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Case Report

Tailgut Cyst with a Neuroendocrine Tumour

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Abstract

Tailgut cysts or presacral cystic hamartomas are rare lesions with an incidence of less than 1 in 40,000 They are more common in female patients and can undergo malignant transformation [1]. Most patients are asymptomatic and the lesions are discovered incidentally.

We report here a case of a male patient with a large tailgut cyst who was relatively asymptomatic and the lesion was discovered on routine sonography. After further appropriate investigations, the lesion was removed by a laparoscopic technique assisted by an open perineal approach. The pathological features were suggestive of a tailgut cyst which also harboured a neuroendocrine tumour.

Keywords: *Tailgut Cyst; Neuroendocrine Tumour; Presacral Cystic Hamartomas*

Introduction

The growing embryos have a tail which contains the terminal part of the hindgut. The tail reaches its maximum length at 35 days and then regresses. Failure of the hindgut to regress leaves behind part of the hindgut which can become cystic. These are termed tail gut cysts or cystic hamartomas [2]. There is a female preponderance of 5: 1 to 8: 1 and around 50% of these patients are asymptomatic. Being a rare condition, there are very few case series and most published cases are single-case reports. Different approaches depending on the size have been described. A transabdominal open or laparoscopic, a perineal and a transrectal [2].

We present here a case of a large tailgut cyst in a male patient removed by a combined laparoscopic and perineal approach.

Case Report

A 26-year-old male patient presented with mucous in his stools and left groin pain for 3 months. He had no other symptoms. No bleeding per rectum, no difficulty in passing stools and no tenesmus.

General examination was normal, and no lump was felt per abdomen. On rectal exam at about 6 to 8 cm from the anal verge, a lump was felt outside the rectum posteriorly. It was firm and the upper limit could not be reached. The rectal mucosa was easily mobile over the growth.

Routine biochemical evaluation was normal, tumour markers were not done. Sonography showed a large retro rectal cyst which was reported as a tailgut cyst. An MRI of the pelvis revealed a large cystic lesion which looked dark on T1 (Figure 1) and bright on T2 images. The lesion had septations and appeared bilobed. At the most caudal part of the lesion, a small mural nodule was identified as shown in figure 2 with the yellow arrow (Figure 2). The lesion extended from S2 to the supra-levator level, measuring 11 x 9 x 8 centimetres. There was anterior compression of the rectum.

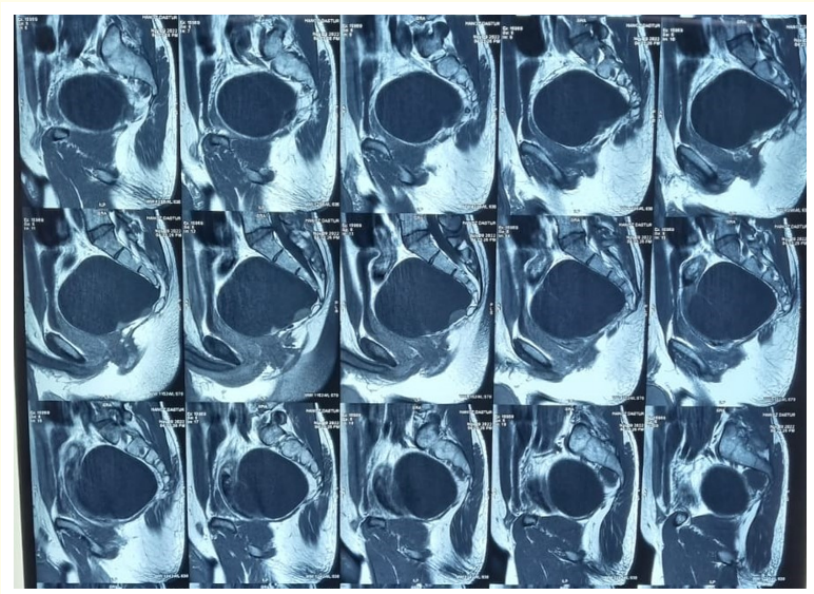

Figure 1

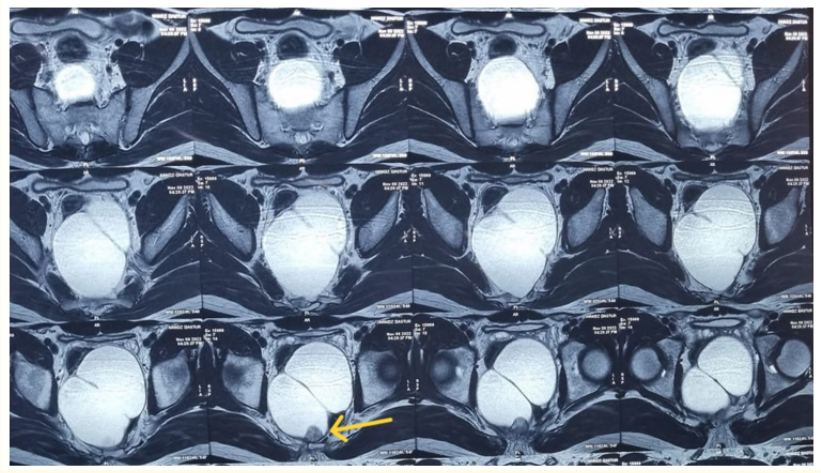

Figure 2

Operative technique: With the patient in a modified lithotomy position five ports were used as in a low anterior rectal resection. The peritoneum on either side of the rectum was incised and the rectum and mesorectum were elevated with an umbilical tape. Gradual dissection downwards revealed the cyst which was separate from the rectum and mesorectum. The cyst was carefully dissected downwards and separated from the mesorectum (Figure 3). The most caudal part of the cyst could not be approached by laparoscopy and this was done by the perineal approach.

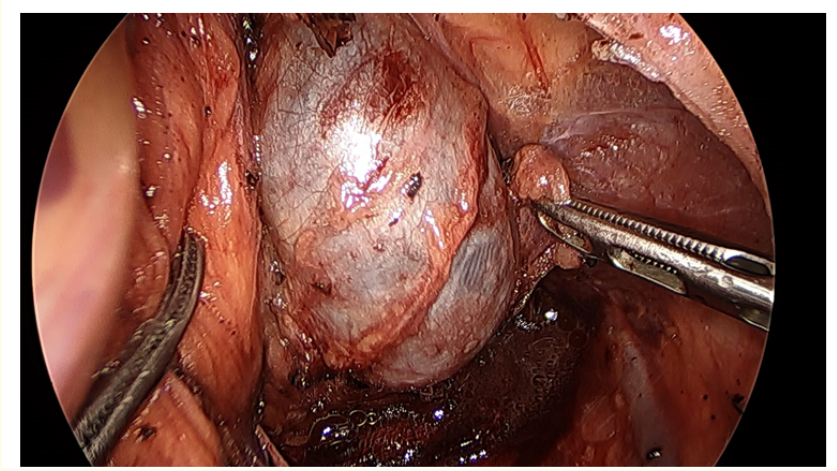

Figure 3

A posterior horseshoe incision was taken and deepened down to the anococcygeal raphe which was divided, the tip of the coccyx was excised and Waldeyer's fascia divided. The lower border of the cyst could be felt. The vision was not ideal but upward dissection was possible. There were some thick fibrous adhesions to the sacrum, when dividing these the cyst opened and the fluid spilt out. This was cleaned out and irrigated. The collapsed cyst was dissected free and brought out through the perineum. A Drain was placed from the abdomen and the perineum was closed without a drain. His post-operative recovery was uneventful. The drain was removed on the 8th day after surgery.

The pathology report showed a keratinised squamous epithelial lined cyst containing a well-differentiated neuroendocrine tumour. On Immunochemistry the tumour expressed positivity for chromogranin A and INSMI. The KI index was < 2%. Two months after surgery a PET DOTATATE scan was done which revealed no neuroendocrine activity.

Discussion

Retro rectal lesions are rare with a reported incidence of 1 in 40,000 [3]. The commonly described ones are desmoids, rectal duplications, neuro enteric, anterior meningocele and tailgut cysts. Tail gut cysts develop when the hindgut fails to regress and leaves a remnant behind. Females are more commonly affected and 5th decade is the most common age of presentation [2]. The lining can have different epithelium [4], in our case, it was a squamous lining. Adenocarcinoma and neuroendocrine tumours are the most associated tumours and are almost always incidental findings. MRI is an ideal modality of investigation to assess the size, location, and morphology of the lesion. In our case, a mural nodule was detected which should have drawn our attention to an underlying neoplasm (Figure 2). The approach to these lesions depends upon the location. The more distal lesions can be removed by a perineal approach, but larger lesions require a dual approach, either by open laparotomy or laparoscopy. If the upper border of the cyst can be felt on per rectal examination, then the

perineal approach is better. We found that the laparoscopic approach was effective and gave excellent vision all the way down to the most distal part of the cyst. By careful separation from the rectum and meso- rectum the cyst could be separated in the retro-rectal area easily. The perineal approach was cramped and vision was inadequate. This could be the reason for the cyst rupture. Kraske has described a perineal approach in a prone position which gives better access to the retro rectal area [5]. Dissection needs to be done with care and several complications have been described including rectal perforation, and hypogastric nerve injury leading to pelvic floor dyssynergia [2]. Surgical excision is mandatory even if patients are asymptomatic as the chance of malignancy is high and, in some reports, up to 30% [1].

Conclusion

In conclusion, tailgut cysts are rare lesions which need surgical excision. Laparoscopy is good to approach these lesions when they lie above the mid-third of the rectum. Larger lesions often need a dual approach as described in this case.

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