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FRANTZ TUMOR - A CASE REPORT

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Abstract: Frantz's Tumor, also known as solid pseudopapillary tumor, is a rare neoplasm (1 to 2% of exocrine pancreas tumors), more prevalent in females, occurring mainly between the second and third decade of life. This is a case report of a 51-year-old patient diagnosed with Frantz's Tumor after performing an abdominal tomography, treated at the University Hospital, Federal University of Sergipe (HU-UFS). The patient in question underwent a surgical procedure (partial body-caudal pancreatectomy and splenectomy), with good postoperative evolution. Her diagnosis was confirmed by immunohistochemistry.
Keywords: Pancreatic neoplasm. Surgery. pancreas. Immunohistochemistry.

INTRODUCTION

Solid pseudopapillary tumor of the pancreas is a rare epithelial tumor with low malignancy potential and undetermined behavior. [1],[2],[3]. This neoplasm occurs mainly in females. [1],[2],[3], with a predominant age group between the second and fourth decade of life[1],[2]. Treatment consists of surgical resection [7], which, due to the excellent prognosis and the low rate of recurrence after the procedure, is usually curative, with a 5-year survival greater than 97%. [1],[2],[4].

CASE REPORT

A 51-year-old female patient with nonspecific and insidious abdominal pain for 2 years. For this reason, an abdominal tomography was performed, which showed a nodular image in the pancreas tail topography, with soft tissue density, lobulated contours, contrast uptake, measuring 4.8 x 4.7 x 4.5 cm. She was submitted to partial pancreatectomy (body-caudal) and splenectomy, with anatomopathological description describing epithelioid and spindle cell neoplasm, measuring 6.4 cm in its longest axis, with mild cellular pleomorphism, absence of necrosis, low mitotic index (1 mitosis|10 CGA), absence of angiolymphatic invasions, neoplasm-free margins.

The material was sent for immunohistochemistry, which revealed expression for b-catenin in nuclear and cytoplasmic pattern, positivity for progesterone receptor and loss of e-catenin expression, with a confirmed solid pseudopapillary tumor of the pancreas (Frantz Tumor).

DISCUSSION

Solid pseudopapillary tumor of the pancreas is a rare epithelial tumor with low malignancy potential and undetermined behavior. [1],[2],[3]. Also known as Frantz Tumor, a reference to the person who first described this tumor in 1959, it received its current name in 1996 from the World Health Organization. (OMS)[2],[3] [4].

In recent years, the incidence of this pathology has increased, it is believed that due to better diagnostic techniques, such as imaging and immunohistochemistry, in addition to a better understanding of the clinical aspects involved. Frantz's tumor comprises 5% of pancreatic cystic tumors [5], 1 to 2% of exocrine tumors of the pancreas [1],[2] and is responsible for 13% of surgical resections of pancreatic lesions. [1]

Solid pseudopapillary tumor occurs mainly in females [1],[2],[3], in a ratio of 10:1 in relation to males, with a predominant age group between the second and fourth decade of life [1],[2]. It is rare for children and adolescents to be affected, but there is a description in the literature, with some studies reporting 20 to 25% of cases. [2] [4],[5]. This patient. A case report is outside this predominant age group, with a slightly older age (51 years old).

Its origin still remains unknown, but it is speculated that this tumor may be related to female sex hormones, due to the predominant

age group and the frequent presence of progesterone receptors.

[1,2]. In addition, some authors argue that this neoplasm originates from epithelial cells, arising from primitive acinar pancreatic cells. [2]. There is usually no association No with specific laboratory tests or tumor markers [56], as well as endocrine and exocrine changes [2].

Patients with Frantz Tumor may present with a nonspecific clinical picture, with complaints of insidious or acute abdominal pain and progressive increase in abdominal volume, with perception of a palpable mass and gastrointestinal symptoms. However, an important percentage of cases are asymptomatic and the diagnosis is made through imaging tests, such as computed tomography or magnetic resonance imaging, performed for other reasons.[1,2,4]. A similar fact happened with the patient in question, who had her diagnosis suspected after performing an abdominal tomography, due to poorly characterized abdominal pain. Obstructive jaundice and hemorrhagic acute abdomen are rare[2].

This neoplasm presents as an encapsulated mass, well delimited in relation to the pancreas, heterogeneous, with solid, cystic and pseudopapillary content. [1,2,4], with varying degrees of hemorrhagic degeneration and calcification [6]. They can be bulky, with an average size of 10 cm (ranging from 2-20 cm), even causing compressive symptoms. In this case report, the tumor size was 6.4 cm in its longest axis, consistent with data found in the literature, without associated compressive symptoms. Some studies associate tumor size with malignancy potential[7]. Its most common topography is the tail of the pancreas, as occurred in this case report, followed by the body and head of that organ. [1,2].

The use of endoscopic ultrasound-guided fine needle aspiration as a diagnostic aid is still controversial in the literature. [1,2]. Some authors argue that it would be a good resource in cases of cystic pancreatic lesions of unknown origin. [3,5,7], helping to differentiate benign from malignant lesions, which may change the therapeutic approach [4]. On the other hand, this practice is questioned by some authors, alleging the risk of tumor implants and dissemination at the puncture site. [1,4]. The aforementioned patient did not undergo this procedure.

Histological findings show solid areas with pseudopapillary and pseudocystic structures, cellular degeneration, cells with exocrine endocrine characteristics [1,4]. Some findings that may suggest a worse prognosis are the presence of extensive necrosis, nuclear atypia, high mitotic rate, high Ki 67 indices, sarcomatous areas, and vascular and perineural invasion. [1,2].

Immunohistochemistry has become a great tool, being responsible for confirming the diagnosis after resection of suspicious lesions. In it, positivity for progesterone receptors, vimetine, alpha1-antirtipsin, neurospecific coils, among others [1,2]. In this report, the immunohistochemistry was in agreement with the literature, showing progesterone and b-catenin receptors.

Treatment consists of surgical resection [7], which, due to the excellent prognosis and the low rate of recurrence after the procedure, is usually curative, with a 5-year survival greater than 97%[1,2,4]. Pancreatoduodenectomy or distal pancreatectomy (with or without splenectomy) is generally chosen, depending on the tumor location; lymphadenectomy is not routinely necessary [1,4,5], although some authors suggest nodal resection in patients with suspicious lymph nodes or in tumors larger than 5 cm [7]. As she had a tumor in body-caudal topography, the aforementioned patient underwent a partial pancreatectomy (body-caudal) and splenectomy.

Furthermore, even in situations of

asymptomatic disseminated disease or unresectable tumors, the literature demonstrates a high survival rate in most cases. [2],[3], despite some studies reporting a median survival of 5.7 years for patients with residual disease [4].

Tumor invasion to adjacent organs is rare [2],[4] and, on these occasions, differential diagnoses must be evaluated, such as the adrenal masses, hepatic cysts, and other pancreatic lesions. [2] The occurrence of metastases is uncommon, with some studies indicating occurrence in 10 to 15% of tumors, being more prevalent in males. [5]. The most common metastatic foci are the hepatic, lymph node, and peritoneal foci. [1],[7], with reports of pulmonary and cutaneous metastasis in the literature [4]. The surgical proposal of the patient in question, similarly to what the literature shows, was curative and the anatomopathological examination showed an anatomical specimen with free margins.

Adjuvant treatment is usually not indicated due to the high resectability of the tumor. [1],[2]. There are reports of the use of chemotherapy and radiotherapy in tumors that were not fully resectable, [1] with benefits still limited to a few clinical cases [4].

CONCLUSION

The solid pseudopapillary tumor is a rare tumor, with a low degree of malignancy and uncertain behavior, with insidious growth and a non-specific clinical picture. Its epidemiology involves young women, and its appearance on imaging tests is very characteristic (heterogeneous, well-delimited mass with solid, cystic and pseudopapillary content), which leads to diagnostic suspicion, confirmed by immunohistochemistry after surgical resection, which is the treatment of choice, with a curative proposal. Other information, such as the etiopathogenesis and the impact of adjuvant treatment of these tumors, still seem uncertain, as well as the role of FNA in diagnosis is controversial. Such questions need further studies for elucidation.

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