Asimmetria addominale in una ragazza di 17 anni

An abdominal asimmetry in a 17 years old girl

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Riassunto

Il tumore pseudopapillare solido pancreatico (TPSP) è una neoplasia rara tipica di giovani donne in età adulta (solo 12 casi pediatrici descritti dal 2000 al 2009). Può metastatizzare e recidivare. La prognosi è generalmente buona dopo asportazione chirurgica radicale. Presentiamo un caso clinico sottolineando l'importanza di pensare al TPSP nella diagnosi differenziale di masse retrogastriche peripancreatiche soprattutto in femmine puberi.
Descriviamo il caso di un'adolescente con riscontro di una massa addominale rivelatosi istologicamente un tumore pancreatico raro.

Abstract

The pseudopapillary pancreatic solid tumor (TPSP) is a rare malignancy typical of young adult women (only 12 pediatric cases from 2000 to 2009), it can recur and metastasize. The prognosis is usually good after radical surgical removal. We emphasize the importance of TPSP in differential diagnosis of retrogastric, peripancreatic masses especially in puberal females.
We describe the case of an adolescent girl with an abdominal mass revealed as a rare pancreatic neoplasia.

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Case report

A 17 th year old girl was referred for an abdominal asymmetry. Her history was remarkable for a dyspepsia dating since 4 weeks, with a concomitant a bulimia started few years before. No trauma history was referred. On admission at the physical examination a firm epigastric abdominal mass was present. An ultrasonography of the abdomen was performed showing a voluminous retrogastic peripancreatic, mass partially cystic . A CT scan (Fig. 1) and diagnostic laparoscopy was performed. The first hypothesis was a gastric duplication or a pancreatic neoformation. An open surgery convertion was needed for multiples pancreatic adherences and for the intraoperative histology suggestion of malignancy. A radical mass enucleation was made (Fig. 2) and the final histology showed a solid pancreatic pseudopapillary tumor (TPSP) (Fig. 3). After two days a post-operative pancreatitis and haemoperitoneum required a second laparotomy. Pancreatitis resolved with medical therapy with a persistence of a small retrogastric liquid collection at discharge. After a week the girl

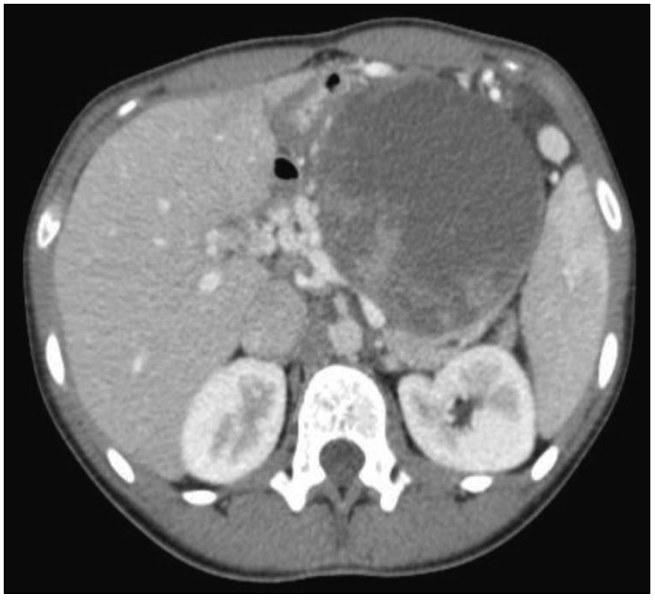

Figura 1.
Pre-operative axial CT scan of the mass

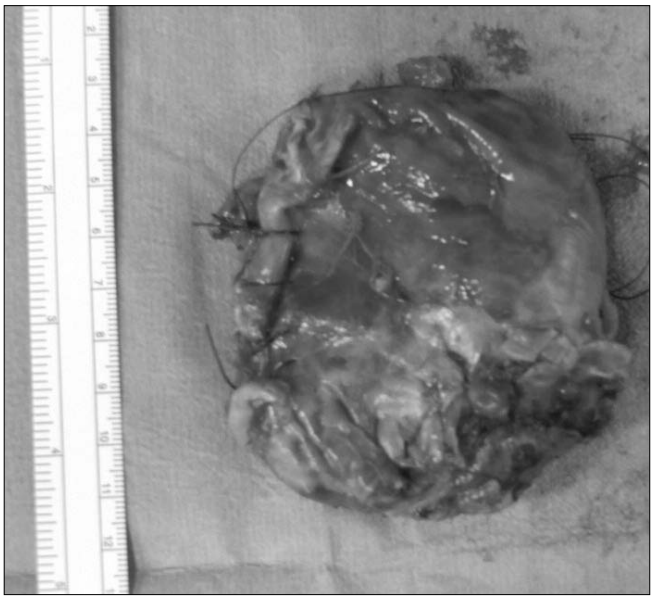

Figura 2.
Macroscopic image of the neoplasia after radical excision

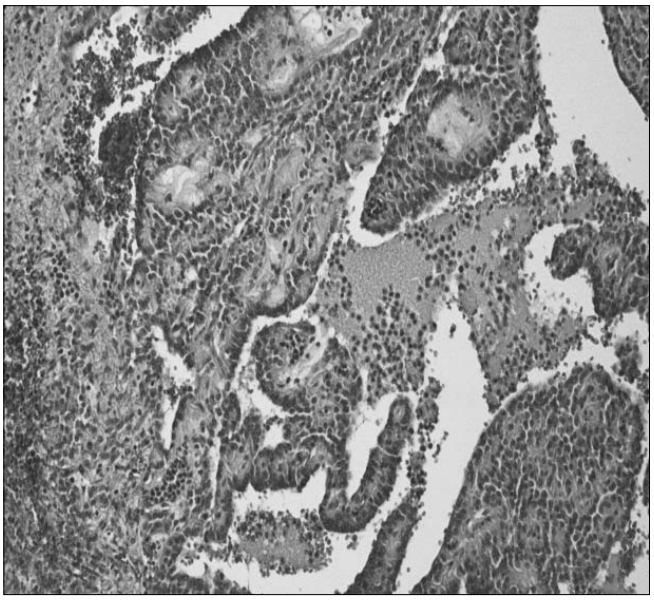

Figura 3.
Histological findings of the biopsy specimen

come back for an abdominal pain and yellow secretions from surgical wound. No metastasis but a 8 cm peripancreatic pseudocystic collection were revealed on abdominal and thoracic scan. Medical therapy with gabesato mesilato, fasting, parenteral nutrition and antibiotic endovenous therapy for 22 days allowed a significant reduction of the mass. At discharge and after 8 months the patient is asymptomtic and no lesions is present at abdominal RMN.

Conclusion

This case underlines the relevance of TPSP in differential diagnosis of retrogastric and peripancreatic lesions in puberal girls. The mass adherences and the dimensions impose a open surgery to radically enucleate the neoplasia according to the oncologic criteria. For this neoplasia, first described by Frantz in 1959[1] , standard treatment consists in radical excision of the mass even to confirm the diagnosis. Although preoperative diagnosis is difficult by computed tomography, magnetic resonance imaging may potentially improve this situation[2]. Prognosis is good after excision even with metastasis[3] or local recurrences After pancreatic surgery it is important to consider postoperative pancreatitis and its complications (haemoperitoneum, late peripancreatic collections) to limit fatal events. A solid, cystic or mixed form of the neoplasia exists and it represents 2-3% of pancreatic primitive tumors occurring at all ages.[4] At least 4-yearly follow-up is mandatory for all patients undergoing surgical resection.[5]

Remarkably only a case of spontaneous regression shrinkage is reported in literature.[6] This is a very rare disease and only 12 pediatric cases were reported from 2000 to 2009 in Italy.[7]

References

1 Cervantes-Monteil F, Florez-Zorrilla C, Alvarez-Martínez I. "Solid-cystic pseudopapillary tumor of the pancreas: acute post-traumatic presentation. Case report and review of the literature". Rev Gastroenterol Mex 2002; 67: 93-6

2 Ng KH, Tan PH, Thng CH, Ooi LL. "Solid pseudopapillary tumour of the pancreas". ANZ J Surg 2003; 73: 410-5

3 Faraj W, Jamali F, Khalifeh M, Hashash J, Akel S. "Solid pseudopapillary neoplasm of the pancreas in a 12-year-old female: case report and review of the literature". Eur J Pediatr Surg 2006; 16: 358-61

4 Vergauwen W, Op de Beeck B, Hagendorens M, Wojciechowski M, Vaneerdeweg W, Ramet J. "A solid pseudopapillary tumour of the pancreas presenting after an abdominal trauma". Acta Chir Belg 2010; 110: 390-3.

5 Yang F, Fu DL, Jin C, Long J, Yu XJ, Xu J, Ni QX. "Clinical experiences of solid pseudopapillary tumors of the pancreas in China". J Gastroenterol Hepatol 2008; 23: 1847-51 Epub 2008 Aug 24

6 Nakahara K et Al. "Solid pseudopapillary tumor of the pancreas showing a remarkable reduction in size over the 10-year-follow-up period". Intern Med 2008; 48: 1335-39, Epub 2008 Jul 15

7 Dall'Igna P, Cecchetto G, Bisogno G, et al. "Pancreatic tumors in children and adolescents: the Italian TREP project experience". Pediatr Blood Cancer 2010; 54: 675-80