Journal of Clinical and Medical Images

Case Report

ISSN: 2640-9615 | Volume 6

Solid Pseudopapillary Tumour of the Pancreas, a Chameleon-like Tumour? High CD138 Expression by a Pseudopapillary and Solid Tumour of the Pancreas Simulating a Plasmacytoma

Dieng N[1*], Diagne NM[1], Diack A[2], Sarr EHS[3] and Diémé E[4]

[1]Medical Oncology and Clinical Haematology Department, Hôpital Principal de Dakar, Senegal

[2]Imaging Department, Hôpital Principal de Dakar, Senegal

[3]Anatomopathology Department, Hôpital Principal de Dakar, Senegal

[4]General Surgical Department, Hôpital Principal de Dakar, Senegal

[*]Corresponding author:

Nata Dieng,
Medical Oncology and Clinical Haematology Department, Hôpital Principal de Dakar, Senegal,
E-mail: natadieng89@yahoo.fr

Received: 20 Nov 2022
Accepted: 01 Dec 2022
Published: 10 Dec 2022
J Short Name: JCMI

Copyright:

©2022 Dieng N, This is an open access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and build upon your work non-commercially.

Citation:

Dieng N, Solid Pseudopapillary Tumour of the Pancreas, a Chameleon-like Tumour? High CD138 Expression by a Pseudopapillary and Solid Tumour of the Pancreas Simulating a Plasmacytoma. J Clin Med Img. 2022; V6(20): 1-6

Keywords:

Solid Pseudopaillary Tumour of the Pancreas; Aetiology; Plasmacytoma

1. Abstract

Solid Pseudopapillary Tumour of the Pancreas (SPTP) is an uncommon tumour that occurs mainly in young women. Its aetiology is unknown with a low potential for malignancy. Its diagnosis is most often difficult by biopsy. Imaging data associated with epidemiological data should be sufficient to conjure up its diagnosis. We report the case of a young woman who presented with abdominal pain and an epigastric mass that had been evolving for two years. We admitted her for consultation purpose in haematology clinic for a plasmacytoma of the pancreas which was evoked after several biopsy examinations of the epigastric mass. The diagnosis of SPTP was made only after histological and immunohistochemical study of the surgical specimen.

2. Introduction

Solid Pseudopapillary Tumour of the Pancreas (SPTP) is a rare, low-grade malignancy that occurs electively in young women. Its histogenesis is still unclear [1,2]. Its prognosis is unremarkable after surgical removal. It often poses a real diagnostic challenge because of its clinical and radiological polymorphism and its highly variable immunohistochemical profile [3]. Usually, tumour cells are labelled with anti-CD10, anti-CD56, anti-CD99, alpha-1antitrypsin, vimentin, E-caderin and betacatenin antibodies [2, 4]. We report a case of a female patient in whom we found a high expression of CD138, which so far was considered as a specific marker for plasma cells. The diagnosis was made on the basis of histological study associated with immunohistochemistry of the corporal-caudal splenopancreatectomy specimen.

3. Observation

We report the case of a 28-year-old woman with no particular medical history, primigravida, primiparous, referred to clinical haematology for the management of a plasmacytoma of the head and body of the pancreas. She was followed in the gastro-enterology department for epigastralgia and an epigastric mass that had been evolving for 2 years. On examination she had a good general condition PS 0 and presented with a firm regular painless epigastric mass. Upper gastro-intestinal endoscopy showed a large outer compression of the stomach and a small antral erosion. Abdominopelvic computed tomography (CT) showed a heterogeneous mass, developed upon the body and tail of the pancreas with a solid cystic appearance containing a peripheral enhanced fleshy portion, a fluid portion and calcifications, measuring 124 x 117 x 76 mm (Figure 1). Abdominal Magnetic Resonance Imaging (MRI) showed a large, heterogeneous T2-hypersignal encapsulated mass containing necrotic-hemorrhagic changes (Figure 2).

The extension assessement showed no loco-regional or distant invasion. The first two cytopunctures performed on the mass were non-contributory. The histological study of the third cytopuncture showed a tumour proliferation of small round cells resembling plasma cells with eccentric, hyperchromatic nuclei and basophilic cytoplasms. They showed a low degree of nuclear atypia and were grouped in a diffuse sheet in places crossed by thin fibrous trabeculae. The appearance was suggestive of a plasmacytoma. The histological study of the last cytopuncture showed areas of plasma cells of varying degrees of differentiation; large cells with basophilic cytoplasm, rounded nuclei with fine nuclear chromatin and central nucleoli; the stroma was poorly marked, quite highly vascularised (Figure 3). Immunohistochemistry showed strong CD138 expression, absence of anti-CD99 labelling, diffuse anti-Lambda labelling and absence of anti-Kappa expression (Figure 3). The histological and immunohistochemical feature was also suggestive of a plasmacytoma. The histology results were from two different pathologists. The plasmacytoma extension work-up did not show bone lysis on standard radiography, nor a monoclonal peak on serum protein electrophoresis. The medullogram was normal. The osteomedullary biopsy showed a normal bone structure. Amylasemia, lipasemia, alphafetoprotein were normal. HIV, HBV and HCV serologies were negative. The indication for surgery was given in a multidisciplinary team meeting (MDTM). A corporal-caudal splenopancreatectomy was performed. On gross examination, the specimen weighed 596g and included a large pancreas tumour measuring 12 x 10 x 6 cm, a residual tail measuring 3 cm, a spleen measuring 8.5 x 6.5 x 4.5 cm, and three lymph nodes included in the curage (Figure 4). Histology showed a well encapsulated, well limited malignant tumour proliferation (Figure 5). This proliferation was made of papillary, pseudo-papillary, trabecular architecture. The cells were medium-sized, monomorphic in appearance and showed rare cytonuclear atypia and images of mitosis (Image 5). The stroma was small. Cholesterolic granulomas were present in some areas, and perineural neoplastic sheathing was present. There were no vascular emboli. All borders were healthy. The adjacent pancreatic parenchyma was discretely fibrous. Spleen samples showed normal splenic parenchyma. The three lymph nodes found in the curage were reactive, not metastatic. On immunohistochemistry, CD138 markers were positive (Figure 6). CD56, vimentin and synaptophysin were also positive (Figure 6). The following markers were negative: CKAE1.AE3, CD20, CD30, CD15, PS100, myogenin, CD99, CK7, CK20, chromogranin and melan A. The proliferation index assessed by ki67 was low (2%). The morphological and immunohistochemical profile was therefore in favour of a pseudopapillary and solid tumour of the pancreas. At 24 months follow-up, the patient is in good general condition with no clinical or scannographic recurrence.

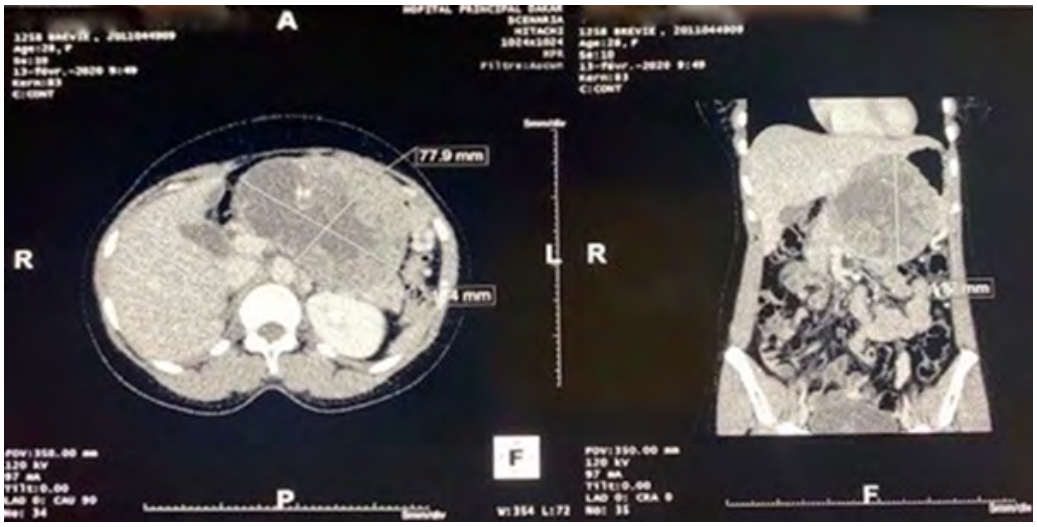

Figure 1: Abdominopelvic CT: heterogeneous mass, developed upon the body and tail of the pancreas with a solid-cystic appearance containing an enhanced peripheral fleshy portion, a fluid portion and calcifications, measuring 124 x 117 x 76 mm; no locoregional or distant lesions identified.

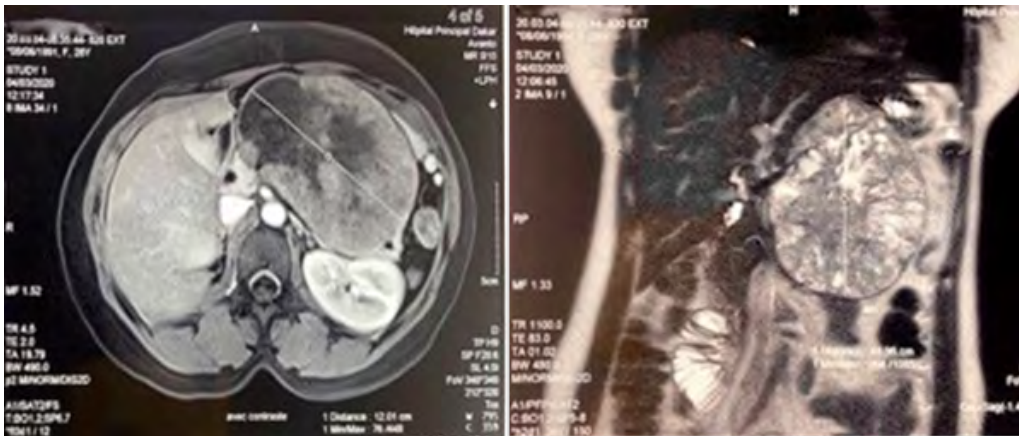

Figure 2: Abdominal MRI: large, partially necrotic pancreatic corporal-caudal mass, well encapsulated, measuring 12 cm, with no signs of aggressiveness towards neighbouring organs

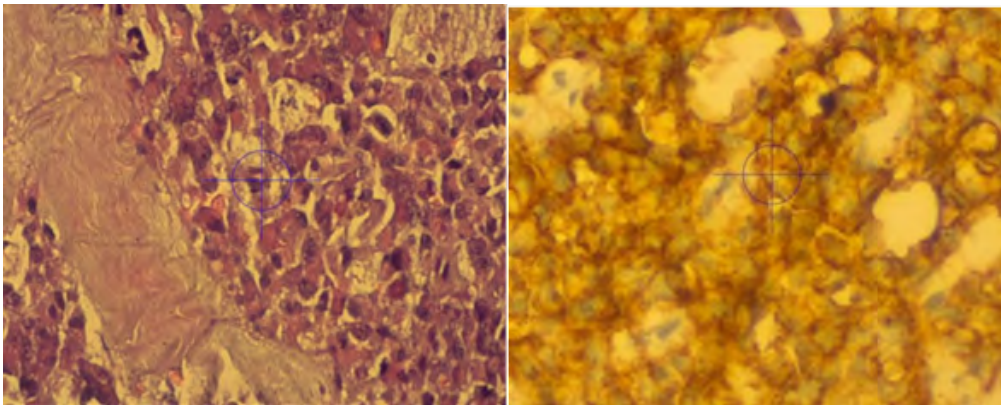

Figure 3: Left, H.E.S. staining, magnification x10) Massive proliferation of cells with abundant basophilic (eosinophilic) cytoplasm and an eccentric nucleus often nucleated. Right, (x10) Membrane labelling of tumour cells with anti-CD38.

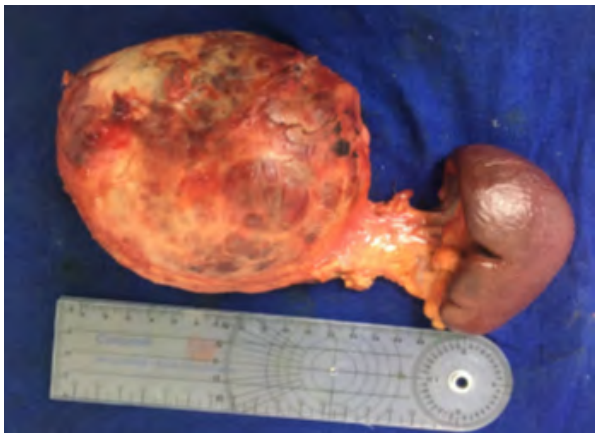

Figure 4: Complete surgical specimen comprising the pancreas with a large tumour measuring 12 x 10 x 6 cm, a residual tail measuring 3 cm, a spleen measuring 8.5 x 6.5 x 4.5 cm and three lymph nodes

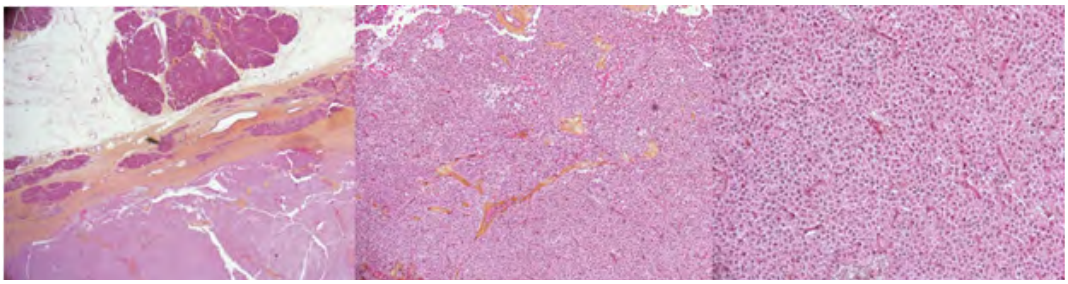

Figure 5: On the left, a well encapsulated, well limited malignant tumour proliferation. Middle and right, high magnification and HES staining.

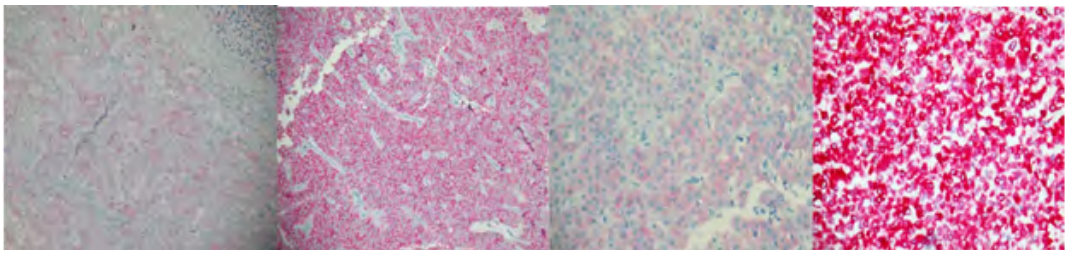

Figure 6: CD138, CD56, synaptophysin, vimentin positivity

4. Discussion

Firstly described by Frantz [5] in 1959 as a benign or malignant papillary tumour of the pancreas, the solid pseudo-papillary tumour of the pancreas is an uncommon anatomicalclinical entity and accounts for 2-3% of pancreatic tumour lesions and 0.9-2.7% of exocrine tumours of the pancreas [1,4]. This tumour has been given several names such as papillary epithelial tumour, cystic and solid tumour, solid epithelial tumour, papillary cystic epithelial tumour and Frantz tumour respectively. These different names reflect both the morphological diversity of the tumour tissue. In 1996, the WHO defined SPTP as a borderline tumour of the pancreas or tumour of uncertain potential malignancy and named it the solid pseudopapillary tumour [6]. It mainly occurs in young women with a mean age of 22 years and a male to female sex ratio of 1:10 for some authors [3,7] and for more recent reviews a mean age of 28 years and a M/F sex ratio of 1:7 [8]. The tumour affects the head, body or tail of the pancreas, but develops preferentially in the corporal-caudal region (64% of cases) [3]. The circumstances of discovery are highly variable and usually non-specific : discovery may be incidental to an imaging examination performed for another reason, or to the appearance of a palpable abdominal mass, or to atypic abdominal pain. Sometimes the tumour, as it increases in size, causes signs of compression of neighbouring digestive, biliary or vascular structures [9]. SPTP can be detected by ultrasound, computed tomography (CT), magnetic resonance imaging (MRI) and positron emission tomography (PET). The ultrasound appearance of the tumour varies according to the size of the cystic areas, but typically, SPTP presents as a well-contained, evenly contoured cystic mass with little or no vascularity, heterogeneous contents and no internal partitions. CT shows a solid, cystic mass with areas of haemorrhage and/or cystic degeneration. Calcifications and enhanced solid areas may be present around the mass. MRI essentially shows a wellcircumscribed lesion with a mixture of high and low signal intensity on T1 and T2 weighted images. The typical appearance is that of a well-circumscribed mass with varying degrees of internal haemorrhage and cystic degeneration, which may be associated with calcifications. Imaging data, guided by the clinical context, should suggest a diagnosis of SPTP [1]. In case of doubt, endoscopic ultrasound should be performed. Although essential for pre-operative diagnosis, the sensitivity and specificity of cytopuncture for pseudopapillary and solid pancreatic tumours is highly controversial due to the rarity of this condition and the lack of quality data [10].In high income countries, cytological examination is usually the first and most used approach in the diagnostic assessment of solid and cystic pancreatic masses, so that the first diagnosis of SPTP is often cytological. Ultrasound-guided endoscopic fine-needle aspiration is the most frequently used procedure, which in skilled hands is a safe, cost-effective and valuable technique [4]. However, percutaneous biopsy is associated with a non-negligible risk of tumour dissemination along its route and complications such as haemorrhage, pancreatic fistula and biliary fistula [11]. CT or MRI data combined with age and sex should be sufficient to indicate surgery, and pre-operative biopsy should be performed when the radiological diagnosis is not clear enough [12]. Macroscopically, the tumour is often large, averaging 10 cm in length, rounded or oval, surrounded by a fibrous capsule [13]. On cross-section, the pseudocystic appearance is associated with necrotic and haemorrhagic changes [14]. Histologically, the tumour consists of peripheral solid patches and central papillary structures. The tumour cells are monomorphic, small, cuboidal or polygonal and often arranged around fibrovascular septa. Mitoses and cytonuclear atypia are exceptional. Clusters of foamy histiocytes and giant cells may be found around cholesterol crystals. The stroma is usually endocrine, rich in blood capillaries. Pathological

criteria for malignancy are found in only 1015% of cases (invasion of adjacent structures, vascular emboli, perineural invasion and lymph node or distant metastases); in these cases, PPSCT is classified as a pseudopapillary solid carcinoma [15]. The classical histopathological appearance usually allows the correct diagnosis, but some pancreatic tumours may show similarities and, for differential diagnostic reasons, various complex immunohistochemical examinations are necessary. In addition, these markers may serve as a tool to the possible histogenesis of SPTP [2]. The immunohistochemical profile of SPTP is variable. Typically, tumour cells are labelled with anti-CD 10, alpha-1 antitrypsin, vimentin, NSE, E-caderin and betacatenin antibodies. There is also labelling with anti-progesterone antibody [16]. Positive immunostaining of tumour cells for some endocrine markers may indicate some endocrine differentiation [17]. In the literature, a large number of antibodies have been tested, but sometimes the results seem contradictory. There is general agreement that virtually all of these tumours are positive for vimentin, β-catenin, cyclin D1, alpha-1-antitrypsin (AAT) or CD56. Loss of E-cadherin expression is also a typical finding. The presence of the progesterone receptor is regularly detected, the androgen receptor is expressed in about 80% of cases, but the oestrogen receptor is most often negative. Cytokeratin expression is highly variable, ranging from 28 to 70%, but CK7 is negative. Most SPTP are CD10 positive, usually with a perinuclear dot pattern, and CD99 has also been reported as a reliable positive marker. Among the neuroendocrine markers, chromogranin-A is usually absent, NSE (although its specificity is arguable) may be negative or consistently positive, while in a small percentage of tumours synaptophysin is detectable. Because the SPTP is not considered a hormonally active tumour, specific pancreatic hormones are not found in the cells. TFE3, a transcription factor promoting several genes involved in cell proliferation and growth, has been shown to be a very sensitive marker (75-96% of cases), and can be used to strengthen the diagnosis of SPTP.There are other apparently unrelated immune positivities (LEF-1, FUS, WIF-1, CD200) that are expressed with high frequency, but their significance and applicability is yet to be clear in this tumour.SPTP, neuroendocrine tumours and acinar cell carcinomas can sometimes present similar histological profiles making their distinction difficult [2].The specificity of our observation is the expression of CD138 in SPTP. However, this finding is not surprising as CD138 is thought to be a promoter of Wnt/beta-catenin signalling in multiple myeloma cells, with beta-catenin expression being an important diagnostic marker for SPTP [18, 19]. The biological relevance of this feature, as raised by Adriana Handra-Luca in her paper [18]., is difficult to determine, whether it is indicative of a plasma cell type, neuroendocrine or mesothelial derivation. Furthermore, various epithelial cells, such as keratinocytes, are labelled with CD138 with high membrane positivity and low cytoplasmic labelling except for superficial differentiated cells [20]. It has also been described that high plasma cell density is associated with a better prognosis in many solid cancers. Indeed, plasma cells are an important component of the anti-tumour response. Their differentiation can take place at the level of a secondary lymphoid organ and/or within the tertiary lymphoid structures (TLS) associated with tumours. The markers used to detect plasma cells are diverse and only partially overlapping in different studies, due to the lack of specificity of some of these markers in certain cancers and/or differences in the roles played by plasma cells depending on the type of tumour [21].

5. Conclusion

Immunohistochemistry is today the basis for the diagnosis of SPTP. Its absence, due to a lack of technical facilities in our countries and its high cost, as it is often sent abroad, could be a source of diagnostic errancy and delays in therapeutic management.

References

1. Dinarvand P, Lai J. Solid Pseudopapillary Neoplasm of the Pancreas: A Rare Entity With Unique Features. Arch Pathol Lab Med. 2017; 141(7): 990–995.
2. Zalatnai A, Kis-Orha V. Solid-pseudopapillary Neoplasms of the Pancreas is still an Enigma: a Clinicopathological Review. Pathology & Oncology Research. 2019; 26: 641-649.
3. Jakhlal N, Njoumi N, Hafid Hachi H, Bougtab A. Tumeur pseudopapillaire et solide du pancréas : à propos d'un cas et revue de la literature. Pan Afr Med J. 2016; 24: 104.
4. La Rosa S, Bongiovanni M. Pancreatic Solid Pseudopapillary Neoplasm Key Pathologic and Genetic Features. Arch Pathol Lab Med. 2020; 144: 829–837.
5. Elkaoui H, Bouchentouf SM, Sall I, et al. Tumeur pseudopapillaire et solide du pancréas. J. Afr. Cancer. 2010; 2(2): 116-119.
6. Yao J, Song H. A Review of Clinicopathological Characteristics and Treatment of Solid Pseudopapillary Tumor of the Pancreas with 2450 Cases in Chinese Population. Biomed Rest Int. 2020; 2020: 2829647.
7. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. Journal of the American College of Surgeons. 2005; 200(6): 965–972.
8. Law JK, Ahmed A, Singh VK, Akshintala VS, Olson MT, Raman SP, et al. A systematic review of solid-pseudopapillary neoplasms are these rare lesions? Pancreas. 2014; 43(3): 331–337.
9. Wang DB, Wang QB, Chai WM, Chen KM, Deng XX. Imaging features of solid pseudopapillary tumor of the pancreas on multi-detector row computed tomography. World J Gastroenterol. 2009; 15(7): 829-35.
10. Reddy S, Cameron JL, Scudiere J, Hruban RH, Fishman EK, Ahuja N, et al. Surgical management of solid-pseudopapillary neoplasms of the pancreas (Franz or Hamoudi tumors): a large single- institutional series. J Am Coll Surg. 2009; 208(5): 950-7.
11. Pettinato G, Di Vizio D, Manivel JC, Pambuccian SE, Somma P, Insabato L. Solid pseudopapillary tumor of the pancreas: a neoplasm with distinct and highly characteristic cytological features. Diagn

Cytopathol. 2002 Dec; 27(6): 325- 34.

12. Ren Z, Zhang P, Zhang X, Liu B. Solid pseudopapillary neoplasms of the pancreas: clinicopathologic features and surgical treatment of 19 cases. Int J Clin Exp Pathol. 2014; 7(10): 6889-6897.
13. Guedira M, Hrora A, Raïss M, El Alaoui M, Kettani F, Tounsi A. Tumeurs pseudopapillaires et solides du pancréas. J Chir. 2006; 143: 275-278.
14. Podevin J, Triau S, Mirallié E, Le Borgne J. Tumeurs pseudopapillaires et solides du pancréas : à propos de cinq cas et revue de la littérature. Ann chir. 2003; 128: 543–548.
15. Hamilton SR, Aaltonen LA. Solid-pseudopapillary neoplasm. World health organization classification of tumors. Pathology and genetics of tumours of the digestive system. Lyon, IARC press, 2000; 246-8.
16. Kim MJ, Jang SJ, Yu E. Loss of E-cadherin and cytoplasmic- nuclear expression of beta-catenin are the most useful immunoprofiles in the diagnostis of solidpseudopapillary neoplasm of the pancreas. Hum Pathol. 2008; 39(2): 251- 8.
17. Ayadi L, Ellouze S, Khabir A, Daoud E. La tumeur de Frantz : une étude anatomoclinique de six cas tunisiens. Rev Med Brux. 2008; 29(6): 572-6.
18. Handra-Luca A. CD138/syndecan–1 in pancreatic solid and pseudopapillary neoplasms. J Clin Pathol. 2019; 72(2): 186.
19. Ren Z, van Andel H, de Lau W, Hartholt RB, Maurice MM, Clevers H, et al. Syndecan-1 promotes Wnt/®-catenin signaling in multiple myeloma by presenting Wnts and R-spondins. Blood. 2018; 131: 982-94.
20. Kaplon H, Dieu-Nosjean MC. Quel avenir pour les lymphocytes B infiltrant les tumeurs solides Marqueur pronostique et/ou cible thérapeutique ? Med Sci (Paris). 2018; 34(1): 72-8.
21. CD138. Référentiel des examens biologie clinique- génétique- anatomie et cytologie pathologiques.