SOLID PSEUDOPAPILLARY NEOPLASM OF THE PANCREAS IN A YOUNG PATIENT WITH PAPILLARY SQUAMOUS CELL CARCINOMA OF THE ORAL MUCOSA

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Abstract

Introduction. Both papillary squamous cell carcinomas (PSCC) and pancreatic solid pseudopapillary neoplasm (SPN) represent rare cases. PSCC is rare variant of SCC of the upper aerodigestive tract mucosa, occurring frequently in the larynx and hypopharynx. SPN is a low-grade malignancy of the pancreas, affecting primarily young females. **Case report.** We present the case of a 27 year-old woman, who was incidentally diagnosed with pancreatic mass during workup of a previously diagnosed PSCC. The patient was further addressed for endoscopic-ultrasound (EUS)-guided sampling by means of fine-needle aspiration (FNA), in order to achieve differential diagnosis. The EUS identified in the neck and body of the pancreas an encapsulated heterogeneous tumor, with solid and cystic components, and mixed elastography pattern. EUS-FNA was performed and the immunochemistry analysis of the specimens confirmed the diagnosis of solid pseudopapillary neoplasm. The patient underwent distal pancreatectomy followed by surgical resection of the oral PSCC without any complications. Two years after, she is free of symptoms or recurrence signs. **Conclusion.** Surgical resection of both tumors alongside with proper follow-up represent the mainstay of the treatment, providing long-term survival.

Keywords: *pseudopapillary tumor, endoscopic ultrasound, fine-needle aspiration, immunohistochemistry, pancreatectomy.*

1. INTRODUCTION

Papillary squamous cell carcinomas (PSCC) are rare types of squamous cell carcinomas of the oral cavity. PSCC is characterized by papillary proliferation of squamous carcinoma tissue, affecting mostly the larynx or hypopharynx, but PSCC of the oral cavity have been also described in the literature [1-3]. Solid pseudopapillary neoplasm (SPN), also known as Frantz's tumor, accounts for less than 3% of all pancreatic tumors [4]. It was classified by World Health Organization as an epithelial low-grade malignant neoplasm arousing from the exocrine pancreas [5]. SPN occurs typically among young females, between the second and third decade of life. Clinical manifestations depend on the location and size of the tumor, and are usually absent or nonspecific, abdominal pain being the most common complaint. Due to its low potential of malignant transformation, SPN has an excellent prognosis after complete surgical resection [6].

2. CASE REPORT

We present the case of a 27 year-old female, with recent history of PSCC diagnosed on routine dental checkup and one abortion for anencephalic fetus. The oral tumor has been detected by oral cavity examination that revealed an exophytic papillary tumor of the oral mucosa of the left check. Tumor biopsy revealed PSCC. The patient was referred for multidisciplinary workup – including gastroenterology (GI) evaluation and digestive endoscopy within the Gastroenterology Department of the Clinical Emergency Hospital of Bucharest. The GI workup of the PSCC found mild upper abdominal discomfort, in the absence of any other symptoms. Physical examination was normal. Abdominal ultrasound examination detected a well-defined 6 cm hypoechoic lesion with cystic content, enclosed by a well-defined capsule, localized in the head of the pancreas. For establishing the histological origin of the tumor, the patient was sent for sampling of the pancreatic lesion by endoscopic ultrasound guidance with fine-needle aspiration (EUS-FNA). The EUS identified in the neck and body of the pancreas a 70/60 mm heterogeneous tumor, with

evenly distributed transonic, cystic areas throughout the tumor. The solid component presented relatively poor Doppler signal and a mixed (soft and hard) pattern at elastography (Figs. 1A and 1B).

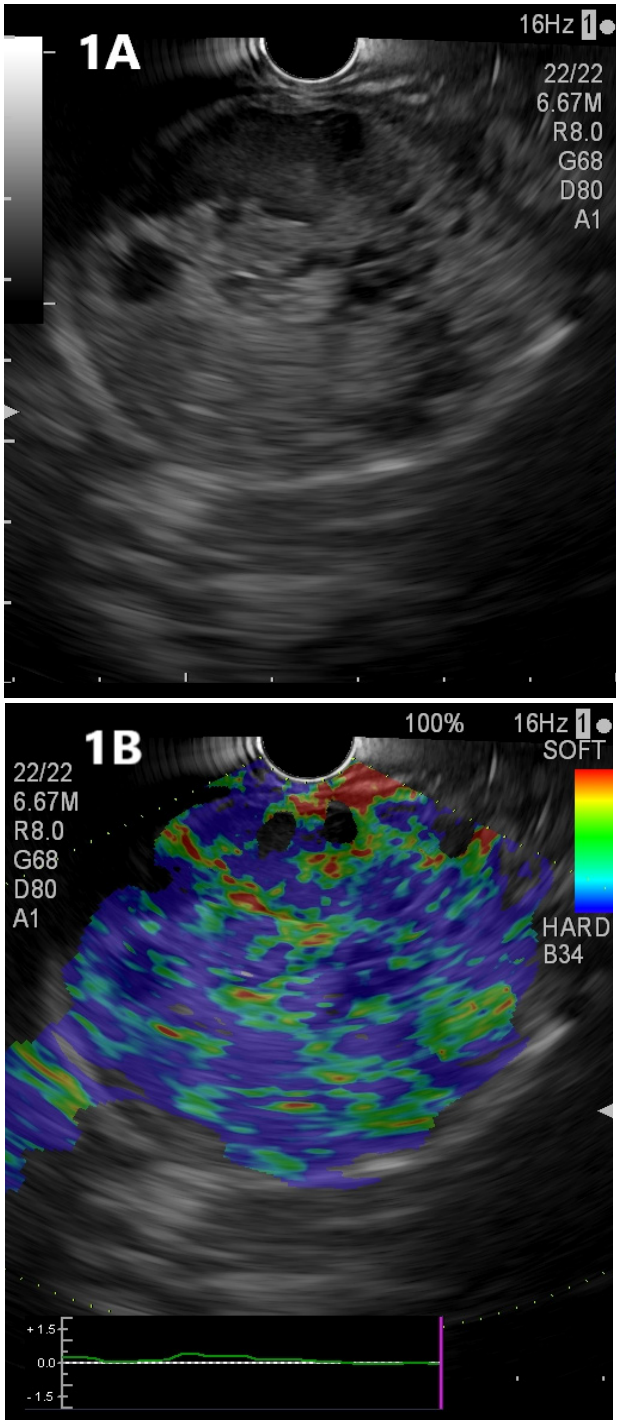


Fig. 1. Endoscopic ultrasound images of the well-defined, solid and cystic pancreatic lesion (1A) with mixed (soft and hard) pattern at elastography investigation (1B)

EUS-FNA with a 22G needle was performed and tissue samples were sent for cytopathological examination. Microscopically, the tumor cells were monomorphic, uniform, round-ovoid, with a palisade cellular architecture along fibro-vascular septs. No significant atypia or mitotic activity was noticed. In between the fibro-vascular stalks, the histopathologist observed microcystic areas. Necrosis was not encountered in the studied smears. Nuclei were round-ovoid shaped (Fig. 2). An immunohistochemistry analysis was additionally conducted (Figs. 3A and 3B, Fig. 4). The tumor was positive for vimentin, beta-catenin, CD10, CD56, chromaffin granule protein A (CgA), synaptophysin and focal receptor of progesterone. Immunohistochemistry also showed a low mitotic Ki-67 index (≤5%). The results were compatible with a solid pseudopapillary tumor of the pancreas.

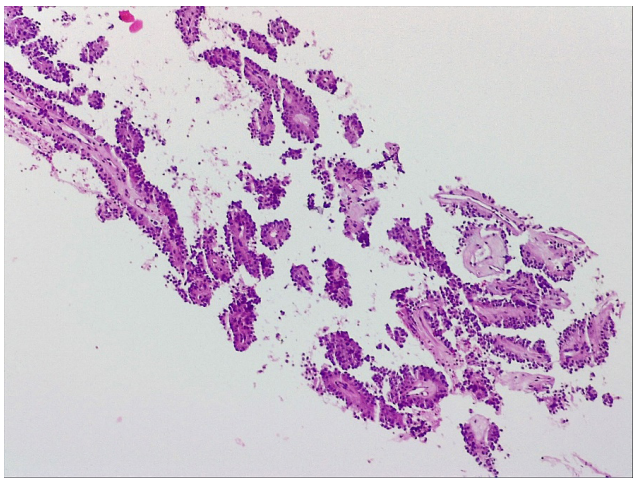
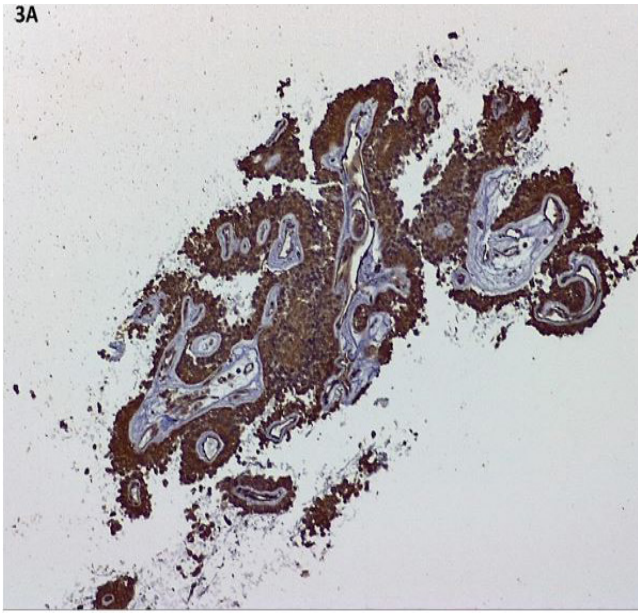


Fig. 2. Hematoxilin-Eosin staining of solid pseudopapillary tumor of the pancreas sample obtained through EUS-FNA (100X)



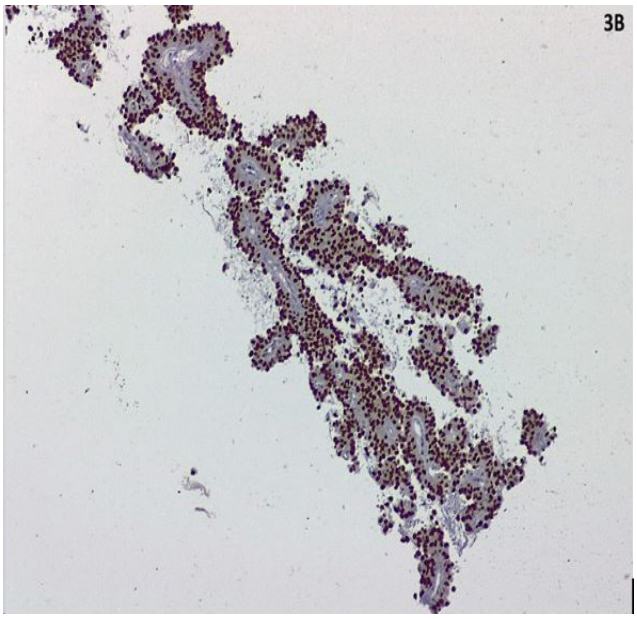


Fig. 3. Histopathologic analysis of the solid pseudopapillary tumor of the pancreas using different immunohistochemical markers (100X): vimentin (3A) and progesterone (3B)

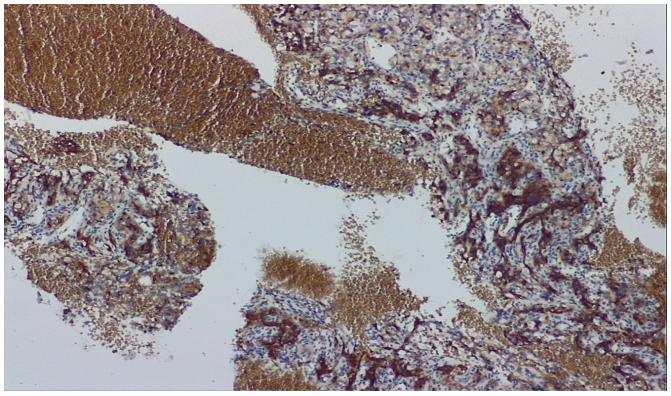


Fig. 4. CD10 immunostaining of solid pseudopapillary tumor of the pancreatic sample obtained through EUS-FNA sampling (200X)

There was no evidence of distant metastasis in the preoperative head-neck-thorax computed tomography (CT) and abdominal magnetic resonance imaging (MRI) secondary to either of the two tumors. Therefore, the patient was sent to surgery for curative resection, decided to be performed prior to oral surgery, due to the increased complexity of the operation. She underwent a body-tail laparoscopic pancreatectomy, with spleen preservation. No perioperative complications were recorded. The pathological features described were those of a solitary mass with cystic and solid components, areas of hemorrhage and cystic degeneration. Histopathological examination confirmed the diagnosis of solid pseudopapillary tumor of the pancreas.

Three weeks after discharge, the patient underwent resection of the PSCC. Two years after both surgical resections, the patient remains asymptomatic, continuing regular follow-up, including clinical examination, routine laboratory tests, abdominal ultrasound and CT or MRI every 6 months.

3. DISCUSSION

Both PSCC and SPN are rare tumors, even rarer in young adults. SPNs could be a challenge for clinicians due to their scarce and nonspecific symptoms. Most patients accuse abdominal discomfort or mild pain, while one-third of them are completely asymptomatic [7]. Pancreatic exocrine insufficiency, malnutrition and weight loss represent uncommon manifestations of SPN and are most likely part of the clinical scenario of patients with aggressive pancreatic malignancy, adenocarcinoma in particular [8,9].

SPN is generally discovered incidentally, as a result of the widespread availability of imagistic modalities. At conventional ultrasound, SPN appears as a hypoechoic mass with heterogeneous or homogeneous content and hyperechoic rim. Contrast CT and MRI have higher specificity and sensitivity compared to ultrasound for identifying the capsule and the intramural bleeding, which represent characteristic features of SPN [10]. Awareness of the imagistic features of SPN is very important for establishing an accurate diagnosis before further surgery. Therefore, preoperative differential diagnosis must be thoroughly assessed by excluding other cystic and/ or solid lesions that may mimic SPN, such as: pseudocyst, cystic neoplasms, adenocarcinoma with cystic degeneration, lymphomas, neuroendocrine tumors, teratoma, pancreatoblastoma [7]. Nevertheless, endoscopic ultrasound with fine-needle aspiration/ biopsy represents nowadays the gold-standard diagnostic approach of pancreatic tumors. Thus, EUS-FNA provides the possibility of tissue sampling and increases

diagnostic accuracy, having low risk of complications [10,11]. Regarding the cytopathological analysis of SPN, worth mentioning is that several histological characteristics distinguish it from other neoplasms: solid and pseudopapillary structures, intensive vascularization, or hemorrhagic pseudocystic structures in various proportions [4]. Moreover, there are specific immunohistochemical markers for SPN which confirm the diagnosis: beta-catenin, vimentin, progesterone receptor, CD56, neuron-specific coil, CD10, cyclin D1, E-membranous and E-cadherin [12]. A high Ki-67 index is an indicator of malignant potential and poor prognosis in patients with SPN [10].

SPN is amenable for complete surgical resction with curative intention, unlike other pancreatic neoplasms which cannot benefit from radical surgery and usually require endoscopic or percutaneous palliation by inserting biliary and duodenal stents to overcome malignant stenosis [13,14]. Patients with SPN have an overall 5-year survival rate higher than 95% following complete en-bloc resection, either by pancreatoduodenectomy or distal pancreatectomy with/ without splenectomy, depending on tumor location [15]. Extensive lymphatic dissection or resection of adjacent structures is not warranted, as lymph node metastasis and recurrence of the tumor are extremely rare [12,15]. Not the least, as shown, digestive endoscopy proved highly useful in a proper systemic workup of oncologic patients [16-20].

4. CONCLUSIONS

Solid pseudopapillary neoplasms of the pancreas are rare, indolent tumors encountered primarily among young females. As shown, such tumors could be incidentally found during workup for other concomitant tumors. The differential diagnosis for primary tumor metastases is important. Therefore, preoperative characterization and sampling of SPN by means of EUS-FNA represents the gold-standard approach offering the possibility of immunochemistry analysis and histologic confirmation. Surgical resection of the tumor represents the treatment of choice, providing an excellent long-term prognosis.

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