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Case Report

Paratesticular metastasis from primary midgut neuroendocrine tumor: A rare initial presentation ☆

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ABSTRACT

Neuroendocrine tumors are malignant neoplasms arising from neuroendocrine cells. These are increasingly recognized with rising incidence and encompass a diverse range of phenotypes. The large majority of these originate in the gastrointestinal tract however primary neuroendocrine tumors have also been reported to arise in a variety of organs such as lung, breast, prostate, and skin. Primary malignant paratesticular masses are often sarcomatous in origin and metastatic spread to the paratesticular region or scrotum is exceedingly rare. We report a fascinating case of a 56-year-old male who had an unusual initial presentation of paratesticular lesions on a background of an undescended testicle and an incidental umbilical nodule. After a combination of radiological and histopathological investigations, he was diagnosed with metastatic midgut neuroendocrine tumor.

Introduction

Neuroendocrine tumors (NETs) constitute a rare and diverse spectrum of cancers originating from neuroendocrine cells. The collective incidence of all neuroendocrine tumors stands at approximately 5 cases per 100,000 individuals, a figure that has exhibited an upward trend in recent years [1]. Predominantly, NETs manifest within the gastrointestinal tract (73%) and the bronchopulmonary system (25%). Notably, their occurrence within the gastrointestinal tract is most prevalent in the small bowel (29%), followed by the appendix (19%) and rectum (13%) [2]. These tumors have the capacity to secrete various peptide hormones, enabling the application of functional imaging techniques such as positron emission tomography computed tomography (PET CT) and Gallium-68 DOTATATE PET CT in conjunction with conventional imaging modalities. This combined approach facilitates the comprehensive characterization of tumor subtypes and assessment of disease extent.

Metastasis occurs in around 30% of cases of Neuroendocrine tumors [2], in which the liver is the most prevalent site for metastases among NET patients, documented as high as 82% [3]. Other common metastatic sites include

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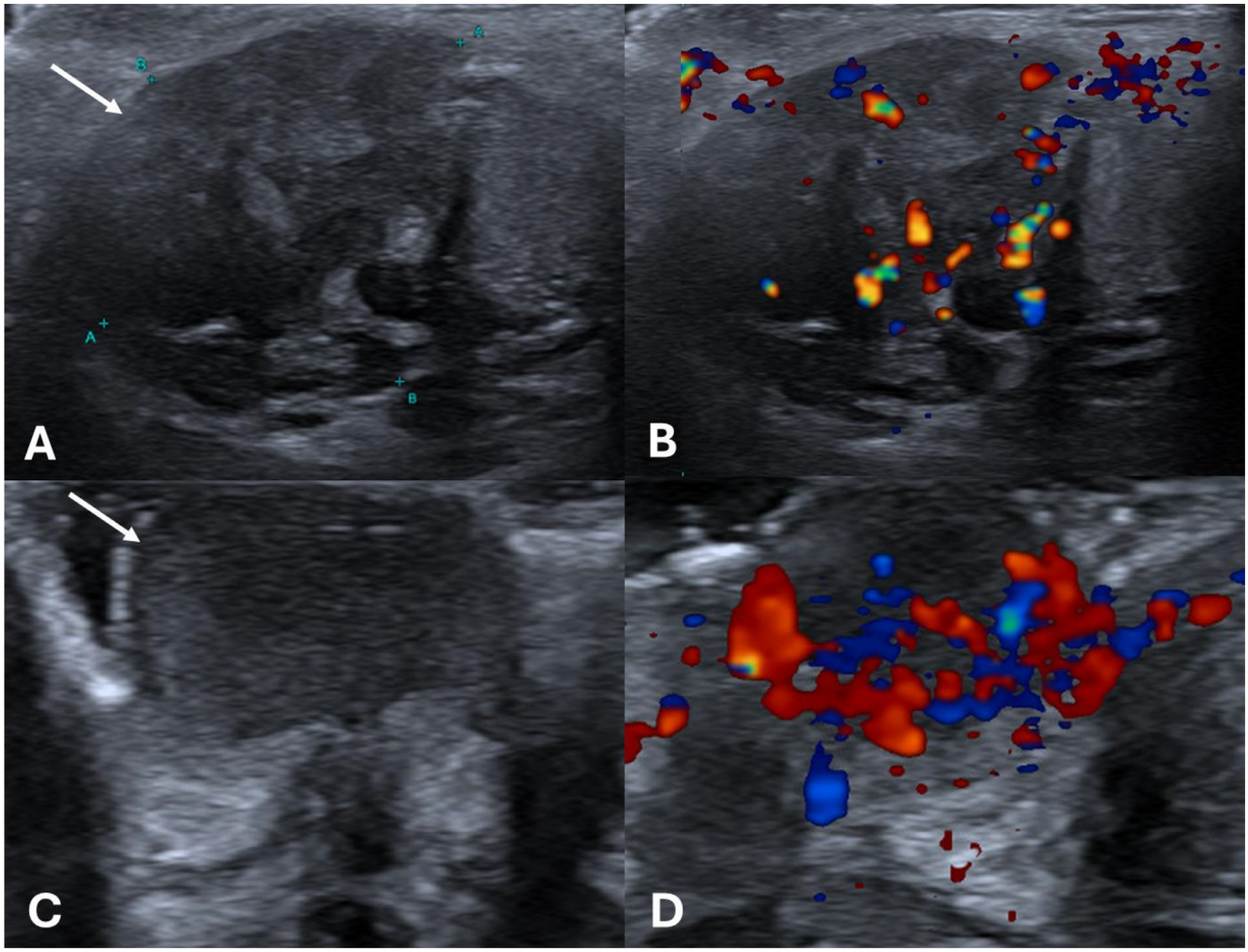

Fig. 1 – Ultrasound of the testes and inguinal region. (A) Hypoechoic solid right paratesticular lesion (white arrow). (B) This demonstrated marked internal vascularity on color doppler imaging. (C) About 12 mm superficial hypoechoic nodule within the umbilicus (white arrow). (D) Significant internal and peripheral vascularity on color doppler.

bones, adrenal glands, lung, brain, and peritoneum [3,4]. Within neuroendocrine tumors of the gastrointestinal tract, approximately 10% of cases result in peritoneal metastasis [5]. While peritoneal disease is not uncommon, occurrences of extra-testicular metastases in neuroendocrine carcinoma remain rare. The most common primary sources for extra-testicular metastases, albeit very uncommon, are from testicular, prostate, and renal origins [3,4].

This is a case of an unusual initial presentation of paratesticular metastasis from primary midgut neuroendocrine tumor in a patient who was also diagnosed with an ipsilateral undescended testis. We describe and discuss the investigations which ultimately led to this rare presentation and discuss the management implications.

Case report

A 56-year-old male was referred to the urology team reporting discomfort due to a high riding right testicle over the past 2 years, notably worsening in the last 3 months. Prior to this the testes were down within the scrotum. On examination there was no evidence of infection, and all the initially performed biochemical tests and tumor markers were withing normal limits.

He was referred for an ultrasound examination which demonstrated a right testicle located within the inguinal canal. The right testicle itself had a normal sonographic morphology and vascularity with no intratesticular mass. However, there were several paratesticular hypoechoic, solid lesions within the inguinal canal which demonstrated pronounced central vascularity, the largest of which measured 27 mm (Fig. 1). The left testicle was normal in appearance and position. During the ultrasound scan, the patient revealed to the operator that he was also concerned about an umbilical lump which had been previously diagnosed in the community as an umbilical hernia. Focused ultrasound of this region demonstrated a 12 mm, solid, round, hypoechoic lesion within the umbilicus with a similar sonographic appearance to the paratesticular lesions and marked central vascularity.

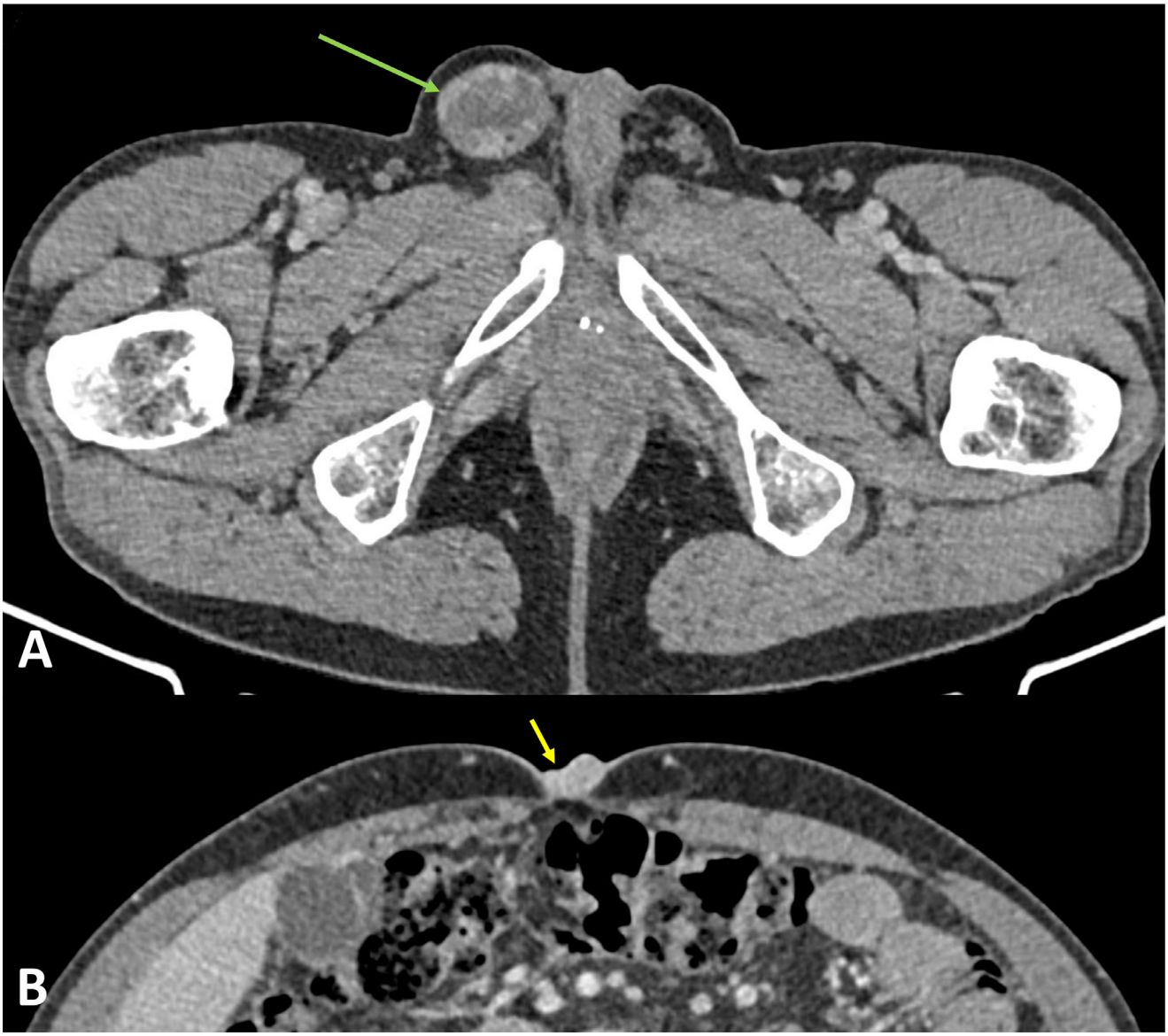

Fig. 2 – Axial slices of CT abdomen and pelvis with contrast. (A) Undescended testes in right inguinal canal with multiple enhancing paratesticular soft tissue nodules (green arrow). (B) Soft tissue nodule in the umbilicus (yellow arrow).

Given the suspicion of a systemic disease process, a CT thorax, abdomen, and pelvis with contrast was performed which corroborated with the ultrasound findings; demonstrating an umbilical soft tissue nodule and multiple nodules within the right inguinal canal surrounding the undescended testis (Fig. 2). However, no clear primary source or further sites of metastases were identified on the CT.

After discussion at the urology multidisciplinary team meeting (MDT), a decision was made to surgically excise the umbilical nodule for tissue analysis. The histopathology of this lesion confirmed a well-differentiated neuroendocrine tumor of intermediate grade.

As per local staging protocols at our institution for neuroendocrine tumors, both FDG PET CT and Gallium DOTATATE PET CT scans were performed.

The FDG PET CT scan demonstrated an intensely avid right paratesticular lesion, a moderately avid 7 mm perivesical nodule and postsurgical uptake within the umbilical region from the excised umbilical lesion (Fig. 3). No abnormal uptake was seen in the small or large bowel.

The Gallium DOTATATE PET CT study demonstrated an additional site of intense uptake within the ileum which was considered the likely primary (Fig. 4) as well as nodal and peritoneal metastases.

A subsequent CT enterography for surgical planning and primary tumor evaluation highlighted an enhancing soft tissue lesion within the ileum (Fig. 5).

Prior to the surgery the patient was commenced on somatostatin analogues. The patient then underwent an elective right hemicolectomy with resection of the mesenteric nodes and peritoneal/spermatic cord deposits.

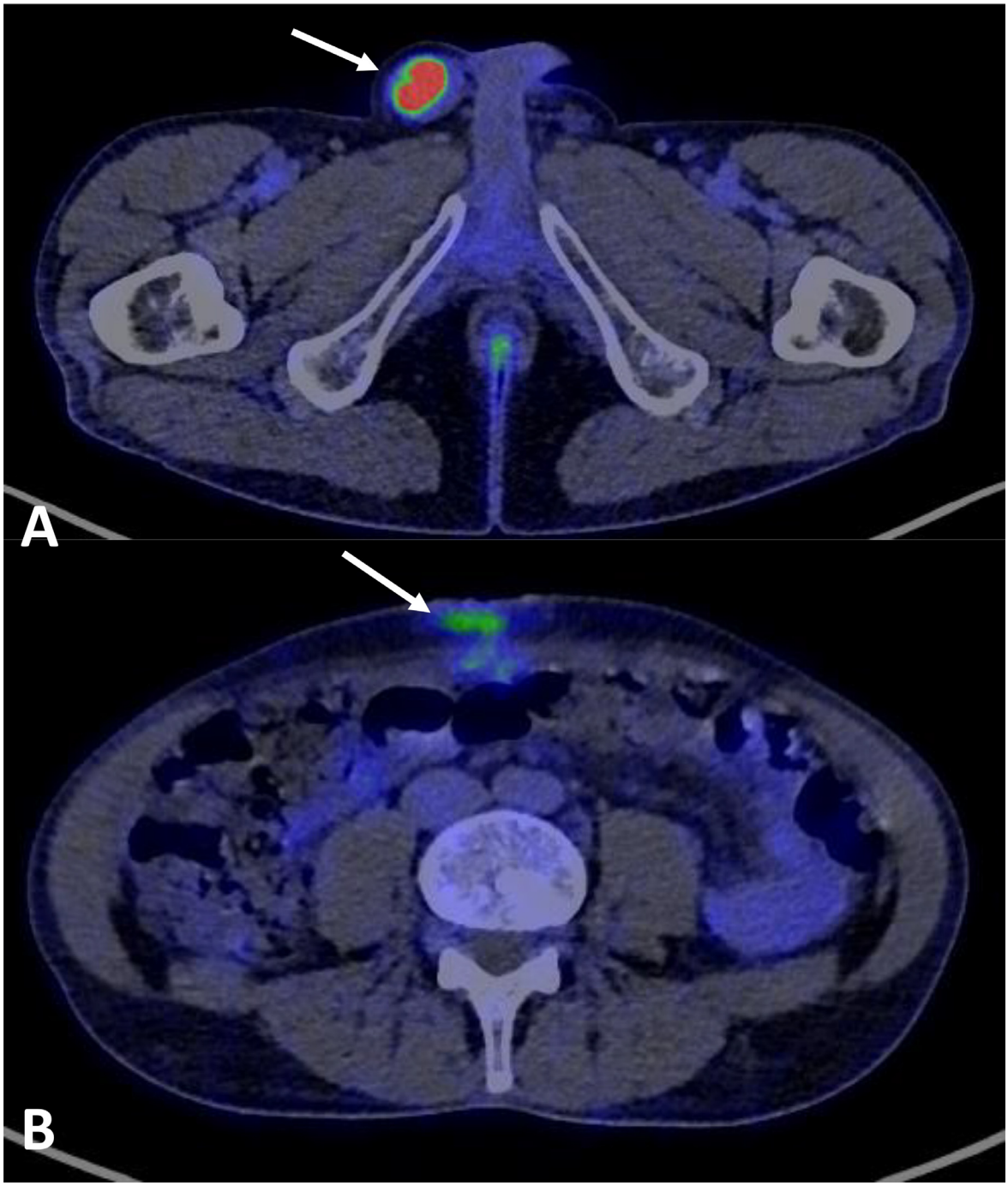

Fig. 3 – Axial reconstruction of FDG PET-CT. (A) Markedly increased tracer uptake within the right inguinal paratesticular soft tissue. (B) Mild to moderate tracer uptake in the soft tissues at the site of the excised umbilical lesion.

Discussion

Paratesticular lesions, in contrast to intratesticular lesions, are usually benign and cystic. Spermatic lipomas are the most frequent solid benign tumors [6] Malignant tumors are very rare, with rhabdomyosarcoma being the most common primary tumor. Metastases usually originate from testicular, prostate, renal and gastrointestinal cancers [3]. Despite the low prevalence of malignant paratesticular lesions, a high index of suspicion must be must be maintained in patients with a background of a primary malignancy.

To our knowledge, only a few cases of gastrointestinal NET have been reported to metastasize to the scrotum or inguinal lymph nodes [7–9]. In one described case report ultrasound revealed a hypoechoic mass surrounding the testis with extension in the spermatic cord [9]. The proposed mechanisms of dissemination include lymphatic spread, retrograde exten-

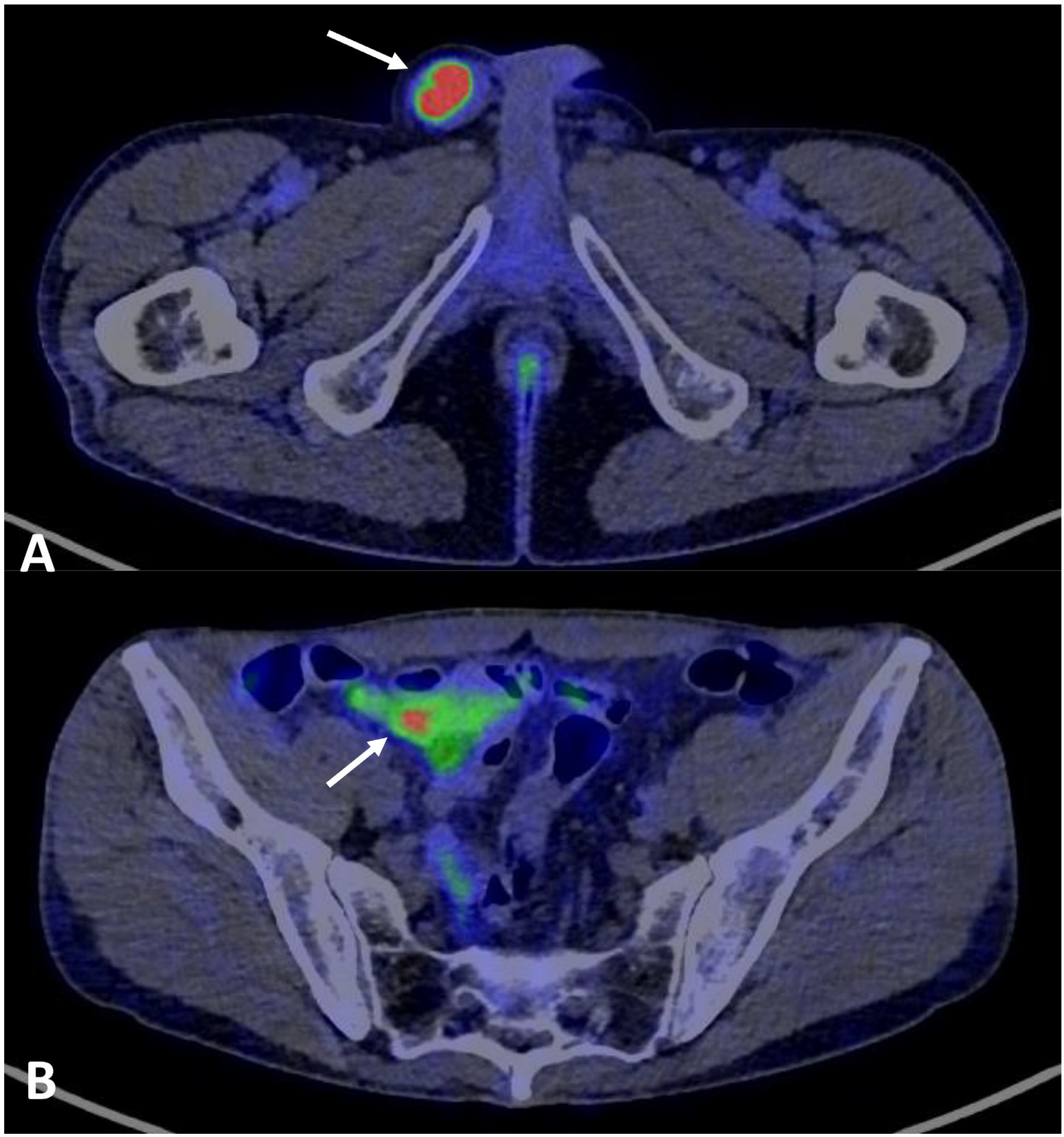

Fig. 4 – Axial reconstruction of Ga-Dotatate PET-CT. (A) Markedly increased tracer uptake within the right inguinal paratesticular soft tissue. (B) New increased focal uptake within the terminal ileum at the site of the primary small bowel neuroendocrine tumor which was not seen on the FDG PET-CT.

sion via the vas deferens or retrograde venous embolism. In cases involving peritoneal metastasis, a patent processus vaginalis emerges as a plausible conduit for metastatic spread.

Ultrasound imaging can be a useful initial modality as it can differentiate extratesticular masses from intratesticular masses and benign cystic lesions from solid lesions. However, further definitive characterization is usually limited in solid lesions, the exception being a well-defined echogenic solid mass which would favor a spermatic cord lipoma. Hence additional imaging is often required for further characterization [3].

In our case, the patient had an undescended testis on the ipsilateral side to the index paratesticular lesions, hence the concern initially was that the patient may have developed a

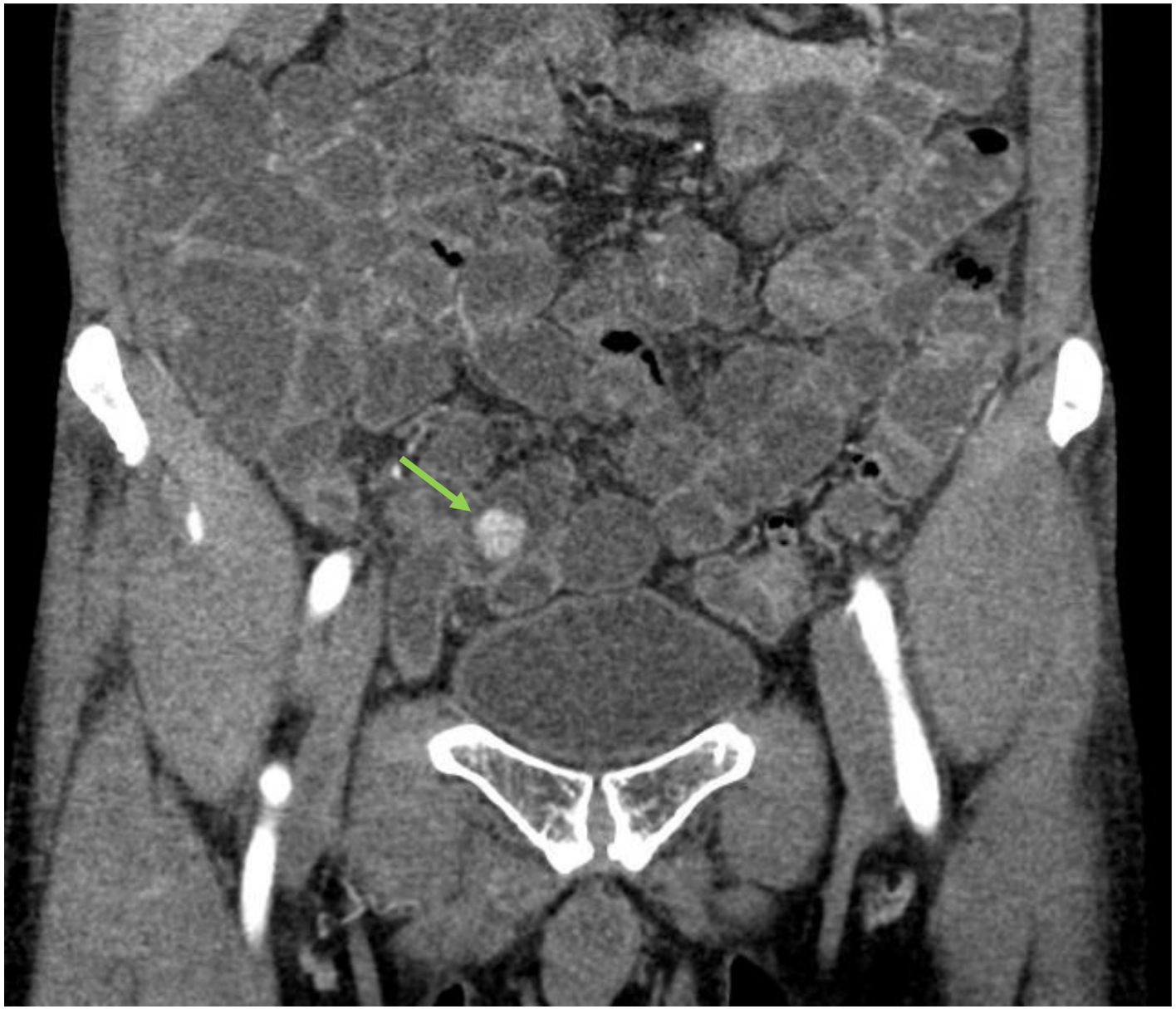

Fig. 5 – Coronal reconstruction of CT enterography illustrating an enhancing soft tissue lesion within the terminal ileum which was the primary neuroendocrine tumor origin.

primary malignant process on a background of undescended testis which is a well-recognized risk factor for carcinogenesis. However, the undescended testis was otherwise of normal sonographic morphology and this in combination with the pathological umbilical nodule, suggested a systemic process to the presentation.

The processus vaginalis represents a cul-de-sac evagination of the abdominal wall that evolves during embryological development and typically closes in early life. Persistence of the processus vaginalis correlates with various pathologies, including congenital indirect inguinal hernias, communicating hydroceles, and undescended testis, and has the potential to serve as a pathway for the dissemination of peritoneal disease [10]. It is therefore likely that transcoelomic disease spread via a patent processus vaginalis on the side of the undescended testes was the likely cause of initial presentation.

FDG PET CT has good clinical utility when attempting to establish a primary site in malignancy of unknown origin in addition to further distant disease. However, like in this case, the use of Gallium DOTATATE PET CT in neuroendocrine disease has been shown to have a greater sensitivity and diagnostic value in identifying a primary source and distant metastases, particularly well-differentiated tumors [11]. Gallium DOTATATE PET CT was able to demonstrate the primary site of disease in the terminal ileum in addition to nodal and peritoneal disease which had a positive impact in the patient's management and targeted treatment.

Conclusion

This case highlights an extremely rare presentation of midgut neuroendocrine tumor metastasis to the paratesticular tissue and inguinal canal as the initial clinical presentation with concomitant undescended testicle ipsilateral to the lesions. It is arguable that without cross-sectional imaging, one may have potentially attributed this to a testicular malignancy due to undescended testicle being a strong risk factor associated with carcinogenesis. It is therefore imperative for clinicians and radiologists to be aware that unusual paratesticular lesions may represent a communicating peritoneal disease process which therefore requires thorough further investigation to identify the primary malignancy.

Patient consent

Formal written consent from the patient has been obtained for the purpose of publishing this case report.

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