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AN UNUSUAL CASE OF PANCREATIC TUMOR; OSTEOCLASTIC LIKE GIANT CELL TUMOR.

Pankreasın nadir rastlanan tümörü; Osteoklastik tip dev hücreli kanser.

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ABSTRACT

The osteoclast-like giant cell tumor of the pancreas, described by Rosai, is a rare pancreatic tumor. Since then, only 40 cases have been reported in the literature. A 51-year-old Turkish woman was admitted because of nocturnal fever, trembling and jaundice. Blood tests on admission showed a liver cytolitic syndrome. The patient had laparotomy which confirmed the presence of a big epigastric mass adherent to the distal portion of stomach and spleen, and the portal vein thrombosis. Total pancreaticoduodenectomy was performed while the obturated portion of the portal vein was resected and replaced by polytetrafluoroethylene vascular graft. The postoperative period was uneventful. Unfortunately, three months after surgery, liver and lung metastases were noted. The patient passed away about four months after the cancer diagnosis. This case is being reported mainly to report an unusual tumor and secondary to discuss about the agressivity observed despite a radical excision of the tumor.

Key words: Giant cell tumor, pancreatic tumor, pancreaticoduodenectomy, portal vein, vascular graft.

ÖZET

İlk kez Rosai tarafından tanımlanan pankreasın osteoklastik tip dev hücreli tümörlerine nadir rastlanmaktadır. Literatürde şimdiye kadar 40 vaka bildirilmiştir. Olgumuz 51 yaşında kadın hastadır. Ateş yüksekliği, sarılık nedeniyle başvurdu. Yapılan kan biyokimya tetkikleri sonucunda hastada tıkanma ikteri olduğu saptanarak hastaneye yatırıldı. Perkutan safra drenajı ve intravenöz antibiyotik tedavisi uygulandı. Ateş yüksekliğinin devam etmesi üzerine operasyona karar verildi. Operasyonda tüm üst karını dolduran, mide ve dalağı kendine doğru çeken, portal vende tromboza neden olan yaklaşık 20 cm boyutunda kitle saptandı. Olguya splenektomi ile birlikte total pankreatikoduodenektomi uygulandı. Ayrıca tromboze olmuş portal ven segmenti çıkartılarak portal ven devamlılığı politetrafloroetilen damar grefti ile sağlandı. Operasyon sonrası izleminde hastada majör cerrahi sorunla karşılaşılmadan taburcu edildi. Takiplerinin 3. ayında yaygın karaciğer ve akciğer metastazları saptandı. Operasyon sonrası 4. ayında metastazlara bağlı tümör yükü nedeniyle gelişen komplikasyonlar sonucu hasta kaybedildi. Bu makalede pankreasın nadir rastlanan kanser türü olan osteoklastik tip dev hücreli kanserli bir olgudaki agresif cerrahi tedaviyi ve sağkalım süresini sunmayı amaçladık.

Anahtar kelimeler: Dev hücreli tümör, pankreas kanseri, pankreatikoduodenektomi, portal ven, vasküler greft.

INTRODUCTION

The osteoclast-like giant cell tumor of the pancreas (OGTP) is a rare pancreatic tumor first described in 1968 by Rosai (1). Since then, only 40 cases have been reported in the literature, mainly as single case reports, with respect to the clinical and pathologic particularities of the entity (2,3). We describe here the case of a patient with abdominal pain associated with jaundice. The histological examination performed in the meantime revealed that

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the mass was in fact an OGTP. This case is being reported mainly to report an unusual tumor and secondary to discuss about the agressivity observed despite a radical excision of the tumor.

Case

A 51-year-old Turkish woman was admitted because of nocturnal fever, trembling and jaundice. In her examination, the patient was icteric, had pain in the right upper hypochondrium and also an epigastric mass was palpated. Blood tests on admission showed a liver cytolitic syndrome. The whole clinical picture strongly evoked cholangiitis. Morphological tests (ultrasonography, magnetic resonance and ERCP) led to the initial diagnosis of an intrapapilary mucinous tumor located at the head of pancreas (Figure 1).

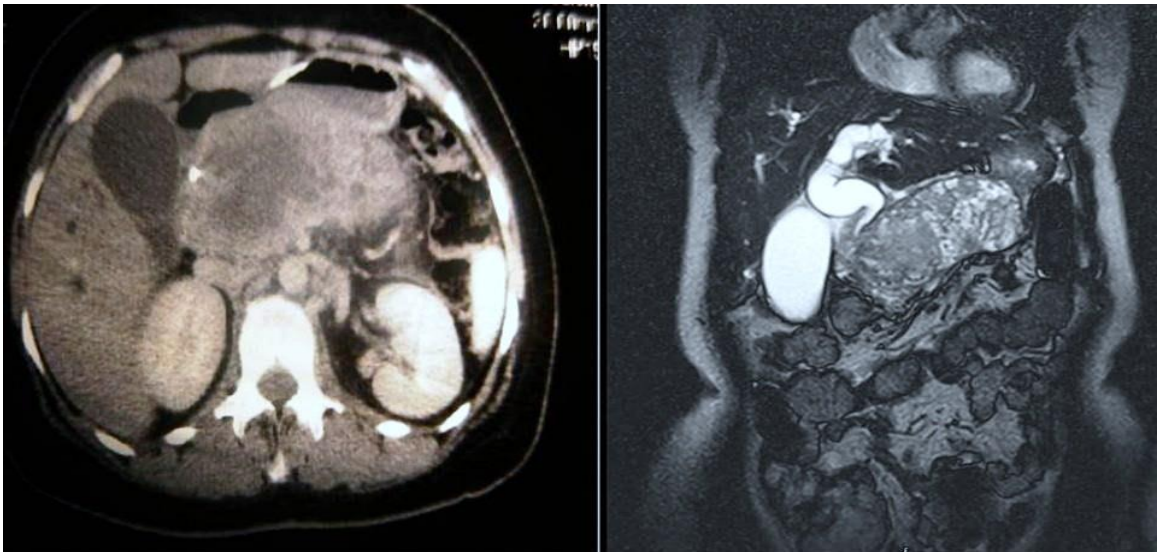

Figure 1: A big epigastric mass adherent to the stomach and spleen and choledochus.

In addition, portal vein thrombosis was found on a small portion, suggesting local invasion. No further tumor extension was identified. As high temperature persisted, positive hemoculture (Pseudomonas aeruginosa) motivated the prescription of adapted antibiotics without significant improvement of the septic state. The patient had consecutively a laparotomy which confirmed the presence of a big epigastric mass adherent to the distal portion of stomach and spleen, and the portal vein thrombosis. Total pancreaticoduodenectomy with splenectomy was performed while the obturated portion of the portal vein was resected and replaced by a vascular reconstruction using the polytetrafluoroethylene vascular graft (Gore-Tex , Flagstaff, AZ) (Figure 2).

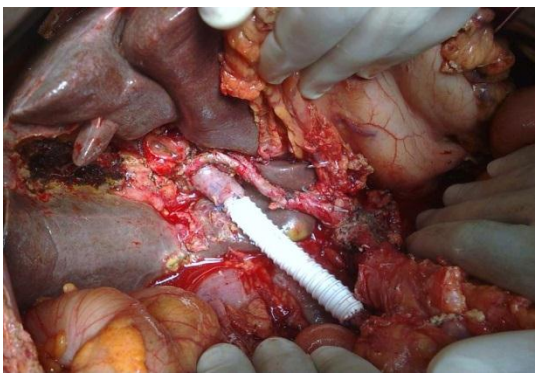

Figure 2: The reconstruction of portal vein after total pancreaticoduodenectomy.

The postoperative period was uneventful and the patient was discharged from the hospital after 14 days free of fever and pain, with functional vascular graft. Surprisingly, the histological analysis of the resected abdominal tumor reported a malignant tumor with osteoclastic giant cell of the pancreas, asteroid formation and sarcomatiod transformation. The patient then benefited chemotherapy with gemcitabine and 5 fluorouracil. Unfortunately, three months after surgery, liver and lung metastases were noted. The patient passed away about four months after the cancer diagnosis.

DISCUSSION

The osteoclast-like giant cell tumor of the pancreas is a malignant pancreatic tumor, which has also been seen in many organs such as breast, thyroid, parathyroid, heart and soft tissue. It is extremely rare and represents less than 1% of the pancreatic tumors. In some reported studies, this represents only 0,2% of the pancreatic carcinomas (4). The tumor appears in the 6th or 7th decade with nearly equal gender ratio (4, 5). The main symptoms and signs are abdominal pain, palpable mass, and loss of weight and jaundice (5). In our case the patients presented with an advanced cholangitis complicated by sepsis due to the compressive manifestations of the mass. Radiologically, OGTP is commonly exhibited as large cystic mass that tends to involve the head and corpus of the pancreas.

The mass has a predilection to local spread and rarely metastases to lenf nodes (6).

The diagnosis is difficult to be suggested before the operation and most of time a pancreatic cystic tumor is firstly considered before resection (6,7). Because of the disease rarity the treatment protocol has not clearly been established. Surgical resection is recommended if possible. Adjuvant chemotherapy or radiotherapy is also being discussed. The outcome after surgery is variable. However, the majority of patients have survival less than one year (7). According to the tumor prognostic factor outcome varies from 4 month to 15 years (7, 8).

In conclusion; the intraoperative findings should be considered as an important prognostic factor. In the present case although the advanced local spread of the mass total resection was possible and any residual tumoral mass was not noted. However the early outcome of metastasis (3 months after the operation) should emphasize about the prognostic of OGTP.

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