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CASE REPORT

Malignant Duodenal GIST in a Patient with Situs Inversus Totalis—a Rare Association and Brief Review of Literature

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Introduction

Gastrointestinal stromal tumors are mesenchymal tumors arising from neoplastic transformation of interstitial cells of cajal. Mutation in the KIT proto-oncogene has been widely linked with the pathogenesis of GIST. The normal arrangement of the human abdominal and chest organs is termed as situs solitus. When these organs are arranged in the exact mirror image of their normal anatomical positions, it is termed as situs inversus. If the heart is swapped to the right side of the thorax, it is known as situs inversus totalis. To the best of our knowledge till date, there are no case reports or published evidence about the association of situs inversus totalis with GIST of duodenum. Ours would be the first case of a duodenal GIST presenting in a patient with situs inversus totalis.

Case Report

A 50-year-old female presented to us with complaints of abdominal pain for 1 month. The pain was diffuse, dull aching, epigatric, and non-radiating. There was no history of vomiting, weight loss, or altered bowel habitus. She had no significant past medical or surgical history. On examination, there was no mass palpable in the abdomen. Baseline blood investigations were normal. Chest x-ray revealed dextrocardia, and upper GI endoscopy was normal. CT abdomen imaging was done which revealed situs inversus totalis and a large hypodense exophytic mass of size 9.5 × 6 × 7.2 cm from the second and third part of the duodenum. This was displacing the ascending colon antero laterally. There were no other masses seen elsewhere. [Figs. 1, 2, 3] After pre operative work up, patient was taken up for diagnostic laparoscopy.

Intraoperatively, there was a huge mass arising from the second and third part duodenum adherent to the ascending colon and Gerota's fascia. Due to large size of tumor and dense adhesions, the procedure was converted to open laparotomy. A sleeve resection of the duodenal mass was done along with a gastrojejunostomy (Figs. 4, 5). An appendectomy was also done to prevent future diagnostic dilemmas in view of situs inversus.

Histopathological exam revealed a tumor of size 10 × 10 × 7 cm weighing 220 g composed of spindle-shaped cells with high Mitotic rate(>5/50 HPF). IHC markers were positive for CD117 and CD34, Tumor was G 2 and resection margins were free. It was a T3 N0 M0 high-grade malignant GIST. Postoperatively, the patient tolerated orals by day 3. She had an uneventful recovery and was discharged by day 6. In view of her high-grade tumor, she was started on imatinib therapy. After 8 months of follow-up, she is doing fine with no tumor recurrences.

Discussion

The only paper till date describing the incidence of situs inversus, by Mayo et al., states that the disease has very low incidence in the range of 1:10,000 to 1:20,000 [1]. Few studies have suggested an autosomal recessive pattern of inheritance along with genomic mutation in DNAH11 gene as the causative factor.

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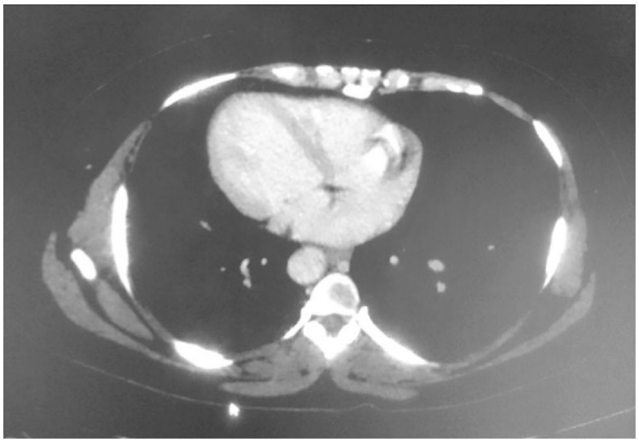

Fig. 1 CT imaging showing dextrocardia

There is high level of association of situs inversus with cardiac anomalies and ciliary dysmotility syndrome especially Kartagner syndrome [2, 3]. However, our patient did not have any of the abovementioned congenital diseases.

Surgical candidates with situs inversus have always posed an intriguing surgical challenge for even the most veteran surgeons. Due to the mirror image anatomy, operating on these patients can be tricky and at times dangerous due to unfamiliar anatomy. A thorough knowledge of surgical anatomy is required for safe surgical outcome in such scenarios.

Laparoscopy would be an ideal approach for these patients, but in our patient, due to the large size of the tumor and dense adhesions, we had to convert to an open procedure intraoperatively.

Many innovative techniques such as operating with a mirror, altering the port placement, have been introduced to tackle the anatomical difficulty.

In a previous case report published by our surgical team, we were exposed to such challenges when operating on a situs inversus patient with cholecystitis [4].

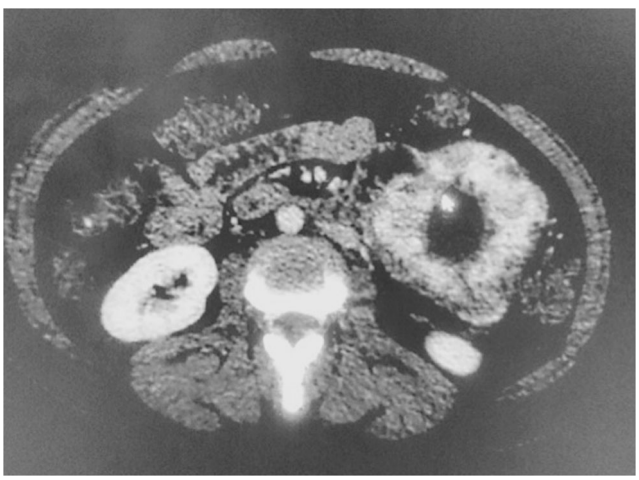

Fig. 2 CT imaging axial view showing tumor arising from the duodenum

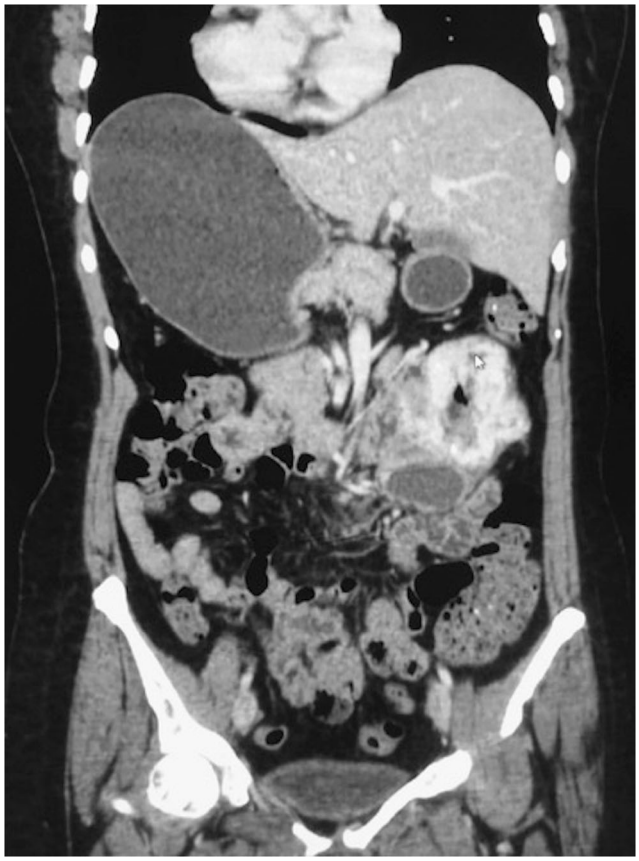

Fig. 3 CT imaging coronal view showing situs inversus totalis, the stomach and liver on the right with the duodenum and the GIST mass on left

Common hurdles that a clinician would face in such patients can be subdivided as follows:

(a) Wrong preoperative diagnosis
(b) Inappropriate surgical incision
(c) False localization of pain can be misleading

To prevent such problems, a thorough physical examination along with careful interpretation of basic investigations like chest x-ray and ECG is required [5].

CT Imaging of the abdomen and thorax is the gold standard for diagnosis. Our patient had a high-grade GIST; hence, surgical resection followed by imatinib adjuvant therapy was given. After extensive online literature search, we concluded that ours was probably the first case that reported about a duodenal GIST presenting in a situs inversus totalis patient. This prompted us to discuss our case and bring to light certain technical aspects of surgery in such patients.

Since this was our initial experience in handling such a complex case, we took certain measure in the work up to the surgery for the patient. Situs inversus patients usually have coexistent cardiac anomalies; hence, a thorough cardiologist

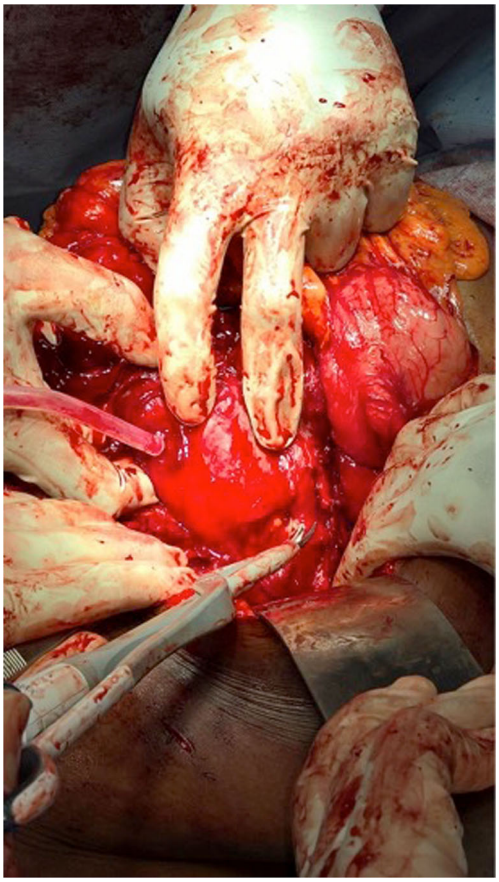

Fig. 4 Releasing the posterior adhesions between the mass and gerotas fascia

examination with a 2D ECHO was done to rule out these. Preoperatively, a virtual 3D model reconstruction was done with the help of a radiologist's help to plan surgical approach and to help the operating surgeon to get familiarized to the altered anatomy.

Appropriate time needs to spend on certain technical details preoperatively like rearranging all the surgical and anesthetic equipment in the operation room. We realized that this altered anatomy best suits a left-handed surgeon. However, since none of our surgeons were left handed, we did experience difficulty when performing some fine precise movements in laparoscopy. Several reports in the literature emphasize the feasibility of the safe laparoscopic cholecystectomy in this challenging situation but no report describes any technique for a duodenal GIST excision. We would like to advise a diagnostic laparoscopy as a first approach for all such patients after extensive preoperative imaging and anatomy reconstruction. As a build-up to the week before the surgery, strengthening the nondominant left hand by doing certain routine activities with the left hand can be a small trick which can help immensely intraoperatively especially in performing laparoscopically fine movements. Another key aspect to the patient work is extensive patient counseling regarding the difficulty of surgery and the possibility for conversion to open technique. All these issues must be discussed preoperatively with the patient and the family.

The use of laparoscopy in scenarios other than cholecystectomy in these patients is still not well established. Innovative techniques and more literature publication of these ideas are required in this field to help surgeons worldwide tackle this surgical complexity.

Conclusion

We hope that our case widens the existing research and knowledge in the field of surgically curable diseases in situs inversus patients. Situs inversus and other congenital anatomical variations can pose a tough challenge to the surgeon. Good

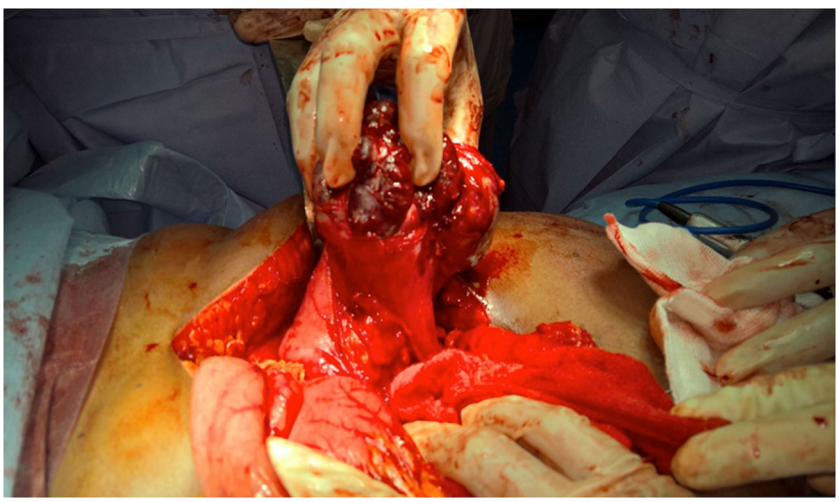

Fig. 5 Mass completely mobilized, ready for sleeve resection

knowledge of surgical anatomy, accurate preoperative imaging, and careful planning can help in tackling these cases.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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