Malignant Hypertension Due to Mesenteric Fibromatosis: A Case Report

Mezenterik Fibromatozise Bağlı Malign Hipertansiyon: Olgu Sunumu

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ABSTRACT

Malignant hypertension (MHT) is an urgent clinical entity with high morbidity and mortality related to progressive renal and cardiac dysfunction. Eye is one of the target organs which is effected due to MHT. MHT should diagnosed and treated immediately to prevent target organ damage and death.

Fibromatosis occurs in shoulders, neck, dorsum and in soft tissues of the extremities. Also 5-10% of fibromatosis located in the mesentery, retroperitoneum and intestinal wall. Mesenteric fibromatosis (MF) exhibits benign pathology but it's locally aggressive and infiltrative. MF should be resected as wide as possible to prevent of tumor recurrence and to increase the survival ratio.

A 18 year-old female patient with epilepsy was admitted to our department of emergency complaining of blurred vision on her right eye (OD) for two days. Best corrected Snellen visual acuity (BCVA) was 4/10 in right eye and 9/10 in her left eye. Fundus examination revealed apparent optic disc swelling, blurred optic disc margins (papilledema), hemorrhages in optic disc, cotton-wool spots and macular edema in both eyes.

In this case report, we aimed to share diagnosis and treatment of a patient developing MHT due to recurrence of MF.

Key Words: Malignant hypertension, mesenteric fibromatosis, papilledema, macular star.

ÖZ

Malign hipertansiyon günler içerisinde progresif renal ve kardiyak fonksiyon bozukluğuna bağlı yüksek morbidite ve mortalite oranına sahip acil bir klinik tablodur. Göz, malign hipertansiyonun etkilediği hedef organlardan bir tanesidir. Hedef organ hasarını ve hayati tehlikeyi önleyebilmek için malign hipertansiyonun erken tanısının konması ve tedavi edilmesi gerekmektedir.

Fibromatozisler vücuttaki tüm kas dokularından köken alabilen en sık olarak baş-boyun, ekstremiteler ve intraabdominal olarak bulunan yumuşak doku tümörleridir. Yaklaşık % 5-10'unu intraabdominal yerleşim gösterir ve genellikle mezenterde, retroperitonda ve intestinal duvarda bulunurlar. Mezenterik fibromatozis benign karakter gösterse de lokal olarak agresif ve çevre dokulara infiltrasyon yapma potansiyeline sahiptir. Bu nedenle nüksü önlemek ve sağkalım oranını artırabilmek için mümkün olduğunca geniş rezeke edilmelidir.

Epilepsi hastalığı olduğu bilinen 18 yaşındaki kadın hasta 2 gündür sağ gözü ile az görme şikayeti ile kliniğimize başvurdu. En iyi düzeltilmiş görme keskinliği Snellen eşeline göre sağda 4/10, solda ise 9/10'du. Fundus muayenesinde sağ gözünde daha belirgin olmak üzere bilateral papilödem, optik disk başında kıymık tarzı hemorajiler ve tek tük atılmış pamuk manzarası mevcut iken makülalar ödemli görünümdeydi.

Bu çalışmada nüks mezenterik fibromatozise sekonder malign hipertansiyon gelişen olgunun tanı ve tedavisinin paylaşılması amaçlanmıştır.

Anahtar Sözcükler: Malign hipertansiyon, mezenterik fibromatozis, papilödem, makuler star.

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INTRODUCTION

Malignant hypertension (MHT), first described by Keith et al., is an urgent clinical entity with high morbidity and mortality related to progressive renal and cardiac dysfunction.[1,2] In MHT, systolic blood pressure (BP) is higher than 200 mmHg, diastolic BP is higher than 120 mmHg.[1,3]

Fibromatosis is a rare fibroblastic proliferation that originates from superficial and deep soft tissue. Also it's the most common primary tumor of intestinal mesentery.[4] Fibromatosis makes 3% of all soft tissue tumors and 0.03% of all neoplasms.[5] Mesenteric fibromatosis (MF) presents a benign clinical behaviour but has metastatic potential, it's locally aggressive, infiltrates the nearby tissues and has a high rate of recurrence.[6] MF mainly developes in females and the fourth decade is the most commonly diagnosed period.[7] The etiology is not known and most of the MF occur sporadically but it's associated with a history of previous abdominal or pelvic surgery, trauma, injury, pregnancy, long term use of estrogen, familial adenomatous polyposis and Gardner's syndrome.[8]

In this case report, we aimed to share diagnosis and treatment of a patient developing MHT due to recurrence of MF.

CASE REPORT

A 18 year-old female patient with epilepsy was admitted to our department of emergency complaining of blurred vision on her right eye (OD) for two days. Light reflexes, Ishihara colour vision were normal both eyes and relative afferent pupillary defect was not detected. Best corrected Snellen visual acuity (BCVA) was 4/10 in OD, 9/10 in her left eye (OS). Bilateral intraocular pressure and anterior segment examination was normal. Fundus examination revealed apparent optic disc swelling, blurred optic disc margins (papilledema), hemorrhages in optic disc, cotton-wool spots and macular edema (ME) in both eyes, which was prominent in the right fundus (Figure 1). Bilateral serous macular detachment (SMD) was observed in optical coherence tomography (OCT), ME thickness was 633 µm in OD, 476 µm in OS (Figure 2). Bilateral hyperfluorescence of the optic disc due to leakage were observed in fundus fluorescein angiography (FFA) (Figure 2).

Provisional diagnosis was focused on increased intracranial pressure, bilateral optic neuropathy, bilateral neuroretinitis. Blood count, sedimentation rate, CRP, thyroid hormone levels were all normal, serological and microbiological tests, antineutrophil cytoplasmic antibodies, antinuclear antibodies, rheumatoid factor were all negative.

The patient was referred to department of neurology but no neurologic pathology was reported. Since the BP was 210/130 mmHg, the fundus picture was attributed to hypertensive retinopathy. The patient was administered oral antihypertensive therapy, laboratory tests and radiological imaging were performed for the most common causes of secondary hypertension such as pheochromocytoma, renal artery stenosis, hyperaldosteronism. When questioned again, the patient's history revealed a history of previous abdominal surgery.

In abdominal computed tomography, a large soft tissue mass in the left retroperitoneal area associated with the tail of the pancreas infiltrating aorta and branches of the aorta was detected. The mass was displacing the left kidney and measured as 139x99 mm (Figure 3). In old records of the patient, distal pancreatectomy, splenectomy and segmental left colon resection operations were performed due to intraabdominal mass in our hospital, in 2013. Thus, the patient was referred to department of general surgery.

Due to infiltration, left radical nephrectomy with left hemidiaphragma repair, piloromyotomy, fundoplication opera-

Figure 1. *Fundus photograph of both eyes.*

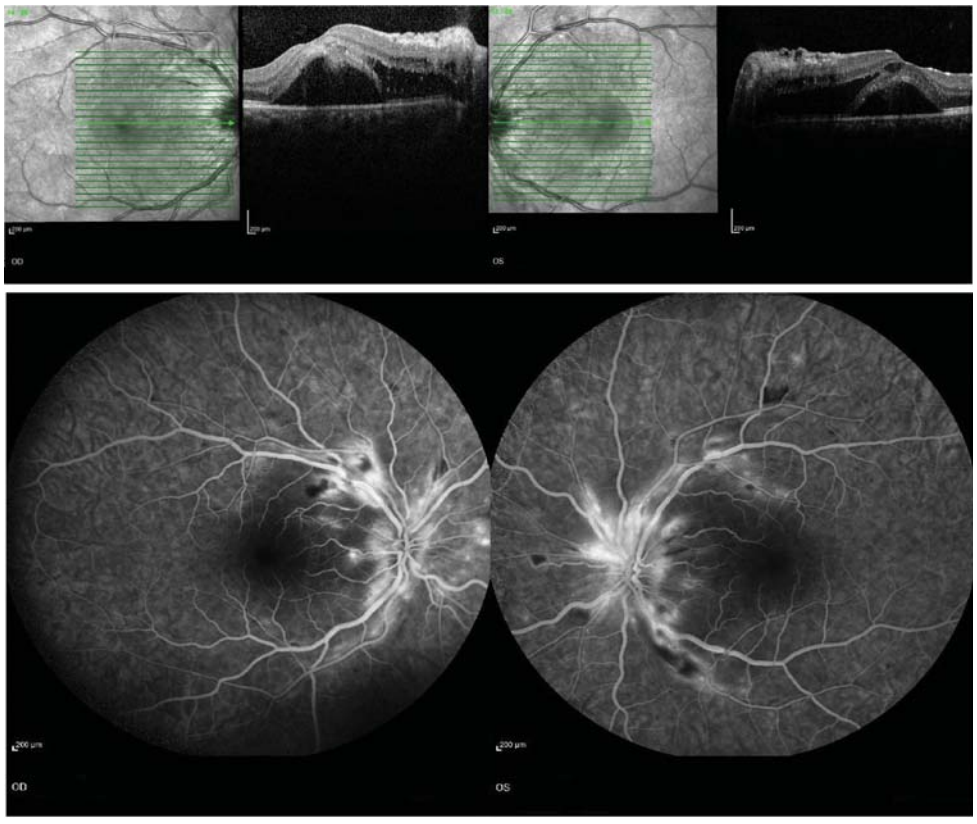

Figure 2. *OCT appearance of serous macular detachment and FFA appearance of hyperfluorescence of the optic disc due to leakage.*

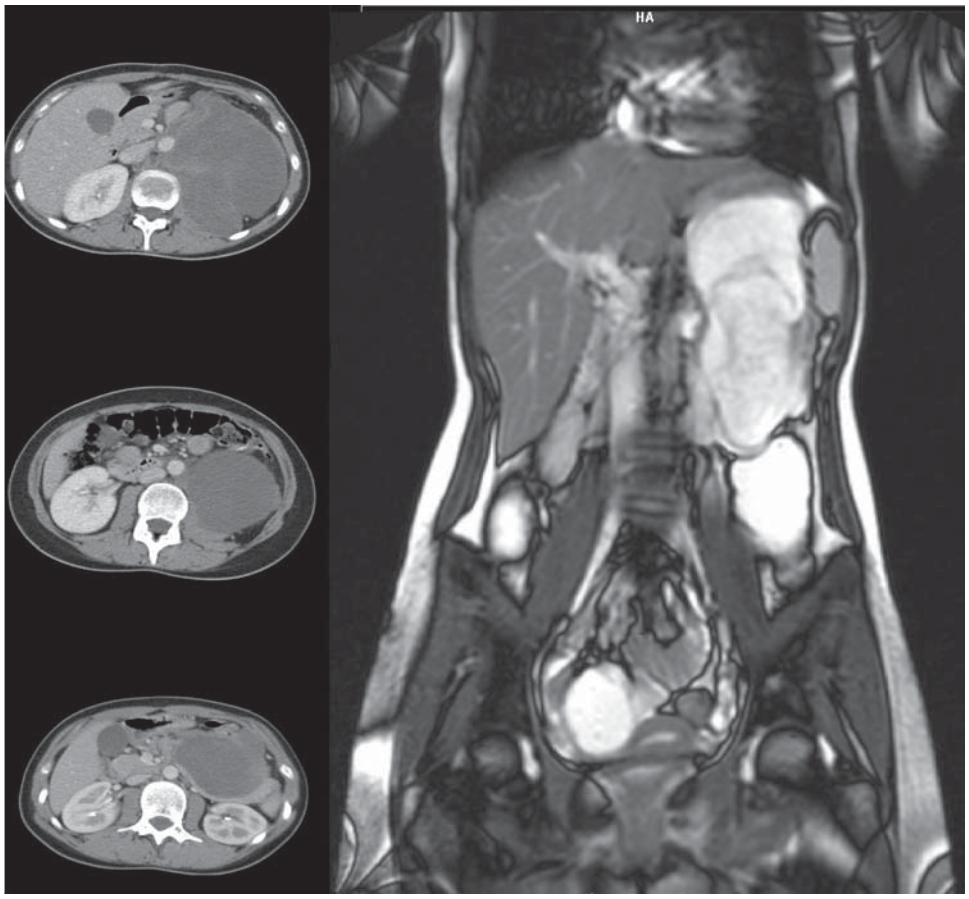

Figure 3. *Abdominal computed tomography showing a large soft tissue mass in the left retroperitoneal area associated with the tail of the pancreas infiltrating aorta and branches of the aorta.*

tions were performed and mass was resected with a wide margin. The final pathologic analysis confirmed the MF diagnosis, immunohistochemical staining showed that negative for CD34 (-), CD117 (-), DOG-1 (-), SMA (-), S100 (-), b-katenin (-) and the Ki67 index of the cells was only 1 %.

One month after first examination, BCVA was 9/10 in both eyes and fundus findings such as papilledema, SMD regressed but bilateral macular star appeared (Figure 4). Retinal pigment epithelium (RPE) abnormalities, intraretinal exudates were observed in OCT. Bilateral ME completely resolved and the macula was reattached (Figure 4). Six months after, BCVA was 10/10 in both eyes, macular star and exudates regressed and there was no ME (Figure 5).

DISCUSSION

MHT is a life-threatening disease, which can effect all body and characterized by progressive target organ damage. Therefore MHT should diagnosed and treated immediately to prevent target organ damage and death. Eye is one of the target organs which is effected due to MHT. Retinopathy, choroidopathy and optic neuropathy can be detected.

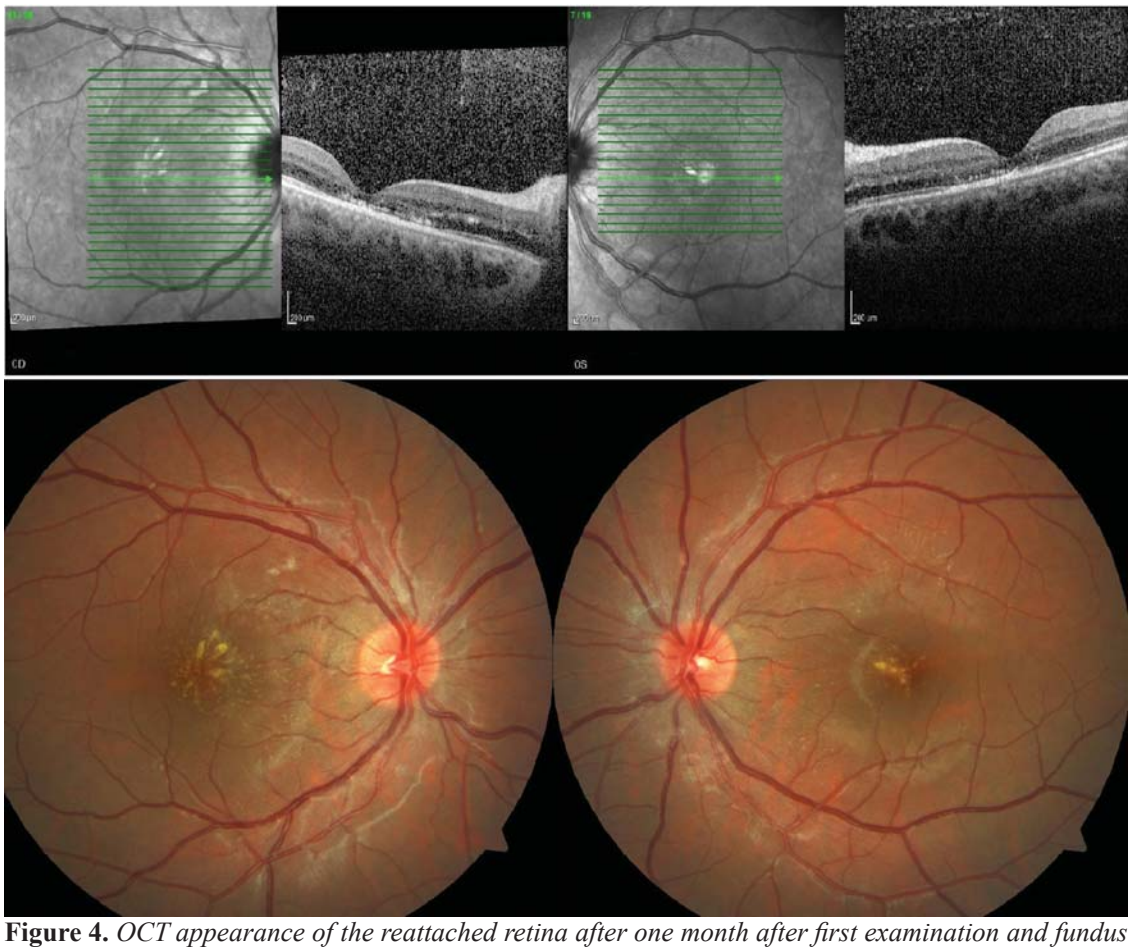

Figure 4. *OCT appearance of the reattached retina after one month after first examination and fundus photograph of macular star.*

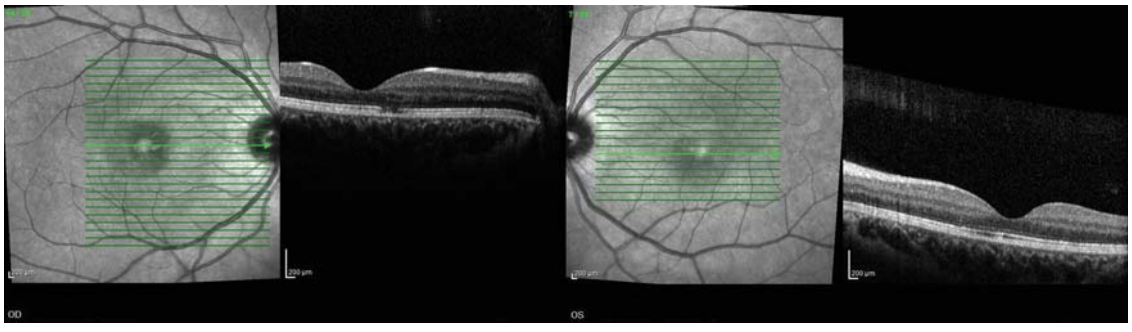

Figure 5. *OCT image showing regression of macular edema, macular star and exudates six months after first examination.*

Ischemia occurs in retinal nerve fiber layer and in inner retinal layer due to vascular occlusion. Cotton-wool spots, ME, SMD can occur. Macular star can detect due to exudates and this entity persists a few months or years.[9,10]

Macular hemorrhages, ME, exudates, RPE abnormalities due to choroidal ischemia, retinal artery–vein occlusion are responsible for vision loss. Papilledema is a sign of the advanced stage of MHT and it's formation has been suggested as a cause of interruption of the axoplasmic flow that developes ischemia and leaking in optic disc.[9,11]

Bilateral papilledema is a cause of metabolic, inflammatory, infectious, toxic disease, increased intracranial pressure or MHT.[9] It's a vital importance to eliminate increased intracranial pressure. In young patients with papilledema, neuroretinitis should be considered as a second diagnosis.

Generally fibromatosis occurs in shoulders, neck, dorsum and in soft tissues of the extremities.[12] Also 5-10% of fibromatosis located in intraabdominal cavity and mostly in the mesentery, retroperitoneum and intestinal wall.[13,14] MF has a slow growth pattern thus, it's usually asymptomatic. The clinical presentation of MF is often nonspecific abdominal pain, nauseation, vomiting and constipation. Also MF can cause intestinal obstruction, gastrointestinal hemorrhage, ischaemia and ureteral compression when its enlarged. Vascular, intestinal, ureteric and neural invasion maybe the first detectable signs.[15] To the best of our knowledge, present case is the only reported recurrence of MF who presented with ophthalmological symptoms.

The diagnose of MF should confirm histopathologically, because there is no pathognomonic sign and also no specific radiological major feature. Estrogen receptor antagonists, anti-inflammatory drugs, cytotoxic agents, radiotherapy may be the conservative treatment options but surgery with wide margins is the primary and successful treatment option[6]. MF exhibits benign pathology but it's locally aggressive, infiltrative and MF tends to recur if incompletely resected. Thus, MF should be resected as wide as possible to prevent of tumor recurrence and to increase the survival ratio.

REFERENCES / KAYNAKLAR

1. Keith NM, Wagener HP, Kernohan JW. The syndrome of malignant hypertension. Arch Intern Med. 1928; 41: 141-88.
2. Richard JG. Current therapy in nephrology and hypertension. The CV Mosby Company. 1984-1985; 324-33.
3. Mitchell RN, Schoen FJ. Blood vessels. In. Kumar, Abbas & Fausto, et al. Robbins and Cotran Pathologic Basis of Disease. Philadelphia, Pennsylvania, USA: Saunders Elsevier, 2010; 487–528.
4. Bar-Maor JA, Shabshin U. Mesenteric fibromatosis. J Pediatr Surg. 1993; 28: 1618-9.
5. Nuyttens JJ, Rust PF, Thomas CR Jr., Turrisi AT. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors. Cancer. 2000; 88: 1517-23.
6. Smith AJ, Lewis JJ, Merchant NB, et al. Surgical management of intraabdominal desmoid tumors. Br J Surg. 2000; 87: 608-13.
7. Kreuzberg B, Koudelova J, Ferda J, et al. Diagnostic problems of abdominal desmoid tumors in various locations. European Journal of Radiology. 2007; 62: 180-5.
8. Vandevenne JE, De Schepper AM, De Beuckeleer L, et al. New concepts in understanding evolution of desmoid tumors: MR imaging of 30 lesions. Eur Radiol. 1997; 7: 1013–9.
9. Murphy HP, Chew EY. Hypertension: In: Ryan SJ, Schachat AP, Murphy RP, Pätz A, ed. Retina. St Louis: The Mosby Company. 1989: 2: 449-55.
10. Walsh JBM. Hypertensive retinopathy. Descriptions, classification and prognosis. Ophthalmology. 1992: 89: 1127-31.
11. Jampol LM. Ocular manifestations of selected systemic disease. In: Peyman AG, Sanders DR, Goldberg MF, ed. Principles and practice of ophthalmology. Philadelphia: WB Saunders Company. 1980: 3: 1633-40.
12. Sutton RJ, Thomas JM. Desmoid tumours of the anterior abdominal wall. European Journal of Surgical Oncology. 1999; 25: 398-400.
13. Reitamo JJ, Hayry P, Nykyri E, Saxen E. The desmoid tumor. Incidence, sex- , age. and anatomical distribution in the Finnish population. Am J Clin Pathol. 1982; 77: 665-73.
14. Miettinen M, Monihan JM, Sarlomo-Rikala M, et al. Gastrointestinal stromal tumors/smooth muscle tumors (GISTs) primary in the omentum and mesentery: clinicopathologic and immunohistochemical study of 26 cases. Am J Surg Pathol. 1999; 23: 1109-18.
15. Chanco G, Rose EF. Mesenteric fibromatosis following colectomy for familial polyposis. Arch Surg. 1972; 104: 851-2.