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Case Report / Olgu Sunumu

Giant retroperitoneal ancient schwannoma: An unusual schwannoma type in the rarest localization

Dev retroperitoneal ancient schwannoma: Oldukça nadir bir lokalizasyonda, nadir tipte bir schwannoma

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ABSTRACT

Schwannomas are tumors originating from Schwann cells in the nerve sheath. Although they can appear on many parts of the body, they occur most frequently in the head and neck. Furthermore, they are very rarely seen in the retroperitoneal region. Ancient schwannoma is a sub-type of schwannomas that is quite rare. In this study, the case of a retroperitoneal ancient schwannoma, which showed cystic degeneration and reached gigantic sizes, is presented. A forty-nine-year-old female patient was admitted to our center with complaints of growing abdominal distention and stiffness for 7 years. In the physical examination, a non-mobile and stiff mass that was approximately 30 cm along its longest axis and extended from the subcostal area to the inguinal region was detected on the right side of the abdomen. A solid retroperitoneal mass containing encapsulated and cystic components was confirmed in the oral and intravenous contrasted abdominopelvic tomography. The size of the mass was 28×20×12 cm, and a total excision of the mass was planned. The mass was excised together with the capsule. In the pathological examination, ancient schwannoma, which showed widespread multifocal cystic degeneration with benign characteristic, was found. In the immunohistochemical staining, S100 was found to be positive. The patient was discharged on the 5th postoperative day. Schwannomas are slowly progressive benign tumors that can reach a large size without any symptoms and rarely settle in the retroperitoneal region. Ancient schwannomas are also a rarely seen sub-type of schwannomas. In this study, the case of a retroperitoneal ancient schwannoma, which showed cystic degeneration and reached gigantic sizes, is presented.

Keywords: Giant, retroperitoneal, ancient, schwannoma

ÖZ

Schwannoma'lar, sinir kılıflarındaki schwann hücrelerinden köken alan tümörlerdir. Vücudun pek çok bölgesinden kaynaklanabilirler. En sık olarak baş boyun bölgesinde görülürler. Retroperitoneal bölgede ise oldukça nadirdirler Ancient schwannoma ise oldukça az görülen bir subtipdir. Bu vaka sunumunda; nadir görülen, kistik dejenerasyon göstermiş ve dev boyutlara ulaşmış retroperitoneal ancient schwannoma olgusu sunulmuştur. Kırk dokuz yaşında kadın hasta, merkezimize 7yıldır karında giderek büyüyen şişlik ve sertlik şikayeti ile başvurdu. Fizik muayenede karnın sağ tarafında; subkostal alandan inguinal bölgeye kadar uzanan sert, mobilitesi olmayan ve uzun ekseni yaklaşık 30cm olan kitle palpe edildi. Oral ve intravenöz kontrastlı abdominopelvik tomografide enkapsüle, kistik komponentler de içeren solid retroperitoneal kitle saptandı. Kitlenin boyutu 28x20x12 cm boyutlarında idi. Kitlenin total eksizyonu planlandı. Kitle, kapsülü ile birlikte total olarak çıkarıldı. Patolojik incelemede yaygın multifokal kistik dejenerasyon gösteren benign karakterli ancient schwannoma tespit edildi. İmmünohistokimyasal boyamada S100 pozitif bulundu. Hasta ameliyat sonrası 5. günde taburcu edildi. Schwannomalar yavaş seyirli benign tümörlerdir. Belirti vermeden dev boyutlara ulaşabilirler. Shwannomalar olukça nadir olarak retroperitoneal bölgeye yerleşirler. Ancient schwannomalar, schwannomaların nadir görülen bir subtipidir.

Anahtar kelimeler: Dev, retroperitoneal, ancient, schwannoma

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INTRODUCTION

Schwannomas are tumors originating from Schwann cells in the nerve sheath. They are generally solitary and encapsulated. Although they can appear on many parts of the body, they occur most frequently in the head and neck, and they are rarely seen in the retroperitoneal region. Although schwannomas are slow growing tumors with benign characteristics, they can reach gigantic sizes over the years without any symptoms (1). Ancient schwannoma, on the other hand, is a sub-type of schwannoma that is quite rare (2). In this study, the case of retroperitoneal ancient schwannoma, which showed cystic degeneration and reached gigantic sizes, was presented.

CASE PRESENTATION

A forty-nine-year-old female patient was admitted to our center with complaints of growing abdominal distention and stiffness for 7 years. She was previously admitted to another health care provider with these complaints, and a surgery was recommended for her, but she refused to have a medical operation at that time. In recent months, some complaints including an increase in abdominal pain, daily life stress, and increased effect of the mass depending on its weight and constipation had gradually increased. In the physical examination, a non-mobile and stiff mass that was approximately 30 cm along its longer axis and extended from the subcostal area to the inguinal region was detected on the right side of the abdomen. The patient's vital signs were normal. There were no abnormal findings except anemia in the tests. Her hemoglobin value was 8.6 gr/dl. Her medical history indicated that she had myoma uteri that caused excessive bleeding, which also caused anemia. The solid retroperitoneal mass containing encapsulated and cystic components was confirmed in the oral and intravenous contrasted abdominopelvic tomography. The size of the mass was 28×20×12 cm. This mass pushed the duodenum and small intestines to the left. The right kidney was pushed back and up. The vena cava inferior was attached to the mass and prominently deviated to the left (Figure 1). Therefore, total excision of the mass was planned. Two units of erythrocyte suspension were transfused to the patient before the surgery. In the surgery, a midline laparotomy incision was performed from the xsifoid to below the navel. Retroperitoneal tumors were observed. The mass extended from the subhepatic region to the level of the right iliac vessels, and its longer axis was 28 cm (Figure 2).

The inferior vena cava on the mass was separated from the mass using a sharp and thin dissection. The mass was excised together with the capsule. The size of the mass was reduced to 5 cm in the macroscopic scale due to the aspiration of liquid in cystic components. The macroscopic dimension of the mass was identified as 23×18×10 cm (Figure 3). Postoperative problems did not emerge. The drain of the patient was removed on the 4th day. The patient was discharged on the 5th postoperative day. After two weeks, the patient did not have any postoperative problems. In the pathological examination, ancient schwannoma, which showed widespread multifocal cystic degeneration with benign characteristics, was found. In the immunohistochemical staining, S100 was found to be positive.

Figure 1. The image of the mass by its coronal section in the computed tomography

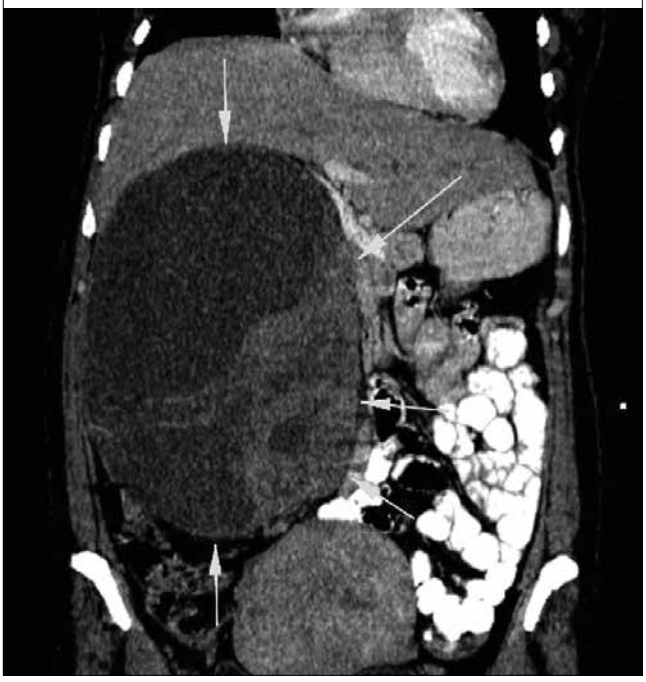
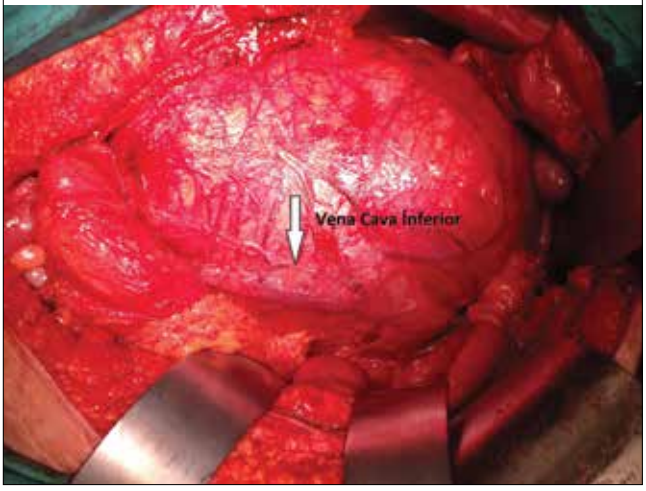


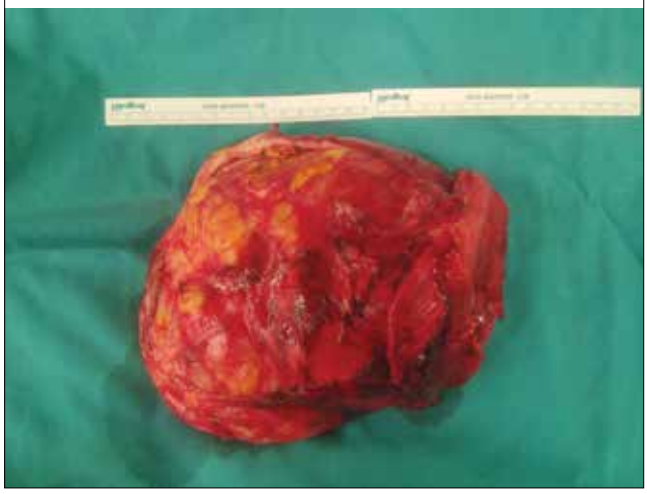
Figure 2. The appearance of the mass during the surgery and its relationship with inferior vena cava



DISCUSSION

Schwannoma was found by Mason in the Schwann cells for the first time in 1932. The term ancient schwannoma was used by Ackerman and Taylor for the first time in 1951 to describe a very rarely seen sub-type of schwannomas containing hypocellular and degenerative areas. These changes are thought to be due to its progression over many years (3). They are slow-growing and benign tumor masses that originate from the peripheral nerve sheath. A malignant type known as neurofibrosarcoma can turn into a tumor at a ratio of 1%. Schwannomas are often seen in the head and neck region of the body. The retroperitoneal schwannomas are seen within a range of 0.7%–5% (1). This case was a case of schwannoma that originated from the retroperitoneal area.

Figure 3. Macroscopic image of the mass that was totally excised



In the literature, the largest retroperitoneal schwannoma was one with the dimensions of 43×40×20 cm published by Foote et al. (4) in 1963, which was followed by the one published by Kuriakose et al. (5) with the dimensions of 42×16×16 cm and the one published by Schindler and Dixon (6) with the dimensions of 35×25 cm. Lastly, a retroperitoneal schwannoma with the dimensions of 32×28×26 cm was recently published by Mahendra et al. in 2014. Our case can also be considered to be among the large retroperitoneal ancient schwannomas that were reported in the literature with a size of 28×20×12 cm.

Imaging methods including ultrasound imaging, computed tomography (CT) and magnetic resonance imaging (MRI) are generally used for cases when an abdominal mass is found in the physical examination. In ultrasound imaging, the localization and structure of the mass is evaluated and other properties of the mass are determined after performing CT by giving intravenous contrast (5). In our case, localization and boundaries of the mass and its relationship with the surrounding organs were assessed in detail. The results of computed tomography showed the significant pressure of the mass on the inferior vena cava. In addition, it was observed that the mass caused a displacement of the right kidney to the superior and posterior region. However, since the mass was encapsulated, it did not have an obvious invasion into the adjacent organs. In our case, a direct excision was decided because of the pressure symptoms since the mass became symptomatic and can negatively affect the quality of life of the patient. Preoperative imaging guided biopsy remains controversial (7).

In these types of retroperitoneal tumors, total excision is recommended. Partial or subtotal resection is not enough, but subtotal resection can be performed in order to avoid injury to the vessel and adjacent organs. Relapse is inevitable in this type of approach (8). In our case, the mass was totally excised together with the capsule. In these types of cases, excessive bleeding from blood vessels adjacent to the mass may occur during the dissection in the surgery. In such cases, packing and re-intervention within 24–48 hours is quite helpful (6). Although another problem for this type of case is bleeding from lumbar arteries, in our case, minimal bleeding from the lumbar venous vessels was observed; however, there was no excessive bleeding. Bleeding was controlled by vascular repair.

In the pathological examination of schwannomas, Antoni A hypercellular, Antoni B hypercellular areas were observed in the microscopy. Antoni A regions form a cluster of cells known as verocoy's bodies that cells are typically stained by S-100 as immunopositive (6). In this case, immunohistochemical staining with S-100 was positive. Retroperitoneal paragangliomas should be considered in the differential diagnosis because of staining the S-100 positive (9). Some cases are misdiagnosed with malignancy pathologically. Nuclear atypia and pleomorphism may be the cause of this wrong diagnosis. In controversial cases, the mitotic index plays a vital role. In malignant schwannoma cases, there is an increase in the mitotic index and irregular infiltrates to the nuclear atypia and surrounding tissues (10, 11). In our case, the malignancy criterion was not observed.

Ancient schwannoma is a term used to describe hyperchromatic lesions containing nuclear atypia like degenerative changes. Ancient schwannomas are extremely rare tumors originated from the peripheral nerve sheath. Retroperitoneal schwannomas are also rarely seen tumors. This case was also a rarely seen retroperitoneal ancient schwannoma with giant dimensions.

CONCLUSION

Schwannomas are slowly progressive benign tumors that can reach huge dimensions without any symptoms and rarely settle in the retroperitoneal region. Ancient schwannomas are a rarely seen sub-type of schwannomas.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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