Urology Case Reports 55 (2024) 102759

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Retroperitoneal adrenal neuroblastoma with bone marrow metastatic activity in a young adult

Norman Alejandro Rendón Mejía [a,*], Iram Ivey Ávila Quiñones [b], Jesús Alfonso Preciado Hernández [c], Karla Dillery García Castillo [d]

[a] Department of Surgery, Hospital General Salvador Zubirán Anchondo, Universidad Autónoma de Chihuahua, Chihuahua, Mexico
[b] Department of Urology, Hospital Central Universitario Dr. Enrique Grajeda Herrera, Chihuahua, Mexico
[c] Department of Urology, Hospital General Regional Ignacio Zaragoza, Estado de México, Mexico
[d] Department of Urology, Hospital General de Occidente, Guadalajara, Mexico

ARTICLE INFO

Keywords:
Neuroblastoma
Nephrectomy
Round cell neoplasm
Retroperitoneal

ABSTRACT

A 31-year-old male came to emergency department with acute abdominal pain from the right flank, diaphoresis and, palpitations. Contrasted abdominal CT revealed a tumor dependent from right adrenal gland and kidney. A laparoscopic radical nephrectomy was made to resect the tumor and pain relief. Pathological analysis reported poorly differentiated neuroblastoma from the right adrenal gland without involvement to kidney and ureter. The patient was sent to oncology clinic to continue with chemotherapy treatment. This is the third case of adult neuroblastoma reported in South America. Adult neuroblastoma is an uncommon cause of malignant neoplasm with an exceptional incidence reported.

1. Introduction

Neuroblastoma (NB) is the most common solid extracranial tumor in children, and more than 95 % of cases are detected at the age of 10.[1] There have been fewer than 100 cases of neuroblastoma in adults reported in medical literature. The incidence of NB in adults older than 30 is 0.2 cases per million inhabitants.[2] The NB originates from the neural crest cells of the sympathetic nervous system. In most cases, it develops from the adrenal gland and mediastinum.[3] NB metastasizes across the hematopoietic system; the most common places of metastasis are the bone, bone marrow, lung, pleura, brain, breast, liver, and lymphatic nodes.[4] In adults, NB occurs more frequently as an advanced metastatic disease. The prognosis of patients with NB depends on age at the moment of diagnosis, stage of disease, localization of the tumor, molecular markers, histology, and chromosomal aberrations.[5]

We present a case report of a young male adult who presented abdominal pain and was diagnosed with a retroperitoneal neuroblastoma that originated from the right adrenal gland.

2. Case presentation

A 34-year-old male adult with no clinical background presents mild to severe abdominal pain on the right flank following irradiation to the lumbar area. The symptoms presented were nausea, vomiting with gastrointestinal content, diaphoresis, dyspnea, and palpitations. He decides to go to the nearest emergency service for a medical evaluation. In the emergency department, medical personnel request laboratory testing and imaging studies. A diagnosis of acute abdominal pain was made. The laboratory results showed values within normal ranges. A retroperitoneal tumor measuring 77 × 44 × 102 mm with affection for the right adrenal gland, renal artery and vein, and inferior vena cava was seen in the abdominal computer tomography. The tumor was located on the superior pole of the right kidney (Fig. 1) and (Fig. 2).

A urology evaluation was performed due to the suspicion of a right adrenal tumor in the retroperitoneum. The urology department opted to admit the patient and perform laparoscopic surgical exploration and resection of the retroperitoneal tumor using minimally invasive method. The urologist surgeon chose during surgery to perform a radical right nephrectomy and resection of the adrenal tumor due to the tumor's

* Corresponding author. Department of Surgery, Hospital General Salvador Zubirán Anchondo, Avenida Prolongación Teófilo Borunda, #510, Chihuahua, 31200, Mexico.
E-mail addresses: norman-rendon@hotmail.com (N.A. Rendón Mejía), drivey_avila@hotmail.com (I.I. Ávila Quiñones), japhdez@gmail.com (J.A. Preciado Hernández), karladillery@gmail.com (K.D. García Castillo).

https://doi.org/10.1016/j.eucr.2024.102759
Received 15 April 2024; Received in revised form 17 May 2024; Accepted 21 May 2024
Available online 21 May 2024

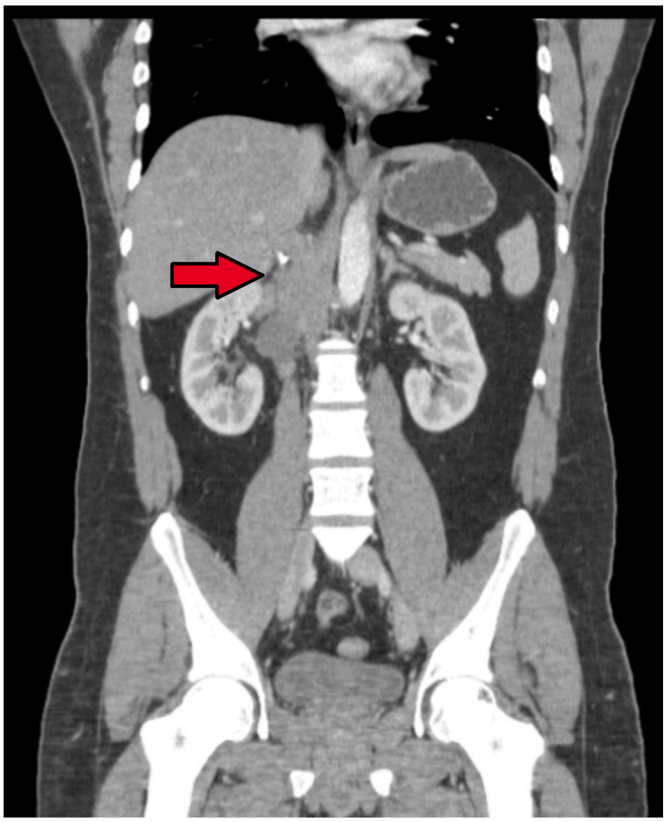

Fig. 1. Abdominal contrasted computed tomography showing a tumor on the superior pole of the right kidney over the right adrenal gland.

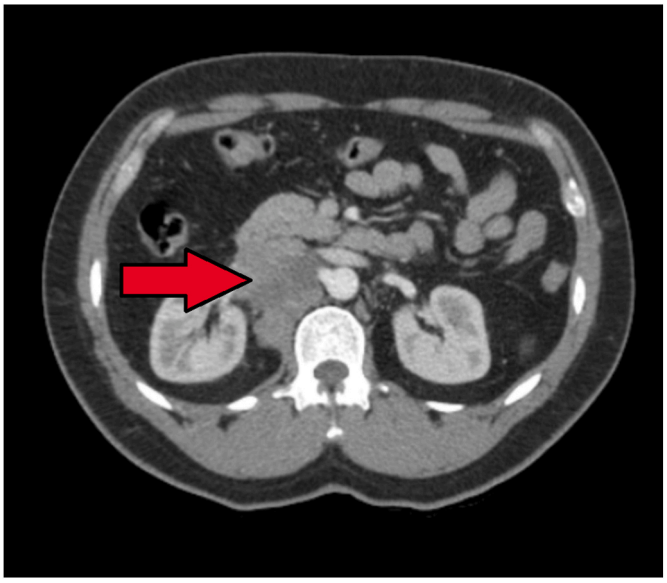

Fig. 2. Abdominal contrasted computed tomography showing an adrenal tumor infiltrating renal parenchyma and renal vessels.

arterial, lymphatic and renal vein involvement. The surgical block obtained from right nephrectomy, measured 16.2 × 6.1 × 5.7 cm; the surgical piece was collected and delivered to the surgical pathology department for histological evaluation and tumor identification (Fig. 3) and (Fig. 4).

Small, round, blue cells were found with direct optical microscopy using hematoxylin-eosin strain; the pathology piece measured 6.5 × 5.2 × 3.1 cm, the adventitia, periurethral fat, and the exterior muscle layer of the ureter and kidney had no evidence lymphatic and vascular infiltration (Fig. 5 a-b). The biopsy's immunohistochemical panel revealed the expression of GATA3, FL1, synaptophysin, neuron-specific enolase, INSM1, and CD56. The right adrenal gland biopsy was found to have a poorly differentiated neuroblastoma.

A cranial, neck, thorax, abdomen and pelvis tomography was requested, in the imageology findings revealed an atypical choroidal plexus with discrete contrast enhancement and internal calcifications, as

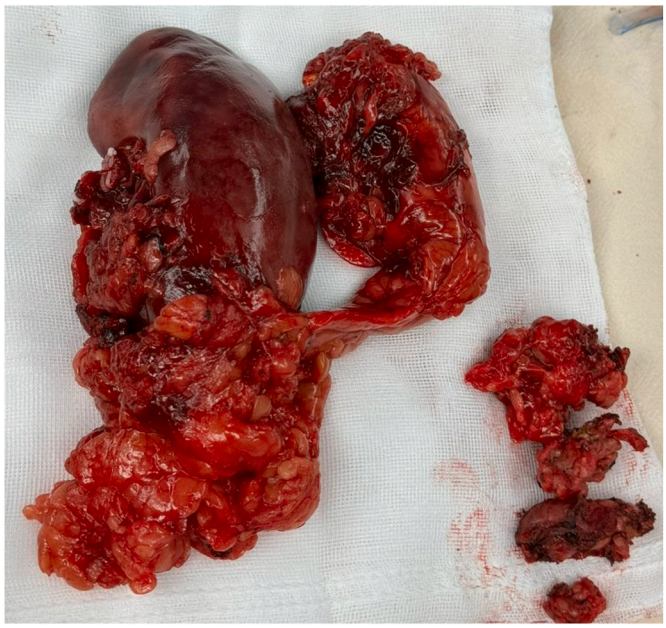

Fig. 3. Macroscopic surgical block from a right laparoscopic nephrectomy.

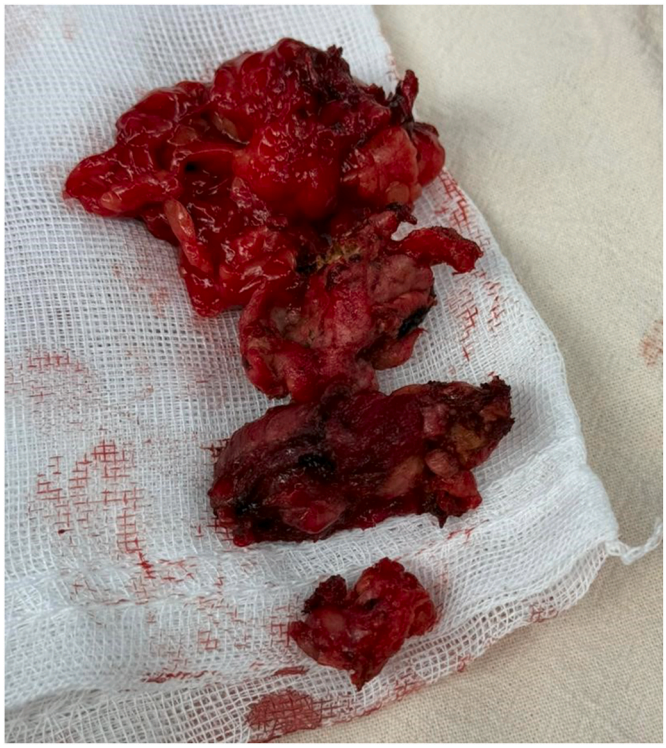

Fig. 4. Macroscopic aspect of a right adrenal tumor with 6.5 × 5.2 × 3.1 cm dimensions.

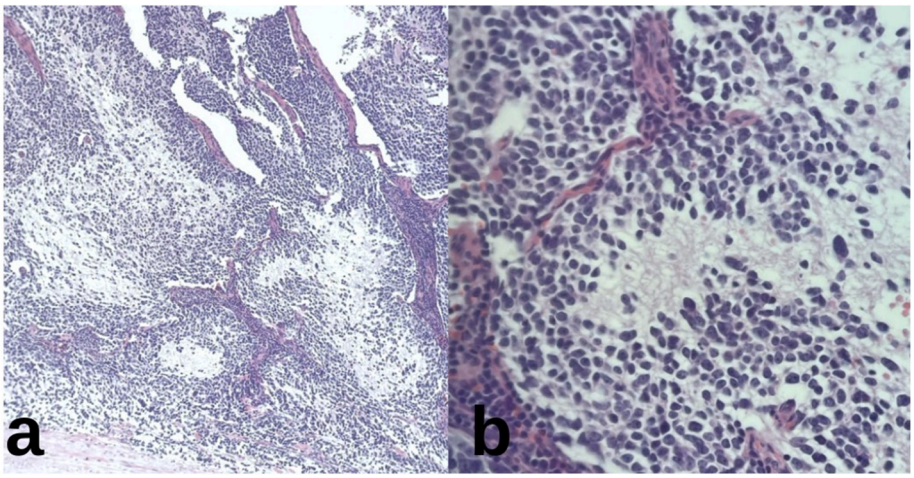

Fig. 5. a–b:
a. H-E strain of the adrenal tumor cells with the appearance of salt and pepper. b. H-E strain of the tumor showing round and blue neoplastic cells without infiltration into renal parenchyma. H-E= Hematoxylin-eosin.

well as sclerotic lesions in the trabecular bone compromising the head, neck, and trochanter on the right femur. Lytic lesions were discovered in the fourth left and ninth right costal arches. In mediastinum, lymphatic nodes had a reactive shape on bilateral inferior paratracheal levels; the right renal fossa had a solid nodular lesion of 28 × 33 × 32 mm with regular and well-defined borders; and the retroperitoneum had an increased size and number of lymphatic nodes.

An oncology consultation was scheduled, and a bone gammagraphy was performed to identify the bone lesions described in prior imaging investigations. The bone grammagraphy scans revealed an aberrant distribution of contrast in the costal arcs, iliac crests, pelvis, and right femur. A hematology medical evaluation was performed, and a bone marrow biopsy from both ileal crests was obtained. The bone biopsy revealed metastasis of a malignant tumor with round and oval cells, nucleomegaly, oval nuclei, and scant cytoplasm.

During his hospital stay, the patient reported being in good clinical and physical condition, with low to light abdominal pain near the surgical wounds. Tolerating enteral feeding. The urologist and oncology departments agreed to continue his treatment at the oncology clinic by initiating chemotherapy to treat the metastatic activity found in bone and bone marrow.

3. Discussion

NB accounts for 7 % of malignant tumors in children, with one instance per 100,000 inhabitants; in adults, it is rare, with just one case per 10 million inhabitants.[6] A retrospective cohort study from 1980 to 2009 comprised 15 adult patients with neuroblastoma, with a median age of 23 and a range of 19–33.[7] Another study anticipated a very low prevalence among adults, with approximately 0.3 cases per million inhabitants per year.[8]

As a small round blue cell tumor, neuroblastoma is generated by abnormalities in the gene control matrix during the neural crest's embryonic development. This leads to imbalanced proliferation and changes in the neuroblastoma's differentiation. NB normally consists round, small, homogeneous cells with sparse cytoplasm and hyperchromatic, dense nuclei..[9] The macroscopic features of neuroblastomas often consist of hard, bland, and nodules. They have a white or gray-pink tint with hemorrhagic regions on their surface. The tumor's dimensions can range from 5.5 to 15 cm, with a median of 8.7 cm. The chromatin with the ¨salt and pepper¨ picture and the nuclei from tumoral cells' varying shapes are the two histopathological features of NB.[10]

The immunohistochemical panel reveals that the NB reacts to CD56, chromogranin-A, neurofilament, synaptophysin, and neuron-specific enolase.[7] In a case series with adults diagnosed with neuroblastoma, showed certain immune reaction to vimentin, CD56 (100 % of cases), chromogranin A (88 %), synaptophysin (88 %), neurofilament (63 %), and presented negative reaction to desmine, smooth muscle alpha-actine, muscle-specific actine, myogenin, CD45, and CD34. The most important positive and negative molecular markers for diagnosing neuroblastoma are CD56 and CD99, respectively. The phenotype of retroperitoneal neuroblastomas is positive for CD56 and negative for the CD99 marker.[10]

Clinical manifestations reported in a case series of patients diagnosed with neuroblastoma: adult patients presented more frequently pain in 7 cases; 3 patients reported the pain occurred on the abdomen, 2 in the dorsum, one in the pelvic region, and one in the inguinal region; and also, 2 patients presented a palpable abdominal mass.[7] In their study, Hasegawa et al. reported 5 patients with retroperitoneal NB with abdominal pain in 2 patients, sensation of abdominal mass, systemic pain, loss of appetite, and constipation.[10] The median time to the presence of symptoms at the moment of the diagnosis was 12 months, and the principal sign in patients with NB was abdominal pain, which was reported in 11 patients.[11]

Contrasted axial tomography is currently used for estimating tumor size, density, local invasion, distant metastases, and surgical planning.[7] Adults are more prone to developing NB as an advanced metastatic disease.[5] In clinical stage IV of NB, metastases were discovered in 10 patients' distant lymphatic nodes, 7 in bone marrow, 7 in bone, and 2 in the liver.[11] In a cohort study of adult neuroblastoma patients, a significant statistical relationship was discovered between tumor anatomic site and genetic and clinical features.[12] In a series of cases, the Evan criteria for NB was used, and 5 patients were classified as stage I, 5 as stage III, and 5 as stage IV. The metastatic sites in the five patients with stage IV disease included bone, bone marrow, lymphatic nodes, liver, and lung. All patients with retroperitoneal neuroblastoma had disease progression and recurrence; five died within three weeks to a year of diagnosis.[11]

The treatment for NB consists of a total surgical resection of the tumor, followed by chemotherapy and radiation. For disseminated and recurrent diseases, chemotherapy should be combined with radiation therapy.[5] Esiashvii et al. found that patients over the age of 18 who were diagnosed with neuroblastoma and treated with surgical resection, radiation therapy, and chemotherapy had low survival rates, with 75 % survival at 2 years, 44 % at 5 years, 37 % at 10 years, and 10 % at 20 years after diagnosis. In adults with NB, chemotherapy treatment only has a palliative effect on the disease, and an aggressive treatment with higher chemotherapy doses combined with surgery and immunological

therapy may result in better survival rates.[8] Adults with NB benefit from a combination of surgical resection, increased chemotherapy doses, and radiation therapy. Surgical treatment must be focused on achieving complete tumor resection while avoiding tumor dissemination or seeding during resection. The most commonly used chemotherapy agents are cyclophosphamide, iphosphamide, vincristine, adriamycin, cisplatin, carboplatin, and etoposide.[4]

There are currently no adult guidelines for the treatment of NB. Nuclear imaging and tumoral markers must be obtained in patients with neuroblastoma as a follow-up or if residual disease is suspected after treatment.[4,5]

4. Conclusion

Great efforts are required to identify and diagnose adult patients with retroperitoneal neuroblastoma. This rare presentation of the disease in this range of age is exceptional, which makes it more difficult to make a diagnosis. Physicians need to make an appropriate diagnosis with an extensive interrogation and physical exploration of the adult patient with abdominal pain and sensation of abdominal mass in the emergency department and make a complete laboratory and imaging workup. Recompilation of cases are needed to improve the survival of adults diagnosed with neuroblastoma, modify the course of the disease, and have a better outcome.

Statement of ethics

Institutional review board approval is not required as it is a case report. Patient consent was obtained from the patient itself.

Funding

None.

Disclosure

The authors declare no conflicts of interest.

CRediT authorship contribution statement

Norman Alejandro Rendón Mejía: Writing – original draft, Investigation, Formal analysis. **Iram Ivey Ávila Quiñones:** Writing – review & editing, Resources, Investigation. **Jesús Alfonso Preciado Hernández:** Investigation, Conceptualization. **Karla Dillery García Castillo:** Methodology, Data curation.

Acknowledgments

None.

References

1. Kushner BH, Kramer K, LaQuaglia MP, Modak S, Cheung N-KV. Neuroblastoma in adolescents and adults: the Memorial Sloan-Kettering experience. *Med Pediatr Oncol.* 2003;41:508–515. https://doi.org/10.1002/mpo.10273.
2. Zhang H, Feng Z. Adrenal neuroblastoma in an elderly adult: a case report and review of the literature. *J Med Case Rep.* 2019;13:284. https://doi.org/10.1186/s13256-019-2204-7.
3. Adrenal neuroblastoma J. *Am J Obstet Gynecol.* 2021;225:B5–B6. https://doi.org/10.1016/j.ajog.2021.06.037.
4. Gupta P, Maiti A, Aich RK, Deb AR. Adrenal neuroblastoma in an adult. *J Cancer Res Therapeut.* 2013;9:96–98. https://doi.org/10.4103/0973-1482.110389.
5. Selcukbiricik F, Tural D, Esatoglu N, Kocak S, Mandel NM. A very rare adult case with neuroblastoma. *Case Rep Oncol.* 2011;4:481–486. https://doi.org/10.1159/000332761.
6. Conter HJ, Gopalakrishnan V, Ravi V, Ater JL, Patel S, Araujo DM. Adult versus pediatric neuroblastoma: the M.d. anderson Cancer Center experience. *Sarcoma.* 2014;2014, 375151. https://doi.org/10.1155/2014/375151.
7. Jrebi NY, Iqbal CW, Joliat G-R, Sebo TJ, Farley DR. Review of our experience with neuroblastoma and ganglioneuroblastoma in adults. *World J Surg.* 2014;38:2871–2874. https://doi.org/10.1007/s00268-014-2682-0.
8. Esiashvili N, Goodman M, Ward K, Marcus Jr RB, Johnstone PAS. Neuroblastoma in adults: incidence and survival analysis based on SEER data. *Pediatr Blood Cancer.* 2007;49:41–46. https://doi.org/10.1002/pbc.20859.
9. Park JR, Eggert A, Caron H. Neuroblastoma: biology, prognosis, and treatment. *Hematol Oncol Clin N Am.* 2010;24:65–86. https://doi.org/10.1016/j.hoc.2009.11.011.
10. Hasegawa T, Hirose T, Ayala AG, et al. Adult neuroblastoma of the retroperitoneum and abdomen: clinicopathologic distinction from primitive neuroectodermal tumor. *Am J Surg Pathol.* 2001;25:918–924.
11. Podda MG, Luksch R, Polastri D, et al. Neuroblastoma in patients over 12 years old: a 20-year experience at the istituto nazionale tumori of milan. *Tumori.* 2010;96:684–689. https://doi.org/10.1177/030089161009600507.
12. Salim A, Raitio A, Pizer B, Mullassery D, Losty PD. Neuroblastoma: the association of anatomical tumour site, molecular biology and patient outcomes. *ANZ J Surg.* 2021;91:1000–1004. https://doi.org/10.1111/ans.16595.

Abbreviations:

NB: Neuroblastoma
H-E:: Hematoxylin-eosin