

## **Afghanistan Journal of Basic Medical Sciences**

ajbms@knu.edu.af

Vol. 1, No.2, July 2024, pp. 127-133

# An Asymptomatic Large Adrenal Ganglioneuroma in a 34-Year-Old Male: A **Case Report**

Mojtaba Behroozirad<sup>1</sup>, Ruhollah Taqavi<sup>1</sup>, Morteza Sedaqat<sup>2\*</sup>

- Department of Surgery, School of Medicine, Shahid Beheshti Hospital, Oom University of Medical Sciences, Oom, Iran
- Medical Student, Student Research Committee, Qom University of Medical Sciences, Qom, Iran

#### ARTICLE INFO ABSTRACT

Received: 9 June, 2024

Accepted: 21 July, 2024

**Introduction:** Right-sided infective endocarditis, a rare cardiac illness, accounts for 5-10% of cases. It's caused by damage to the heart's endothelium, inflammatory cytokines, tissue factors, and increased fibronectin expression. Chronic drug use can damage and seed bacteria on a healthy tricuspid valve.

\*Corresponding Author: Morteza Sedaqat Address: Medical Student, Student Research Committee, Qom University of Medical Sciences, Qom, Iran

E-mail address:

sedaqatmorteza1375@gmail.com

Case Report: A 34-year-old male patient was diagnosed with an adrenal mass during routine sonography. Despite being asymptomatic, further investigations were conducted due to the mass's size. Laboratory findings were normal, and an ECG showed no abnormalities. Detailed imaging studies confirmed the presence of a large, homogenous mass measuring 103x77mm in the left adrenal gland. The patient underwent open surgery and the mass was removed without complications. Histopathological examination confirmed the mass as an adrenal ganglioneuroma.

**Discussion:** This case highlights the importance of thorough investigation and management of incidentally discovered adrenal masses, even in asymptomatic patients. It also underscores the significance of diagnostic imaging studies in the diagnosis and surgical planning of adrenal tumors. The patient's progression after surgery was uneventful, demonstrating the effectiveness of surgical intervention in such cases.

Keywords: Endocarditis, Adrenal Ganglioneuroma, Case Report.

To cite this article: Behroozirad M, Taqavi R, Sedaqat M. An Asymptomatic Large Adrenal Ganglioneuroma in a 34-Year-Old Male: A Case Report. Afghanistan Journal of Basic Medical Sciences . 2024 Jul;1(2):127–33. https://doi.org/10.62134/ajbms/v2.i2.khatamuni.12



#### 1. Introduction

Ganglioneuromas (GNs) are uncommon tumors that originate from the neural crest tissue. They are most frequently found in the posterior mediastinum and retroperitoneum. However, they are seldom seen in the adrenal gland (1, 2). These tumors are situated in the retroperitoneal space in 32-52% of cases, the posterior mediastinum in 39-43% of cases, and the cervical region in 8-9% of cases. However, their occurrence in the adrenal gland is infrequent (3). Detecting most adrenal ganglioneuromas (AGNs), which are nonfunctional, can be notably difficult. These tumors are often found unexpectedly during CT scans, MRIs, or ultrasounds that are conducted for various reasons (4). If AGNs are identified as incidental findings in the adrenal gland, then all patients should undergo screening tests before any surgical procedures (5). Laparoscopic adrenalectomy is considered the best treatment for symptomatic AGNs. However, a pathological examination is essential for a definitive diagnosis (6, 7). Here, we aim to present the case of a 34year-old man who was asymptomatic at the time of presentation and was incidentally diagnosed with AGN during an examination.

#### 2. Case Report

A 34-year-old male, with no known medical history, presented to the emergency department carrying a sonography report that suggested a potential adrenal mass. The patient denied experiencing any symptoms such as abdominal pain, flank pain, vomiting,

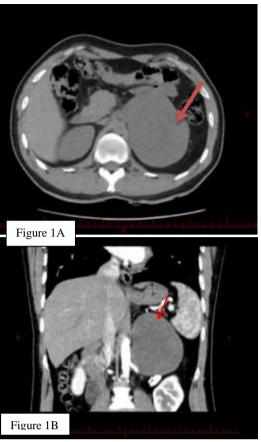
nausea, weight loss, fever, diarrhea, headache, chest pain, or dyspnea.

On admission, the physical examination revealed a blood pressure of 116/84 mmHg, a pulse rate of 72 beats per minute, a respiratory rate of 19 breaths per minute, a temperature of 36.3 °C, and a pulse oximetry measurement of 97% blood oxygen saturation in an ambient room. Jugular venous distension was not detected. A cardiac examination revealed regular S1 and S2 sounds and a soft systolic murmur. Lung auscultations were vesicular. The abdomen did not appear deformed or distended upon examination and was smooth to the touch. There was no tenderness, rebound tenderness, rigidity, or guarding. Therefore. the physical examination was concluded to be entirely normal. Laboratory findings showed a red blood cell count of  $4.86 \times 106 \mu L$  (normal range 4.5-6.1 million/µL for men), a white blood count of 5,600/µL, and a neutrophil percentage of 85% (normal range: 4,500-11,000 per µL). hemoglobin was 14 g/dL (normal range: 13-17 g/dL for men), platelets were 171,000/µL (normal range: 145,000–450,000 per μL), creatinine was 0.9 mg/dL (baseline creatinine was 0.8-1.4 mg/dL), sodium was 136 mmol/L (normal range: 135–145 mEq/L), and potassium was 3.9 mmol/L (normal range: 3.5-5.5 mEq/L).

The electrocardiogram (ECG) was normal. In the sonography performed on the patient about two weeks ago, the radiologist reported a relatively round mass with clear boundaries, hypoechoic compared to the adjacent splenic and renal parenchyma, with approximate dimensions of  $103 \times 75 \times 68$  millimeters. In addition, the mass was accompanied by internal calcified foci that

originated from the left adrenal gland. Also, in the color Doppler sonography examination performed on the patient, no clear arterial flow was observed.

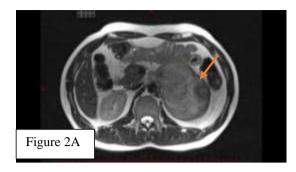
It should be mentioned again that the patient had no symptoms, and the mass was accidentally discovered during a check-up. The radiologist recommended a more detailed examination by performing an abdominal and pelvic CT scan, both with and without contrast. A spiral CT scan of the abdomen and pelvis, taken with and without contrast, demonstrates evidence of a large, well-defined, homogenous density mass lesion in the left adrenal, measuring  $103 \times 77$  mm and containing some calcified flecks, with a compressive effect upon the left renal and left adrenal vein. The contralateral adrenal gland is intact. No prominent perilesional lymphadenopathy is seen (Figures 1A and 1B). Considering all the above findings, the left adrenal tumor mass was the suggested diagnosis for the patient. An abdominal and pelvic MRI was conducted on the patient, which confirmed the findings of the CT scan, and no evidence was presented in favor of metastasis (Figures 2A and 2B). Based on the imaging findings and the reported large size of the mass, the patient underwent open surgery. Following the establishment of sterile conditions, a midline incision was initiated. The fascia and peritoneum were subsequently opened. The initial step involved the mobilization of the splenic flexure of the colon. The spleen was liberated by medially displacing the colon. The posterior peritoneum was then opened, granting access to the retroperitoneal space. A mass measuring 11×9×4 centimeters was carefully dissected from the surrounding structures and ultimately excised (Figures 3A and 3B).

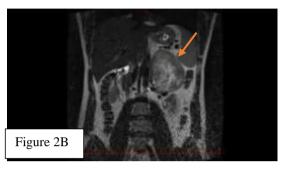


**Fig. 1A.** Axial CT scan showing a large adrenal ganglioneuroma. The tumor, measuring 103 \* 77 millimeters, is visible as a well-defined mass contrasting with the surrounding tissues. **Fig. 1B.** Coronal CT scan of the same patient, further highlighting the size and location of the adrenal ganglioneuroma.

This procedure delineates the open surgical resection of an adrenal incidentaloma. The operation lasted for 2 hours. A biopsy sample was taken from the mass for histopathological examination. The histopathological examination of the sample reported ganglioneuroma. The patient was discharged without any complications three days after the open surgery.

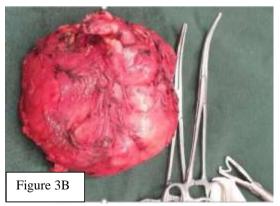
During his follow-up visit, he reported no issues or discomfort post-operation. His recovery was smooth and without incident, indicating a successful outcome from the surgery.





**Fig. 2A.** Axial MRI scan showing a large left adrenal ganglioneuroma. The tumor is visible as a well-defined mass. **Fig. 2B.** Coronal MRI scan of the patient, emphasizing the dimensions and position of the adrenal ganglioneuroma, with no signs of metastasis detected.





**Fig. 3A**: Intraoperative photograph showing the large adrenal ganglioneuroma. **Fig. 3B**: Close-up view of the resected adrenal ganglioneuroma, measuring 11×9×4 centimeters and weighing 340 grams

#### 3. Discussion

Ganglioneuroma (GN) is a type of tumor originating from neural crest cells, encompassing the adrenal glands and sympathetic ganglia. It is comprised of various components, such as Schwann cells, ganglion cells, neurites, and fibrous tissues. This tumor predominantly affects children and young adults, with 60% of cases developing before the age of 20. It has been observed that females are more susceptible to this condition than males. Most GNs are typically found in the thoracic and retroperitoneal regions, whereas the occurrence of GNs in the adrenal glands is uncommon (8). The clinical signs of GNs are often vague, primarily hormone-inactive, associated with their size and location. Although GNs are typically benign, they can become noticeable due to their pressure adjacent structures (9). Adrenal gangliogliomas usually exhibit a gradual growth rate and are often asymptomatic (10). In the case of our patient, the relatively large adrenal ganglioneuroma was also asymptomatic, with no reported symptoms such as flank pain.

Adrenal ganglioneuromas (AGNs) are inactive hormonally typically nonfunctional. However, they can produce catecholamines and their metabolites (11). Before any surgical procedures, all patients with adrenal incidentalomas should undergo hormone screening tests. Furthermore, approximately 4% of adrenal incidentalomas have been detected in crossabdominal sectional imaging (5).Considering the limitations our diagnostic capabilities and the likelihood that the relatively large mass in our patient's case could exert pressure effects on other organs, hormonal testing was not prioritized and was not conducted.

The diagnosis of differential ganglioneuroma includes adenoma. nodular hyperplasia, pheochromocytoma, myelolipoma, hamartoma, granulomatosis, metastatic cancer. and ganglioneuroblastoma. While the imaging features of GN on CT and MRI scans have been thoroughly documented, accurately diagnosing adrenal **GN** through radiological assessments before surgery remains challenging (12). Qing et al. have indicated that the rate of incorrect diagnosis for AGN using CT and MRI before surgery stands at 64.7% (13). AGN is characterized as a well-defined, lobular-shaped tumor with low signal intensity. According to various studies, calcification within the tumor has been observed in anywhere from 0% to 29% of cases (13, 14). In MRI scans, AGN typically presents with a uniformly low or medium signal intensity in T1weighted images and a slightly elevated, varied signal intensity in T2-weighted images (15). In our patient's case, given the homogeneous mass and well-defined nature, the likelihood of malignancy was less probable.

Ganglioneuroma (GN) is a benign tumor that originates from the neural crest cells of the adrenal medulla, or sympathetic ganglia. When observed macroscopically, an adrenal GN presents as a solid, uniform, grayish-white mass that is encapsulated and has a firm texture. The histopathological examination reveals the presence of mature ganglion cells and Schwann cells within fibrous connective tissue (15). Ultimately, based on the biopsy sample taken from the mass during the operation, the pathologist

reported ganglioneuroma as the definitive diagnosis.

Recent research suggests that nonfunctioning adrenal incidentalomas that are larger than 6 cm, or those that show signs of potential malignancy in imaging tests, should be managed with adrenalectomy (13, 14, 16-18). There are no medical therapies available for these types of tumors. Despite being benign, GN can exhibit aggressive growth. Patients who undergo surgical intervention for a benign neurogenic tumor generally have a very favorable outcome. Papavramidis et al. have stated that AGN should be removed via adrenalectomy, while retroperitoneal GN can be excised without the need for adrenalectomy (19). The utilization of laparoscopic adrenalectomy in treating patients with large adrenal masses or possible cancer is still a matter of debate. However, the shorter hospitalization period and lower complication rates have led to laparoscopic adrenalectomy being the preferred surgical method for removing most small-sized adrenal lesions (those less than 6 cm in size) (1, 16, 18). Considering that our patient's adrenal mass was relatively large (more than 6 centimeters), it was not possible to remove the mass laparoscopically. We removed the mass using open surgery. The prognosis for an adrenal ganglioneuroma after surgical removal is generally favorable, and it usually does not require any further treatment. However, there have been a few instances where the condition has recurred.

#### Conclusion

This case report presents a 34-year-old male patient, asymptomatic but diagnosed with adrenal ganglioneuroma (AGN), a rare

benign tumor. The tumor was found incidentally and surgically removed due to its large size. The case emphasizes the importance of considering AGN in the differential diagnosis of adrenal incidentalomas and the need for surgical intervention for larger tumors, even if they are asymptomatic. It also highlights the lack of clear guidelines for managing asymptomatic AGN, suggesting further research in this area for improved patient outcomes and efficient healthcare resource This case contributes to AGN knowledge and identifies areas for potential research.

#### Conflict of Interest

The authors have no conflicts of interest relevant to this article.

#### Authors' Contributions

All of the authors contributed equally to the present study. All authors read and approved the final manuscript.

#### Ethics approval

Informed consent was obtained from the patient to access medical records.

### **Funding**

This research did not receive any grants from funding agencies in the public, commercial, or non-profit sectors.

#### References

 Geoerger B, Hero B, Harms D, Grebe J, Scheidhauer K, and Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. Cancer: Interdisciplinary International Journal

- of the American Cancer Society. 2001;91(10):1905–13.
- 2. Erem C, Ucuncu O, Nuhoglu I, Cinel A, Cobanoglu U, Demirel A, et al. Adrenal ganglioneuroma: report of a new case. Endocrine. 2009;35:293-6.
- 3. Rha SE, Byun JY, Jung SE, Chun HJ, Lee HG, and Lee JM. Neurogenic tumors in the abdomen: tumor types and imaging characteristics. Radiographics. 2003;23(1):29-43.
- Carsote M., Ghemigian A., Terzea D., Gheorghisan-Galateanu AA, and Valea A. Cystic adrenal lesions: focus on the pediatric population (a review). Clujul Medical. 2017;90(1):5.
- 5. Santosh K, Das MK, Ayyanar P, Mandal S, Nayak P, and Kumar S. Laparoscopic excision of a large adrenal ganglioneuroma masquerading as pheochromocytoma: A case report and review of the literature Urology Case Reports. 2020;33:101276.
- Program NCD. NIH state-of-thescience statement on management of the clinically inapparent adrenal mass ("incidentaloma"): National Institutes of Health, Office of the Director; 2002.
- 7. Rao RN, Singla N, and Yadav K. Composite pheochromocytomaganglioneuroma of the adrenal gland:

  A case report with immunohistochemical study. Urology Annals. 2013;5(2):115–8.
- 8. Sucandy I, Akmal YM, and Sheldon DG. Ganglioneuroma of the adrenal gland and retroperitoneum: A case report. North American journal of medical sciences. 2011;3(7):336.
- 9. Ann Hayes F, Green AA, Rao BN. Clinical manifestations of ganglioneuroma. Cancer. 1989;63(6):1211-4.
- Kacagan C, Basaran E, Erdem H, Tekin A, Kayikci A, and Cam K. Case report: Large adrenal ganglioneuroma.

- International Journal of Surgery Case Reports. 2014;5(5):253-5.
- 11. Diab DL, Faiman C, Siperstein AE, Zhou M, and Zimmerman RS. Virilizing adrenal ganglioneuroma in a woman with subclinical Cushing syndrome. Endocrine Practice. 2008;14(5):584–7.
- 12. Adas M., Koc B., Adas G., Ozulker F., and Aydin T. Ganglioneuroma presenting as an adrenal incidentaloma: a case report. Journal of Medical Case Reports, 2014, 8:1–5.
- 13. Qing Y, Bin X, Jian W, Li G, Linhui W, Bing L, et al. Adrenal ganglioneuromas: a 10-year experience in a Chinese population. Surgery. 2010;147(6):854-60.
- 14. Maweja S., Materne R., Detrembleur N., de Levai L., Defechereux T., Meurisse M., and Hamoir E. Adrenal ganglioneuroma. neoplasia to exclude in patients with adrenal incidentaloma. Acta Chirurgica Belgica. 2007;107(6):670-4.
- 15. Sasaki S, Yasuda T, Kaneto H, Otsuki M, Tabuchi Y, Fujita Y, et al. Large adrenal ganglioneuroma. Internal Medicine. 2012;51(17):2365–70.
- 16. Yamaguchi K, Hara I, Takeda M, Tanaka K, Yamada Y, Fujisawa M, and Kawabata G. There are two cases of ganglioneuroma. Urology. 2006;67(3):622. e1–e4.
- 17. Grumbach MM, Biller BM, Braunstein GD, Campbell KK, Carney JA, Godley PA, et al. Management of the clinically inapparent adrenal mass (incidentaloma). Annals of internal medicine, 2003;138(5):424–9.
- 18. Joshi VV. Peripheral neuroblastic tumors: pathologic classification based on recommendations of the International Neuroblastoma Pathology Committee (Modification of the Shimada classification). Pediatric

- and Developmental Pathology. 2000;3(2):184–99.
- 19. Papavramidis TS, Michalopoulos N, Georgia K, Kesisoglou I, Valentini T, Georgia R, Papavramidis ST. Retroperitoneal ganglioneuroma in an adult patient: a case report and literature review of the last decade. South Med J. 2009;102(10):1065-7.