Case Report

Retroperitoneal Schwannoma: A Case Report and Review of The Literature

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Abstract

A 32 year-old woman admitted to our clinic with right flank pain. Her abdominal computed tomography showed a huge mass in the right adrenal area, compressing the right kidney. The mass had a regular surface and consisted of cystic and necrotic areas. The left kidney and the adrenal gland had a normal appearance. The magnetic resonance imaging demonstrated that the lesion originated from neural foramina of the lumbar area. The mass was freed from the spinal cord and was completely resected using posterior and retroperitoneal approach. Histopathological examination of the specimen revealed a degenerate schwannoma. Patient was discharged on the 4th day after the operation without any complication. The patient has been uneventful until the 6th month of follow up.

Keywords: Schwannoma, Retroperitoneal tumor, Nerve sheath tumor

INTRODUCTION

Schwannoma is also called to as neurinoma, neurilemmoma, and perineural fibroblastoma, and is a tumor originating from the Schwann's cells derived from the neuroectoderm. In 1980, Verocay reported for the first time a tumor that occurred in the neurons histologically. Masson has described that these tumors arise from Schwann's cells and are termed as schwannoma. Most schwannomas are benign, though malignant schwannomas are usually seen associated with Von Recklinghausen's disease. The majority of the schwannomas arise from cranial nerves or nerves of upper extremities but rarely they can arise from mediastinum and retroperitoneum. Urinary tract is seldom involved, most commonly testis, penis, spermatic cord, and tunica vaginalis. Retroperitoneal schwannoma is also rare and is commonly diagnosed by the urologists, secondary to...
symptoms produced. Most common presentation of the retroperitoneal schwannoma is abdominal distention with vague or dull abdominal ache(21,26,32).

We report on a case with benign retroperitoneal schwannoma and describe the diagnostic and management options.

CASE PRESENTATION

A 32-year-old woman was referred to our clinic with a chief complaint of right flank pain for one year. Past medical history and review of systems was otherwise unremarkable. A thorough physical and abdominal examination were made and were normal. Blood work for kidney and liver function tests and urinalysis and urine culture were normal. Computed tomography (CT) of the abdominal area showed a huge mass in the right adrenal area. Upper abdominal magnetic resonance imaging (MRI) confirmed the presence of a right retroperitoneal mass which had a heterogeneous reflectivity in T1- and T2-weighted images, with cystic and necrotic components on T2-weighted images. After gadolinium injection, the mass had a heterogeneous contrast but there was no contrast seen at the central necrotic area. Lesion dimensions were 9.5 x 7 x 8 cm and had a regular surface. The lesion originated from the right paravertebral distance at T12-L1 and L3-4 levels and also from the neural foramina at L1-2 levels. The mass was dumbbell shaped and compressed the liver, kidney and the right adrenal gland (Figure 1).

\[\text{Figure 1: The mass was dumbbell shaped and was compressing the liver, kidney and the right adrenal gland (A: Coronal image, B: Axial image, T1 and T2 sequence).}\]

In the pre-operative period we didn't take CT guided biopsy. Because this mass was mimicking adrenal tumor and in preoperative period we didn't correctly estimate the mass originated from adrenal tissue or nerve sheath. So we started with a posterior approach in the beginning and we tried to cut off the connection of the mass with medulla spinalis.
The patient underwent open surgery and the mass freed from the spinal cord by a posterior approach. L1 hemilaminectomy, L2 foraminectomy was performed. Additionally some of the lateral fibers of L2 nerve root were sacrificed because of tumor involvement. Frozen section was taken during the surgery before complete resection of the mass. Frozen section of the mass confirmed the diagnosis of schwannoma. Thereafter the mass (9 cm × 7 cm × 8 cm) was completely resected by a retroperitoneal approach, (Figure 2). In the early and long-term postoperative period there was no neurological deficit in our patient, as confirmed on neurological examination. On the 4th postoperative day, patient was discharged symptom-free. After 6 months, physical examination and control abdominal CT was normal. The pathology report concluded the final diagnosis as degenerate schwannoma.

DISCUSSION
Schwannomas are usually benign tumors arising from the schwann cells of the peripheral nerve sheath(33). The benign schwannomas produce symptoms are nonspecific and depend on the location and size of the mass and results in compression and irritation of the nerve roots, gastrointestinal system, urinary system, impingement of the major vessels, and even severe destruction of the adjacent bone. Most common symptoms are abdominal distention and abdominal ache/pain(21,26,32). More atypical presentations include flank pain, hematuria, headache, secondary hypertension, recurrent renal colic pain and Horner’s Syndrome (29,33). In the presented case, the main symptom was also flank pain.

Retroperitoneal schwannomas occur most commonly between 40 and 60 years of age, with a male/female ratio of 2:3(24,35). The diagnosis of patients with retroperitoneal schwannoma was usually delayed(6,13,21). Because the retroperitoneal cavity is flexible, unless the tumor becomes a substantial size, symptoms are not developed. At the time of diagnosis, the size of a retroperitoneal schwannoma is usually more than 8 cm in diameter as seen in our case(18).

Though schwannomas are found in peripheral nerve fibers in the limbs, head, and neck, they are the cause of 0.5 to 12% of retroperitoneal masses(17). In reviewing the recent 20-year English literature, 51 cases of schwannoma from spinal nerve root were reported. The high incidence areas were lower cervical and lumbosacral region. Authors claim that chronic irritation of the nerve roots in these degenerative processes of the spine may play a role(4).

Keep in mind the differential diagnoses for a retroperitoneal schwannoma include, epithelial cyst, abscess, plexiform malignant peripheral nerve sheath tumor (MPNST), sacral meningioma, ependymoma, chordoma, chondrosarcoma, giant cell tumor, aneurysmal bony cyst, osteoblastoma, sacrococcygeal teratoma, lymphoma, and the malignant
transformation of a benign tumor. These disease categories can be distinguished from schwannoma by consideration of the clinical symptoms, age, the radiologic image, subcutaneous paracentesis, and chromosomal study\(^{(23)}\). Ganglioneuroma is a neurogenic tumor which should be considered in the differential diagnosis. It is shown more commonly in the retroperitoneum than schwannoma and has similar findings on CT and MRI\(^{(27)}\).

Macroscopically, schwannomas are solitary, well circumscribed, firm, smooth-surfaced tumors. Because of their large size, these tumors are likely to manifest degenerative changes such as cysts and calcification\(^{(5)}\). Microscopically, the benign variant has a variation of Antoni A and B areas, with a diffuse positivity for S100 protein in the cytoplasm of the tumor cells\(^{(5,33)}\).

Most schwannomas are benign and malignant degeneration of schwannomas is extremely rare. Malignant degeneration particularly occurs in association with Von Recklinghausen’s disease, as occurs in 5% to 18% of cases\(^{(25)}\). To date, no standard diagnostic criteria or radiologic feature of malignant schwannomas have been described. Malignant schwannomas are commonly larger in size and they act as high-grade sarcomas with the possibility of producing local recurrence and distant metastasis. Typically, malignant schwannomas are diagnosed histopathologically after the surgical excision of a mass, with features of high mitotic rate, pleomorphism, and blood vessel infiltration\(^{(3)}\).

Appropriate radiological evaluation is important both for diagnosis and management. Computed tomography typically shows well-defined mass with low or mixed attenuation and cystic and/or necrotic central areas. Cystic changes occur more commonly in retroperitoneal schwannomas (up to 66%) than in other retroperitoneal tumors\(^{(36)}\). Other degenerative changes, such as calcification and hemorrhage can be seen on CT. On MRI, schwannomas are seen as masses of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images\(^{(12,24)}\). However these radiologic findings are characteristic but not specific of schwannomas and was noted in only 57% of the cases\(^{(12)}\). Depending on the cell density, the signal intensity on T2-weighted images may vary. Tumors with microscopic findings of hypercellular Antoni type A tissue have intermediate signals, while tumors with Antoni type B tissue have a bright signal on T2-weighted images. However, based solely on imaging, the percentage of misdiagnosis was found very high (%84). Common misdiagnoses are hepatic tumors, pancreatic cystic tumors, and psoas abscess\(^{(18,27,28)}\). CT-guided core biopsy and fine needle aspiration are not sufficient for the diagnosis of retroperitoneal schwannoma\(^{(17)}\). Degenerated areas can hinder the correct diagnosis and malignancy may be missed because of cellular pleomorphism and the presence of degenerative cells. Additionally, these procedures have the risks of hemorrhage, infection, and tumor seeding. Thus, many authors do not recommend CT-guided biopsy\(^{(5,22)}\).

Treatment depends solely on surgery. Laparoscopic resection may be useful because a retroperitoneal schwannoma, which is commonly localized and hypovascular, can easily be dissected from the adjacent tissues\(^{(14,37)}\). Considerable controversy exists over management of negative soft tissue margins. Since local recurrence rate ranges from 16% to 54% after conservative approach, sacrifice of adjacent tissues and viscera may be considered for complete surgical resection\(^{(1,5,35)}\). Dominguez et al.\(^{(7)}\) have reported recurrence in 16% in partial resection\(^{(7)}\). Abernathey et al. did not approve partial resection of the tumor because of the consideration of local recurrence\(^{(2)}\). Giglio et al. proposed that even if the tumor were determined to be
benign using frozen biopsy, the possibility of malignancy can not be excluded accurately\(^{(10)}\). On the other hand, some authors believe that a simple enucleation or partial excision of the tumor is sufficient. Because the benign nature of the disease, no increase in the size of schwannoma during a 6- and 14-year period and have been reported\(^{(28)}\). Is the size of the residual tumor after surgery the greatest factor that influence on the recurrence of schwannoma. Therefore, maximal removal as completely as possible of the operable part of tumor without severe hemorrhage and anticipated neurological injury level after surgery are two factors considered during the surgery\(^{(8,10,16,17,34)}\).

Definition of the originating nerve may not always be possible and a minor degree of neurological impairment is therefore to be anticipated\(^{(30)}\). Kim et al. demonstrated that neurological deficit was detected in 23% of the patients after completely killing a specific lumbar nerve root for tumor resection, but none of the deficits were debilitating to the patient\(^{(16)}\). The reason why only minor neurological deficit was detected in some cases after root resection is that the involved nerve may had became functionally compensated by the neighboring roots before operation\(^{(16,31)}\). There are a few reported cases in which metastases occurred after resection of a histologically benign schwannoma\(^{(24)}\). The prognosis for retroperitoneal benign schwannoma is good. On the other hand, in malignant cases, extraction of the tumor and adjuvant radiation therapy or chemotherapy are required, and even in cases in which the appropriate treatments are administered, prognosis is poor, and it has been reported in one series that 62% of patients died. Furthermore in these cases diagnosed using histologic results, the extraction is often insufficient and the local recurrence rate is high. White has reported that 60% died within 2 months, and Ghosh et al. have reported that in cases with a single lesion, the 5-year survival rate is 72.5%, and in cases concomitant with von Recklinghausen's disease the 5-year survival rate is 30\%\(^{(9,14,38)}\). Single therapy modality of malignant schwannomas has shown poor results.

In summary, retroperitoneal schwannomas often present as unrecognized slow growing masses. Keep in mind potentially severe bleeding and neurological deficit risk of surgical intervention without away from oncologic principle. Therefore, careful preoperative evaluations and postoperative monitoring is necessary.

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