

Case Report



Primary Paraganglioma of the Pancreas: Review of Literature and a Case Report

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ABSTRACT

Extra-adrenal paragangliomas are very rare tumors which arises from the extra-adrenal chromaffin cells. Pancreatic paraganglioma is extremely rare tumor. It grows slowly, so radical resection is recommended to achieve curability with good prognosis. In this report, we present a 51 year old woman patient with pancreatic paraganglioma which was completely removed by surgery and review all previously reported cases.

Key words: Paraganglioma, pancreas, surgery

ÖZET

Primer Pankreas Paraganlioması: Vaka Sunumu ve Literatür İncelemesi

Adrenal bezi dışında yer alan paragangliomalar, adrenal dışı kromafin hücrelerden köken alan nadir görülen tümörlerdir. Pankreatik paragangliomalar ise daha nadir görülen tümörlerdir. Bu tümörler yavaş büyüme özelliğine sahiptirler ve radikal cerrahi rezeksiyon iyi prognoz için önerilen küratif tedavi yöntemidir. Bu vaka sunumunda 51 yaşında pankreatik paraganglioma tanılı cerrahi uygulanan hasta literatür eşliğinde tartışılacaktır.

Anahtar Sözcükler: Paraganglioma, pankreas, cerrahi

Extra-adrenal paragangliomas are very rare tumors which arise from the extra-adrenal chromaffin cells. Pancreatic paraganglioma is extremely rare tumor. This tumor is encountered only as case reports. In this report, we present a case of paraganglioma of pancreas which was completely removed by surgery and review all previously reported cases.

CASE REPORT

A 51-year-old woman was admitted to the hospital with epigastric pain in December 2006. There was no palpable abdominal mass in the physical examination and laboratory data on admission were within normal limits including tumor markers. Abdominal ultrasound showed a mass in pancreas and gallstones. The tumor was identified as a 4,5x4 cm low intensity mass on T1-weighted and high intensity on T2-weighted upper abdominal magnetic resonance images (MRI) (Figure1). Her thorax and abdominal computerized tomography (CT) as well as bone scintigraphy scan showed no

abnormalities in other organs. She was accepted as having operable pancreatic cancer. At laparotomy, the tumor was observed on the anterior of the corpus of the pancreas. It was elastic, tense and capsulated. The tumor was resected with ligation of multiple arterial and venous branches in January 2007.

Pathologically, the tumor was identified as being a paraganglioma. It was 5.0x3.5x3.0 cm in diameter, fleshy, reddish brown and surrounded by thick capsule. Microscopically, the tumor showed the classical zellballen pattern and irregular anastomosing sheets around a delicate vasculature (Figure 2). The cells have an abundant homogenous or finely granular cytoplasm that can be eosinophilic. The nuclei were round to oval and showed atypia with rare mitotic figures. Immunohistochemical staining was positive for neuron specific enolase (NSE), synaptophysin, S-100 protein and chromogranin (Figure 3). They were negative EMA, CEA, CK7, CD117, CD31, CD34, F VIII, CD10 and P53.

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Figure 1. Sagittal and axial magnetic resonance images of the lumbar spine showing L3-L4 intervertebral spondylodiscitis in 1999.

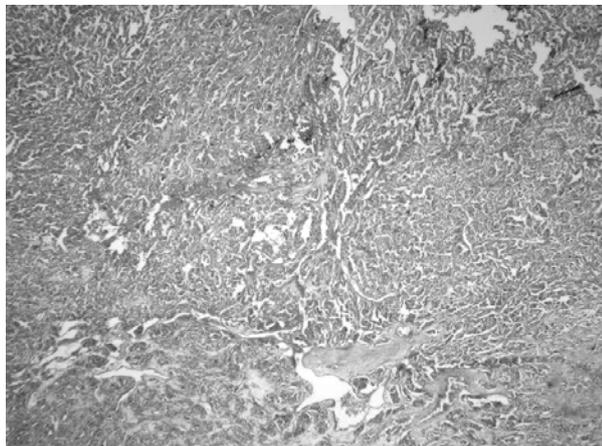


Figure 2. Anterior-posterior radiograph of the lumbar spine showing bilateral symmetric sacroiliitis.

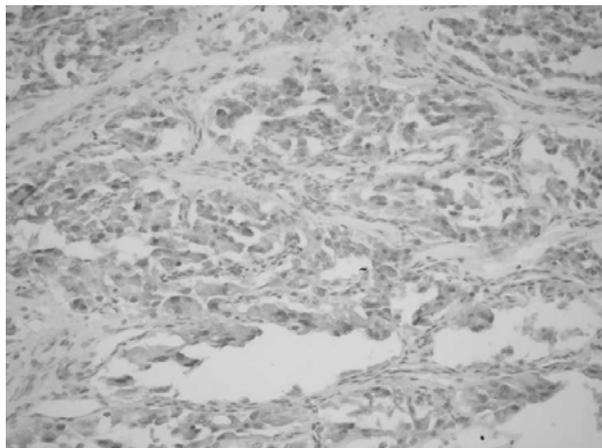


Figure 3. Sagittal and axial magnetic resonance images of the lumbar spine showing L3-L4 intervertebral spondylodiscitis in 2002.

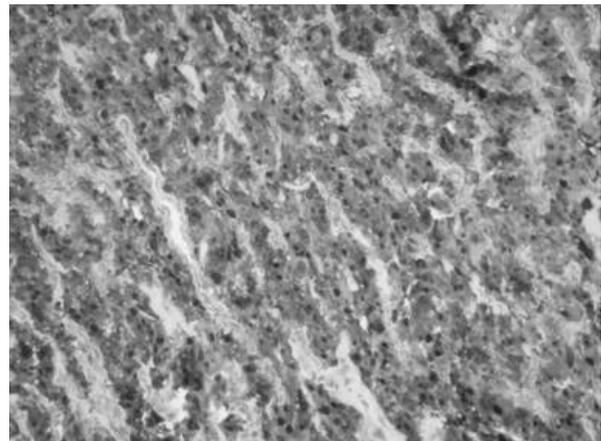


Figure 4. Computerized Tomography images of the sacroiliac joints showing bilateral symmetric sacroiliitis.

DISCUSSION

Paragangliomas are rare neuroendocrine tumors, which arise from the extra-adrenal chromaffin cells. They are found in the tissues such as the adrenal medulla, carotid and aortic body, organs of Zuckerkandl, and paraganglia of the sympathetic and parasympathetic neurons. They represent 10-18% of all chromaffin tissue related tumors. The head, neck, and retroperitoneum are the most common sites for paragangliomas. Other less common locations for abdominal paragangliomas include the gallbladder, urinary bladder, prostate, spermatic cord, uterus and duodenum (1, 2).

Histologically, all paragangliomas present a similar appearance regardless of their site of origin. They consist of clusters of cells separated by a highly vascular, reticular network forming the characteristic zellballen pattern. The malignant potential of these tumors cannot be determined from their histological appearance (3, 4).

Paraganglioma of the pancreas is also rare and only 14 patients including our case have been reported up to now (1, 3, 5-12) (Table 1). Eight cases were considered to be benign from pathological and clinical findings, while the frequency of malignant paragangliomas of the retroperitoneum ranged between 20% and 42% (3, 10, 13). The mean age of these 14 cases was 63 years. the male to female ratio was 0,75/1. In the nine patients, the tumor was located in the head of the pancreas. In the three cases, tumors were located in the corpus of pancreas and in the one, it was located in the tail of pancreas. In our case, tumor was located in the corpus of pancreas.

Paraganglioma, like other benign tumors, usually does not present any symptoms (especially nonfunctional tumors) and is often found incidentally (11), In our case, presented epigastric pain. In functional tumors, urinary catecholamines are elevated, usually with predominance of norepinephrine (1, 6). Abdominal ultrasonography and CT scan generally demonstrate a well defined mass. Paragangliomas are characterized by highly vascular and well-enhanced tumors with a cystic area in CT scan (3).

The main therapy is surgical resection. In general, the resection of paraganglioma of the pancreas is technically difficult due to the anatomical complexity around the

pancreas and the possibility of sudden catecholamine release during the operation (10). In our case, tumor was resected. The local resection was performed in 9 of the 14 patients in the literature. Pancreatectomy was the another resection

procedure used in 4 of the 14 patients. All of these patients showed equally good outcome after surgery. Our case is still in complete remission also.

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