CASE REPORT

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Retrorectal Cystic Hamartoma

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ABSTRACT Retrorectal cystic hamartoma (Tailgut cyst) is a rare congenital lesion mostly located in the retrorectal space and is a postnatal remnant of the hindgut. Tailgut cysts may be asymptomatic or cause symptoms related to mass effect on the neighboring organs. These cysts are frequently diagnosed in fourth decade of life. Tailgut cysts should be distinguished from other retrorectal lesions like teratomas, cystic schwannomas, chordomas and abscesses. In surgical approach, mostly retrorectal masses are resected in an en bloc fashion with any tethered structures, including the sacrum, coccyx, or rectum. This is a report about the prenatal diagnosis of a tailgut cyst and its complete removal after birth.

Keywords: Hamartoma; sacrococcygeal region

Retrorectal cystic hamartomas, which are also known as 'tailgut cysts' are rare congenital lesions located in retrorectal space.[1] Embryologically, they are thought to be derived from the postnatal remnants of the hindgut. The tailgut normally regresses by the 8th week of fetal life.[2] Cystic structure is formed if those mucous secreting remnants fail to regress. Tailgut cysts are predominantly asymptomatic but they might present with infections or obstruction. Treatment is based on the information obtained by imaging modalities. Surgical resection of the mass provides a specimen for the definitive diagnosis and exclusion of possible malignant transformation. This is a report about the prenatal diagnosis of a tailgut cyst, excised completely in en bloc fashion after delivery.

CASE REPORT

A 28-year-old primigravid woman was referred to the department of obstetrics for routine second trimester ultrasonography scan. The pregnancy had been uneventful and previous sonograms were reported as normal. At 24th week of gestation, a detailed sonography was performed for screening congenital anomalies and a large sacrococcygeal mass was found. Despite the tumoral width, no increased vascularization was detected. Fetal magnetic resonance imaging (MRI) was carried out and showed a cystic multilobular mass was visualized in presacral space. Radiology department interpreted the mass as a 'tailgut cyst'. A cesarean section was performed due to the risk of perinatal delivery trauma and dystocia and a healthy neonate except for a cystic mass in retrorectal area was delivered (Figure 1). The newborn was then referred to the department of pediatric surgery. Both the ultrasonography and MRI demonstrated a cystic thin-walled mass of

6x3x3 cm within the sacrococcygeal region (Figure 2). The mass had no solid component and it was excised without difficulty (Figure 3). The macroscopic specimen consisted of a multilocular, 5.8 x 3.4 x 2.2 cm cyst with steady borders and an irregular inner aspect. Microscopically, the cyst walls were lined by various epithelia including squamous, ciliar and columnar cells. Associated inflammation was present and specific tissue fragments of significance were not found. The histopathological examination revealed 'retrorectal cystic hamartoma' and there was no evidence of malignant transformation. The one-week-old newborn had an uneventful postoperative course.

DISCUSSION

Tailgut cysts are rarely observed congenital cystic lesions which are predominantly located in retrorectal space. This space is formed posteriorly by the sacrum and coccyx, anteriorly by the rectum and extends superiorly to peritoneal reflection and inferiorly to the rectosacral fascia and the supralevator space. The lateral boundaries consist of the ureters and iliac vessels. The differential diagnosis of masses within this space includes dermoid and epidermoid cysts, cystic sacrococcygeal teratomas, meningoceles, lymphangiomas, abscesses, chordomas and tailgut cysts.[3]

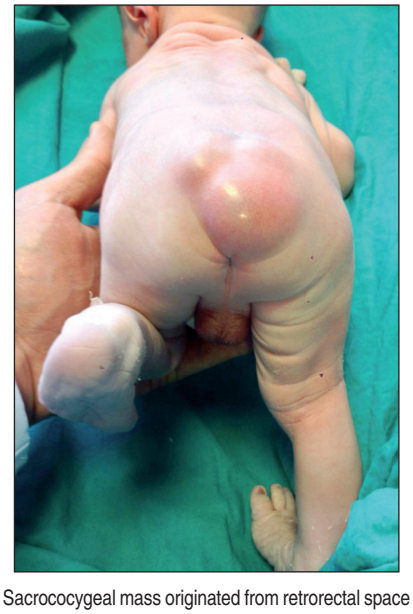

FIGURE 1: Sacrococcygeal mass originated from retrorectal space in neonate.

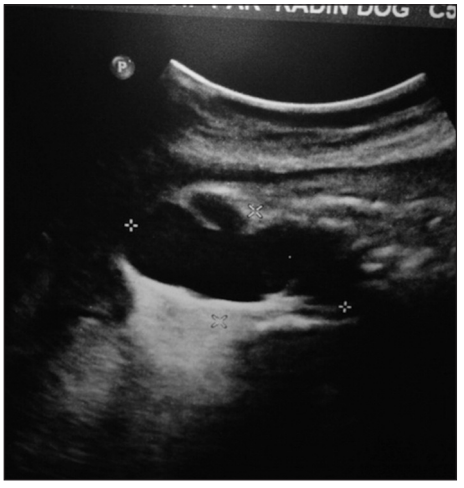

FIGURE 2: Ultrasound image of the cystic, thin walled mass located in retrorectal space of the fetus.

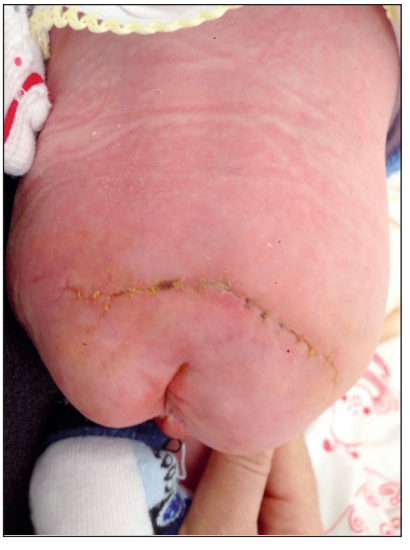

FIGURE 3: Postoperative image of neonate.

Tailgut cysts arise from the remnants of hindgut which normally regress during pregnancy.[2] Although these developmental lesions can be diagnosed in utero by ultrasonography, they are most frequently diagnosed in middle aged women.[4] Female to male ratio is approximately 3:1, but the association with gender remains unclear.[5]

Half of the cases with tailgut cysts are discovered incidentally. However, patients may present with symptoms resulting from local mass effect or related complications.[6] The most important complications of these cysts are infections,

fistulization and malignant degeneration. It is also worth mentioning that tailgut cysts, when infected, may present with symptoms similar to pelvic abscess.

The diagnosis can be made based on the prenatal ultrasonography findings. Computerized tomography (CT) scan and MRI are other imaging methods for making a preoperative diagnosis and choosing the appropriate technique for surgery.[5] Preoperative biopsy should not be attempted as there is a risk of spreading dysplastic cells throughout the thin cystic walls.[7] Instead of a preoperative biopsy, complete excision is necessary to prevent recurrence, infection, and possible malignant transformation. Malignancy criteria for these masses are nodular wall thickening, indistinct margins, lymphadenopathy, superior extension above third sacral vertebra (S3) and intracystic vegetations.

Different methods have been defined for excision of the mass, but en bloc fashion removal is the recommended approach for tailgut cysts. The most appropriate technique for excision is dependent on the size and location of the mass and the experience of the surgeon. Complete excision is necessary to prevent recurrence, infection, and rule out possible malignant transformation. Posterior approach is suitable especially for small lesions below the level of S3 while the cysts that have their lowest border above the fourth sacral vertebra should be treated with a transabdominal approach.[8]

The postoperative follow up period is determined according to the clinical history, physical findings and imaging scans. It has been advised to perform digital exam for evaluating recurrence risk and CT scan in the first postoperative year.[8] Most of the retrorectal tumors are benign and disease free survival rates are high. In a series of 47 patients with retrorectal tumors, 5-year survival rate was 75% for patients with malignant tumors and 93% for patients with benign disease.[5] Tumor markers like CEA and CA 19-9 may be used to detect recurrences during the postoperative follow-up period.[9]

Although tailgut cysts are encountered rarely, they should be considered in the differential diagnosis of sacral masses. These cysts deserve careful evaluation especially in the neonates. Complete surgical excision is the appropriate treatment as it provides a specimen for the ultimate diagnosis of cystic hamartomas and exclusion of possible malignant degeneration.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Fırat Tülek, Özgür Kan;* ***Design:*** *Fırat Tülek;* ***Control/Supervision:*** *Ali Gemici, Ayşegül Alkılıç;* ***Analysis and/or Interpretation:*** *Ali Gemici;* ***Literature Review:*** *Ayşegül Alkılıç, Özgür Kan;* ***Writing the Article:*** *Özgür Kan, Fırat Tülek;* ***Critical Review:*** *Ali Gemici, Ayşegül Alkılıç;* ***References and Fundings:*** *Ayşegül Alkılıç;* ***Materials:*** *Fırat Tülek.*

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