

Jejunal Gastrointestinal Stromal Tumor Associated with Synchronous Periampullary Adenocarcinoma – A Case Report

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Abstract Gastrointestinal stromal tumors (GIST) are rare mesenchymal neoplasms of the gastrointestinal tract with a malignant potential and unpredictable behavior. In the literature very few cases of synchronous development of a GIST with other neoplasia of different incidence, etiology, evolution and prognosis have been described.

Keywords: *gastrointestinal stromal tumors, pancreatic adenocarcinoma, synchronous tumors*

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1. Introduction

Gastrointestinal stromal tumors (GIST) are rare mesenchymal neoplasms of the gastrointestinal tract with an incidence of 1.5 / 100,000 / year, typically described in adults, with a peak incidence in the sixth and seventh decades.

Gastrointestinal stromal tumors comprise a recently defined entity of the most common mesenchymal tumors of the gastrointestinal (GI) tract. In the 1940s, Stout et al. first described stromal tumors arising from the smooth muscle of the GI tract which are known as leiomyoma, leiomyosarcoma, leiomyoblastoma and bizarre leiomyoma. In the late 1960s, with the use of electron microscopy and in the early 1980s with the introduction of immunohistochemistry, the Schwannian differentiation was identified in some neoplasms of this kind. On basis of these findings in 1983 Mazur and Clark introduced more generic term 'stromal tumor' [1].

It is believed that these tumours arise from interstitial cells of Cajal (ICCs) or their precursors that regulate gut motility. This presumption is related to the observation that both GISTs and ICCs express tyrosine kinase receptor kit.

In the era of targeted therapy GISTs are defined as C-kit (CD117) positive mesenchymal spindle, epithelioid or mixed-type cell neoplasms that may originate from the wall of esophagus to anal canal. GISTs strongly express (90-95%) the C-kit (CD117) protein, a type III tyrosine kinase receptor encoded by C-kit protooncogene. Approximately 75% of GISTs are positive for CD34, 32-40% are positive for smooth muscle actin (SMA), 8% for

S-100 protein and < 5% for desmin. As mesenchymal tumors, GISTs also show strong positivity for vimentin, which is in accordance with their origin from the ICCs that present a C-kit +, CD34 +, Vim + immunophenotype [2]. They account for approximately 0.3%-3.3% of all primary GI neoplasms and for 6%-12% of all sarcomas. GISTs typically occur in older individuals and the majority of them (55-71%) comprise primary neoplasms of the stomach, followed by small intestine (21-26%), colon and rectum (4%), and esophagus (< 4%).

Benign tumors much more common than the malignant, however, at present every case of GIST is considered as potentially malignant. High mitotic index and larger tumor size are associated with risk of malignancy [3,4].

Gastrointestinal stromal tumors (GISTs) are an uncommon mesenchymal tumor affecting GI tract. The synchronous occurrence of mesenchymal tumors with other primary gastrointestinal malignancies is quite rare. Even with extensive search we found only few case series related to it. Here we are reporting first case of periampullary adenocarcinoma in association with synchronous jejunal GIST.

2. Case Report

An 50 years old female with known hypertensive was admitted to our hospital with complaints of passing black, tarry, foul smelling and sticky stool with associated history of easy fatigability and weight loss of about 20 kgs during the last three months. No history of any fresh bleeding. History of multiple blood transfusion present. On physical examination pallor was present and other laboratory tests were unremarkable. Esophago gastroscopy

revealed an ulcero proliferative mass in the second part of duodenum to third part of it, scope going easily beyond growth. Pathology report of the endoscopic biopsies revealed a well differentiated intestinal type gastric adenocarcinoma. Computed tomography (CT) revealed a discrete mass 7.8 x 4.2 x 4.9 cm seen along lateral wall of 2nd and 3rd part of duodenum projecting intraluminally and causing its obliteration and pancreas was atrophic, with no evidence of distant metastasis. (Figure 1) PET-CT showed heterogeneously enhancing asymmetrical mural thickening in second and third part of duodenum (SUV-5.9) with no evidence of any distant metastasis. (Figure 2) The patient underwent classical pancreaticoduodenectomy. During laparotomy a hard nodular bulky tumor 8 x 10 cm found in head of pancreas which was infiltrating medial wall of D2 and extending to the body of pancreas with a second nodule was palpated of 1.5 x 1.5 cm over proximal part of jejunum. (Figure 3, Figure 4) Pathology examination confirmed the presence of a well differentiated papillary type of adenocarcinoma (Figure 5) and jejunal tumour was a very low grade GIST with no mitosis (Figure 6) and strongly positive for C-kit immunostain. (Figure 7) Patient had smooth postoperative recovery. She was regularly followed and had no recurrences after 3 years.

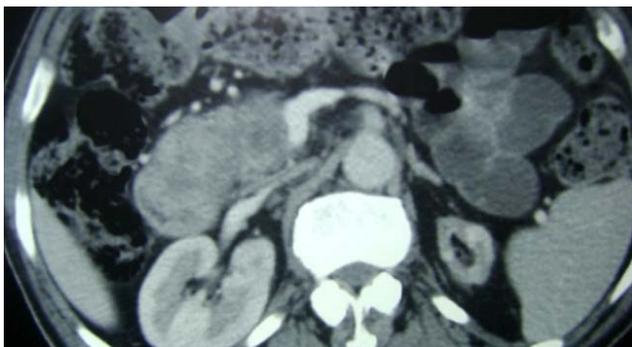


Figure 1. Computed tomography revealed a discrete mass 7.5 x 5.2 x 3.9 cm seen along lateral wall of 2nd and 3rd part of duodenum projecting intraluminally and causing its obliteration

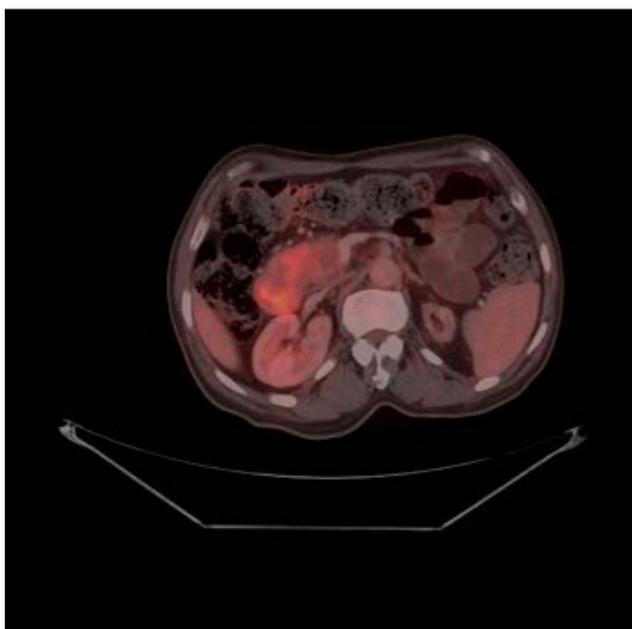


Figure 2. PET-CT showed heterogeneously enhancing asymmetrical mural thickening in second and third part of duodenum (SUV-5.9)

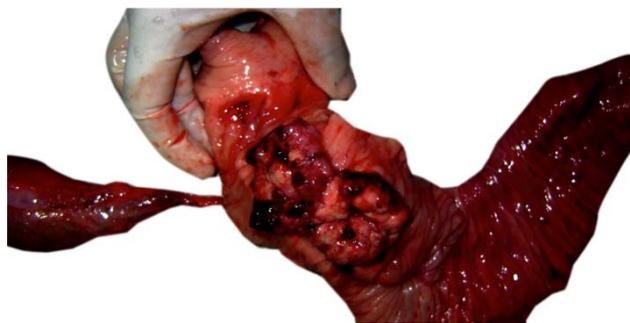


Figure 3. Intraoperative picture of hard nodular bulky tumor 8 x 10 cm found in head of pancreas which was infiltrating medial wall of D2 and extending to the body of pancreas

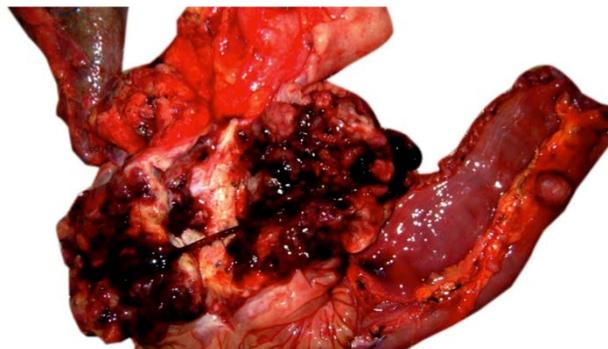


Figure 4. Second nodule was palpated of 1.5 x 1.5 cm over proximal part of jejunum

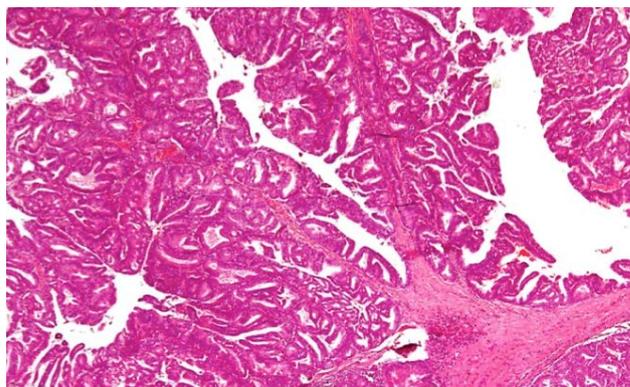


Figure 5. Well differentiated papillary type of adenocarcinoma on H & E staining at magnification of 400X

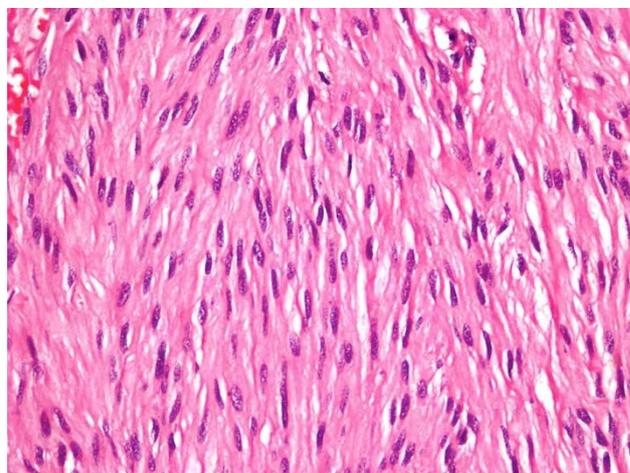


Figure 6. Very low grade GIST with no mitosis on H & E staining at magnification of 400X. (Jejunal Tumour)

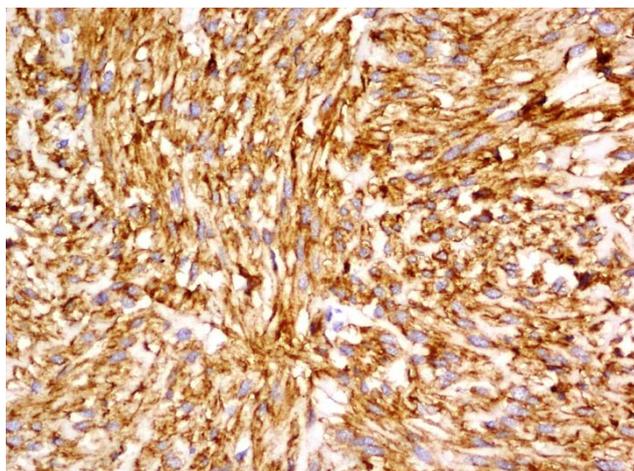


Figure 7. Jejunal GIST showing C-kit positivity

3. Discussion

GIST and adenocarcinomas represent distinct oncogenic entities. GISTs are the most common mesenchymal tumors of the GI tract and this group of tumors represents about 0.3% to 3.3% of all GI neoplasms. Synchronous occurrence of a gastrointestinal stromal tumor with a tumor of different origin is rare and very few such associations are reported in literature so far. Slightly above 32 cases of the synchronous occurrence of mesenchymal tumors (including GIST) and other gastrointestinal malignancy have been reported in the literature.

The largest published series by Maiorana et al. 11.5% of gastric GISTs (6 cases gastric adenocarcinoma in 5 cases and carcinoid in 1) were associated with other gastrointestinal malignancies [5].

In a recent series (4 cases) published by Wronski et al. also showed that GIST as most commonly associated synchronous tumour related with adenocarcinoma [6]. But in a study by Liu et al (54 cases of incidental GIST), the most common tumors associated with GIST was esophageal squamous cell carcinomas (1.13%), followed by gastric (0.53%), pancreatic (0.38%) and colorectal (0.03%) adenocarcinomas [7]. In most of the reported cases of synchronous gastric adenocarcinoma and GIST, the preoperative biopsy reported as adenocarcinoma. GISTs are usually seen incidently following laparotomy for some other reason. In our case classical pancreaticoduodenectomy was done for periampullary adenocarcinoma and small GIST of very low risk was found incidentally on proximal jejunum which was confirmed on histopathological examination.

The synchronous occurrence of GIST and other abdominal malignancy seems to be just a coincidence, the development of these tumors may involve common carcinogenic agents. Various hypotheses have been proposed regarding the simultaneous occurrence of GIST and adenocarcinoma [8].

In study by Maiorana et al suggests that coincidence alone could easily account for such association, particularly in countries that exhibit high incidence rates of gastric adenocarcinomas, such as Japan, northern Italy. On the other side Liu et al. reported that most common epithelial tumor associated with GIST were esophageal

squamous cell carcinoma which may be again because this study was done in Sichuan Province is a key area with highest esophageal cancer.

Another interesting hypothesis is that a single carcinogenic agent might interact with two neighboring tissues, inducing the development of tumors of different histiotypes. Sugimura et al. N-methyl-N'-nitro-N-nitrosoguanidine induces the development of gastric adenocarcinomas after oral administration in rats. However, when the same compound is combined with agents that alter the gastric mucosal barrier, such as aspirin or stress, leiomyosarcomas develop in conjunction with epithelial tumors. Equally compelling, although also experimental, are reports on the induction of gastric tumors in rats after 9, 10-dimethyl-1, 2-benzanthracene (DMBA) injection. Whereas administration of DMBA alone induces the development of adenocarcinomas, treatment with DMBA and cellophane plate causes mainly the induction of gastric sarcomas [9,10].

GISTs that occur simultaneously with other neoplasms were usually smaller and of low or very low risk of malignancy compared to a single GIST and this finding is in accordance with the literature [11]. In our case we also found a very low risk GIST in jejunum while operating a case of periampullary adenocarcinoma.

The problem of the coexistence of GISTs with other neoplasms is important from an oncological, surgical and histopathological point of view. As many GISTs are clinically silent and detected incidentally during surgery for a different reason, it is essential that a thorough examination during surgery must be performed. Another problem arising from the incidental discovery of GISTs is that they can be mistaken for a metastasis of the coexistent primary tumor, thus precluding its staging.

4. Conclusion

The synchronous occurrence of GISTs and other gastrointestinal malignancies particularly with adenocarcinomas is more common than it has been considered. The concomitant GIST is usually discovered incidentally during the operation done for other reasons. Surgeons are advised to be alert against possible primary GIST accompanying other tumors and if found it must be resected as it has malignant potential.

Competing Interests

None.

Funding

None.

Ethical Approval

Written informed consent to publication was obtained from the patient or next of kin.

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