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Surgical Resection Combined with Preoperative Radiotherapy for a Retroperitoneal Giant Schwannoma

Retroperitoneal Dev Şıvannoma'nın Preoperatif Radyoterapi ile Birlikte Cerrahi Rezeksiyonu

CASE REPORT
OLGU SUNUMU

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ABSTRACT
ÖZET

Retroperitoneal schwannoma is a very rare tumor, originating from Schwann cells in the peripheral nerve sheath. Complete surgical resection is considered the best treatment, but the control of bleeding can be difficult because of the tumor's hypervascularity. Therefore, angiography is recommended before surgical resection. Here, we report on a 40-year-old male patient who was successfully treated with total tumor resection after radiotherapy. This case indicates that preoperative radiotherapy may be beneficial in bleeding control in hypervascular schwannomas

Retroperitoneal şıvannova, periferik sinir kılıfının Şıvann hücrelerinden kaynaklanan oldukça nadir görülen tümörlerdir. Tümörün tam olarak çıkarılması en iyi tedavi seçeneği olarak kabul edilir, fakat kanama kontrolünün sağlanması tümörün hipervasküler olması nedeniyle zor olabilir. Bu nedenle cerrahi rezeksiyon öncesi anjiografi önerilmektedir. Burada radyoterapi sonrası total olarak rezeke edilen retroperitoneal yerleşimli şıvannom olgusu sunulmuştur. Bu vaka preoperatif radyoterapinin hipervasküler şıcannom cerrahi tedavisi sırasında, kanama kontrolünün sağlanmasında etkili olabileceğini göstermektedir.

Key words: Schwannoma, hemorrhage, radiotherapy

Anahtar kelimeler: Şıvannoma, kanama, radyoterapi

Introduction

Schwannomas can be benign or malignant tumors that arise from the neural sheath Schwann cells. They may occur nearly anywhere in the body, but the retroperitoneal location is much less common than other regions, such as the cranial or peripheral nerves (1). Only 0.3-3.2% of benign schwannomas are found in a retroperitoneal location (2). They generally reach large sizes before producing symptoms due to mass effects. The preoperative diagnosis of retroperitoneal schwannomas is difficult and surgical treatment is the first choice (2). Radical surgical excision may be rather difficult due to tumor localization and vascularization (3). We report a case with a retroperitoneal giant schwannoma that was treated by surgical resection combined with preoperative radiotherapy.

Case Report

A 40-year-old man presented to the hospital with a three month history of lower abdominal pain. His medical history and physical examination findings were unremarkable. The routine blood count and biochemical tests were within normal limits. Contrast-enhanced computed tomography revealed a well-circumscribed, oval-shaped lesion measuring 12 cm in diameter located posterior to the bladder (Figure 1A, B).

Angiography could not be performed prior to surgery because of technical deficiencies. An exploratory laparotomy revealed a mass located in the pelvis and posterior to the bladder. The biopsy of the mass was reported as a benign schwannoma. Hemorrhage can be a serious problem for cases with major vessels and structures localized nearby the tumor. During the biopsy, despite the application of hemostatic stitches, the bleeding was extremely difficult to control, because the tumor was excessively vascular and fragile. Therefore, surgical excision was deferred to a second surgery.

Radiotherapy, with a total dose of 66 Gy, was administered to the mass lesion in 33 fractions over a period of 6 weeks, to reduce the tumor vascularity. Seven weeks after radiotherapy, the tumor size had reduced by approximately 9% of the initial size. In the following period, approximately two months after radiotherapy, the mass was totally resected. No complications were encountered during surgery. The patient's postoperative course was uneventful.

Histological examination revealed that the tumor was composed of spindle cell proliferation with hypercellular areas. The presence of histiocytes among the tumor cells was noted (Figure 2A). There was no cellular atypia or mitosis. Thick-walled and hyalinized blood vessels were also noted (Figure 2B). The tumor was stained positively

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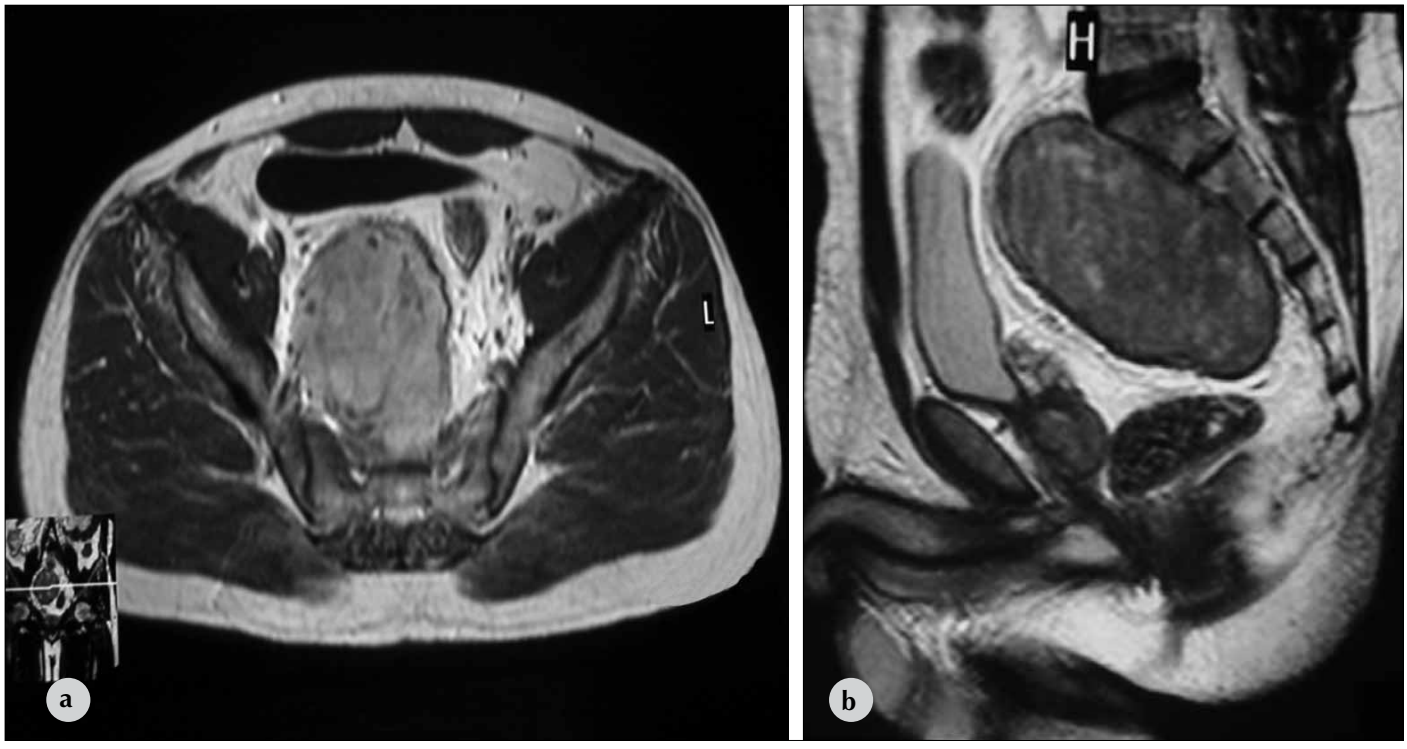

Figure 1. Computerized tomography (CT) images of the patient. (a) Axial CT scan showing an 11 cm enhancing mass with cystic areas arising from the retroperitoneum. (b) Sagittal CT scan showing a large, heterogeneous, well-defined mass, lying within the pelvis and posterior to the bladder. The mass is in close relationship to the rectum and bladder but with no obvious bony destruction

for S-100 protein (Figure 2C) and negatively for CD117, CD34, actin, and desmin (not shown). The final pathological diagnosis was reported as retroperitoneal cellular benign schwannoma. The patient is being followed up with three-monthly controls for 18 months.

Discussion

Retroperitoneal schwannomas comprise 3% of all schwannomas. Although they are mainly in benign nature, malignant tumors have been reported (1). Retroperitoneal benign schwannomas are solitary and slow-growing; they may remain asymptomatic in the retroperitoneum until they reach a large size (2). Patients are usually diagnosed in the third through fifth decades. Benign schwannomas may arise along the course of any myelinated nerve.

The typical schwannoma is a well-circumscribed mass with a capsule of epineurium (3). The hallmark pattern of the benign schwannoma is an alternation of Antoni A and B areas, with a diffuse positivity for S100 protein in the cytoplasm of tumor cells (1-3). Antoni A regions are characterized by dense aggregation of spindle-shaped cells arranged in parallel configurations, palisades, or whorls. Antony B regions manifest as hypocellular areas with a predominantly loose myxoid matrix (4). Also, positive staining for S100 protein and negative staining for CD34 are helpful in the diagnosis and differentiation of a benign schwannoma from a malignant peripheral nerve sheath tumor and from other benign spindle cell tumors (5). In our case, the tumor had Antoni A-dominated areas that were S-100-positive, but CD117, CD34, actin, and desmin negative (Figure 2A, C).

The mainstay treatment is complete excision; as well, there are also some reports of laparoscopic resections (6). However, the surgery can be dangerous, especially if the tumor is adherent to peripheral tissue and organs. Severe bleeding complications due to damage to vascular structures can occur (7). Carpenter reported one intraoperative death related to uncontrollable hemorrhage from severing the right common iliac artery during a difficult dissection. Also, hypervascularity of the tumor can further complicate its excision (8). Therefore, preoperative angiography has been recommended to assess tumor vascularity, operability, and the origin of the tumor blood supply (9).

Radiotherapy may be given to reduce the tumor vascularity and size, but the malignant form appears to be unresponsive (10). Once completely excised, the recurrence of benign schwannomas is not expected. In our case, the tumor was very hypervascular and the excision was deferred to a second surgery because of the difficulty in controlling hemorrhage during the biopsy. Following wound healing, the patient underwent 33 scans of radiotherapy in order to reduce tumor vascularization by inducing fibrosis. In the following period (approximately two months after radiotherapy), the patient was operated on and the mass was totally excised without any complications.

Fibrosis is a late response of tissues to radiation. The loss of vascular endothelium and thrombosis leads to capillary necrosis. Capillary destruction begins at 40 Gy, while arterial damage begins at 50 Gy of radiotherapy; veins are less sensitive to radiotherapy. As the cell cycle period is prolonged in venular smooth muscle cells, the loss of these cells takes a long time. The process of apoptosis begins in

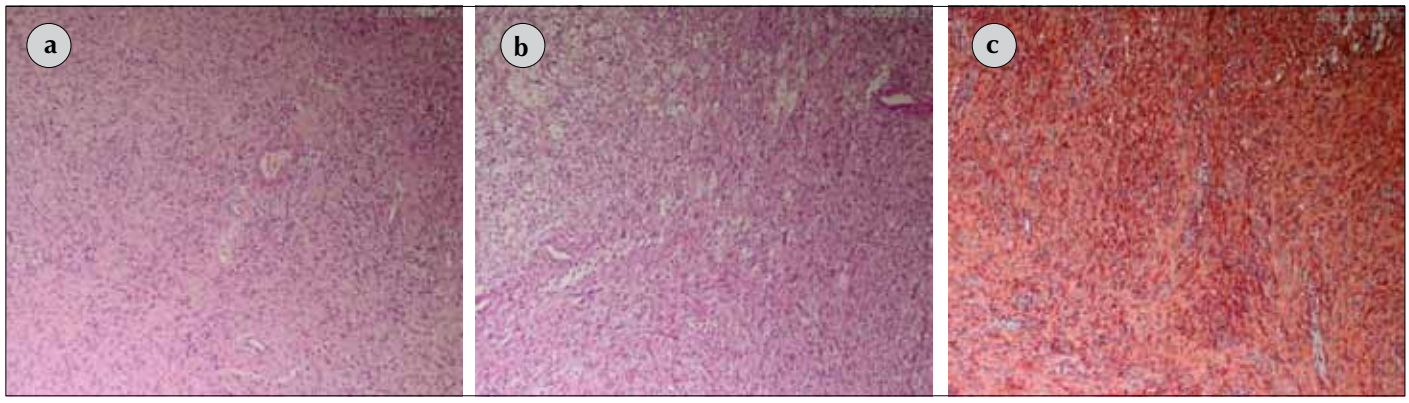

Figure 2. Microscopic view of the specimen showing the typical appearance of a cellular benign schwannoma. (a) (left-top): Spindle cells in Antoni type A areas without nuclear palisading (HE, x100). (b) (right-top): Hyalinized blood vessels and histiocytes were observed between tumor cells (HE, x100). (c) (left-bottom): Positive staining for S-100 protein in a cytoplasmic pattern (S-100, x100)

endothelial cells 4-8 hours after the start of radiation therapy and is more significant after 48 hours. Stress, hypoxia, and apoptosis due to radiotherapy induce angiogenesis inside the tumor and this may induce bleeding. In the late period, collagen synthesis increases in the tissue, leading to fibrosis and the obstruction of blood vessels and the surrounding tissue (11).

Conclusion

This case indicates that preoperative radiotherapy should be recommended in the treatment of schwannomas, particularly in hypervascular ones.

Conflict of Interest
No conflict of interest was declared by the authors.

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Authors' contributions: Conceived and designed the experiments or case: SC, MT, FB. Examination and follow-up of the patient: SC, MFD, ET. Analysed the data: SC, MNY. Wrote the paper: SC. All authors have read and approved the final manuscript.

Çıkar Çatışması
Yazarlar herhangi bir çıkar çatışması bildirmemişlerdir.

Hakem değerlendirmesi: Bağımsız hakemlerce değerlendirilmiştir.

Yazar katkıları: Çalışma fikrinin tasarlanması: SC, MT, FB. Hastanın muayenesi ve takibi: SC, MFD, ET. Verilerin analizi: SC, MNY. Yazının hazırlanması: SC. Tüm yazarlar yazının son halini okumuş ve onaylamıştır.

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