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A Rare Complex of Pancreatic Adenocarcinoma, Sarcoma and Osteosarcoma was Confirmed as a Ploy-Clonal Entity

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Introduction

The pancreatic carcinosarcoma was a mixed adenocarcinoma with sarcoma or osteosarcoma components. It was considered as a rare tumor, which was mostly described in a number of cases reports [1,2]. However, this diagnostic term to this tumor was revised according to the recent nomenclature system of WHO classification of tumors for the digestive system. A new terminology of undifferentiated carcinoma with osteoclast-like giant cells (UCOGs) replaced the old terminology of carcinosarcoma used before. Although in the mixed pancreatic neoplasms, the term of carcinosarcoma was seldom used now, and by now the diagnosis of undifferentiated or anaplastic pancreatic ductal adenocarcinoma was mostly preferred instead of carcinosarcoma. Meanwhile some authors still favored pancreatic carcinosarcoma as a real entity [3], thus it still arose a question: Is the term of carcinosarcoma a real entity? Herein we provided one case with more proof of immunohistochemical and genetic data.

Case

The patient was a 44 years old woman with discomfort in the upper abdomen. The tumor was sized 2.9 cm × 1.6 cm in the pancreas head. It was a heterogenous and bone-like high-density lesion in CT scan. After a pancreaticoduodenectomy, the tumor revealed three distinct histological components, which was consisted of adenocarcinoma, sarcoma and osteosarcoma. In immunohistochemistry, every component had its own distinct phenotype. The laser-capture method was applied to separate three components respectively in the following genotype analysis.

The K-ras and P53 gene mutations were compared their variations as the paper did [3-5]. The whole exon 1-2 of K-ras and exon 4-9 of P53 of three components were sequenced. The results showed that the ductal carcinoma was identical to sarcoma without mutations in the target exons, however the osteosarcoma had two mutations in P53 and one mutation in K-ras. Thus this tumor should be a pancreatic carcinosarcoma.

Discussion

In papers More than 90 similar cases were reported, which included 72 cases in English papers and another 18 cases in Chinese papers (Based on the search of Medical Wanfang Database). However the statistic of UCOGs must be undervalued due to a number of terms used to describe this type of tumor or some cases unpublished [6]. For instance, we also found another two similar cases from unknown case seminar. Several sarcomoid mesenchymal components were reported in the tumors, which were included Leiomyosarcomamalignant peripheral nerve sheath tumor (MPNST) malignant fibrous histiocytoma, osteosarcoma, chondrosarcoma and other sarcomas at the same time [7]. Okamura firstly confirmed a heterologous osteosarcoma originating from an intraductal papillary-mucinous carcinoma (IPMN) by phenotype. Resch also reported a case of extra-skeletal osteosarcomas in the pancreas head without any ductal carcinoma.

There was much debate in the literature as to the origin of the UCOGs, with many authors favoring epithelial origin, and only a little favoring mesenchymal pathway. Most UCOGs harbored activating point mutations in the codon 12 of K-ras oncogene. As to the pancreatic tumors with ductal and sarcomoid components, both components reveal identical point mutation of the K-ras oncogene, indicating that the anaplastic carcinoma may arise from the ductal carcinoma. This was based on the genotyping analysis that the K-ras and P53 mutations were identical in carcinoma and osteosarcoma in the Okamura's paper [4]. However the phenotype and genotype of the osteosarcoma were distinct from that of pancreatic carcinoma in our case [4]. Thus our case had presented an unusual instance to this question and this is why we preferred to the term of carcinosarcoma colliding osteosarcoma. Interesting, the similar counterpart of UCOGs even with osteosarcoma occurred in liver had been reported either [5].

Although thought as a high aggressive malignant epithelial neoplasm, the prognosis of UCOGs was noticeably better than pure pancreatic adenocarcinoma in some reports [8,9]. Our patient was staged B and the follow-up showed she has survived for more than 80 month by now, and the pancreatic UCOGs or carcinosarcoma might have a better prognosis were also usually compared to the pancreatic adenocarcinoma. In this point,

UCOGs with osteosarcoma could be considered as a relatively differentiated tumor rather than anaplastic malignancy.

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