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# ROBOTIC ENUCLEATION OF A SOLID PSEUDOPAPILLARY NEOPLASM IN A 9-YEAR-OLD GIRL

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**Abstract – *Objective:*** *Here we reported the case of a 9-year-old girl with no relevant past medical or surgical history. Solid pseudopapillary neoplasms (SPN) is a rare pancreatic tumor, which is often observed in females in their second or third decades, but it can also be found in pediatric age. It is considered a low-grade malignant epithelial neoplasm with low metastatic rate and surgical resection represents the treatment of choice with an excellent long-term prognosis.*

***Case report:*** *A 9-year-old girl presented at our Emergency Department with a three days history of vomiting and vague abdominal pain. US showed an epi-mesogastric solid mass of 8 cm, diagnosis confirmed by the CT scan. A US-guided Tru-cut biopsy was performed obtaining the histologic diagnosis of Solid pseudopapillary neoplasm (Gruber-Frantz tumor). A Robotic enucleation of the neoplasm was performed, and histopathology confirmed an SPN with complete resection. In the case of low-malignant neoplasms like SPNs, enucleation, when feasible, represents the best surgical approach. Robotic procedures offer some technical and oncological advantages over minimally invasive techniques due to the stability of the operative field, the 3D and magnified vision and the articulated robotic arms.*

***Conclusions:*** *Robotic enucleation of solid pseudopapillary pancreatic tumors, when feasible, represents an excellent minimally invasive technique with a favorable long-term prognosis.*

**KEYWORDS:** *Gruber-Frantz tumor, Pancreatic tumor, Tru-cut biopsy, Robotic surgery, Tumor enucleation, Solid pseudopapillary tumor.*

## INTRODUCTION

Solid pancreatic pseudopapillary tumor, also known as Gruber Frantz tumor, is a rare epithelial pancreatic neoplasm, representing less than 3% (0.2 to 2.7 %) of pancreatic tumors [1] . It typically occurs in young females, around the second or third decade, but it can also affect children in 20-25% of cases [2]. The most frequent localizations are the tail and the head of the pancreas[2]. It is a low-grade malignant neoplasm that rarely metastasizes, in less than 15% of the cases[3]. The treatment of choice is surgical, and the approach depends on the localization and the eventual infiltration of surrounding organs. The prognosis is excellent in most cases with a 10-year survival rate approaching 100 %[1].

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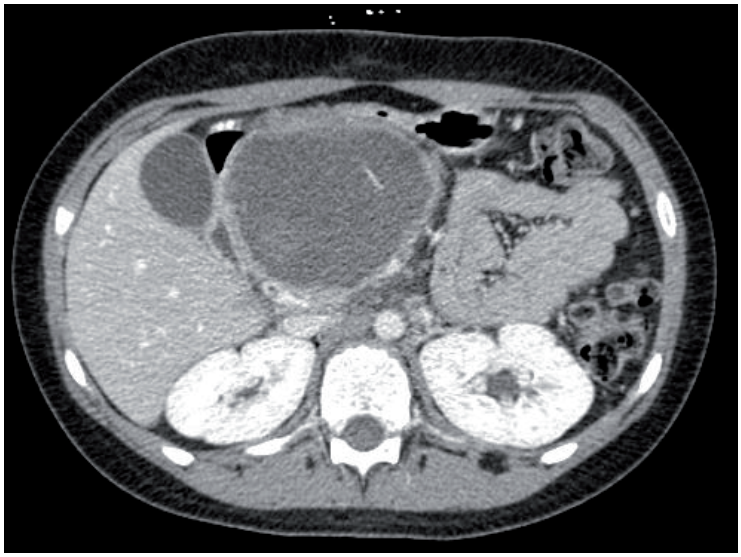

**Fig. 1.** Portal phase CECT scan: large mass in the cephalic portion of pancreas. The lesion has a cystic aspect - linear calcification, intralesional blood and capsular enhancement; it displaces and compresses vascular structures and there is no invasion.

## CASE-REPORT

A 9-year-old girl, with no relevant past medical or surgical history, presented to a nearby Emergency Department with a three days history of vomiting and vague abdominal pain. The patient underwent an abdominal US that showed an epi-mesogastric solid mass of 78x73x63 cm. The patient was transferred to our Department and underwent a CT scan. The CT showed a voluminous heterologous lesion in pancreas cephalic portion with irregular margins; the lesion contained corpuscular fluid, thin septa, linear calcifications and hyperdense areas (blood content) and it showed peripheral capsular enhancement, after iodinate contrast was injected (Figure 1). This neoplasm compressed some near vascular structures, such as spleno-mesenteric confluence, portal vein, superior mesenteric vein and anteriorly displaced the gastroduodenal artery, coronary vein and pyloric vein. It also impinged upon other structures like antral-pyloric region, first duodenal portion and gallbladder wall (Figure 2).

**Fig. 2.** MPR reconstruction of CT scan: in this image the relationships between the lesion and the surrounding anatomical structures can be well evaluated.

To obtain a histopathologic diagnosis, the patient underwent an Ultrasound-guided percutaneous biopsy, performed under anesthesia by the interventional radiologist who biopsied the known mass localized in the right abdominal median-paramedian area. Multiple samples of tissue were placed in special containers in formalin to perform histopathologic analysis. Histopathology was suggestive of Solid Pseudopapillary Neoplasm or SPN (Gruber-Frantz's tumor).

A few days later the patient underwent elective robotic assisted enucleation of the neoplasm. The Da Vinci XI Surgical System (Intuitive Surgical, Sunnyvale, CA, USA) was used. The patient received prophylactic antibiotic therapy to prevent site infection. The patient was in supine position, the robotic cart was located on the patient's right side and the assistant on the left side and the trocars' ports were positioned as shown in Figure 3.

Tissue dissection and tumor's enucleation were achieved by robotic bipolar forceps and monopolar scissors (Figure 4), paying attention to respect the main pancreatic duct and the main abdominal vessels. The fourth robotic arm allowed a stable retraction of the structures. A small tear from the pancreatic parenchyma was repaired with robotic hand-sewn stiches. The mass was removed through a Pfannenstiel incision, allowing a good cosmetic result. Two drainage tubes were positioned.

Post-operative course was uneventful with no significant raise in pancreatic amylase and lipase.

12 days after surgery, the patient underwent an US study, that did not show any lesions or tumor recurrence or abdominal effusion.

Seven months after surgery, a MRI was performed. It demonstrated a significant decrease in pancreatic volume, in the body-tail region, while the main pancreatic duct and secondary ducts were

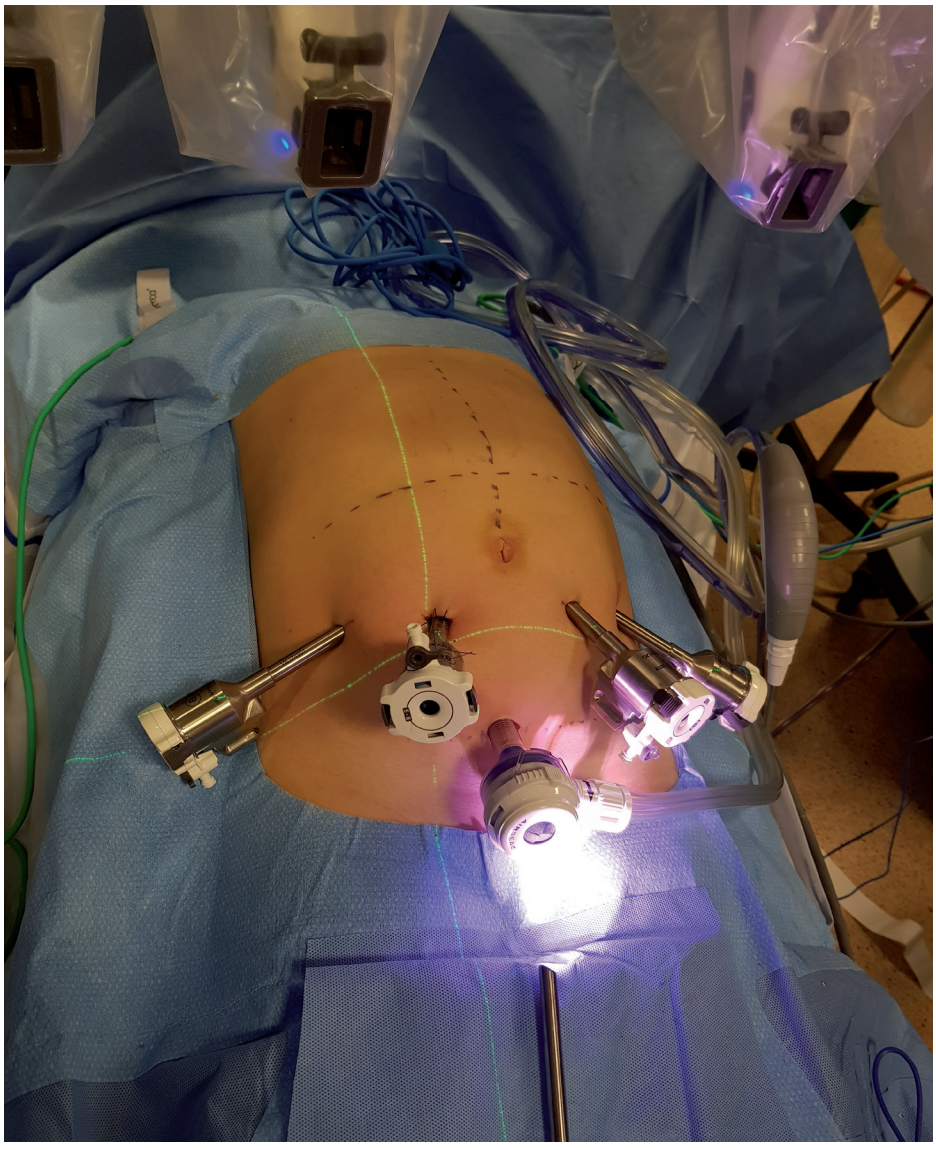

**Fig. 3.** Robotic trocars' ports position.

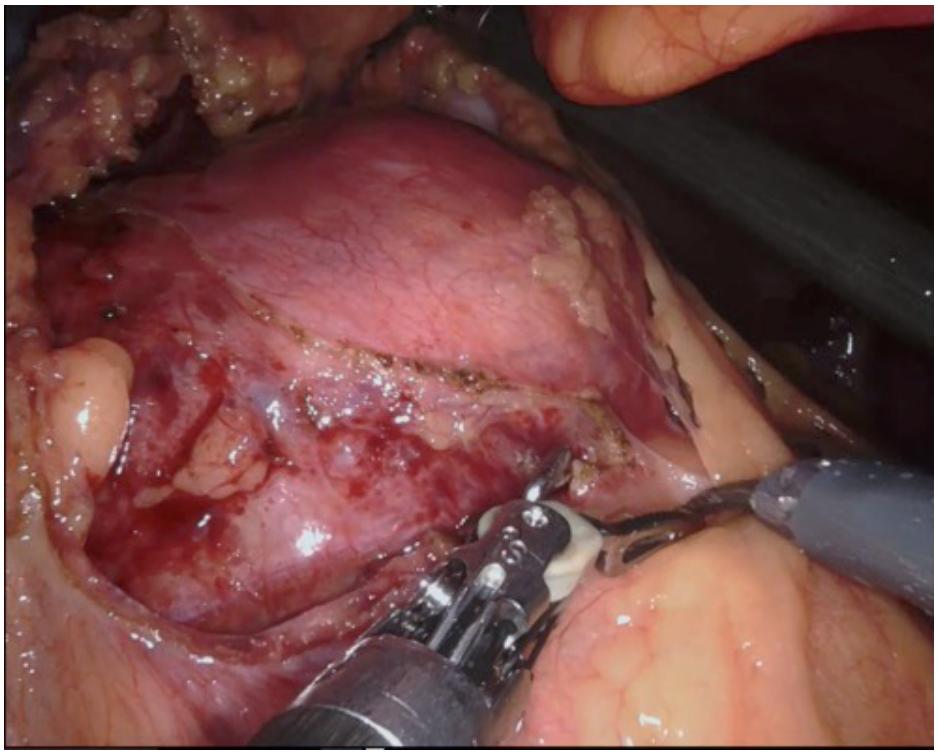

**Fig. 4.** Dissection of pancreatic mass with bipolar forceps and monopolar scissors.

mildly dilatated. There was no sign of tumor recurrence in the surgical site and the main vascular and parenchymal structures were not compressed.

After 9 months of follow-up neither signs of tumor recurrence neither endocrine nor exocrine pancreatic insufficiency are present.

## DISCUSSION

Solid pseudopapillary tumor of the pancreas is extremely rare, accounting for less than 1% of pancreatic neoplasms[1,2]. It occurs most often in females in their second or third decades, but it can also be found in pediatric age[1,2,4].

It is a low-grade malignant tumor with a low metastatic rate that tends to grow slowly, and the majority of patients are asymptomatic. When symptomatic, the most commonly reported complaint is diffuse nonspecific abdominal pain. Vomiting appears as a mass effect of the tumor[1,2,4,5].

There are no specific tumor markers for this entity and diagnosis relies mostly on imaging[6].

CT and MR imaging, in the appropriate clinical context, are able to provide a differential diagnosis of SPN. In particular, in young women and children, the presence of a well-defined mixed solid-cystic pancreatic mass, which presents a well-defined capsule and, sometimes, could show intratumoral hemorrhage or peripheral calcifications, should raise the suspicion of SPN [7,8]; after contrast injection, this kind of neoplasm presents early enhancement ("hemangioma-like") and the capsule and solid portion of SPN often enhance like the normal pancreatic parenchyma, in both arterial and venous phases [9]. Furthermore, it is important to remember that SPN will typically displace adjacent structures rather than invading them[10].

These features allow differential diagnosis with pancreatic adenocarcinoma (which appears hypoattenuated in the venous phase), neuroendocrine tumors (hyperattenuated in arterial phase) and non-hyperfunctioning islet cell tumors, that are different from SPN for the margins morphology (oval/round or smoothly lobulated in 95% of SPN *vs.* focal/eccentric lobulation in 50% of carcinoma), and for the tumor capsule morphology (complete encapsulation or no visible capsule in 94% of SPN *vs.* focal capsule discontinuity in 58% of carcinoma)[11].

Once the diagnosis is suggested by imaging, histological identification can be done by Tru-cut biopsy: usually this procedure is not performed before surgery, but it is very useful to make the right diagnosis and choose the right surgical technique. Ultrasound-guided Tru-cut needle biopsy is a well-tolerated and reliable procedure for providing a tissue diagnosis of malignancy before definitive treatment. It is performed with needle sizes from 14G to 18G, in our case with a 18G needle.

The mainstay of therapy is surgical resection with an improved prognosis and overall survival rate of more than 95% [4,5,12]. Radical resection is generally performed, but pancreatic parenchyma sparing operations are an excellent option if a complete curative resection is achievable [4,5].

Minimally invasive approaches are reported to be feasible, safe and effective[13-17]. Moreover, robotic surgery seems to be feasible and at least equal if not superior to conventional laparoscopic surgery for pancreatic resections[13,18]. Many are the advantages of robotic surgery, such as the stability of the operative field, the 3D and magnified vision and the articulated robotic arms that allow an accurate dissection.

## CONCLUSIONS

Solid pseudopapillary neoplasm is a rare pancreatic tumor with a long-term survival rate after surgical resection[1,2]. Pre-operative imaging represents the first step to guide you toward the diagnosis of Frantz' tumor via typical tumor's features [6]. Moreover, Tru-cut biopsy should be considered the gold standard because it allows obtaining the definitive histological diagnosis with a minimally invasive approach. Once obtained the diagnosis of Gruber-Frantz tumor, known its very-low malignant nature, it is possible to approach it, when feasible, in a conservative way with enucleation. Robotic surgery represents the best surgical approach thanks to its minimally-invasive nature (guaranteeing an excellent cosmetic result) and its highly superior performances when compared to laparoscopic surgery[13,18].

**Consent:**
Written informed consent was obtained from the parents of the patient for publication of this case re-port and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Data statement:**
The raw data supporting the conclusions of this manuscript will be made available by the authors, without undue reservation, to any qualified researcher.

**Conflict of interest:**
The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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