Annals of Surgical Case Reports

Case Series
Published: 21 Nov, 2022

Huge Adrenal Schwannoma: A Rare Type of Adrenal Tumor, Misconceived for Adrenocortical Carcinoma

*Han JH[1], Lee BC[1], Park YM[1], Jung HJ[1], Kim D[2] and Choi JB[1]**

[1]Department of Surgery, Pusan National University Hospital, Busan, South Korea

[2]Department of Surgery, Pusan National University Yangsan Hospital, Yangsan, South Korea

Abstract

Schwannoma is benign and uncommon neoplasm arising from the neural crest cells. The most common tumor sites are the skin and subcutaneous tissue of the head and neck. Retroperitoneal schwannomas account for 1% to 3% of all schwannomas and are predominantly female. In particular, adrenal schwannoma is very rare, and due to its large size at the time of diagnosis, adrenal schwannoma is frequently misdiagnosed as adrenocortical carcinoma or pheochromocytoma. Adrenal schwannoma is difficult to distinguish it from other adrenal diseases based on imaging findings alone. In this report, we introduce the case of left huge adrenal schwannoma.

Keywords: Adrenalectomy; Schwannoma; Adrenal surgery; Adrenal tumor

Introduction

Schwannoma is benign and uncommon neoplasm arising from the neural crest cells. The most common tumor sites are the skin and subcutaneous tissue of the head and neck, but sometimes schwannoma originates from the pancreas, stomach, kidney and liver [1]. Retroperitoneal schwannomas account for 1% to 3% of all schwannomas and are predominantly female [2]. In particular, adrenal schwannoma is very rare, and due to its large size at the time of diagnosis, adrenal schwannoma is frequently misdiagnosed as adrenocortical carcinoma or pheochromocytoma [3]. Adrenal schwannoma is classically well encapsulated, hypervascular and heterogeneous, so it is difficult to distinguish it from other adrenal diseases based on imaging findings alone. Histopathological and immunohistochemical studies of surgically removed specimens are the only and essential means of confirming the diagnosis [2,4].

In this report, we introduce the case of left huge adrenal schwannoma in an 87-year-old woman presented with abdominal pain and discomfort.

Case Presentation

An 87-year-old, female, with no past medical history, was admitted emergency center in our tertiary hospital (Pusan National University of Hospital, Busan, Korea) with a 1-month history of abdominal pain and discomfort. Initial work up using Abdominopelvic Computed Tomography (APCT) presented 16 cm sized heterogeneous tumor in left adrenal gland (Figure 1). She was referred to the endocrine department for further evaluation. The results of laboratory tests, including those for measuring plasma levels of adrenocorticotrophic hormone, cortisol, aldosterone, epinephrine, norepinephrine, metanephrine, dehydroepiandrosterone sulfate as well as urine levels of epinephrine, norepinephrine, metanephrine, vanillylmandelic acid and homovanillic acid were within normal limits. Plasma renin activity was also normal. The levels of serum markers for carcinoma were within normal limits Radiolabeled Metaiodobenzylguanidine Single Photon Emission Computed Tomography (I-123 MIBG SPECT) presented huge mass with mild peripheral uptake of left adrenal gland (Figure 2). Finally, Positron Emission Tomography (PET) was done for evaluation of distant metastasis. PET showed the accumulation of Fluorodeoxyglucose (FDG) in the left adrenal tumor with [maximum standardized uptake value (SUVmax: 7.4)] and lymph nodes in para-aortic area (SUVmax: 2.4) (Figure 3). We initially expected adrenocortical carcinoma with lymph nodes metastasis.

Radical adrenalectomy with para-aortic lymph nodes dissection was performed with patient's consensus. Grossly, a well-demarcated and solid mass measuring 16.8 cm × 11.8 cm × 11.5 cm was seen in the left adrenal gland (Figure 4).

OPEN ACCESS

Correspondence:
Jung Bum Choi, Department of Surgery, Pusan National University Hospital, Busan, South Korea, Tel: +82-10-6309-4480;
E-mail: kamata82@hanmail.net

Received Date: *26 Oct 2022*
Accepted Date: *17 Nov 2022*
Published Date: *21 Nov 2022*

Citation:
Han JH, Lee BC, Park YM, Jung HJ, Kim D, Choi JB. Huge Adrenal Schwannoma: A Rare Type of Adrenal Tumor, Misconceived for Adrenocortical Carcinoma. Ann Surg Case Rep. 2022; 5(2): 1064.

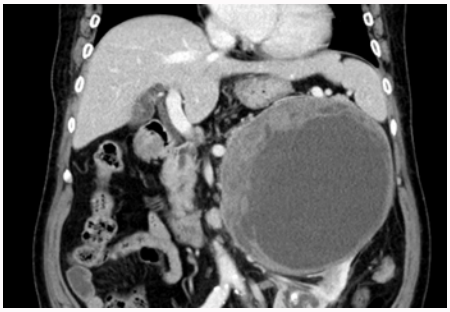

Figure 1: APCT showed 16 cm sized heterogeneous tumor in left adrenal gland.

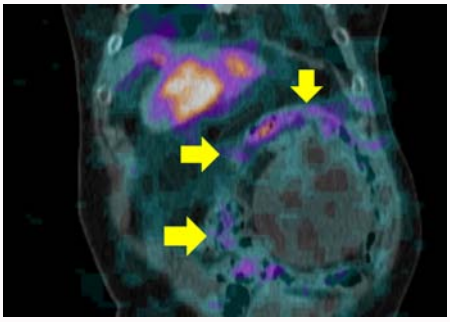

Figure 2: I-123 MIBG SPECT showed huge mass with mild peripheral uptake of left adrenal gland.

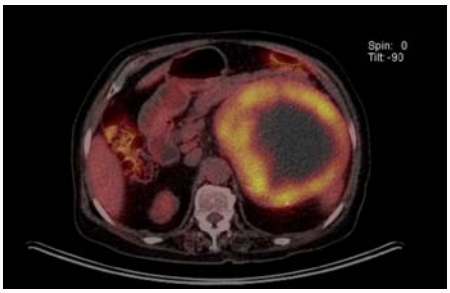

Figure 3: PET showed the accumulation of FDG in the left adrenal tumor and lymph nodes in para-aortic area.

The final pathologic result of tumor at left adrenal gland was 16.8 cm sized cellular schwannoma. Histologically, the mass mostly consisted of an interlacing arrangement of spindle cells (Figure 5A). Immunohistochemical analysis of tumor revealed that the tumor cells were diffusely strong positive for S100 protein (Figure 5B) with weakly positive for Smooth Muscle Actin (SMA), EMA and negative for PANCK, desmin, Human Melanoma Black-45 (HMB45), p53 and CD34. The Ki-67 labeling index was 2%.

Postoperative course was uneventful and patient was discharged on the 15th postoperative day. He is free of disease 5-month post-surgery.

Discussion

In our case, we treated the patient with 16 cm huge left adrenal schwannoma. Schwannomas arise from the Schwan cells surrounding the peripheral nerves and mostly they originate from the nerves of the head and neck or the peripheral extremities [2,5]. They are rarely found in the retroperitoneum; less than 3% of benign schwannomas are seen in a retroperitoneal site [6]. Also, only approximately 6% of

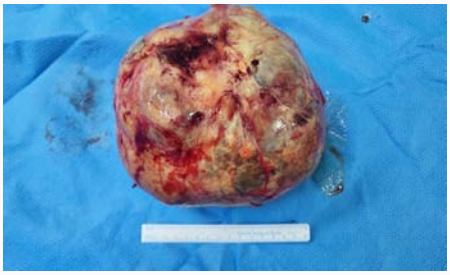

Figure 4: A well-demarcated and solid mass measuring 16.8 cm × 11.8 cm × 11.5 cm.

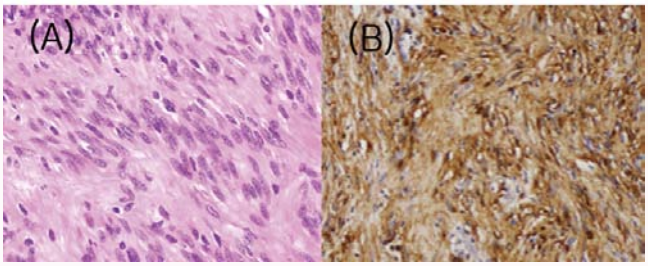

Figure 5: Histologic and immunohistochemical features of tumor. (A) Spindle cells arranged in fascicles with intervening stroma. (B) Cells showed strong S100 immunoreactivity.

primary retroperitoneal tumors are found to be schwannomas [7]. With only about 80 cases reported in worldwide, adrenal schwannoma is a very rare disease [3,8]. A recent study reported 33 cases of adrenal schwannomas with a median patient age of 49 years old, median tumor size of 5.5 cm and a predominance in women [9]. Some studies reported that schwannoma is more common in old women, with approximate male to female ratio of 2:3 [4,10], which is consistent report with our case. The risk factors of adrenal schwannoma were not known well because of its rarity, but some studies reported that adrenal schwannoma is associated with neurofibromatosis and prior ionizing radiation is only known risk factor [4]. In our case, the patient had no such medical history.

Adrenal schwannoma is usually asymptomatic and is found incidentally with large size using imaging study [11]. Because of slow-glowing nature of the tumor, patients are often asymptomatic until it grows large enough to cause mass effect including abdominal pain or discomfort [12]. For this reason, adrenal schwannoma is mostly diagnosed in a large size and it is very rare to be diagnosed in a small size with any symptoms. In our case, the patient was asymptomatic for a long time and complained of abdominal pain and discomfort as its size grew to 16 cm.

Due to the degeneration, cystic change, hemorrhage and calcification within adrenal schwannoma, it often appears heterogeneous on delayed-phase CT and Magnetic Resonance Imaging (MRI). For this reason, adrenal schwannoma is often misdiagnosed for the tumors of the retroperitoneum such as adrenocortical carcinoma, pheochromocytoma, ganglioneuroma, neuroblastoma and adrenal metastatic lesion. In our case, adrenocortical carcinoma was strongly suspected due to suspicious para-aortic lymph nodes seen on PET-CT. Shen et al. suggested that schwannoma can be ruled out through the arterial phase of CT scan showing necrosis and minimal degree of tumor enhancement [13]. But because schwannomas have non-specific or uncommon findings

on imaging studies, the definitive diagnosis must be determined through histologic and immunohistochemical methods. The histology of schwannoma shows spindle-shaped cells having elongated to wavy nuclei with alternating hypercellular and hypocellular regions called Antoni A and Antoni B respectively [14] and immunohistochemically, schwannomas have positive S-100 antibodies [5]. These findings were consistent with our case.

Adrenal schwannoma is usually diagnosed in very large size. For this reason, Adrenal schwannoma sometimes cause significant displacement of nearby structures. Petrucciani et al. reported that adrenal schwannoma can present in close to the great vessels and with several feeding vessels draining from the tumor into the vena cava [15]. Also, some studies reported that tumor can displace the recto-sigmoid colon, ureter, uterus, and psoas muscle [11,16]. In our case, the tumor presented many and large feeding arteries arising from aorta. Due to too many feeding arteries, there was a large amount of bleeding on para-aortic area after tumor removal and meticulous hemostasis was required.

Adrenal schwannoma are typically benign tumors and has a good prognosis with complete surgical resection. Zhou et al. concluded that the patient had favorable survival with no evidence of recurrences after follow-up from 7 to 115 months [3]. Our patient has a good outcome with no specific complication after surgery and is free of disease 5-month post-surgery.

Conclusion

We report a case of huge schwannoma arising from the left adrenal gland. Despite adrenal schwannoma is rare and typically benign tumor, since it cannot be distinguished from other malignant diseases on preoperative imaging, it should always be considered in any adrenal tumors.

References

1. Cury J, Coelho RF, Srougi M. Retroperitoneal schwannoma: case series and literature review. Clinics (Sao Paulo, Brazil). 2007;62(3):359-62.
2. Timilsina S, Joshi SP, Sharma S, Kharel S, Karki S, Tiwari SB, et al. Adrenal schwannoma: A case report of an unusual incidentaloma. Int J Surg Case Rep. 2021;83:106018.
3. Zhou J, Zhang D, Wang G, Li W, Xu J, Ma Y, et al. Primary adrenal microcystic/reticular schwannoma: clinicopathological and immunohistochemical studies of an extremely rare case. Int J Clin Exp Pathol. 2015;8(5):5808-11.
4. Wollin DA, Sivarajan G, Shukla P, Melamed J, Huang WC, Lepor H. Juxta-adrenal Ancient Schwannoma: A Rare Retroperitoneal Tumor. Rev Urol. 2015;17(2):97-101.
5. Shabana W, Raslan W, Hassan S, Al-Tartir T. Schwannoma masquerading as adrenocortical tumor: A case report and review of literature. Urol Case Rep. 2019;26:100926.
6. Schindler OS, Dixon JH, Case P. Retroperitoneal giant schwannomas: Report on two cases and review of the literature. J Orthop Surg (Hong Kong). 2002;10(1):77-84.
7. Rha SE, Byun JY, Jung SE, Chun HJ, Lee HG, Lee JM. Neurogenic tumors in the abdomen: Tumor types and imaging characteristics. Radiographics. 2003;23(1):29-43.
8. Yorita K, Naroda T, Tamura M, Ito S, Nakatani K. Adrenal Schwannoma in an elderly man: A case report and literature review. Intern Med (Tokyo, Japan). 2022;61(1):65-9.
9. Mohiuddin Y, Gilliland MG. Adrenal schwannoma: A rare type of adrenal incidentaloma. Arch Pathol Lab Med. 2013;137(7):1009-14.
10. Ohigashi T, Nonaka S, Nakanoma T, Ueno M, Deguchi N. Laparoscopic treatment of retroperitoneal benign schwannoma. Int J Urol. 1999;6(2):100-3.
11. Theodosopoulos T, Stafyla VK, Tsiantoula P, Yiallourou A, Marinis A, Kondi-Pafitis A, et al. Special problems encountering surgical management of large retroperitoneal schwannomas. World J Surg Oncol. 2008;6:107.
12. Carpenter WB, Kernohan JW. Retroperitoneal ganglioneuromas and neurofibromas. A clinicopathological study. Cancer. 1963;16:788-97.
13. Shen Y, Zhong Y, Wang H, Ma L, Wang Y, Zhang K, et al. MR imaging features of benign retroperitoneal paragangliomas and schwannomas. BMC Neurol. 2018;18(1):1.
14. Jakowski JD, Wakely PE, Jr., Jimenez RE. An uncommon type of adrenal incidentaloma: A case report of a schwannoma of the adrenal medulla with cytological, histological, and ultrastructural correlation. Ann Diagn Pathol. 2008;12(5):356-61.
15. Petrucciani N, Sirimarco D, Magistri P, Antolino L, Gasparrini M, Ramacciato G. Retroperitoneal schwannomas: Advantages of laparoscopic resection. Review of the literature and case presentation of a large paracaval benign schwannoma (with video). Asian J Endosc Surg. 2015;8(1):78-82.
16. Chiang ER, Chang MC, Chen TH. Giant retroperitoneal schwannoma from the fifth lumbar nerve root with vertebral body osteolysis: A case report and literature review. Arch Orthop Trauma Surg. 2009;129(4):495-9.