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Ancient Schwannoma of Abdominal Wall: A Case Report

Karın Duvarının Antik Schwannomu: Olgu Sunumu

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Abstract

Schwannomas are mostly asymptomatic benign tumors originating from peripheral or cranial nerve sheath cells. They are rare and slow-growing tumors that may present with compression-related symptoms such as a palpable mass or incidentaloma. Schwannoma, which is frequently located on the flexor surfaces of the head, neck, and extremities, is usually diagnosed by radiological and pathognomonic microscopic and immunohistochemistry features. Herein, we report the case of a 31-year-old woman who presented with a palpable mass on the lateral abdominal wall.

Keywords: Schwannoma, Ancient schwannoma, Neurogenic tumor, Surgical treatment

Öz

Schwannomlar periferik veya kranyal sinir kılıf hücrelerinden köken alan, çoğunlukla asemptomatik olan benign tümörlerdir. Ele gelen kitle veya insidentaloma gibi basıya bağlı semptomlar gösterebilen nadir ve yavaş büyüyen tümörlerdir. Sıklıkla baş, boyun ve ekstremitelerin fleksör yüzlerinde yerleşen Schwannom'un tanısı genellikle radyolojik görüntüleme bulguları ve patognomonik mikroskobik ve immünohistokimyasal özellikleri ile konulur. Burada karın yan duvarında ele gelen kitle şikayeti ile başvuran 31 yaşındaki kadın hastayı sunuyoruz.

Anahtar Kelimeler: Schwannom, Antik Schwannom, Nörojenik tümör, Cerrahi tedavi

Introduction

Schwannoma, or neurilemmoma with its former definition, is a soft tissue tumor that is mostly benign, tends to grow slowly, has a capsule, and originates from Schwann cells of peripheral, cranial, and autonomic nerves. Schwannomas constitute up to 5% of all benign soft tissue tumors and are the most common type of peripheral nerve-derived tumors. They are also more common in young women (1-10). The most common site of origin is the head and neck region, but reports on the flexor surfaces of the extremities, retroperitoneal area, and other regions are present in the literature [2,6]. In addition, schwannomas may present with a palpable mass or as incidentaloma. Ancient schwannoma is a rare subtype of schwannoma that has a macroscopic appearance with degenerated areas and associated

cystic areas [5-10]. In this study, we aimed to report a case who complained of pain in the right side of the abdomen and right leg; a mass was detected in the right lateral abdominal wall by examination and imaging methods. After the complete excision of the tumor, the final pathological examination confirmed ancient schwannoma.

Case Report

A 31-year-old woman with no history of chronic or familial disease presented to our outpatient clinic with right flank, back, and leg pain accompanied by swelling on the right flank that has been ongoing for almost a year. On physical examination, on the junction of the right flank and back, a slightly mobile, soft, and well-circumscribed mass was palpated. Laboratory findings



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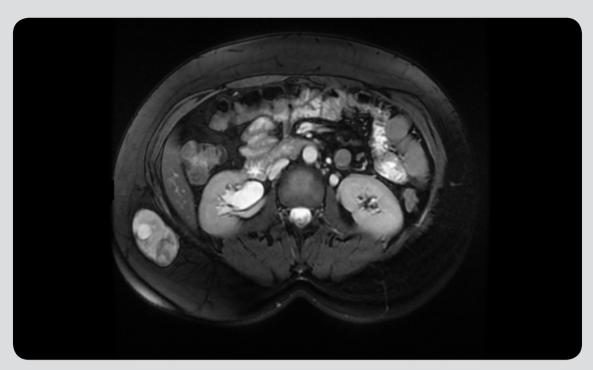


Image 1. The mass shows hyperintense signal intensity on axial T2-weighted image with fat suppression.

were unremarkable. The ultrasonographic evaluation revealed a 6 cm mass with well-circumscribed edges accompanied by calcific areas. On magnetic resonance imaging, the lesion was located on the posterolateral abdominal wall in the subcutaneous fatty tissue and appeared as a nodular lesion with indistinct contrast enhancement in post-contrasted series. Also, no diffusion restriction was detected (Image 1). She underwent surgery for resection, and the lesion was resected with clear macroscopic margins. There were no firm, dense adherences between the lesion and adjacent structures (Image 2). Subsequently, postoperative immunohistopathologic findings were consistent with ancient schwannoma. Macroscopically, the lesion was hemorrhagic and locally calcified with cystic areas. On microscopic examination, the tumor was encapsulated with a large cystic space and hyalinization accompanied by solid cellular and hypocellular areas. Tumor cells were spindle-shaped with twisted nuclei and had indistinct cytoplasmic borders arranged in short bundles and palisades. Degenerative changes, including nuclear atypia and hyalinization, were present. The immunohistochemical examination detected diffuse

and strong positives starting with S-100 (Image 3). The postoperative course was uneventful, and the patient is disease-free in her first year of follow-up.



Image 2. A well-defined subcutaneous mass, enucleated with a single incision through a direct approach.



Discussion

Ancient schwannomas are rare sporadic ectodermal neoplasms arising from the surrounding axons of the nerve sheath (3). After the first description of schwannomas by Verocay in 1908, ancient schwannomas were described by Ackerman and Taylor in 1951 as a subtype (1,4). Degeneration and hypocellularity within the tumor are pathognomonic features. The term "antique" refers to tumors with a slow growth rate, and cystic changes represent areas of Antoni type B that occupy the vast majority of the tumor (4).

Clinical presentation alters by the localization of the lesion. Peripheral nerve tumors like schwannoma or neurofibroma may present with neurologic deficits such as pain, paresthesia, and less frequent motor weakness (2). In addition, ancient schwannomas may sometimes present with muscle atrophy due to the denervation of related nerves (7).

Diagnosing schwannomas is difficult due to imaging methods' lack of characteristic appearance. Magnetic resonance imaging is the most frequent radiologic tool for diagnosis, but yet distinguishing ancient schwannoma from intramuscular classic schwannoma or neurofibroma is challenging (7). On T2-weighted images, hemorrhagic, cystic, and necrotic areas may appear as heterogeneity and may appear as the target or split fat signs (7,8). When detected, the presence of a nerve with entry and exit to the mass is pathognomonic for peripheral nerve sheath tumors to distinguish them from neurofibroma (6).

Histopathological ancient schwannomas consist of areas with hyper and hypocellular regions called Antoni A and Antoni B areas. Cystic changes are seen in Antoni type B areas, defined as hypocellular, covering most of the tumor. With the presence of

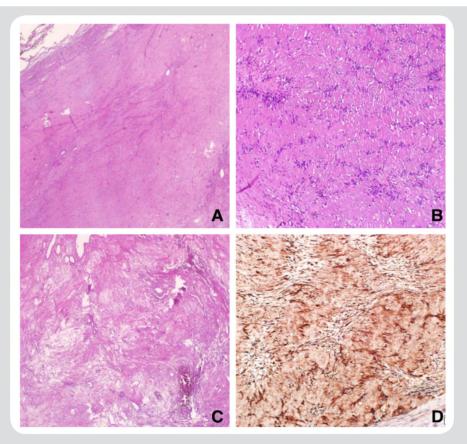


Image 3. Diffuse hyalinization and cystic changes (A). Cellular areas (Antoni A) with focal nuclear palisading (Verocay bodies) and hypocellular areas (Antoni B) (B, C). Diffuse and strong positive staining with S-100. (A: Hematoxylin-eosin x 40; B,C: Hematoxylin-eosin x 100; D: Immunohistochemical analysis x 100)



degenerative changes such as cyst formation, bleeding, calcification and hyalinization, these tumors are called ancient schwannomas (7). Strong positive staining with S100 protein is pathognomonic and aids in differentiating schwannomas from malignant peripheral nerve sheath tumors (9). However, complete surgical resection with a negative margin is warranted due to its potential to recur (1-10).

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