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Urinoma Secondary to Aggressive Mesenteric Desmoid

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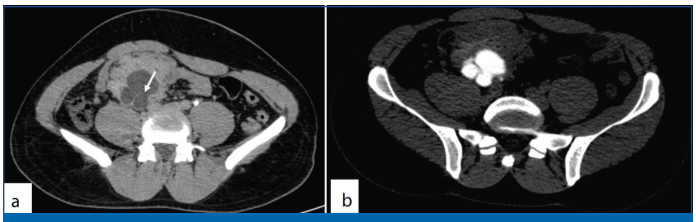
A 29-year-old male presented with right iliac fossa pain and vomiting for three days. Notably, there was no history of fever or alteration in bowel or bladder habits. There was no significant family history of malignancy or prior surgical history. On examination, a palpable mass with well-defined borders was noted in the right iliac fossa, measuring approximately 6×6 cm, and was tender on palpation.

At the time of admission, laboratory parameters were unremarkable. The patient underwent a Contrast-Enhanced Computed Tomography (CECT) scan of the abdomen for evaluation of the mass, which revealed an ill-defined, heterogeneously and progressively enhancing soft-tissue density lesion measuring 6.5×8.4×12.5 cm. The lesion involved the root of the small bowel mesentery and occupied the right iliac fossa [Table/Fig-1a] and umbilical region, extending from the L2 to L5 vertebral levels.

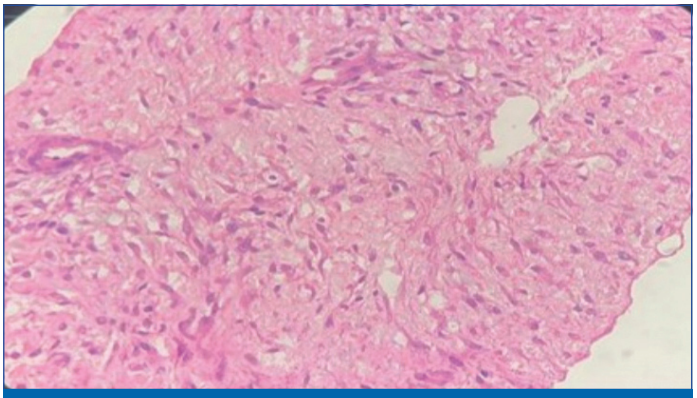
Loss of fat planes was observed between the lesion, the ileocecal junction, and the terminal ileum. The lesion was seen infiltrating the right distal ureter, causing moderate hydroureteronephrosis. Additionally, a focal discontinuity in the right ureteric wall was noted, with a fluid collection adjacent to the ureter within the mass. On delayed imaging, extravasation of contrast into the aforementioned collection was observed, suggestive of a urinoma [Table/Fig-1b]. The lesion also encased the superior mesenteric artery [Table/Fig-2] and caused near-complete luminal occlusion of the superior mesenteric vein.



[Table/Fig-1]: (a&b) Axial sections of the venous (a) and delayed (b) phase of CECT abdomen showing an ill-defined, heterogeneous enhancing soft-tissue density mass lesion in the right iliac fossa. The lesion is seen to cause a desmoplastic reaction in the form of the tethering of the adjacent small bowel loops and the right proximal ureter. At the level of the upper border of the L5 vertebra, there is a focal discontinuity in the anterior wall of the ureter(arrow). (b) The contrast was seen to extravasate into the collection within the tumour, suggestive of urinoma.

Based on the imaging features, the mass was suspected to be an invasive mesenteric desmoid tumour or, less likely, an inflammatory pseudotumour of the mesentery. The patient underwent an ultrasound (USG)-guided biopsy of the lesion, followed by USG-guided right percutaneous nephrostomy and pigtail drainage of the urinoma. Histopathological analysis revealed spindle cells [Table/Fig-3] in the linear core tissue samples, with focal positivity for SMA and negativity for β-catenin. The percutaneous nephrostomy tube and pigtail catheter were subsequently removed, and a right DJ stent was placed. On follow-up after six months of sorafenib therapy, there was a reduction in the lesion size to 4.5×6.9×6.8 cm. The size and extent remained stable on subsequent follow-up.

[Table/Fig-2]: Sagittal reformatted sections of the arterial phase of CECT abdomen showing complete encasement (arrow) of the superior mesenteric artery (from the lower border of the L2 vertebra to the upper border of the L3 vertebra) by the lesion causing significant luminal narrowing.



[Table/Fig-3]: Photomicrographs of hematoxylin and eosin stained biopsy specimen at 100x magnification showing oval to spindleshaped cells with abundant cytoplasm amongst dense collagen.

Desmoid tumours represent less than 3% of all mesenchymal tumours and can arise in any musculoaponeurotic structure. Musculoaponeurotic fibromatosis is classified into extra-abdominal, intra-abdominal, and abdominal fibromatosis [1]. The typical age range for the occurrence of this abdominal mesenchymal tumour is 15 to 60 years, with a peak incidence between 30 and 40 years, and a female predilection attributed to hormonal influence [2]. An increased incidence is observed in young patients, particularly those with a history of familial adenomatous polyposis, who tend to develop desmoid tumours at surgical sites [3].

A wide spectrum of clinical presentations is observed in desmoid tumours, ranging from indolent and stable to rapidly progressive forms. Bowel involvement may present as obstruction [4], fistula formation, or perforation. Involvement of the excretory system typically manifests as hydroureteronephrosis and, in rare casessuch as in the present case can lead to urinoma formation.

A urinoma is a collection of extravasated urine resulting from disruption of the urinary collecting system, most commonly secondary to trauma

or iatrogenic injury. Non-traumatic ureteric rupture due to obstruction is primarily caused by calculi and, in rare cases, by tumours. The most common malignancies associated with urinoma formation include prostate, cervical, and infiltrative bladder cancers [5].

Desmoid tumours are characteristically positive for nuclear β-catenin, vimentin, and actin, and negative for desmin, S-100, and c-KIT [2]. However, the absence of β-catenin positivity does not exclude the diagnosis [6]. Other differential diagnosis for mesenteric masses include inflammatory pseudotumour, metastatic deposits, gastrointestinal stromal tumour (GIST), and sclerosing mesenteritis. These were considered less likely based on the specific imaging findings in this case. Inflammatory pseudotumour was considered a possible differential diagnosis. It most commonly affects the lungs of children and young adults. However, the absence of central calcifications and necrosis, along with its relatively rare occurrence in the mesentery, makes this diagnosis less likely in our case [7].

This case illustrates a rare presentation of an aggressive mesenteric desmoid tumour in a young male, complicated by spontaneous urinoma formation resulting from ureteric infiltration. Histopathological analysis was crucial in refining the differential diagnosis and confirming the fibroblastic characteristics of the tumour, thereby excluding possibilities such as GIST or inflammatory pseudotumour. Treatment with sorafenib led to a reduction in tumour size and maintained disease stability, highlighting its role as an effective non-surgical therapeutic option when complete resection poses significant risk.

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