Adenocarcinoma within a tailgut cyst

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DESCRIPTION

We present a case of a 53-year-old woman with painful defaecation and lower abdominal pain. Her medical history and laboratory testing were unremarkable. An abdominopelvic CT scan was performed. Axial non-enhanced CT image (figure 1) and postcontrast CT image (figure 2) revealed a well-defined, slightly lobular soft tissue density mass in the retrorectal/presacral space. It shows a thin peripheral calcification and internal enhancing components after administration of intravenous contrast material. These features suggest a rare developmental retrorectal lesion, with enteric cysts (tailgut cyst and cystic rectal duplication) being the more common. Differential diagnosis comprises lymphangioma, epidermoid cyst, dermoid cyst, endometrioma, rectal duplication cyst, anal gland cyst and anterior meningocele.

The internal heterogeneity and enhancing components of the lesion are worrisome features.

Surgical excision was performed (posterior approach—Kraske procedure). The specimen revealed an adenocarcinoma arising within a tailgut cyst. The tumour shows focal invasion of the removed sacrum (S4 and S5) and perineurial and vascular permeation. There was no evidence of tumour persistence in postsurgical magnetic resonance study (figure 3). The multidisciplinary team decided to perform adjuvant radiotherapy and chemotherapy regimen.

Tailgut cysts (retrorectal cystic hamartoma) are developmental cystic lesions occurring in adults in the retrorectal/presacral space, most frequently in middle-aged women, with a female to male ratio of 3:1. They are often asymptomatic, although patients may present symptoms resulting from local mass effect, such as constipation, rectal fullness, painful defaecation, lower abdominal pain, and dysuria or urinary frequency.¹

Figure 1 Axial non-enhanced CT image showing a heterogeneous nodular mass of tissue density with a tiny peripheral calcification in the presacral space.

Figure 2 Axial and sagittal postcontrast CT images showing heterogeneous enhancement of the nodular lesion.

Tailgut or retrorectal cyst is typically a multi-loculated cystic mass with a thin wall filled with mucoid material. There are only limited reports on the imaging features of these cysts. CT shows a well-margined, thin-walled, unilocular or more

Figure 3 Axial T1, T2 and postgadolinium (T1 FS Gadolinium) MRI showing postsurgical signal changes due to inflammation. There is no evidence of nodular enhancing nodules.

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Images in...

frequently multilocular, hypoattenuating, non-enhanced lesion in the retrorectal space. In rare cases there are associated thin calcifications.²

Tailgut cysts may complicate with infection (with or without fistulisation), bleeding and malignant degeneration.

Malignant degeneration occurs in 7% of tailgut cysts and results in adenocarcinoma or squamous carcinoma. Rarely carcinoid and sarcoma may also arise within the cyst. MRI or CT imaging typically will show suspicious features, namely irregular wall thickening or a polypoid mass with intermediate signal intensity on both T1-weighted and T2-weighted MRIs and

enhancement after the intravenous administration of contrast material. Of note, if secondarily infected, the tailgut cyst may present thick walls with surrounding inflammatory changes.³

Excision of a tailgut cyst is advised for symptomatic patients but also for asymptomatic patients mainly to rule out the possibility of malignancy or future malignant transformation.

Complete excision for benign lesions comprises disease-free expectancy. In the malignant counterparts excision of the rectum and bony sacrum and coccyx may be necessary.

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Learning points

- Tailgut cysts are retrorectal developmental cystic lesions in adults and are often asymptomatic, although patients may present symptoms resulting from local mass effect.
- Tailgut cysts occur mostly in middle-aged women.
- Infection, bleeding and malignant degeneration are the major complications of these developmental cysts.
- Malignant degeneration of the tailgut cysts occurs in 7% of cases; CT and MRI may show suspicious features such as irregular mass contours and contrast enhancement.
- Owing to the risk of malignant transformation of these cystic lesions, surgical excision is advised.

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