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Safra kesesi paraganglioma olgusu: Nadir bir olgu sunumu

Muharrem Oztas(*), Nail Ersoz(**), Ismail Hakki Ozerhan(**), Armagan Gunal(***), Aytekin Unlu(**), Mehmet Fatih Can(**), Yusuf Peker(**)

SUMMARY

Gallbladder Paraganglioma: Report of a Rare Case

In this study, we aimed to share our clinical experience with gallbladder paraganglioma. Paraganglioma of the gallbladder is the most rare neoplasm. This case was diagnosed incidentally during laparoscopic exploration for suspicion of gallbladder cancer. The patient's data were retrospectively collected and presented with literature. Irregular and solid mass of the gallbladder were observed while a contrast-enhanced computerize tomography on a 61 years old female patient. The main reason for contrast-enhanced computerize tomography was former kidney malignancy. After that malignant neoplasm could not be excluded because of its appearance and she operated with a suspicion of gallbladder cancer. Laparoscopic exploration and cholecystectomy were performed and frozen section of mass demonstrated gallbladder paraganlioma. The definitive diagnosis of patients was made intraoperatively. Cholecystectomy was enough and not required further treatment such as liver resection or lymphatic dissection. Postoperatively, the patient was investigated for any association with other endocrine tumors and was not found. The patient recovered without complications. Even though primary gallbladder paraganglioma is a rare case, it may occur and it should be considered in the differential diagnosis of mass of the gallbladder. A careful search must be carried out for the possible association with MEN syndrome.

Key words: *Gallbladder, paraganglioma, extra-adrenal neoplasm, unusual localization*

ÖZET

Bu çalışmada, safra kesesi paraganglioma olgusu ile ilgili klinik deneyimimizi paylaşmayı amaçladık. Safra kesesi paragangliması çok nadir bir tümördür. Bu olgu safra kesesi kanseri şüphesi ile yapılan laparoskopik eksplorasyonda tesadüfen teşhis edildi. Hastanın verileri retrospektif olarak değerlendirilerek literatür eşliğinde sunuldu. 61 yaşında bayan hastada eski böbrek tümörü nedeniyle yapılan kontrastlı bilgisayarlı tomografide safra kesesinde düzensiz ve solid kitle saptandı. Malignite dışlanamayan hasta safra kesesi kanseri şüphesi ile ameliyat edildi. Laparoskopik eksplorasyon ve kolesistektektomi yapıldı. Frozen inceleme kitlenin safra kesesi paragangliması olduğu ortaya koydu. Hastanın kesin tanısı intraoperatif olarak kondu. Kolesistektomi tedavi için yeterli olarak değerlendirildi ve karaciğer rezeksiyonu veya lenfatik diseksiyon gibi ek işleme gerek kalmadı. Postoperatif dönemde hasta diğer endokrin tümörler açısından da araştırıldı ve ve başka patoloji saptanmadı. Hasta komplikasyonsuz iyileşti. Primer safra kesesi paraganglioma nadir bir durum olsa da, gözlenebilmektedir ve safra kesesi kitlelerinin ayırıcı tanınısında akılda tutulmalıdır. Ayrıca eşlik edebilecek MEN sendromu açısından da dikkatli inceleme yapılmalıdır.

Anahtar Kelimeler: *Safra kesesi, paraganglioma, ekstra-adrenal tümör, nadir lokalizasyon*

Introduction

Paragangliomas are rare extra-adrenal neoplasms of neural crest origin and they are found in association with sympathetic and parasympathetic nervous system (1). Although paragangliomas can occur in a variety of anatomical locations, the majority are seen in the retroperitoneum (1-4). Particularly, their presence in the gallbladder is extremely rare.

The majority of reported gallbladder paragangliomas were discovered and diagnosed incidentally during operations for other diseases, rather than symptoms arising from paragangliomas. Our case was also incidentally discovered on computarized tomography (CT) scan images of the abdomen acquired from a patient with complaints not indicative of gallbladder disease

Case Report

Our patient was a 61 years old female and she was admitted to our clinic as the CT scan of the abdomen revealed a solid, 3.5x2.5 cm mass in the gallbladder (Figure 1). Her blood pressure was 145/90 mm Hg, temparature was 36.5 ºC and heart rate was 80 beats/ minute. Physical examination of the abdomen and on admission blood tests were normal. Under the suspicion of a gallbladder cancer, laparoscopic intervention was performed. During the operation a highly vascular mass that appeared like a hemangioma was noted in the gallbladder (Figure 2). Cholecystectomy was performed laparoscopically and frozen section of the specimen was reported as the mass could possibly be a gallbladder paraganglioma. Macroscopically, a submucosal mass of 2 cm was observed on the wall and there weren't any gallstones (Figure 3). Microscopically, the tumor consisted of tight clusters of monotonous cells with hyperchromatic small round nuclei (Zellballen pattern) (Figure 4).

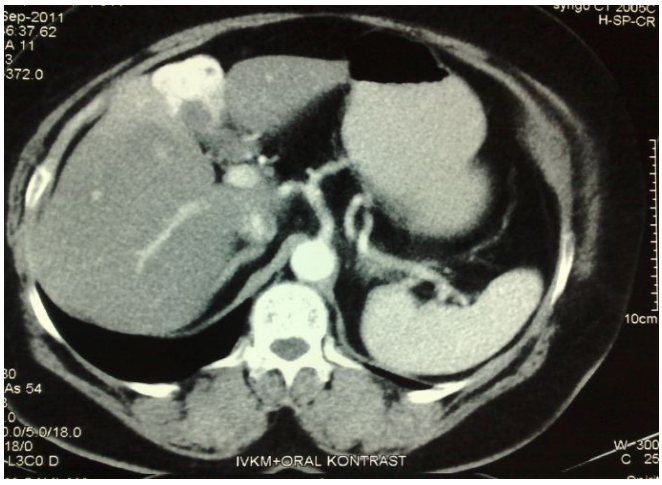

Figure 1: A 3.5x2.5 cm in size, irregular shaped, solid mass in the gallbladder was observed on contrast enhanced CT scan of the abdomen.

Department of Surgery, Ankara Mevki Military Hospital, Ankara, TURKEY;
Department of Surgery, Gulhane School of Medicine, Ankara, TURKEY;
Department of Pathology, Gulhane School of Medicine, Ankara, TURKEY;

Ayrı basım isteği: Muharrem ÖZTAŞ
Adres: Ankara Mevki Military Hospital, Ankara, TURKEY
e-mail: mmoztas@hotmail.com

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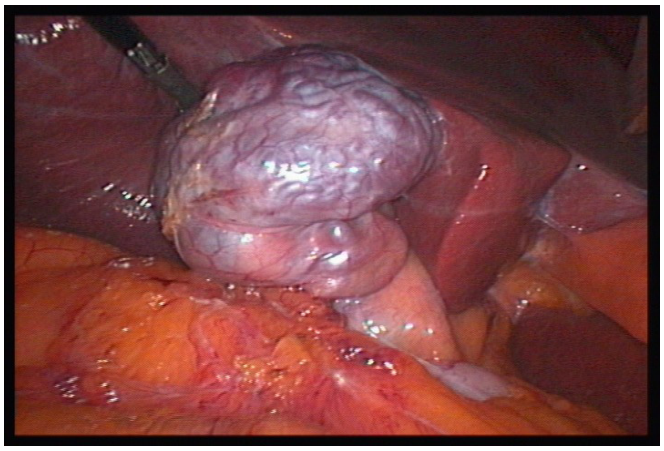

Figure 2: Image of a highly vascular mass that appeared like an hemangioma in the fundus of the gallbladder

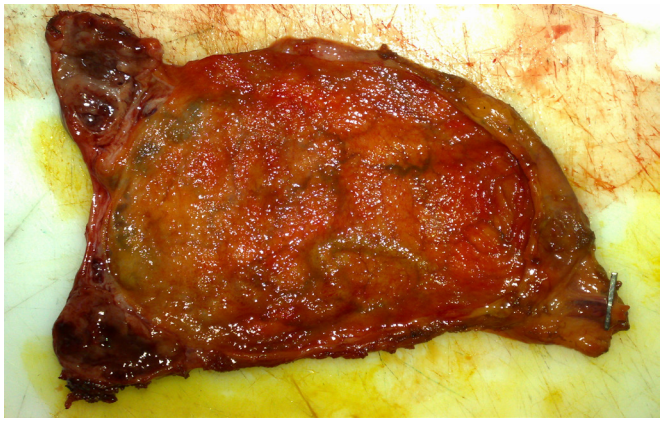

Figure 3: Macroscopically, a submucosal mass of 2 cm was observed on the wall and there weren't any stones in the gallbladder.

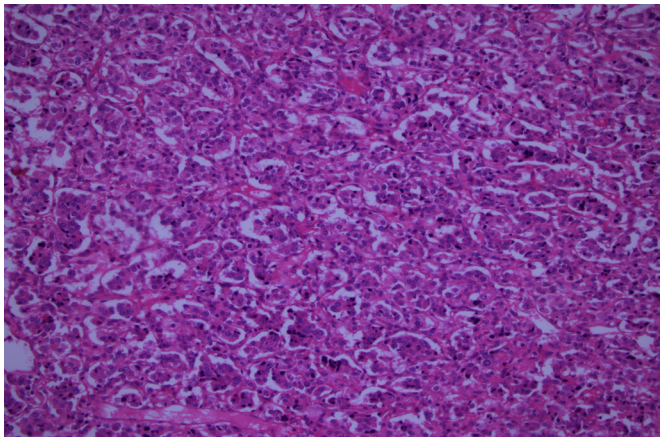

Figure 4: Zellballen pattern of the tumor cells was seen (H-Ex100).

No atypia, necrosis or mitotic activity were detected. Immunohistochemically, tumor cells were chromogranin and synaptophysin positive. Sustentacular cells around the cell nests were also positive with S-100 antibody (1/500 ZYMED, CA, USA) staining. Final pathological diagnosis was paraganglioma of the gallbladder. Family history of patient was re-evaluated and didn't reveal multiple endocrine neoplasia. The patient's postoperative course was uneventful and as no further interventions were appreciated, she was discharged on the 3rd postoperative day.

Discussion

Paragangliomas are rare neoplasms of neural crest origin arising from chemoreceptor tissues (3,5). Although they can be seen in the first decade of life, these tumors most commonly occur after the third decade. Histologically, they are characterized by an organoid arrangement in which clusters or islands of cells are surrounded by sinusoidal vascular channels and peripheral nerves (2). These tumors are classified as pheochromocytoma for tumors located in the adrenal gland and as paraganglioma for extra-adrenal tumors. Although most of these tumors are settled on in the retroperitoneum, exceptionally atypical sites have been reported, such as the carotid body, vagus nerve, larynx, mediastinum, nasopharynx, thyroid gland, duodenum, ileal mesentery, glomus jugulare of the middle ear, cauda equina, lung, kidney, thigh, stomach and urinary bladder (1-4).

The adrenal medulla is derived from nests of paraganglionic tissue that have migrated from the celiac plexus. Since the human gallbladder is innervated by the branches of sympathetic and parasympathetic fibers of the left vagus nerve and celiac trunk, paraganglionic cells could have migrated along these branches to the gallbladder, accounting for the presence of paraganglioma in this unusual location. The confirmation of this hypotheses depends on the identification of paraganglionic tissue within the normal gallbladder (2,6). Extra-adrenal tumors are more commonly malignant than adrenal tumors. Hence, gallbladder paraganglioma should not be misinterpreted as a secondary deposit, specifically in the presence of MEN syndrome. So far, only one biliary system paraganglioma with a familial association has been reported (6). Our patient had no family history of endocrine neoplasy.

Distinguishing a malignant tumor from a benign paraganglioma is difficult, especially in the absence of metastases (3,6). Distant metastasis is the most reliable and definitive indicator of malignancy (6).

Gallbladder paraganglioma is a very rare tumor, and so far only ten cases have been reported in the literature (1-8). Most of these cases were discovered incidentally during cholecystectomy for cholelithiasis and some of these presented with right upper quadrant pain or discomfort. One case manifested with gastrointestinal bleeding (3). In our case, the asymptomatic gallbladder mass was discovered on CT scan images of the abdomen.

None of the reported cases had symptoms or signs resulting from the hypersecretion of catecholamines (1-8). In the postoperative period, our patient's blood pressure remained within the range of preoperative values and there were no blood pressure fluctuations during the operation which indicated that it was a non-functional paraganglioma.

In the previous reports, the size of tumors ranged from 1.3 to 3 cm in diameter (1-8). When found in the gallbladder, they were most commonly located in subserosa and all were nonfunctioning. Recommended treatment of choice is cholecystectomy (4).

Primary gallbladder paraganglioma, although rare, should be considered in the differential diagnosis of gallbladder tumors. A careful search also should be carried out for the possible association with MEN syndrome.

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