

Pancreatic hemangioma manifesting as variceal gastroesophageal bleeding during pregnancy: case report

Hemangioma pancreático, manifestando com hemorragia digestiva alta varicosa durante gestação: relato de caso

LETÍCIA DE CAMPOS FRANZONI,¹ CLARISSA RIBEIRO VILLAR,² FERNANDA PLAZZI CARRARETO,³ FABIO DA SILVA YAMASHIRO,⁴ CÁSSIO VIEIRA DE OLIVEIRA,⁵ LIGIA YUKIE SASSAKI,⁶ FERNANDO GOMES ROMEIRO⁷

SUMMARY

There are only 10 reported cases of pancreatic hemangiomas in adults, only one of them causing digestive bleeding. We present a case of variceal bleeding and portal hypertension caused by a pancreatic hemangioma. The patient had 19 year-old and was received at her 16th week of pregnancy. She had massive hematemesis, controlled after variceal band ligation. Her image exams revealed a cystic lesion of 164 cm³ in the pancreas tail and signs of portal hypertension. Two months after, the ultrasonographic exam documented the lesion growth, achieving 200 cm³ at that time. The patient was submitted to distal pancreatectomy, and the histopathological analysis revealed a pancreatic hemangioma of 11 x 9 x 8 cm. Therefore, we report the second case of digestive bleeding caused by a pancreatic hemangioma, which had a well documented growth during the pregnancy. Additionally, we review the previous reports of pancreatic hemangiomas and discuss the hypothesis of hormonal influence on the natural history of these tumors.

Keywords: Pancreatic Hemangioma, Portal Hypertension, Esophageal Varices, Gestation.

RESUMO

Os pseudotumores inflamatórios são um conjunto de lesões raras, caracterizadas histologicamente por prolifera-

ção de fibroblastos e células inflamatórias permeadas por estroma fibroso. Nos casos de localização hepática, estas massas são difíceis de diferenciar de outras lesões, especialmente de neoplasias primárias ou secundárias. Descrevemos um caso em que os achados clínicos-laboratoriais e as imagens da ultrassonografia e da tomografia computadorizada de abdomen não foram suficientes para afastar uma neoplasia, e só foi possível estabelecer o diagnóstico através da biópsia direta, mediante ressecção cirúrgica da lesão. Diante de um paciente sem antecedentes dignos de nota, com massa hepática e marcadores tumorais negativos, o pseudotumor inflamatório de fígado deve ser lembrado sempre como um possível diagnóstico diferencial.

Unitermos: Hemangioma Pancreático. Hipertensão Portal, Varizes Esofágicas, Gestação.

INTRODUCTION

Hemangiomas are vascular tumors commonly seen in the liver but rarely find in the pancreas. Together with other non-epithelial tumors, they compose only 0.1% of all pancreatic neoplasias.¹ There is some controversy about the growth factors involved in hemangiomas development, but one of the proposed hypotheses is the tumor stimulation by sexual hormones. The hemangiomas proliferation is more pronounced at childhood, and in

1-7. Faculdade de Medicina da Universidade Estadual Paulista (UNESP), Department of Internal Medicine, Gastroenterology Division, Botucatu/SP, Brazil. **Corresponding author:** Letícia de Campos Franzoni - Depto. de Clínica Médica - Distrito de Rubião Jr. s/n Zip code 18 618 970 - e-mail: leticiafranzoni@hotmail.com **Recebido em:** 10/10/2012. **Aprovado em:** 14/11/2012.

the adult phase they tend to have a slow regression. Being so, these tumors are prevalent in children but infrequent in adults. In the clinical practice, these tumors are rarely considered as a cause of disease, because its symptoms are inespecific. As a result, the majority of cases is diagnosed incidentally as a cystic pancreatic lesion in imaging exams.

Nowadays there are only 10 reports about pancreatic hemangiomas in adults. Most of them had abdominal pain, and only one had gastrointestinal bleeding. We report the first case noticeable as a variceal gastrointestinal bleeding in a pregnant woman with segmental portal hypertension caused by a pancreatic hemangioma. Additionally, we discuss the impact of the hormonal influence on the se tumors.

INTRODUÇÃO

Hemangiomas são tumores vasculares comuns no fígado e raros no pâncreas, representando junto com os demais tumores não-epiteliais 0,1% de todas as neoplasias pancreáticas.¹ Além disso, são prevalentes em crianças e infrequentes em adultos, pois tendem a proliferar durante a infância e a involuir lentamente na fase adulta. Seus fatores de crescimento são controversos, sendo uma das hipóteses a estimulação por hormônios sexuais. Tais tumores são raramente suspeita clínica devido aos seus sintomas inespecíficos. Como resultado, a maioria é diagnosticada incidentalmente como massa pancreática cística em exames de imagem.

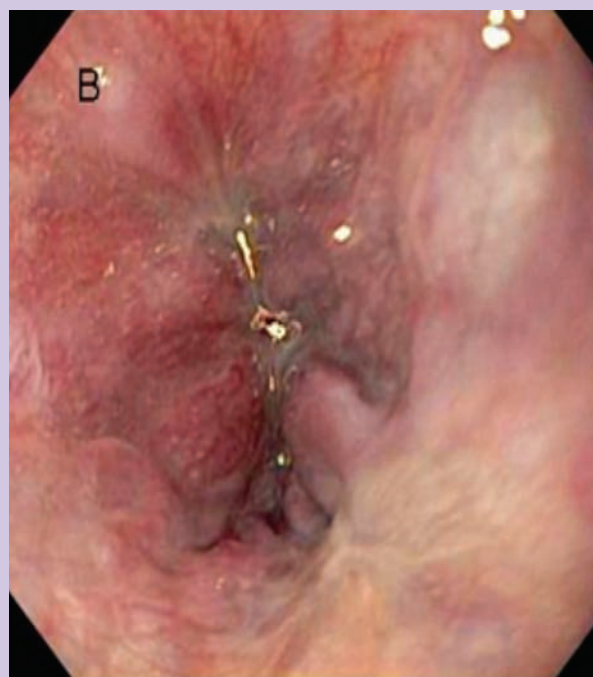
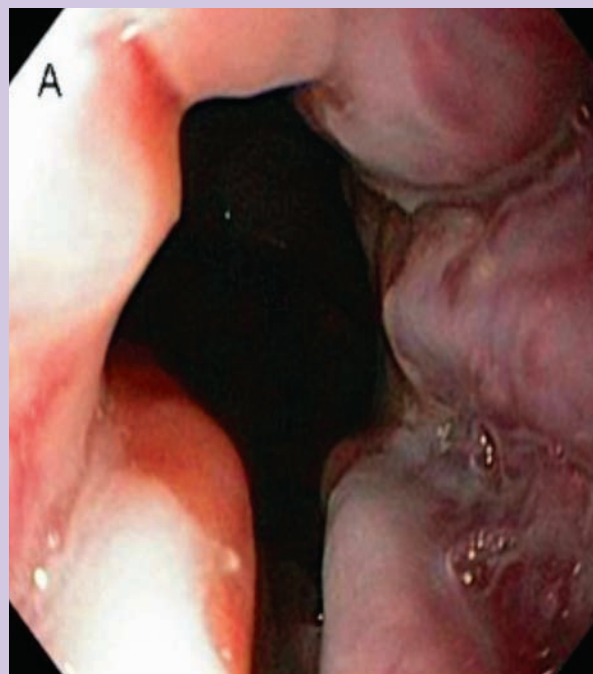
Existem apenas 10 casos relatados na literatura de hemangioma pancreático em adultos. A maioria apresenta dor abdominal, e somente um caso manifestou-se com hemorragia digestiva. Relatamos o primeiro caso que se manifestou com hemorragia digestiva alta varicosa, ocorrido em gestante que apresentava hipertensão portal segmentar secundária a hemangioma cavernoso em cauda pancreática. Discutimos também a influência hormonal nos hemangiomas.

CASE REPORT

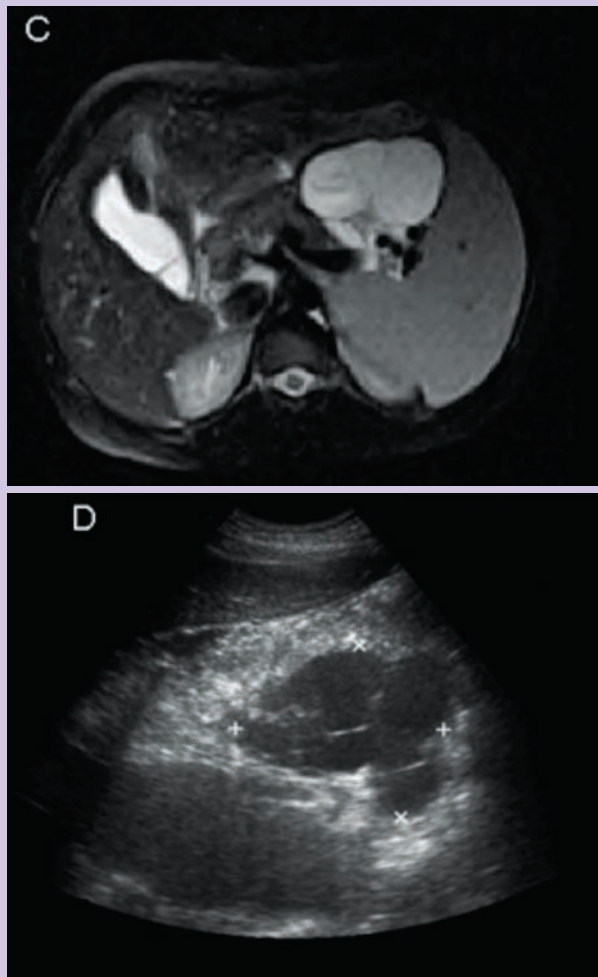
The patient was a previously healthy 19 years old woman who presented a large hematemesis during her 16th week of pregnancy. Until this moment, she never used any medications. There were no signals of liver injury in her laboratorial exams.

At upper gastrointestinal endoscopy (UGE) 4 esophageal vessels of large diameter were found, with “red spots” and “cherry red spots” (figure 1A). In the stomach there were gastroesophageal varices (GOV), enlarged vessels at the gastric body and portal hypertensive gastropathy.

Figuras: A - Endoscopic esophageal image at the hospital admission showing 4 large varices with red spots. B - Endoscopy 4 weeks after the band ligation showing reduction of the vessels size.



Figuras: C - MRI (0.5 T - T2 image) at the 18th week of pregnancy revealing the cystic lesion in the pancreas tail, with 10.0 x 6.8 x 6.6 cm. D - Ultrasonographic image at the 24th week of pregnancy confirming the lesion growth, at that time with the estimated volume of 200 cm³.



The patient received blood transfusions and rubber band ligation was performed to control the esophageal bleeding. The initial ultrasonographic images showed a cystic lesion in the pancreas tail, without detectable flux at the Doppler study. The lengths were 7.9 x 6.1 x 6.5 cm (estimated diameter of 164 cm³).

The patient had no signs of splenomegaly or any liver disease at present. Ten days after, a magnetic resonance imaging exam (MRI) was performed, now showing homogeneous splenomegaly and a pancreatic cystic mass. The lesion had delicate septa and lobulate contours, but at this time the measures were 10.0 x 6.8 x 6.6 cm (figure 1C). Considering the cystic aspect on MRI T2 images, the initial diagnostic hypothesis was serous pancreatic cystadenoma.

As the variceal bleeding was controlled, a beta-blocker was initiated and the patient was maintained in endoscopic follow up to avoid further bleeding.

Four weeks later, a new endoscopic exam showed reduction in the esophageal vessels diameter (figure 1B), but the gastric varices were growing. The ultrasonographic images were repeated at the 24th week of pregnancy, showing a significant growth of the pancreatic mass, which now had 200cm³ (figure 1D).

Considering the rapid growth of the pancreatic lesion and the worsening of the segmental portal hypertension, distal pancreatectomy and splenectomy were performed. The histopathological analysis revealed a pancreatic hemangioma of 11 x 9 x 8 cm with 249.7 grams, located at the pancreas tail. The surgery was performed after the birth delivery, which occurred at the 33rd week of pregnancy without any other problems.

DISCUSSION

The vascular pancreatic neoplasias include hemangiomas, lymphangiomas, hemolymphangiomas, hemangioblastomas and hemangiosarcomas. They are cystic lesions that together compose 0,1% of all pancreatic tumors and must be distinguished of the common cystic lesions seen in this organ, as the pancreatic pseudocysts after pancreatitis.¹

Others differential diagnoses are the serous cystadenomas and the mucinous cystadenomas. Hemangiomas are histologically formed by the aggregation of blood vessels irregularly arranged. The lesions of more than 5 cm can cause complications, as the extrinsic compression of other structures, rupture after traumatic lesions, and the Kasabach-Merritt syndrome (microangiopathic anemia, thrombocytopenia and consumptive coagulopathy), which need treatment even with their histological benign characteristics.

The pancreatic hemangiomas in adults are very rare, and until to now there are only 10 published cases.² The most of these patients was women (70%), and the age at diagnosis varied between 30 and 79 years. Most cases showed unspecific symptoms, but the abdominal pain was present in 70% of them. Few patients had nausea and thrombocytopenia (10%) and only one had gastrointestinal bleeding. In two thirds of these patients the lesions were located at the pancreatic head, and the others had the lesions in the body or in the tail.¹ The clinical data, location and diameters of these lesions are presented in the table 1.

Table 1. Demographic aspects, initial presentation, location and sizes of pancreatic hemangiomas of the 10 reports published. * = without available information.

Year	Author	Gender/age	Initial presentation	Location	Size (cm)
1939	Ranström ³	F/61	Incidental (autopsy)	Head	7,0 x 7,0
1961	Ringoir ⁴	F/61	Hematemesis/melena	Head	15,0
1972	Colardyn ⁵	F/61	Abdominal pain	Body/ tail	*
1985	Mangin ⁶	F/61	Nausea/thrombocytopenia	Head/body/ tail	20,0 x 7,0
1991	Kobayashi ⁷	F/61	Abdominal distention	Head	20,0
1991	Dageförde ⁸	F/61	Abdominal pain	Body/ tail	6,0 x 3,0
1991	Chang ⁹	F/61	Epigastric discomfort	Body/ tail	4,0 x 3,2
1991	Plank ¹⁰	F/61	Abdominal pain	Head	3,0
1991	Mundinger ¹	F/61	Epigastric and dorsal pain	Head	6,2 x 5,3
1991	Weidenfeld ²	F/61	Abdominal (left flank) pain	Uncinate process	5,5 x 4,0

Our case is the first report of variceal esophageal bleeding secondary to segmental portal hypertension caused by a pancreatic hemangioma. Furthermore, we documented adequately the lesion growth associated to the hormonal stimulus during pregnancy. Until to now, the hormonal influence on these tumors was documented only in few published reports. In one of these papers, the authors evaluated prospectively 94 women with hepatic hemangiomas during a medium time of 7.3 years. The lesions growth was documented in 23% of the patients who received hormonal therapy, but only in 10% of the patients that not received it.

Hemangiomas were too reported among pregnant women who had received medications to stimulate the ovulation (human chorionic gonadotrophin or clomiphene citrate).¹¹ Even with this probable association between the tumors and the estrogenic stimulus during pregnancy, a clear documentation associating the hemangiomas growth to the influence of the female sexual hormones is still lacking, because few

imaging exams can be used during pregnancy. The adequate documentation in case reports could be the initial step to find new conclusions about this issue in the future.

In conclusion, we report the first case of pancreatic hemangioma in an adult patient, which caused variceal gastrointestinal bleeding secondary to segmental portal hipertension. We showed the rapid tumoral growth during pregnancy, suggesting an important hormonal influence in the hemangiomas, according to the hypothesis of previous studies.

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