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Case report

Solid pseudopapillary tumors in the pathology department of the Universidad de Antioquia: Series of cases

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Abstract

Introduction: Solid-pseudopapillary tumors represent 1 %-2 % of all the pancreatic solid tumors, with low malignant potential. **Series of cases:** In the department of pathology at the Universidad de Antioquia were diagnosed sixteen cases between January 2004 and July 2019. Two of those cases were pediatric patients, one male with subsequent hepatic metastasis. Two additional cases were represented by two females older than 40 years old and the rest of the cases were females between 17 and 26 years old. One of the cases had sarcomatoid aspect cells, two others revealed multinucleate giant cells, and one last case of severe pleomorphism and presence of atypical mitoses. Capsule invasion was observed in 6 cases: one of the cases with lymphovascular invasion and the other 3 cases with perineural invasion. All cases showed hemorrhage or necrosis, and the immunohistochemical profile was positive for β-catenin, CD10, progesterone receptors, and CD56. Synaptophysin was focally positive.

Keywords

Solid-pseudopapillary tumor, Neoplasia.

INTRODUCTION

The pseudopapillary solid tumor, initially described by V.K. Frantz in 1959, was known by the eponymous until 1996 when the World Health Organization (WHO) designated it as a *solid pseudopapillary neoplasm*. It is a low-grade malignancy with uncertain cellular differentiation, which corresponds to 1% -2% of all solid tumors of the pancreas. It has a benign course with a 5-year survival of 95%, which is why it is considered resolved with surgical treatment; they metastasize in 10% to 15% of cases, almost exclusively to the liver and peritoneum, and are located mainly in the body and tail of the pancreas. It mainly affects women with an average age of 30 years[1-4].

SERIES OF CASES

In the Department of Pathology of the Universidad de Antioquia, between January 2004 and July 2019, 16 cases of pseudopapillary solid tumors of the pancreas were diagnosed. Two of the cases were in pediatric patients, one of them a male patient who, 7 years after diagnosis and surgery, attended the institution again having abdominal pain with tumor progression in the pancreas and liver metastases, capsular invasion and perineural invasion in the initial study. Two cases were women over 40 years old, and the rest were women between 19 and 26 years old. One of the cases was diagnosed in a woman at 21 years old after sur-

gery for abdominal trauma and evolution of more than 20 years with multiple seedings in the peritoneum.

Histologically, all cases presented a solid and pseudopapillary pattern, with epithelioid-like cells. One case had sarcomatoid-like cells, and another two had multinucleated giant cells. Nuclear pleomorphism was absent or mild in most cases. There was only one case with severe pleomorphism and the presence of atypical mitosis, which corresponds to a 56-year-old female patient. In the rest of the tumors, mitotic activity was scarce or null without atypical figures (**Figures 1-9**).

Regarding the morphological characteristics considered predictors of aggressive behavior, the invasion of the capsule was the most common, present in 6 (37%) of the cases; only one of them presented additional lymphovascular and perineural invasion in a male pediatric patient. All cases had hemorrhage or necrosis. Regarding immunohistochemical markers, all cases (100%) presented nuclear positivity for β-catenin, other positive markers were CD10 and CD56 antigens in 43% of all cases, and progesterone receptors were positive in 100% of the cases in which this marker was performed, which correspond to 25% of the total cases. Synaptophysin was positive in 4 cases, corresponding to 25% of all cases and 50% of cases in which it was performed; chromogranin A and C-kit were negative in all cases. Only three cases had the complete immunohistochemistry panel (β-catenin, CD10, progesterone receptors, C-kit, synaptophysin, and chromogranin A). The positivity of the previously exposed markers strongly correlates with what is described in the literature.

The immunohistochemical study was carried out in the immunohistochemical laboratory of the Department of

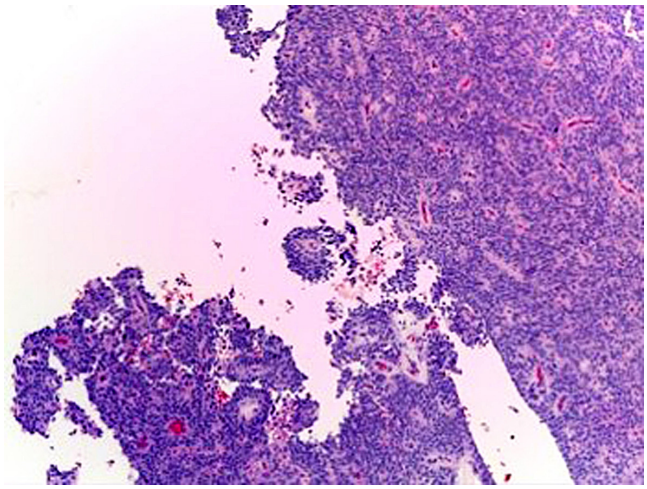

Figure 1. Solid and pseudopapillary pattern. Zoom: 4X.

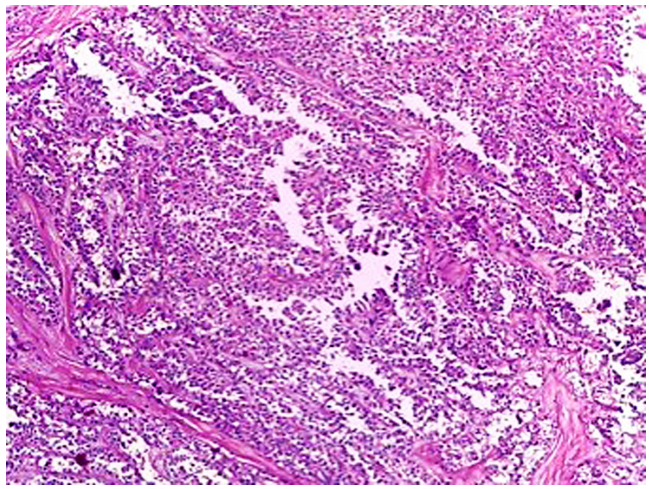

Figure 2. Pseudopapillary structures. Zoom: 10X.

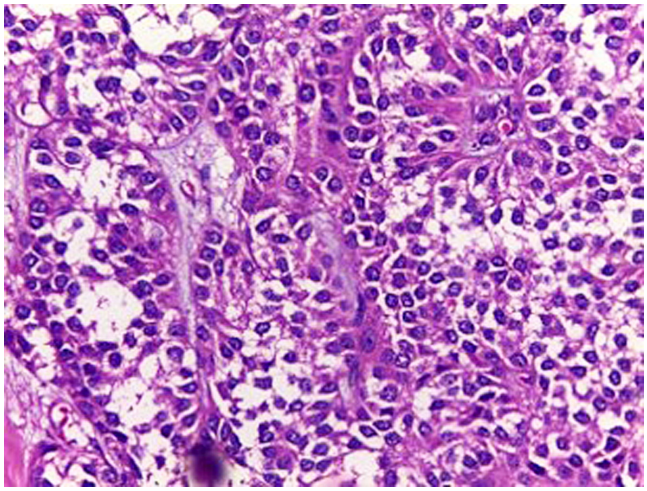

Figure 3. Pseudopapillary structures. Zoom: 40X.

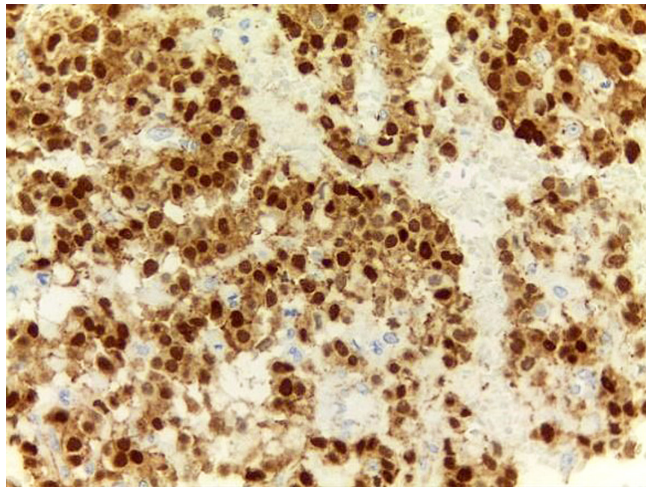

Figure 4. Nuclear and cytoplasmic positivity for β-Catenin. Zoom: 40X.

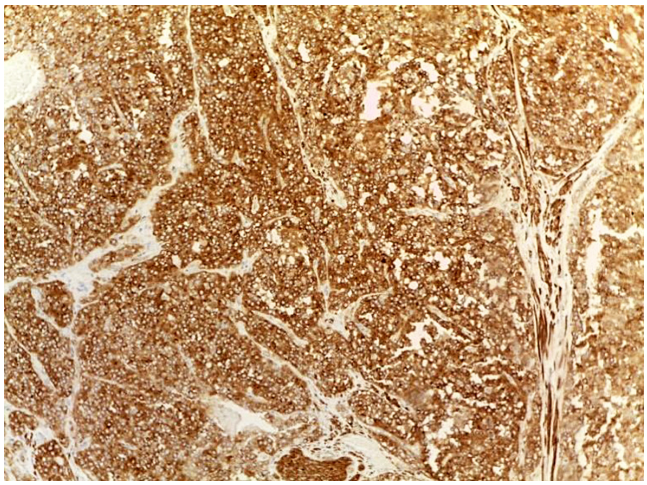

Figure 5. Cytoplasmic positivity for CD10. Zoom: 40X.

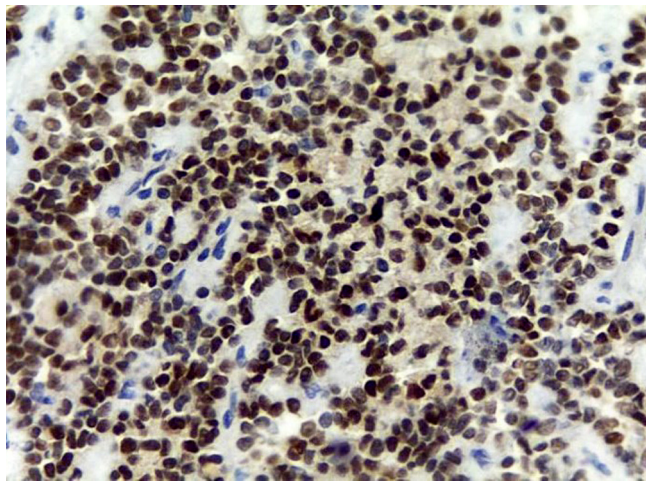

Figure 6. Nuclear positivity for progesterone receptors. Zoom: 40X.

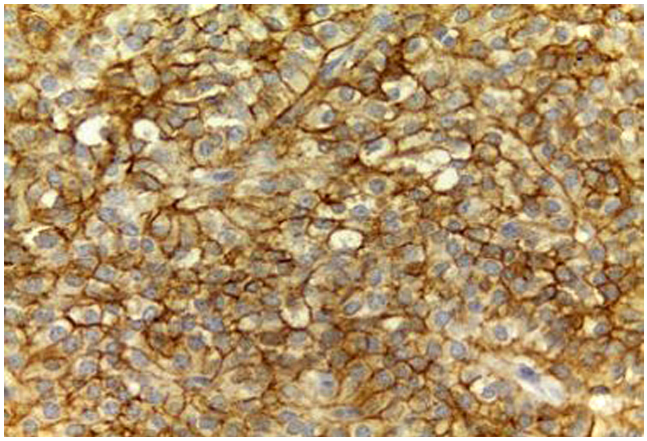

Figure 7. Membrane positivity for CD56. Zoom: 40X.

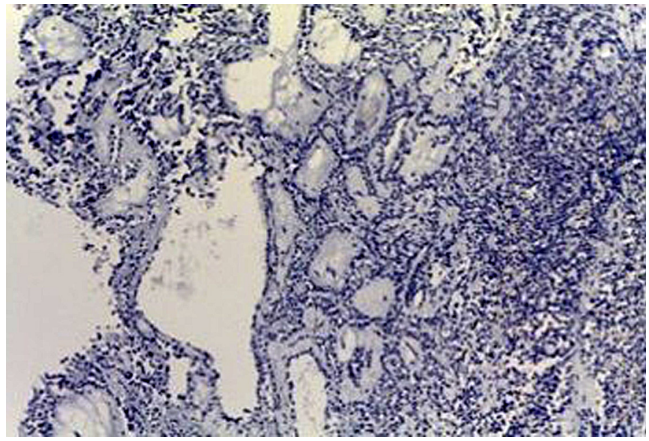

Figure 8. Negativity for synaptophysin. Zoom: 10X.

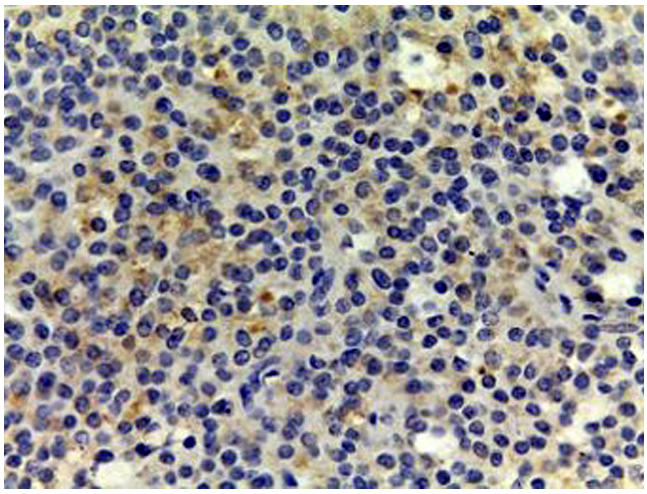

Figure 9. Negativity for chromogranin A. Zoom: 40X.

Pathology of the Universidad de Antioquia, with a manual technique. Due that the marker was not available at the institution at the time of diagnosis, there are cases that do not have a complete immunohistochemical profile because that (**Tables 1** and **2**).

DISCUSSION

Solid pseudopapillary tumor mainly affects young women, with a 9:1 ratio. It causes nonspecific abdominal symptoms, such as acute abdomen in case of rupture of the lesion due to trauma or an incidental finding in radiological images[1,4]. The computed axial tomography (CAT) and magnetic resonance imaging (MRI) of the abdomen shows a well-encapsulated mass with heterogeneous densities or intensities, reflecting cystic degeneration and hemorrhage within the

Table 1. Histopathological characteristics

Case	Architecture	Cell morphology	Nuclear pleomorphism	Atypical mitotic/mitosis activity	Intracytoplasmic hyaline blood cells	Capsule invasion	Perineural invasion	Necrosis/hemorrhage	Others
1	Solid and pseudopapillary	Epithelioid	Mild	Scant/no atypical mitosis	Present	No	No	Yes/No	Hemosiderophagues
2	Solid and pseudopapillary	Epithelioid	No	No	Absent	No	Yes	No/yes	Stroma sclerosis
3	Solid and pseudopapillary	Epithelioid	No	Scant/no atypical mitosis	Absent	Yes	Yes	No/No	*
4	Solid and pseudopapillary	Epithelioid	No	No	Absent	N/A	N/A	Yes/No	Myxoid degeneration
5	Solid and pseudopapillary	Epithelioid	No	Scant/no atypical mitosis	Present	No	No	Yes/yes	Hemosiderophagues
6	Solid and pseudopapillary	Epithelioid	No	Scant/no atypical mitosis	Present	Yes	No	Yes/No	Macrophages
7	Solid and pseudopapillary	Epithelioid	Mild	No	Absent	Yes	No	Yes/yes	Macrophages
8	Solid and pseudopapillary	Epithelioid and multinucleated giant cells	Mild	No	Absent	No	No	Yes/yes	Fibrosis, cholesterol crystals, and foamy macrophages
9	Solid and pseudopapillary	Epithelioid and multinucleated giant cells	Severe	Scant/atypical mitosis	Present	Yes	No	Yes/yes	*
10	Trabeculae, nests and organoid	Epithelioid and fusiform	Moderate	No	Absent	No	No	No/No	Sclerosis
11	Solid and pseudopapillary	Epithelioid	Mild	Scant/no atypical mitosis	Absent	No	No	No/yes	*
12	Solid and pseudopapillary	Epithelioid	No	No	Present	No	No	No/No	Hemosiderophagues
13	Solid and pseudopapillary	Epithelioid	Mild	Scant/no atypical mitosis	Absent	No	No	Yes/yes	Cholesterol crystals
14	Pseudopapillary	Epithelioid	No	No	Present	Yes	No	Yes/yes	Cholesterol crystals
15	Solid and pseudopapillary	Epithelioid	Mild	No	Absent	Yes	No	Yes/yes	Myxoid degeneration
16	Solid and pseudopapillary	Epithelioid	Mild	No	Absent	No	Yes	Yes/yes	Stroma sclerosis

*No other features were found.

tumor[5]. Macroscopically, they reach measures of 10 cm or more; they are well-circumscribed, solid and cystic masses; solid areas have a friable consistency, and cystic areas show necrosis and hemorrhage [5-7]. Microscopically, they have a solid, pseudopapillary architecture around thin blood vessels[8]. Neoplastic cells are discohesive, epithelioid-like with round nuclei and clefts, there is no significant nuclear atypia, and mitoses are rare. The cells tend to move away from the blood vessels and degenerate, resulting in extensive areas of necrosis; the cells that remain around the fibrovascular stems form one or multiple layers that give the characteristic pseudopapillary appearance of the lesion. The stroma can have varying degrees of hyalinization. There may be small groups of epithelioid histiocytes accompanying the lesion[1,4-8].

Table 2. Immunohistochemistry markers

Case	Vimentin	CD10	CD117/C-KIT	RP	β-catenin	Chromo-granin A	Synaptophysin	CD56	Cocktail CK
1	NO	NO	NO	NO	NO	NO	NO	NO	NO
2	NO	NO	NO	NO	+ nuclear and cytoplasmic	*	*	+ diffuse	*
3	+ diffuse	+ diffuse	NO	NO	+ nuclear and cytoplasmic	*	*	+ diffuse	*
4	NO	NO	NO	NO	NO	NO	NO	NO	NO
5	NO	NO	NO	NO	NO	NO	NO	NO	NO
6	NO	+ *dot-like*	*	NO	+ nuclear and cytoplasmic	*	+ focal	+ strong/ diffuse	NO
7	NO	+ *dot-like*	NO	+ strong/diffuse	NO	NO	NO	NO	NO
8	NO	+ diffuse	*	+ strong/diffuse	+ nuclear and cytoplasmic	*	*	NO	+ focal
9	+ focal	+ focal	*	+ strong/diffuse	+ nuclear and cytoplasmic	*	+ focal	+ diffuse	+ focal
10	*	+ diffuse	*	+ strong/diffuse	+ cytoplasmic	*	*	+ diffuse	+ focal
11	NO	+ diffuse	NO	NO	+ nuclear and cytoplasmic	*	+ diffuse	+ diffuse	NO
12	NO	NO	NO	NO	+ nuclear and cytoplasmic	*	+ focal	+ diffuse	NO

*Not performed (not requested by the pathologist at the time of the study).

Three histological variants have been described: Clear cell, pleomorphic, and oncocytic; none of them has been associated with a worse prognosis[4,9]. Histologic features that predict aggressive behavior have been described as a diffuse growth pattern with capsule invasion, lymphovascular and perineural invasion, extensive necrosis, high mitotic rate, and presence of sarcomatoid areas[1,6,10,11]. Electron microscopy has observed zymogen-type electrodense intracytoplasmic granules[3]. So far, the source cell has not been defined; some authors suggest that it is a pluripotential cell of the pancreas, while others suggest that it is an extrapancreatic cell that is introduced into the pancreas during organogenesis since tumors with similar characteristics have been described in the ovaries and testes[12-14]. Its pathophysiology consists of a loss of cell adhesion capacity due to mutations in the CTNND1 gene in exon 3 and alterations in the Wnt/ β -catenin signaling pathway[5,7,13].

For immunohistochemistry markers, β -catenin and progesterone receptors are characteristically positive in 100% of cases. They are positive for CD56 in 98%, synaptophysin can reach a focal positivity of up to 62%, while chromogranin A is positive only in 13% of cases, CD10 in 93% of cases, C-kit in 81%, and cyclin D1 in 71%; E-cadherin is positive only in 13% of cases; cytokeratins can be positive in 30% to 70%. Inhibin, carbohydrate antigen 19-9 (CA 19-9), mucin 6 (MUC6), caudal *homeobox* protein 2 (CDX2), octamer-binding transcription factor 4 (OCT4), and CD30 antigen are negative [1,4,8,15,16].

Differential diagnoses include some adenocarcinomas, neuroendocrine tumors, melanoma, PEComa, adrenal cortex tumors, and granulosa or sex cord cell tumors[1,2,4].

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