presacral space (arrows).

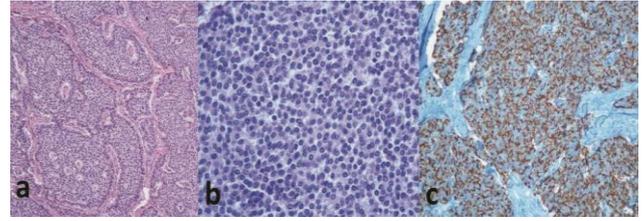


Figure 2:

- a) Tumor with nesting pattern in fibrous stroma (HEx100)
- b) Uniform, round tumor cells within conspicuous cytoplasmic borders (HEx400)
- c) Cytoplasmic positivity with Chromogranin A (DAK-A3x200)

DISCUSSION

The tailgut or postanal gut is the most caudal part of the hindgut, in the future it forms distal region of anus. It normally involutes by the eighth week of gestational age (1). If a tailgut rest proceeds, it may give rise to a tailgut cyst in the retrorectal or presacral space (1). Retrorectal cystic hamartoma and retrorectal cyst are also used for describing the tailgut cyst (2). It is more common in female and usually presents in middle age, but it can appear at any period (3). Our patient's age was 35. It is usually detected as an asymptomatic mass. When symptomatic, it presents with rectal pain or fullness, low back pain, or recurrent retrorectal abscesses and fistulas (4). Malignant transformation of the tailgut cyst is very rare. It has been reported that adenocarcinoma, neuroendocrine carcinoma, and sarcoma with in the tailgut cyst in the literature (4). Tailgut cyst is a multiloculated cystic mass with a thin wall and bright lining and is filled with a mucoid material (1,3). It's usually measured several centimeters in diameter. Infection or inflammation may cause fibrosis of the cyst wall and break down of the cyst lining (1). CT shows a discrete, well-marginated, presacral mass changes from water to soft-tissue density, depending on the contents of the cyst (3). CT can give important details concerning lesion location, size and shape; the presence and thickness of a wall; the presence of septa, calcifications or fat; and involvement of contiguous structures (5). Calcifications may be seen in the cyst wall. If concurrent infection or malignant transformation occurs, CT can shows loss of discrete margins and involvement of adjacent structures (2). In our case there were discrete margins despite malignant transformation and adjacent structures were normal. If the cyst is infected, it is often misdiagnosed as pilonidal cyst,

anorectal fistula or recurrent retrorectal abscess (6). Many differential diagnoses should be considered when a presacral cystic mass is determined, including epidermoid cyst, dermoid cyst, rectal duplication cyst, anal gland cyst, cystic lymphangioma, and anterior meningocele (7). Discrimination of tailgut cyst and other presacral cysts is important because of the malignant potential of a tailgut cyst. However, because substantial overlap exists in the imaging findings of the presacral cysts, it is hard to differentiate the imaging appearance of tailgut cyst from that of many other presacral cysts. Because of that histologic analysis is necessary to establish a right diagnosis of tailgut cyst (8). The unilocular or multilocular characteristic is important. Among the presacral cystic masses, epidermoid cyst, dermoid cyst, rectal duplication cyst, and anterior meningocele are mostly unilocular (7). On the other hand, tailgut cyst and cystic lymphangioma are usually multicystic (7). Our case was also multilocular. Malignant transformation incidence is 2% within tailgut cyst from a single retrospective case series (9). Ballantyne first reported malignant transformation of a tailgut cyst (10). As a result it should be kept in mind the differential diagnosis of presacral masses because of the malignant potential of a tailgut cyst.

REFERENCES

1. Marco V., Fernandez-Layos M., Autonell J., Doncel F, Farre J. Retrorectal cyst-hamartomas: report of two cases with adenocarcinomas developing in one. *Am J SurgPathol* 1982; 6: 707-14.
2. Mathis KL., Dozois EJ., Grewal MS. et al. Malignant risk and surgical outcomes of presacral tailgut cysts. *British Journal of Surgery* 2010; 97:575-9.
3. Johnson A., Ros P., Hjermstad B. Tailgut cyst: diagnosis with CT and sonography. *Ajr* 1986; 147: 1309-11.
4. Mouloupoulos LA., Karvouni E., Kehagias D et al. MR Imaging of Complex Tail-gut Cysts Clinical Radiology 1999; 54: 118-22.
5. Yang DM, Jung DH, Kim H et al. Retroperitoneal Cystic Masses: CT, Clinical and

Pathologic Findings and Literature Review *Radiographics* 2004; 24:1353–65.

6. Schwarz Re., Lyda M., Lew M., Paz Ib. A carcinoembryonic antigen secreting adenocarcinoma arising within a retrorectal tailgut cyst: clinicopathological considerations. *Am J Gastroenterol* 2000; 95:1344–7.

7. Dahan H., Arrive L., Wendum D., et al. Retrorectal developmental cysts in adults: clinical and radiologic-histopathologic review, differential diagnosis and treatment. *Radiographics* 2001; 21:575-84.

8. Yang DM, Park CH, Jin W et al. Tailgut Cyst: MRI Evaluation *Ajr* 2005; 184:1519-23.

9. Hjermstad B., Helwig E. Tailgut cysts. Report of 53 cases. *Am J ClinPathol* 1988; 89:139-147

10. Mathieu A., Chamlou R., Le Moine F., Maris C., Van de Stadt J., Salmon I.: Tailgut cyst associated with a carcinoid tumor: case report and review of the literature. *Histol Histopathol*, 2005; 20:1065–9.