Diagnostic Challenge of a Cystic Solid Pseudopapillary Tumor in Pancreas

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Abstract

Solid Pseudopapillary Tumors of the Pancreas (SPTPs) are rare tumors with non-specific presentation which makes them a difficult diagnostic challenge. the morphologic features of the cells were similar to the cells seen in neuroendocrine tumors. Immunohistochemistry cleared up the doubts and made the diagnosis of SPTP the definitive diagnosis.

Diagnostic Challenge of a Cystic Solid Pseudopapillary Tumor in Pancreas: A Case report.

Clinical key message:

A solid pseudopapillary tumor should be included in the differential diagnosis of every pancreatic cystic lesion, and both morphology and immunohistochemistry lead to the definitive diagnosis.

Introduction:

Solid Pseudopapillary tumors of the pancreas (SPTPs) are cystic and solid neoplasms [1]. They are rare pancreatic neoplasms occurring most commonly in females in the second or third decade and account about 0.17–2.7% of all pancreatic tumors [1, 2]. Many studies from 1961 until 2012 report that the most frequent symptom is abdominal pain, but there are no symptoms in the rest of the cases and the diagnosis is made through routine examination [3]. The monomorphic and bland morphology of SPTP cells make it difficult to be differentiated from other pancreatic tumors, especially Neuroendocrine tumors. Immunohistochemistry (IHC is a crucial factor in making the accurate diagnosis. Here, we present a difficult-to-diagonse (SPTP) case due to the almost subtotal cystic degeneraton and only scanty residual tumor nests in the wall.

Case report:

A 36-year-old non-alcoholic female with a history of smoking for 15 years, presented with abdominal pain radiating to the back. The pain was not relieved by NSAIDS.

The patient mentioned that she had experienced many episodes of non-bilious vomiting, nausea and intermittent non-bloody diarrhea. On physical examination, a mass was palpated in the epigastric region. Lab tests were normal except for a mild anemia. The radiological findings on Ultrasound (US and non-contrast Computed tomography (CT revealed multiple cystic lesion in the tail of the pancreas attached to the spleen. **Fig.1**

The patient underwent distal pancreatectomy with splenectomy and the specimen was sent to the pathology department.

Grossly, the specimen was composed of the distal part of the pancreas adherent to the spleen, where a cystic mass measuring about 8 cm in diameter was found, the rest of pancreas tissue measured 4x6 cm. The spleen measured 6×10 ×15 cm **Fig. 2** . In addition, three regional lymph nodes were detected. The pathologist's first impression was a pancreatic pseudocyst, but also other cystic neoplasms of pancreas could not be excluded.

Microscopically, In the wall of the cyst there were nests of neoplastic histiocyte like cells without significant cellular atypia **Fig. (3 4.** No evidence of vascular invasion was found. The lymph nodes, the spleen and the surgical margins were tumor free.

Beside Hematoxylene and Eosin (H&E stains, IHC was recommended to figure out the diagnosis. The IHC revealed positivity of the tumor cells for CD56 **Fig.5** and Cyclin D1. In addition, NSE was weak positive, whereas pan Cytokeratin (CK, CK7, CK20, Chromogranin A, synaptophisin, CD68, S100, Vimentin, Epithelial membrane antigen (EMA , and smooth muscle actin (SMA were negative.

Based on H&E stain and IHC, the diagnosis was limited between SPT and NET of pancreas. The morphologic features of the cells and the lack of β-catenin stain lead to signing out this case as a well-differentiated endocrine tumor (G1-NET) with almost total cystic degeneration. After a year β-catenin and E-Cadherin stains were performed and the tumor cells showed a positivity for β-catenin and negativity of E-Cadherin**Fig. (6 7.** Therefore, the diagnosis of solid pseudopapillary tumor of pancreas was the final diagnosis.

Discussion: Solid pseudopapillary tumors are rare low-grade malignant neoplasms, commonly located in the tail of pancreas[4]. Necrosis and cystic degeneration are common features [5, 6]. The ultrasound and the CT reveals well-defined solid masses with cystic components [7](2).

Clinically, SPTP are usually non-symptomatic and discovered as an abdominal mass by accident or during physical examination [3]. Complete lab tests are usually normal. Surgery is the gold standard treatment with curative results if the lesion is completely resected [4].

In our case, microscopic examination of a pancreatic cystic lesion revealed nests of round monomorphic cells surrounded by a scant fibrovascular stroma in the cyst wall. The cytoplasm of the cells was clear to granulated and the nuclei were uniform round to oval with finely and evenly distributed chromatin. No mitotic figures were identified. Depending on the morphologic findings, the final differential diagnosis was SPTP and NET.

The IHC results were as following: Chromogranin A was negative, as many other immune stains, and the only positive stains were CD56, NSE, and cyclin D1. At that time, E-cadherin and β-catenin were not available in our lab. Therefore, we favored the diagnosis of low-grade NET.

One year later, E-cadherin and β-catenin were available. Because the tumor cells were negative for E-cadherin with nuclear positivity for β-catenin, we redeemed our diagnosis from NET to SPTP.

SPTs can show significant morphological overlap with NETs but the unclear invasion, clear cytoplasm and nuclear groove in tumor cells support the diagnosis of SPT more than PanNETs [4] Although morphological characteristics of the tumor cells and IHC stains play an essential role in distinguishing between these two neoplasms, their rule is not absolute and an overlap between the two tumor was also documented immunohistochemically. Some studies, demonstrate that E-cadherin and β-catenin are the most useful immunostaining markers to differentiate between them[8]. Our case supports this fact.

Eighteen months following the surgery, Ultrasound revealed no recurrence or metastases. Thus, proving the good prognosis of SPTP.

Conclusion: Solid pseudopapillary tumor should be included in the differential diagnosis of every pancreatic cystic lesion and both morphology and IHC lead to the definitive diagnosis. These tumors and Neuroendocrine tumor (NET) of the pancreas should be distinguished from each other. SPT-specific markers such as β-catenin are recommended, even if the tumor has a NET-like morphology.

Authors' Contributions:

Ebrahim Makhoul and Zeina Alabbas: Drafted the article and collected the patient's data. Ali Adra and Alexey Youssef: Participated in collecting the data and drafting the article. Emad Ayoub: Performed the surgical biopsy and participated in revising the article. Rana Issa: The guarantor and supervisor, critically revised the article, performed the pathologic examination and approved the final manuscript.

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Conflict of interest:

None.

Ethical Approval:

Informed consent was obtained from the patient regarding the report of her clinical scenario data in an anonymous way.

References:

1. Utturkar A, Yu Z, Lawrence C: Solid pseudopapillary tumor of the pancreas: A difficult presentation of an uncommon tumor. Radiol Case Rep 2018, 13(6):1104-1107.

2. Yagmur Y, Yigit E, Gumus S, Babur M, Can MA: Solid cystic pseudopapillary tumor of pancreas with splenic metastasis: Case report and review of literature. Int J Surg Case Rep 2015, 14:50-52.

3. Law JK, Ahmed A, Singh VK, Akshintala VS, Olson MT, Raman SP, Ali SZ, Fishman EK, Kamel I, Canto MI et al: A systematic review of solid-pseudopapillary neoplasms: are these rare lesions? Pancreas 2014, 43(3):331-337.

4. Lanke G, Ali FS, Lee JH: Clinical update on the management of pseudopapillary tumor of pancreas. World J Gastrointest Endosc 2018, 10(9):145-155.

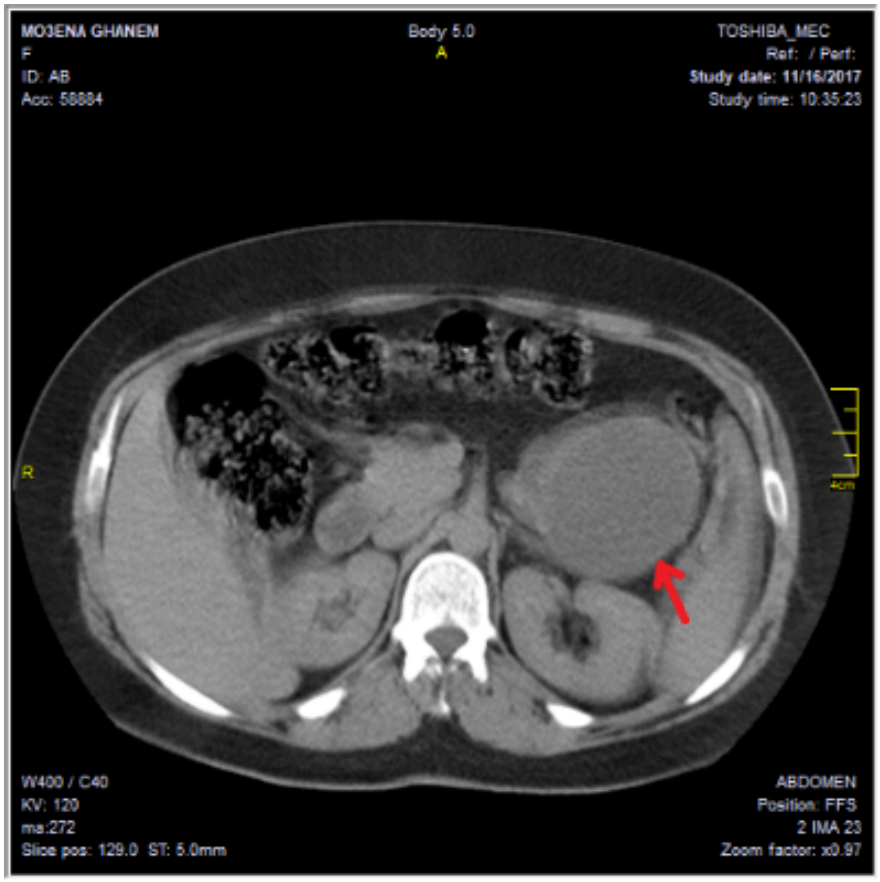
5. Basturk O, Coban I, Adsay NV: Pancreatic Cysts: Pathologic Classification, Differential Diagnosis, and Clinical Implications. Archives of Pathology & Laboratory Medicine 2009, 133(3):423-438.

6. Parra-Herran CE, Garcia MT, Herrera L, Bejarano PA: Cystic lesions of the pancreas: clinical and pathologic review of cases in a five year period. JOP 2010, 11(4):358-364.

7. Ahmed TS, Chavhan GB, Navarro OM, Traubici J: Imaging features of pancreatic tumors in children: 13-year experience at a pediatric tertiary hospital. Pediatr Radiol 2013, 43(11):1435-1443.

8. Ohara Y, Oda T, Hashimoto S, Akashi Y, Miyamoto R, Enomoto T, Satomi K, Morishita Y, Ohkohchi N: Pancreatic neuroendocrine tumor and solid-pseudopapillary neoplasm: Key immunohistochemical profiles for differential diagnosis. World J Gastroenterol 2016, 22(38):8596-8604.

Figures :



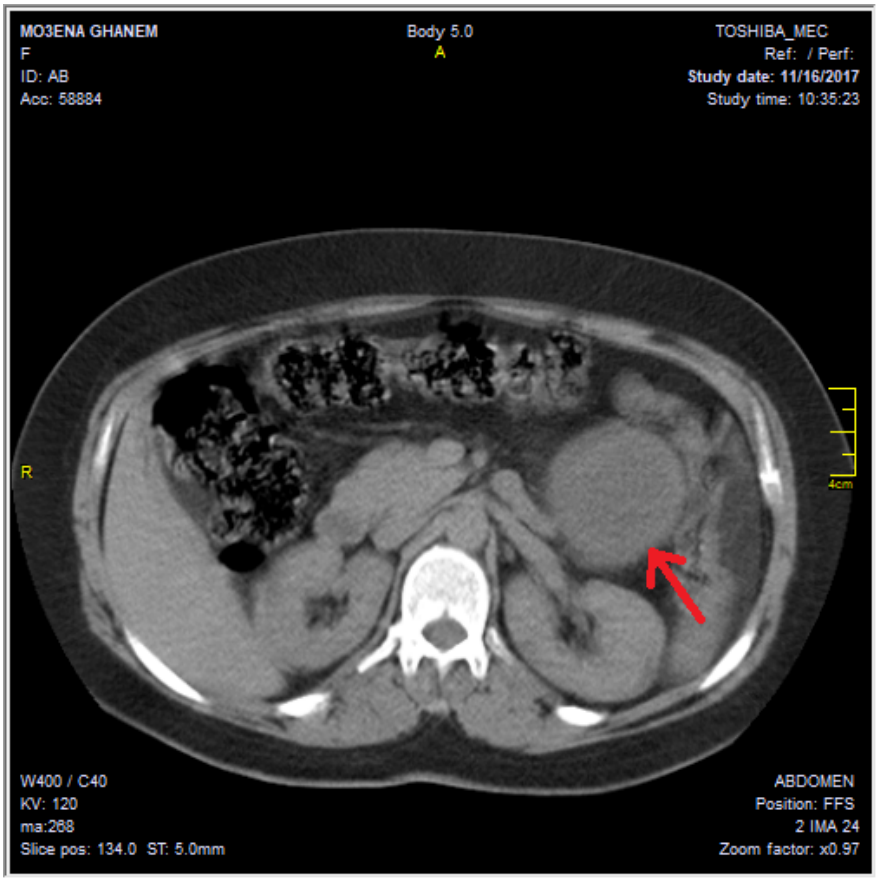


Figure 1. Non-contrast CT scan revealed a mixed solid and cystic lesion in the tail of the pancreas attached to the spleen

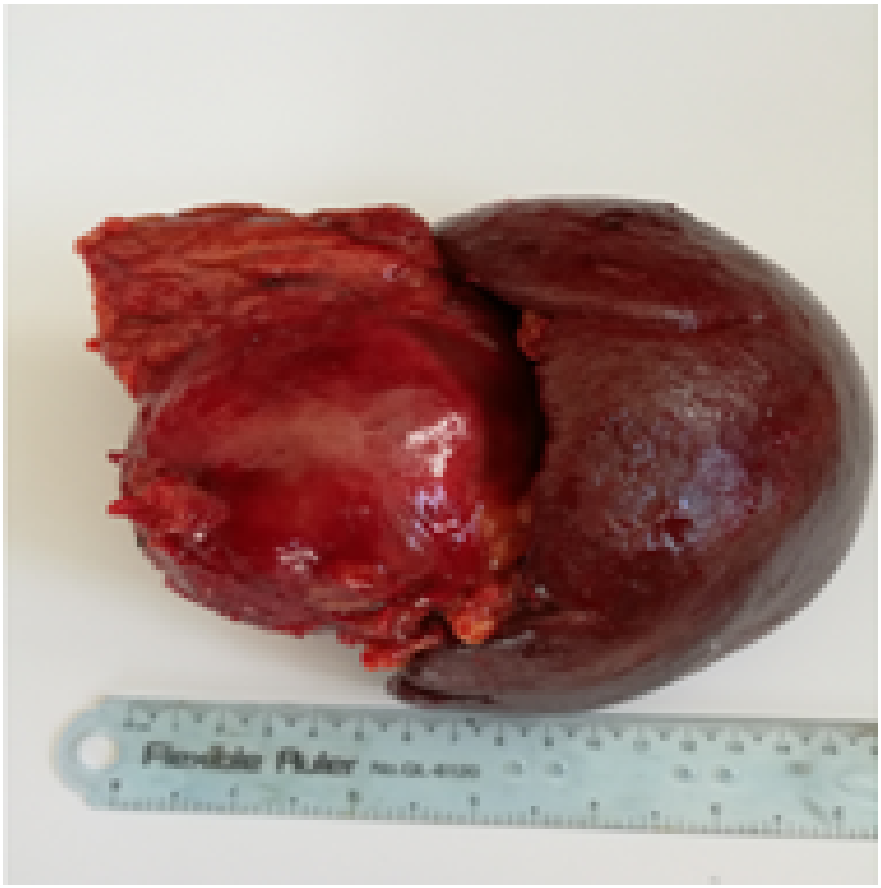
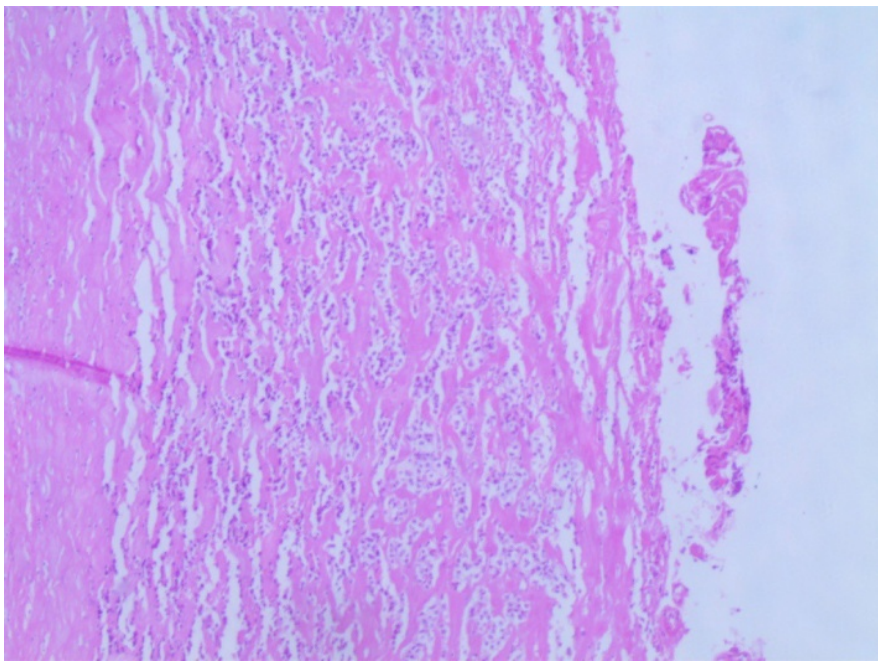


Figure 2. A Cystic mass in the distal part of the pancreas adherent to spleen.



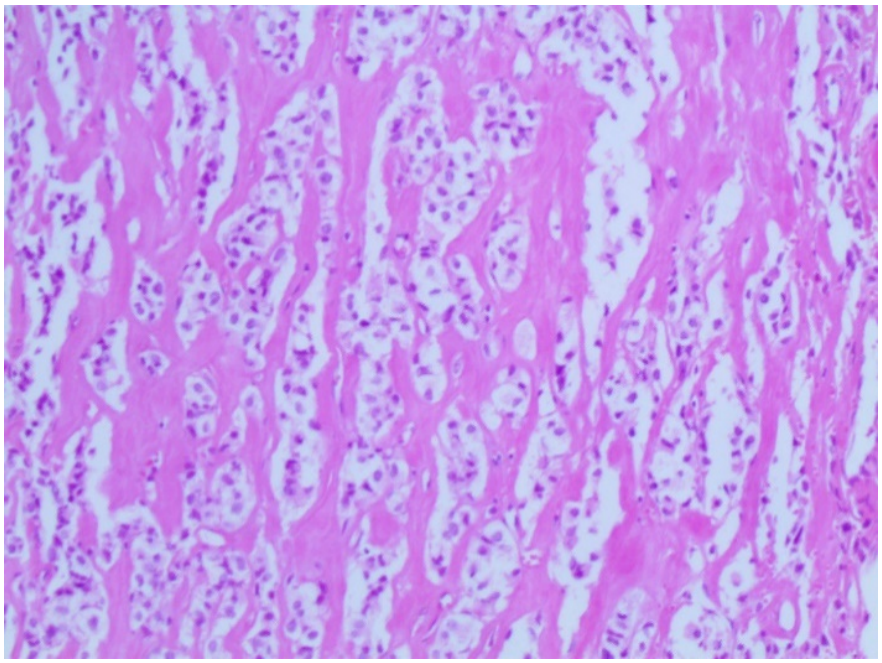


Figure3 . Nests of neoplastic cells within the fibrous wall of the cyst (x40) and (x200)

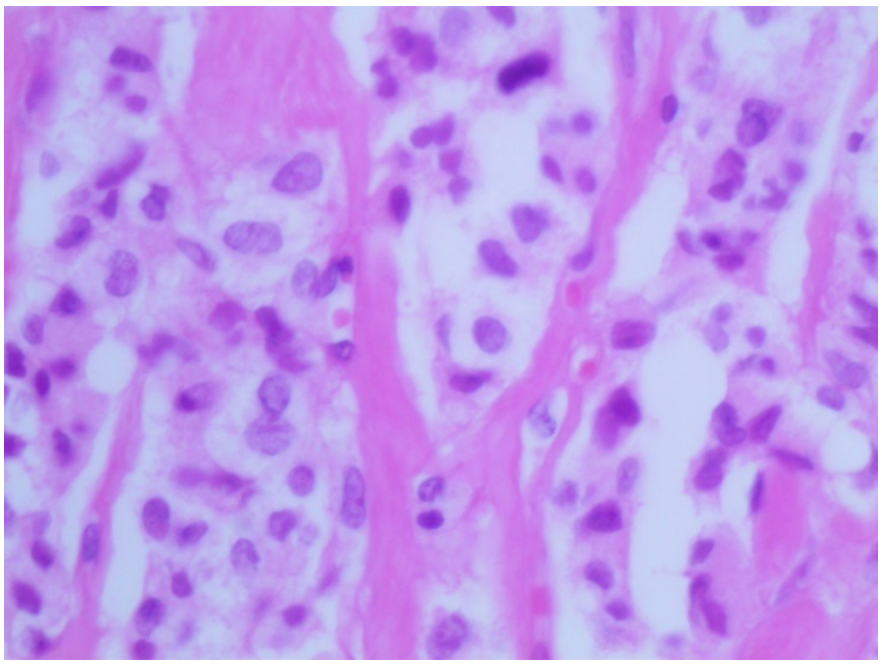


Figure 4. The tumor cells are monomorphic round to oval cells with clear to granulated cytoplasm surrounded by a scant fibrovascular stroma (x400)

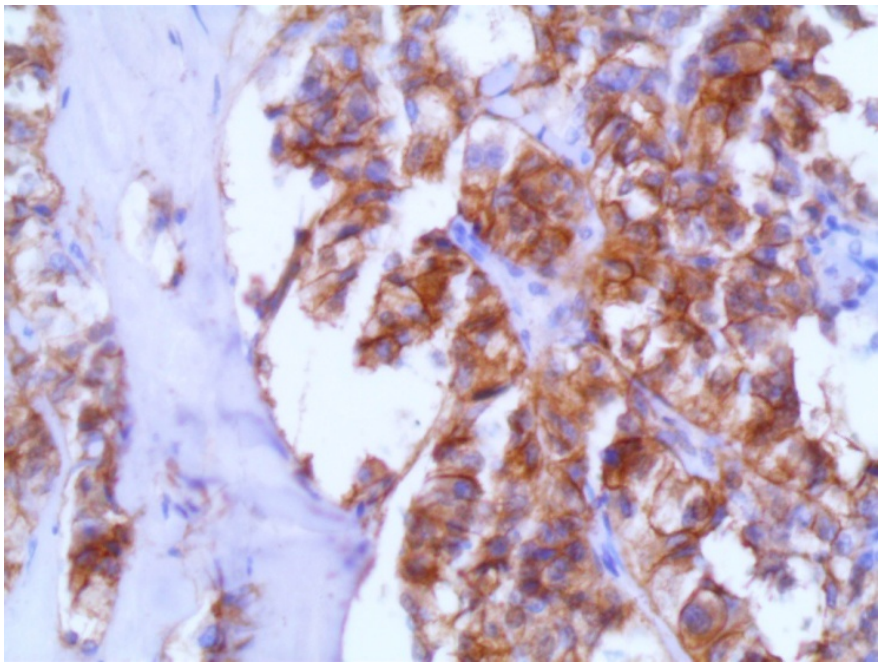


Figure 5. Tumor cells show positivity for CD56 (x400)

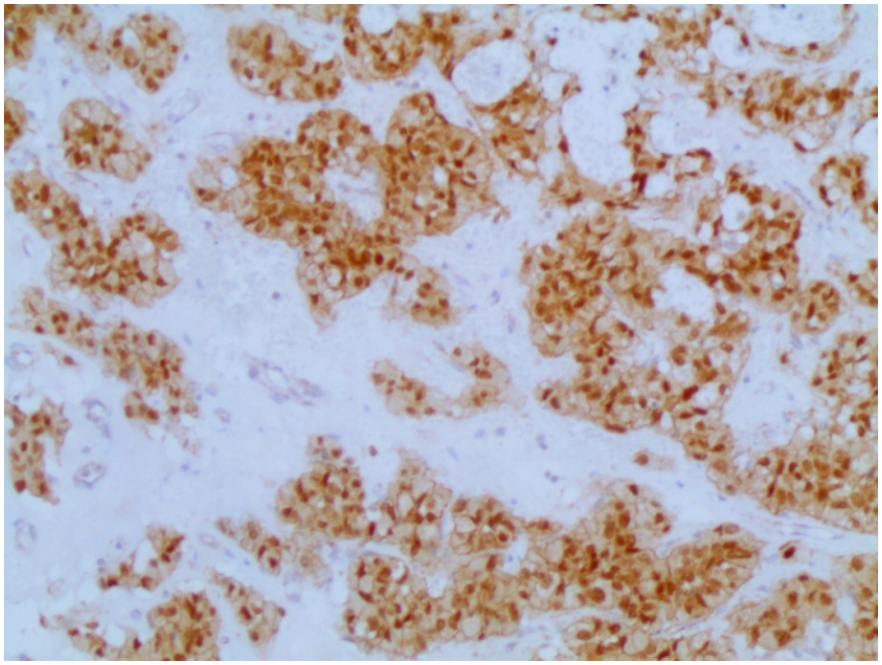


Figure 6. β-catenin nuclear staining of the tumor cells (x200)

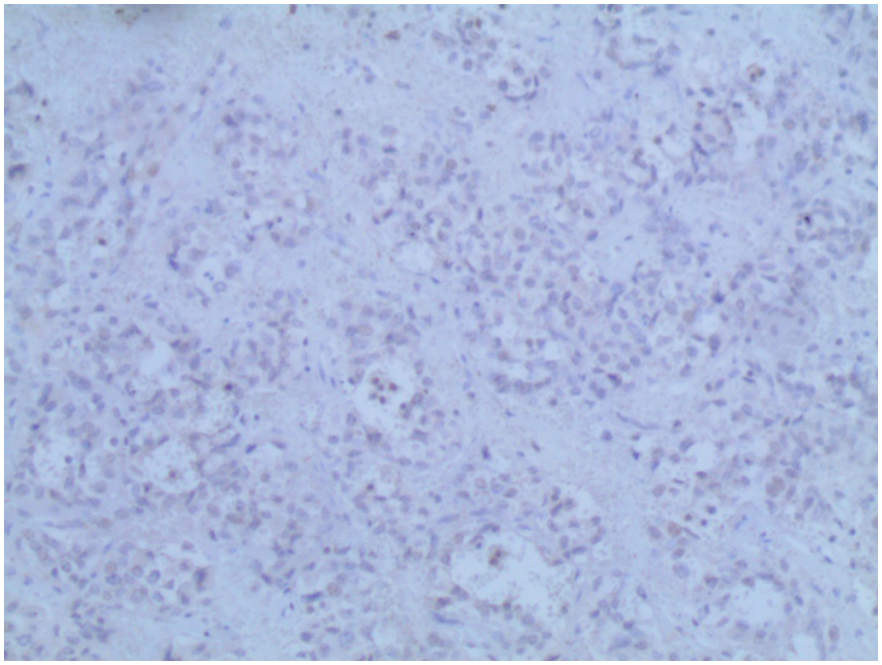


Figure7. Negativity of E-cadherin in the tumor cells (x200)