

Retroperitoneal Schwannoma: A Case Report

Retroperitoneal Schwannom: Bir Olgu Sunumu

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Abstract

Schwannomas are benign tumors originating from Schwann cells of the neural sheaths. Only 0.3% to 3% of the schwannomas are located the retroperitoneal space. Most schwannomas are asymptomatic and often found incidentally. The ideal treatment is complete surgical excision. Here, we report a case of a 49-year-old woman with retroperitoneal schwannoma.

Keywords: Schwannoma, retroperitoneum, tumor

Öz

Schwannomlar sinir kılıfının Schwann hücrelerinden köken alan benign tümörlerdir. Schwannomların sadece %0,3 ile %3'ü retroperitoneal alanda bulunur. Schwannomların çoğu asemptomatiktir ve sıklıkla insidental olarak saptanır. Komplet cerrahi eksizyon ideal tedavi yöntemidir. Bu olgu sunumunda, retroperitoneal schwannomlu 49 yaşında bir kadın hastayı sunuyoruz.

Anahtar Kelimeler: Schwannom, retroperiton, tümör

Introduction

Schwannomas are usually benign tumors arising from Schwann cells of the peripheral nerve sheath (1). Schwannomas can be isolated sporadic lesions or associated with genetic syndromes such as schwannomatosis or neurofibromatosis (2,3). In the absence of genetic syndromes, only 0.3% to 3.2% occurs in the retroperitoneum (1,2). Schwannomas are often found incidentally, or present with vague and non-specific symptoms (4). Here, we describe a case of a 49-year-old woman with retroperitoneal schwannoma.

Case Presentation

A 49-year-old woman had left suprarenal mass found on lumbar magnetic resonance imaging (MRI) for acute lumbar pain and was referred to our institution for further evaluation. Her medical and family history was unremarkable. To further characterize the mass, renal MRI was performed. MRI showed a left retroperitoneal paraaortic mass near the renal hilus,

44x40x49 mm in dimension. The mass was hypointense on T1-weighted images, and hyperintense with central cystic component on T2-weighted and fat-saturated T2-weighted images (Figure 1).

The patient underwent open retroperitoneal mass excision during which the surface of the tumor was found to be smooth and not adherent to the adjacent structures. The resected specimen was spherical and firm and it measured 5x4.5x4 cm. On microscopic examination, the mass consisted of a proliferation of fusiform cells which formed a palisade pattern (Antoni A type) and the regions were composed of mixoid and degenerated tissue with fewer cells and a gelatinous substance (Antoni B type) (Figure 2). Immunohistochemically, the tumor cells were strongly positive for S-100 protein expression and negative for CD-34 (Figure 3).

The postoperative course was uneventful, and the patient was subsequently discharged on the sixth post-operative day. At 6 months follow-up, the patient remained asymptomatic with no evidence of recurrence.

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Discussion

Schwannomas, previously referred to as neurilemmomas, typically derive from Schwann cells of the peripheral nerve sheath (1). They can develop in any nerve trunk in the entire body (except the cranial nerve I and II, which are not enveloped in a Schwann cell sheath) but are found most commonly in cranial and peripheral nerves of the upper limb (2). The majority of retroperitoneal schwannomas are benign in nature although malignant ones have also been reported (5). Schwannomas comprise approximately 4% of all retroperitoneal tumors and only 0.3-3.2% occurs in the retroperitoneum (2,6). They usually affect adult patients aged 20 to 50 years (7,8).

Since the retroperitoneum is rather large, flexible and nonrestrictive space and schwannomas are almost invariably slow growing, the diagnosis of retroperitoneal schwannomas is often delayed or incidental and they can reach a significant size at the time of diagnosis (4,9).

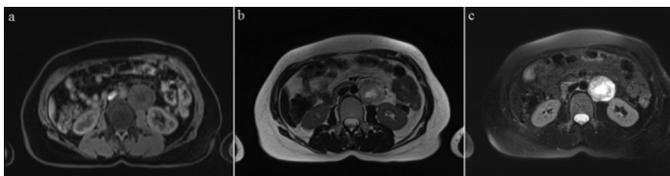


Figure 1. Axial magnetic resonance imaging images of the patient showing well-defined spherical mass which is hypointense on T1-weighted imaging (a), hyperintense with central cystic component on T2-weighted imaging (b) and fat-saturated T2-weighted imaging (c)

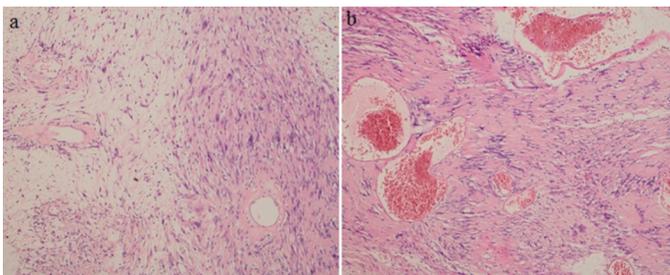


Figure 2. Histopathological examination with hematoxylin and eosin staining showing Antoni A and less cellular Antoni B areas (a) and palisade pattern (b) of organized spindle cells (x100)

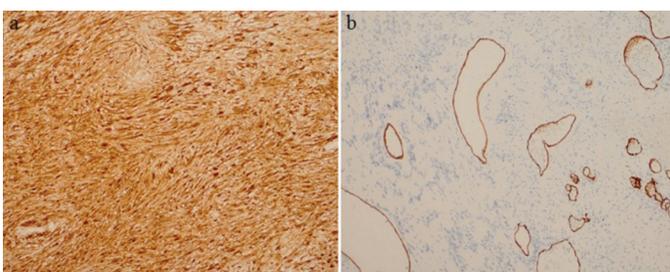


Figure 3. Immunohistochemical-examination showing negative CD-34 staining (a) and positive S-100 staining (b) (x100)

Although patients with a retroperitoneal schwannoma are usually asymptomatic, some patients present with vague and nonspecific symptoms (abdominal or back pain, abdominal distention) (5,9). Depending on the location of the tumors, secondary hypertension, hematuria and renal colic have also been reported (10).

Ultrasonography and computed tomography (CT) are helpful in approximating the size, location, presence of invasion and involvement of adjacent organs (7). MRI allows better visualization of its origin, vascular architecture and involvement of other organs (9). Typically, benign schwannomas appear as tumors with smooth margins that are isointense with muscle on T1-weighted images and hyperintense on T2-weighted images. However, as with the other imaging modalities, there is no characteristic or specific finding for schwannomas (9). The differential diagnosis of retroperitoneal schwannoma includes paraganglioma, neurofibroma, ganglioneuroma, tumors of mesodermal origin and retroperitoneal malignancies (malignant fibrous histiocytoma, lymphoma and liposarcoma) (3).

Although CT-guided biopsy may be helpful for diagnosis if the sample contains sufficient Schwann cells to visualize microscopically, it is usually unreliable. Therefore, many authors do not recommend this modality as a diagnostic tool (9).

The ideal treatment for retroperitoneal schwannomas is complete surgical excision (1,9). However, one should be cognizant that the procedure can be technically challenging, especially, if the tumor is adherent to adjacent structures or hypervascular (9).

Macroscopically, schwannomas are solitary, well-circumscribed, firm and smooth surfaced encapsulated tumors (7,9). Furthermore, large schwannomas may show cystic degeneration, hemorrhage and central necrosis (4). The definite diagnosis is made by histopathological examination and immunohistochemistry (3). Microscopically, they demonstrate Antoni A areas (well-organized spindle cells in a palisade pattern) and Antoni B areas (less cellular, loose pleomorphic cells with an abundant myxoid component). Being positive for S-100 and negative for CD-34 are another two features supporting a correct diagnosis (3,8). In our case, the tumor had Antoni A dominated areas that were S-100 positive, but CD-34 negative (Figure 3).

The prognosis of benign schwannomas is good and recurrence is rare. Recurrences are probably due to incomplete excision which is reported in 5% to 10% of cases (11). In the absence of other schwannomas and with a lack of family history, which excludes schwannomatosis or neurofibromatosis, further investigations are not necessary (3).

In conclusion, retroperitoneal schwannomas are rare tumors that are difficult to diagnose preoperatively. Radiologic findings are usually nondiagnostic. Diagnosis is based on histopathology and immunohistochemistry. The mainstay treatment is complete surgical excision.

Ethics

Informed Consent: Consent form was filled out by the participant.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.R.G., B.H., G.E., Concept: C.Ö., Design: C.Ö., Data Collection or Processing: C.Ö., M.R.G., B.H., G.E., Analysis or Interpretation: C.Ö., M.R.G., Literature Search: C.Ö., Writing: C.Ö., M.R.G.

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