**Isolated lumbar intradural tailgut cyst: A case report and review of the literature**

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**ABSTRACT**

Tailgut cysts are rare developmental cysts arising from remnants of the embryological postnatal gut. Despite being frequently located in the presacral space, isolated cases of aberrant locations have been reported, including, perirenal, perianal, and subcutaneous sites, with only two cases of subdural tailgut cysts reported to date. The clinical course is often marked by linear growth, causing compression of the adjacent structures, however malignant transformation with carcinomatous features has been previously described. Hereby the authors describe a case of an intradural extramedullary tailgut cyst in a 33-year-old man presenting with progressive low back pain and signs of autonomic dysfunction, including urinary retention and bowel incontinence. Whole-spine MRI revealed an intrathecal cystic lesion located at L2-L3 level exhibiting hyperintensity on T2-weighted images not enhancing when contrast was administered. Laminectomy followed by tumor excision was performed and pathological analysis confirmed the diagnosis of tailgut cyst.

1. Introduction

Tailgut cysts, also known as cystic hamartomas, are rare mucous-secreting congenital lesions that are thought to be derived by a developmental nondisjunction of the embryonic hindgut \cite{1, 2, 3}. These benign lesions tend to be located in the presacral space \cite{4}, lying anterior to the sacrum and posterior to the rectum. Occasionally perirenal, perianal, and subcutaneous involve ment have been described \cite{5, 6, 7} with intradural tailgut cysts remaining an extremely rare condition with only two cases reported in the literature \cite{8, 9}. Whereas almost exclusively exhibiting a benign behavior, cases of malignant transformation have been described \cite{8, 10, 11}. Given the abundance of epithelioid cells, frequently associated malignant transformation displays carcinomatous features. Hereby an unusual case of a patient with low back pain unresponsive to symptomatic pharmacotherapy and bladder and bowel incontinence is described. Spinal MRI and microscopic analysis confirmed the diagnosis of intradural tailgut cyst without evidence of malignant transformation.

2. Case report

The authors present a case of a 33-year-old man with progressive worsening of low back pain, unresponsive to symptomatic pharmacotherapy without any other relevant past medical history. At admission, the patient referred reduced touch sensation in the lower limbs, more pronounced in the right leg, associated with diffuse paresthesia of the L2-L3 corresponding dermatomal area. Neurological examination showed marked weakness (MRC 3/5 and 4/5 in the left and right leg, respectively), with a slight but present response to deep tendon reflexes stimulation. Anal sphincter insufficiency and urinary retention were also detected. No other neurological deficits were noticed, therefore, a whole-spine magnetic resonance imaging was performed (Figure 1). The scan showed a cystic lesion of the conus medullaris measuring $16 \times 10 \times 15$ mm located at L2-L3 level exhibiting homogeneous hyperintensity on T2-weighted images. A solid nodule located in the posterior region of the cyst did not demonstrate enhancement when gadolinium was administered. A posterior laminectomy of the L2-L4 segment followed by tumor excision was performed. At the opening, the cystic portion of the lesion contained a brownish fluid and the posterior nodule appeared strictly...
adherent to the thin dural sac. Pathological analysis revealed cystic lining of epithelial-like cells showing mucin secretion capability, confirming the diagnosis of tailgut cyst (Figure 2). The post-operative course was uneventful, and the patient did not experience any surgical complications. He was then transferred to a neurorehabilitation institution and his neurological condition progressively improved. At 6-month follow-up examination, no evidence of lower limbs hyposthenia was noticed and DTR stimulation showed a brisk response. Control spinal MRI did not demonstrate signs of recurrence.

3. Discussion

The tailgut or postanal gut is the distalmost part of the embryonic gut, caudal to future anus. It normally reaches its largest diameter on the 35th day of gestation and involutes by the eighth week of embryonic development [2, 3]. Given the proximity of the tailgut to the neural structures, an intra-axial cyst arising from developmental nondisjunction is possible, however, only two cases of intradural formation of tailgut cysts have been reported to date (Table 1) [8, 9]. Developmental cysts are almost invariably located in the presacral space, lying anterior to the sacrum and posterior to the rectum but have also been described in perirenal, perianal, and subcutaneous locations [4, 6, 7]. Due to the rarity of this malformation, real epidemiological data are difficult to obtain since only case reports and small case series are described in the literature [12]. In general tailgut cysts become clinically evident by the fifth decade, showing a marked female preponderance with a 3:1 ratio [13, 14]. However, this condition has also been described in infants and in the fetus, strengthening the theory of a congenital origin [15]. Hjermstad et al. [4] reported the largest series of tailgut cysts to date. They described 53 cases of tailgut cysts localized to the presacral space. Common clinical manifestations included discomfort and pain when sitting, change of stool caliber, fistula formation, infection, and bleeding. However, in approximately half of the cases, the diagnosis was incidental since these patients presented with no evident symptomatology. Although significantly rare, a review of the papers describing intradural tailgut cysts demonstrates peculiar clinical features that these may exhibit [8, 9]. For instance, common manifestations include autonomic dysfunction, especially of the sacral plexus that supplies the hindgut and the pelvic area, weakness and paresthesia in the lower extremities causing gait disturbances.

Grossly, tailgut cysts are soft, multiloculated, and usually well-circumscribed, exhibiting surrounding adherent fibroadipose tissue. Microscopically, these lesions are usually multicystic and lined by a variety of epithelial types, including transitional, squamous, ciliated columnar, and mucinsecreting epithelium; the latter was also apparent in the present case. Disorganized bundles of smooth muscle cells could be seen interposed within the multilocular cysts along with dense mucoid

![Figure 1. T2-weighted magnetic resonance image showing intradural cystic lesion with an intrinsic nodular component causing compression of the spinal cord.](image1)

![Figure 2. Microphotography revealing mucoid secreting epithelial cells displaying multicystic architecture.](image2)
fluid [12]. Although tailgut cysts include all three germ layers several authors consider this developmental abnormality a separate entity from teratoma due to the absence of neural features or bone encroachment [4, 8]. Whereas the majority of tailgut cysts are primarily benign in both behavior and growth, there have been reports of malignant transformation [8, 10, 11]. Because of the abundance of epithelial cells that organize in cystic formations, it is not surprising that carcinomatous features are frequently observed in malignant tailgut cysts. Niazi et al. [8] described a case of intradural tailgut cysts with carcinoid features, in a patient who did not harbor the typical symptoms of carcinoid syndrome, therefore prompting urgent decompressive surgery followed by tumor excision. The pathological analysis confirmed the diagnosis of tailgut cyst with mucin secreting epithelial cells.

The patient's written informed consent was collected prior to data collection and available on reasonable request.

4. Conclusion

In the present report, a rare case of intradural extramedullary tailgut cyst is described. As a result of its intra-axial location, the lesion caused atypical clinical manifestations secondary to spinal cord and nerve root compression. The patient presented with marked hyposthenia and diffuse paresthesia and was unresponsive to symptomatic pharmacotherapy for low back pain. Furthermore, autonomic dysfunction was noticed during examination with signs of urinary retention and anal sphincter insufficiency, therefore prompting urgent decompressive surgery followed by tumor excision. The pathological analysis confirmed the diagnosis of tailgut cyst with mucin secreting epithelial cells.

Declarations

Author contribution statement

All authors listed have significantly contributed to the investigation, development and writing of this article.

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Data will be made available on request.
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The authors declare no conflict of interest.

Additional information

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References

[1] H. Dahan, L. Arrive, D. Wendum, H. Docou le Pointe, H. Djouhari, J.M. Tubiana, Retrorectal developmental cysts in adults: clinical and radiologic-histopathologic review, differential diagnosis, and treatment, Radiographics 21 (3) (2001 May-Jun) 575–584.

[2] D.M. Yang, C.H. Park, W. Jin, S.K. Chang, J.E. Kim, S.I. Choi, D.H. Jung, Tailgut cyst: MRI evaluation, AJR Am. J. Roentgenol. 184 (5) (2005 May) 1519–1523.

[3] F.I. Lukas, M.E. De Paepe, M. DiLorenzo, C. Mercier, J.G. Desjardins, Tailgut remnant or teratoma? Eur. J. Pediatr. Surg. 3 (3) (1993 Jun) 182–185.

[4] B.M. Hjermstad, E.B. Helwig, Tailgut cysts. Report of 53 cases, Am. J. Clin. Pathol. 89 (2) (1988 Feb) 139–147.

[5] S.H. Jang, K.S. Jang, Y.S. Song, K.W. Min, H.X. Han, K.G. Lee, S.S. Paik, Unusual prerectal location of a tailgut cyst: a case report, World J. Gastroenterol. 12 (31) (2006 Aug 21) 1083–1085.

[6] J. Ohn, S. Jang, S.J. Jo, K.H. Cho, A case of tailgut cyst as a subcutaneous nodule at the coccygeal area, Ann. Dermatol. 28 (5) (2016 Oct) 641–642.

[7] M.T. Sung, S.F. Ko, C.K. Niu, C.S. Hsieh, H.Y. Huang, Perirenal tailgut cyst (cystic hamartoma), J. Pediatr. Surg. 38 (9) (2003 Sep) 1404–1406.

[8] T.N. Niazi, L.M. Shah, S.S. Chin, M.H. Schmidt, Isolated intradural lumbosacral tailgut cyst with carcinoid features, J. Neurosurg. Spine 14 (3) (2011 Mar) 382–387.

[9] J. Kemp, M.A. Guzman, C.M. Fitzpatrick, S.K. Elbahaa, Holocord syringomyelia secondary to tethered spinal cord associated with anterior sacral meningocele and tailgut cyst: case report and review of literature, Childs Nerv Syst 30 (6) (2014 Jun) 1141–1146.

[10] M.G. Horenstein, R.A. Erlandson, D.M. Gonzalez-Cueto, J. Ronai, Presacral carcinoid tumors: report of three cases and review of the literature, Am. J. Surg. Pathol. 22 (2) (1998 Feb) 251–255.

[11] Z. Krivokapic, I. Dimitrijevic, G. Barisic, V. Markovic, M. Krsic, Adenosquamous carcinoma arising within a retrorectal tailgut cyst: report of a case, World J. Gastroenterol. 11 (39) (2005 Oct 21) 6225–6227.

[12] D. Paszouras, N. Pawa, H. Osmani, R.K. Phillips, Management of tailgut cysts in a tertiary referral centre: a 10-year experience, Colorectal Dis. 17 (8) (2015 Aug) 724–749.

[13] A. Saka, H.S. Kim, Y.D. Han, M.S. Cho, H. Hur, B.S. Min, K.Y. Lee, N.K. Kim, Single-center experience of 24 cases of tailgut cyst, Ann Coloproctol 35 (5) (2019 Oct) 268–274.

[14] G. de Castro Gouveia, L.Y. Okada, B.P. Paes, T.M. Moura, A.H. da Conceição Júnior, R.N. Finheiro, Tailgut cyst: from differential diagnosis to surgical resection-case report and literature review, J. Surg. Case Rep. 2020 (7) (2020 Jul 23), rjaa205.

[15] M. Nakagawa, M. Haru, H. Oshima, M. Kiitse, S. Shihamoto, Radiological findings of tailgut cyst in a fetus, J. Comput. Assist. Tomogr. 32 (2) (2008 Mar-Apr) 210–213.

[16] J.J. Liang, S. Alrawi, G.N. Fuller, D. Tan, Carcinoid tumors arising in tailgut cysts may be associated with estrogen receptor status: case report and review of the literature, Int. J. Clin. Exp. Pathol. 1 (6) (2008 Jan 1) 539–543.

[17] A.R. Johnson, P.R. Ross, B.M. Hjermstad, Tailgut cyst: diagnosis with CT and sonography, AJR Am. J. Roentgenol. 147 (6) (1986 Dec) 1309–1311.

[18] G. Lissi, S. Cesari, M. Pavanello, R. Butini, Tailgut cysts: CT and MR findings, Abdom. Imag. 20 (3) (1995 May-Jun) 256–258.

[19] E. Au, O. Anderson, B. Morgan, L. Alarcon, M.L. George, Tailgut cysts: report of two cases, Int. J. Colorectal Dis. 24 (3) (2009 Mar) 345–350.