Ganglioneuroma Mimicking A Retroperitoneal Sarcoma: Differential Diagnosis

RETROPERİTONEAL BÖLGEDE SARKOMU TAKLİT EDEN GANGLİONÖROMA OLGUSU

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Abstract

Ganglioneuroma is the most common tumour of the sympathetic nervous system in adults. It originates from the neural crest cells. It occurs most commonly in the mediastinum and retroperitoneum. They are highly differentiated benign tumors and are compatible with long term disease free survival even though surgical treatment is unsatisfactory. A 44-year-old female patient was admitted to the hospital with the complaint of upper abdominal pain and 8 kg loss in weigth in last 3 months. Ultrasonography and computed tomography (CT) of the abdomen revealed a retropancreatic mass with a diameter of 6 x 8 cm which was invading the vena cava and displacing the left renal vein anteriorly. Intraoperatively, we observed a large solid mass with a diameter of 10 cm behind the pancreas surrounding inferior vena cava. The mass was in close relation with the left renal vein. The infrarenal vena cava and distal portion of left renal vein were clamped and the tumour resected successfully. Repair of the vena cava was achieved with lateral venography with 5-0 prolene suture. The left renal vein was anastomosed to the superior mesenteric vein end-to-side with a 5-0 prolene suture. Postoperative period was uneventful and the patient was discharged from the hospital 5 days after the surgery. A control doppler ultrasonography obtained 2 weeks after the operation demonstrated normal appearance of the inferior vena cava and left renal vein.

Key Words: Ganglioneuroma, sarcoma

Özet

Ganglionöroma erişkinlerde en sık görülen sempatik sinir sistemi tümörüdür. Nöral krest hücrelerinden köken alır ve sıklıkla mediastende ve retroperitonda gözlenir. İyi diferansiye benign tümörlerdir ve cerrahi tedavi yetersiz olduğunda dahi uzun dönem hastalıksız yaşamdan bahsedilebilir. 44 yaşında bayan hasta kliniğimize üst abdominal ağrı ve son 3 ayda 8 kg kilo kaybı şikayeti ile başvurdu. Batın ultrasonu ve tomografisinde vena kava inferioru invaze eden ve sol renal veni anteriora iten 6 x 8 cm'lik retroperitoneal kitle saptandı. Ameliyat esnasında pankreas arkasında vena kava inferioru saran yaklaşık 10 cm'lik solid kitle lezyonu palpe edildi. Kitle ile sol renal ven ara planı seçilmiyordu. İnfrarenal vena kava ve sol renal ven distali klemplenerek tümör tümüyle rezeke edildi. Vena kava inferior 5-0 prolenle lateral venografi tekniği ile onarıldı. Sol renal ven 5-0 prolen sütür ile uç-yan olarak superior mezenterik vene anastomoze edildi. Postoperatif dönem sorunsuz geçti ve hasta postoperatif 5. günde taburcu edildi. Operasyondan 2 hafta sonra kontrol amaçlı yapılan doppler ultrasonda inferior vena kava ve sol renal ven akımları normal olarak gösterildi.

Anahtar Kelimeler: Ganglionöroma; sarkom

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Ganglioneuroma is the most common tumour of the sympathetic nervous system in adults. It originates from the neural crest cells. They are highly differentiated benign tumors and are compatible with long term disease free survival even though surgical treatment is unsatisfactory.[1]

Here, we present a case of ganglioneuroma arising in the retroperitoneum at the level of retropancreatic region, invading the vena cava and radiologically giving the false impression of a retroperitoneal sarcoma.

A 44-year-old female patient was admitted to the hospital with the complaint of upper abdominal pain and 8 kg loss in weigth in last three months. Systemic physical examination was normal.

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Biochemical investigation and blood cell count was in normal ranges. Serum tumour markers could not reveal any abnormality. Ultrasonography and CT of the abdomen revealed a retropancreatic mass with a diameter of 6 x 8 cm which was invading the vena cava and displacing the left renal vein anteriorly (Figure 1 A, B, C).

Surgical treatment was indicated after the retroperitoneal mass was diagnosed.

Intraoperatively, we observed a large solid mass with a diameter of 10 cm behind the pancreas surrounding inferior vena cava. The mass was in close relation with the left renal vein. The infrarenal vena cava and distal portion of left renal vein were clamped and the tumour resected succesfully.

Repair of the vena cava was achieved with lateral venography with 5-0 prolene suture. The left renal vein was anastomosed to the superior mesenteric vein end-to-side with a 5-0 prolene suture.

Postoperative period was uneventful and the patient was discharged from the hospital 5 days after the surgery. A control doppler ultrasonography obtained 2 weeks after the operation demonstrated normal appearance of the inferior vena cava and left renal vein.

Pathologic Findings

Grossly, the tumour was encapsulated and measured 8 x 5 x 4.5 cm. On cut section it had solid, firm, greyish white surface. Microscopically, the tumour consisted almost entirely of well-differentiated ganglion cells and Schwann cells within a neurofibrillar matrix (Figure 2). The tumour was reported as ganglioneuroma.

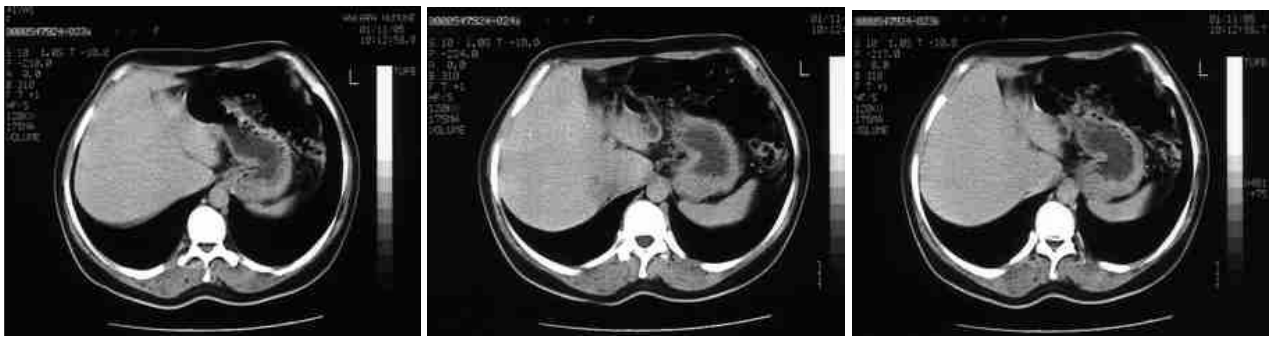
Ganglioneuromas are infrequently occuring tumours and are especially located in the mediastinum and retroperitoneum.[2] They represent the benign end of the spectrum of primitive neuroepithelial tumours, which also includes neuroblastoma (highly malignant) and ganglioneuroblastoma (intermediate). Most do not have secretory activity, and the clinical manifestations are related to the growth of the mass, which leads to compressive symptoms.[1]

They may be found in association with a number of syndromes including Beckwith-Wiedemann, von Recklinghausen's disease, opsoclonus-myoclonus, Hirschprung's disease, watery diarrhoea or Cushing's syndrome.[3]

Diagnosis of the mass is normally made with CT or nuclear magnetic resonance imaging (MRI), and histologic diagnosis may be made with CT guided biopsy.[2,4] But preopertative diagnosis of retroperitoneal ganglioneuroma is often difficult and the diagnosis is usually based on histopathological findings after surgical excision of the tumor.

Surgery is the choice of treatment because of tumor growth, possibility of poorly differentiated components in the mass and malignant transformation.[1]

Preoperative or postoperative chemotherapy or radiotherapy have no value in the treatment.[1] In the



A **B** **C**

Figure 1 A, B, C. A 6 x 8 cm sized, regular contoured retroperitoneal mass, posterior to pancreatic head, right to the aorta and compressing the renal artery and vein.

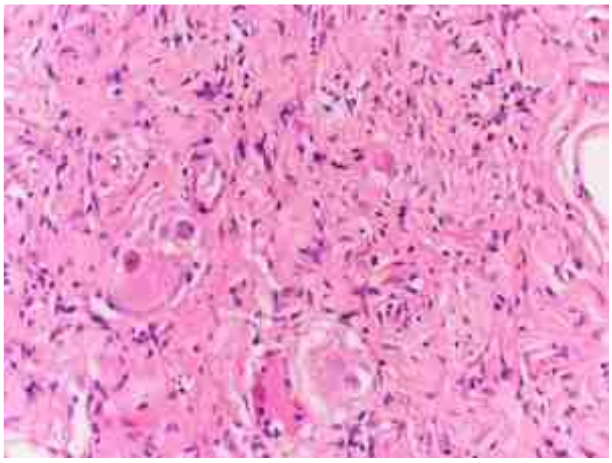


Figure 2. Ganglion cells deposited in a neuromatous stroma (HE, x 40).

present case we reported a case of ganglioneuroma which invades inferior vena cava. Preoperative evaluation is important to find out the relationship of tumoral mass and major vascular structures and to give the descision for resection of the tumor. According to our knowledge, there is only one case of ganglioneuroma in which tumoral mass is in close relation with major vascular structures.[5]

Ganglioneuroma is a rare benign tumour of the mediastinum and retroperitoneum, which can grow to a massive size and present in a varied manner. Management involves total excision if possible. Even with residual tumor, cessation of other treatments and a close follow-up may be adequate.

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