Early pregnancy masquerading as a marker for malignancy in a young woman with curable neoplasm of the pancreas

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The so-called solid or cystic papillary neoplasm of the pancreas is a rare type of tumour occurring almost exclusively in young women. This neoplasm is not associated with any endocrine dysfunction, and typically it has assumed considerable size when discovered. Since most cases are curable by surgery, their distinction from other types of neoplasms with less favourable prognosis is important. The case reported here illustrates a potential pitfall in this context.

Case report

A 19-year-old previously healthy woman was admitted to hospital because of intermittent abdominal pain of 2 weeks duration. A pregnancy test was positive and she was examined by ultrasound with regard to ectopic pregnancy. The uterus had normal size and uniform echo distribution, but in the right upper quadrant of the abdomen an 8 cm non-homogeneous partially cystic mass was demonstrated. Considering the history given, this lesion was suspected to represent an ectopic pregnancy. At subsequent exploratory laparotomy the mass was found to be located within the head of the pancreas. A biopsy was taken and pathological examination revealed an islet-cell neoplasm. The pelvic organs looked quite normal, and the elevated serum chorionic gonadotrophin (HCG) level was now interpreted as an expression of malignancy although no metastases were evident. Tumour resection was not attempted, but the patient was referred to the University of Michigan Medical Center for further evaluation and treatment. Repeat serum analyses showed levels of β-HCG at 775 milliunits/ml (normal <10 milliunits/ml) and α-HCG at 4ng/ml (normal <1.2 ng/ml). Angiographically the tumour appeared hypervascular with the exception of supposedly necrotic central areas.

There was no apparent invasion of the surrounding major vessels, and neither angiography nor computed tomography (Figure 1) showed any evidence of liver metastases. The patient was reoperated on and the tumour was found to be removable by a Whipple procedure. The HCG values did not fall after the operation as expected but continued to rise, and a week later (3 weeks after initial admission) repeat ultrasound examination revealed an intrauterine pregnancy with measures that suggested an age of approximately 7 weeks. Histological examination of the resected tumour did not verify the previous diagnosis of islet-cell carcinoma, but showed a picture characteristic of the so-called solid or cystic papillary neoplasm of the pancreas (Figures 2 and 3). The patient, who subsequently underwent abortion, is doing well 1 year after the operation without any evidence of tumour recurrence.

Discussion

Studies of patients with endocrine pancreatic neoplasms have indicated that serum levels of HCG and its subunits are specific markers for malignancy. Likewise, immunocytochemical demonstration of α-HCG has been found to be associated with malignant behaviour in a large series of islet-cell tumours. Thus, early and unexpected pregnancy is an apparent pitfall in the clinical setting presented here where a pancreatic tumour was discovered in a young woman.

Although endocrine tumours of the pancreas are usually small lesions (unlike the so-called solid or cystic papillary...
Case report

The fact that most solid or cystic papillary neoplasms of the pancreas occur in young women indicates that hormonal factors are of pathogenic importance. This assumption is supported by findings in two cases examined for steroid hormone receptors. Both showed a rich content of progesterone receptors and one of them also had oestrogen receptors. Thus, it is possible that pregnancy might have growth-promoting or other effects on this kind of tumour, explaining the concurrent onset of abdominal pain in our patient.

Given the surgical curability of the solid or cystic papillary neoplasm, its distinction from a malignant islet-cell tumour is important in determining the choice of treatment, and ultimately to the well-being of the patient. It is a differential diagnosis to keep in mind whenever a pancreatic tumour occurs in a woman of childbearing age.

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References