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Rare case of solid pseudopapillary pancreatic tumour (Gruber-Frantz tumour)

Rijetki slučaj solidnog pseudopapilarnog tumora (Gruber-Frantz tumor)

Alma Mekić-Abazović, Hakija Bečulić, Senad Dervišević, Lejla Hantalašević-Bečić*

Summary

In this paper we presented a patient with a very rare form of pancreatic tumour. The disease began with very non-specific abdominal pain, loss of appetite, and vomiting, mostly mucus. Proximal endoscopy was performed which revealed hiatal hernia and ulcer in the duodenum. Computed tomography of the abdomen showed a tumour formation in the tail of the pancreas, which was surgically removed. Histopathological verification confirmed that it was a solid pseudopapillary tumour of the pancreas. The patient is on regular oncological control.

Ključne riječi: pancreas, pseudocyst, solid pseudopapillary tumour

Sažetak

U radu smo prikazali pacijenta s vrlo rijetkom formom tumora pankreasa. Bolest se manifestirala nespecifičnim abdominalnim bolom, gubitkom apetita, te povraćanjem sluzavoga sadržaja. Uradili smo proksimalnu endoskopiju kojom su dijagnosticirani hijatalna hernija i duodenalni ulkus. Kompjuterizirana tomografija je pokazala tumorsku formaciju u predjelu repa pankreasa. Tumor je kirurški uklonjen. Patohistološka analiza je pokazala da se radi o solidnom pseudopapilarnom tumoru pankreasa. Pacijent je pod redovitom kontrolom onkologa.

Key words: pankreas, pseudocista, solidni pseudopapilarni tumor

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Introduction

Gruber-Frantz tumour (Solid pseudopapillary tumour – SPPT) is a very rare neoplasm of the pancreas. The incidence is very low. It accounts for 0.2% - 2.7% of primary pancreatic tumours with slow growth rate and excellent prognosis.[1] In literature it is presented with about 150 cases in the world.[2] SPPT is usually seen in young females and is usually misdiagnosed as pancreatic pseudocyst, especially when symptoms are absent.[3]

The first classification of these tumours was performed by Robson and Moynihan in 1903 and completed by Beust in 1915.[2] Surgical resection is the treatment of choice for these tumours.[4] In 1996, the World Health Organization (WHO) renamed this tumour as SPPT for the international histological classification of tumours of the exocrine pancreas.[4]

In this paper we presented a rare case of SPPT of the pancreas. It was presented with unspecific symptoms. Extended diagnostic evaluation was also unspecific and could mislead an adequate therapeutic approach.

* **Cantonal hospital Zenica,** Bosnia and Herzegovina, Department of oncology, haematology and radiotherapy, (Ass. prof. Alma Mekić-Abazović, M. D.)**,** Department of neurosurgery (MSc Hakija Bečulić, M. D.), Department of surgery (Primarius Senad Dervišević, M. D.), Department of internal medicine (PhD Lejla Hantalašević-Bečić, M. D.)

Correspondence address / Adresa za dopisivanje: Alma Mekić-Abazović, Kantonalna bolnica Zenica, Odjel onkologije, hematologije i radioterapije, Crkvice 67, 72000 Zenica, Bosnia and Herzegovina, E-mail: dr.alma.kbz@gmail.com

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Case report

A 21-year-old female was admitted to the Department of Surgery in December, 2011. She complained of upper abdominal pain for the last four months and vomiting of slime. She had loss of appetite and weight of a few kilograms. She visited the family doctor who referred her to diagnostics. Proximal endoscopy was performed which showed duodenal ulcer and hiatus hernia.

Contrast enhanced computed tomography (CT) scan revealed a well-defined, non-enhancing solid pancreatic mass lesion in the tail region which measured 42 x 37 x 60 mm. Liver, spleen and kidneys were normal. There was no evidence of abdominal lymphadenopathy (Picture 1). Thoracic radiography and laboratory findings were normal.

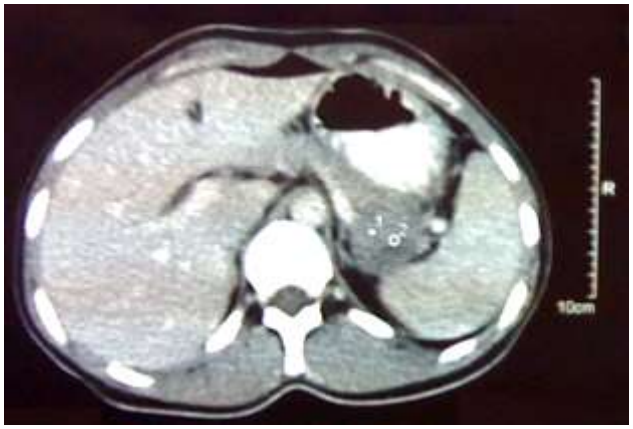
After preoperative evaluation, distal pancreatectomy and splenectomy were performed in the middle of December, 2011. In the postoperative period the patient was admitted to the Intensive Care Unit (ICU) for five days. The postoperative course was uneventful and on the ninth postoperative day she was discharged from hospital.

Histopathology of resected lesion revealed an encapsulated tumour in the tail of the pancreas of about 5 cm diameter, which had cystic and solid components. The cystic contents were pulpy fluid. The tumour was located on the edge of a thin connective capsule.

Histologically, the tumour had two components. The first component was formed from monomorphic cells, oval to elongate which showed round to oval nuclei with dispersed chromatin. The tumour cells from the second component were arranged in pseudorosetes.

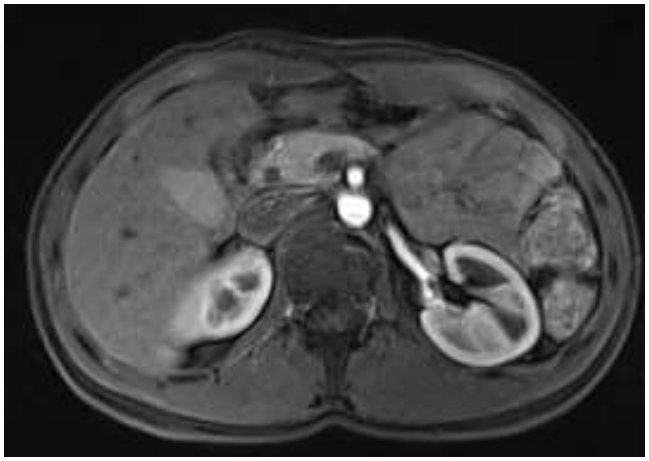
The imunohistochemical profile of the tumour was: vimentin (+), CD 10 (+), alfa1-antitripsine (+), glucagon (+), sinaptophysin (+), neuron specific enolase (NSE) (+), chromogranin (+) and insulin (-). The final diagnosis was Gruber-Franz tumour or solid pseudopapillary tumour (SPPT) of low malignant potential.

The patient was presented at the Oncological Consilium in January, 2012. A decision was made for regular oncological control every three months with abdominal MRI and tumour markers in the first year (Picture 2). Until now, after two years, the control diagnostic has been normal. At the last control the patient was pregnant.



Picture 1 Abdominal CT: well defined no enhancing solid pancreatic mass lesion in tail region of pancreas

Slika 1. Abdominalni CT: dobro izražena, ne pojačana čvrsta lezija pankreatične mase na repu pankreasa



Picture 2 Abdominal MRI (axial plane): postoperative MRI of upper abdomen (three months after tumour resection)

Slika 2 Abdominalna MR (aksijalna ravnina): postoperativna MRA gornjeg abdomena (tri mjeseca nakon tumorske resekcije)

Discussion

Gruber-Frantz tumour (solid pseudopapillary tumour – SPPT) is a very rare cystic neoplasm of the pancreas. SPPT is usually seen in young females between the 2nd and 3rd decades of life.

It is usually misdiagnosed as pancreatic pseudocyst, especially when symptoms are absent.[3] The most frequent symptom of SPPT is upper abdominal pain seen in nearly half of patients. The classic CT features of SPPT are a large well encapsulated mass with varying solid and cystic components caused by

haemorrhagic degeneration.[5,6] SPPT may occur anywhere in the pancreas but is most frequently found in the head or tail.[7]

We reported a case with a rare pseudopapillary pancreatic tumour in a very young woman. On the of the clinical presentation diagnostics was performed which showed an expansive lesion in the tail of the pancreas. Distal pancreatectomy and splenectomy were performed. The postoperative course was normal and the patient discharged. Histopathological analysis of the tumours showed solid pseudopapillary tumour of the pancreas of low malignancy. The patient has completely recovered. She has had a regular oncologic control.

In this case unspecific symptoms and CT were able to refer to another gastrointestinal disease.

The differential diagnosis of SPPT includes pseudocyst of the pancreas and another pancreatic tumour. Diagnosis of SPPT is usually made only after operative biopsy or resection.

The patient is completely cured if the tumour is completely resected without histological evidence of malignancy. Such patients have good prognosis with overall 5 year survival rate of 100%.

From the presented case, we can conclude that good medical history is always needed, which is half of the diagnosis, and detailed diagnostics performance which will direct us to the correct therapy. This is one example more of how in literature there are extremely rare tumours beside us that just need to be recognized.

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