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# Neuroblastoma Masquerading as Pheochromocytoma

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## Introduction

- Peripheral neuroblastic tumors (PNTs) are a group of tumors arising from sympathetic ganglion cells. It is a malignancy of childhood and rare in adults. The incidence in adults is only 0.12-0.2 cases per million per year.
- In two-thirds of the cases, PNTs arise in the adrenal gland or the retroperitoneal paravertebral ganglia with the adrenal gland being the primary tumor site in 38% of cases <sup>(1)</sup>.
- Metastatic dissemination occurs in about 40% of patients. It usually involves bone and bone marrow.
- Catecholamine secretion is documented in more than 70% of cases and hence the tumors may light up on Iodine 123-metaiodobenzylguanidine (I 123-MIBG) scanning.
- There is no distinctive imaging characteristics for neuroblastoma.
- Diagnosis is often unsuspected until we obtain the final histopathologic examination.

## Case Presentation

- A 37 year old male presented with acute exacerbation of low back pain which started months prior to admission. MRI of the lumbar spine revealed a 3.6 x 3.4 cm lobulated heterogeneous mass-like lesion involving his right adrenal gland.
- He was a non-smoker without any significant past medical or surgical history and was not on any medications. He did not have any Pallor or sweats, pain was dominating the clinical picture.

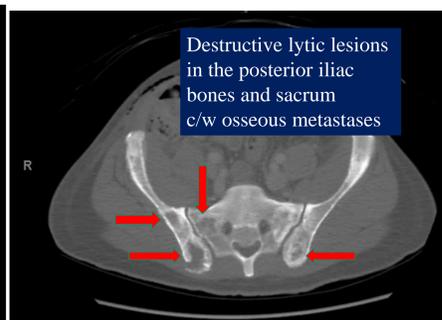


Figure 2 CT abdomen and pelvis on admission

Figure 1: Axial T1 MRI of the lumbar spine 3 months prior to presentation

- Physical examination:** He was normotensive and appeared to be in moderate distress due to severe back pain. He had tenderness over the right sacroiliac joint.
- Laboratory evaluation:** Normal renal and liver functions.

	Value	Reference range
Serum Metanephrines	<57	< 57 pg/mL
Serum Normetanephrines	501	< 148 pg/mL
Serum Total Metanephrines	501	< 205 pg/mL
Chromogranin A	1379	0-95 pg/mL
DHEAS	1064	700-5690 ng/mL
17-hydroxyprogesterone	45	32-332 ng/dL
Androstendione	48	40-190 ng/dL
ACTH	41	< 46 pg/mL

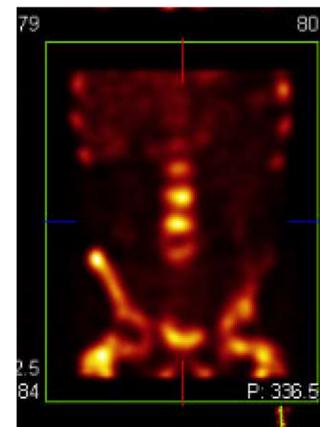
## Diagnosis and Treatment

- Imaging:**
- CT of his abdomen and pelvis with contrast showed a lobular heterogeneous mass involving the right adrenal gland with an increased size of 4.9 x 4.3 cm measuring 55 Hounsfield Units (HU).



Figure 3: CT abdomen and pelvis with contrast

- Histopathology:**
- Right iliac core needle biopsy** was done to evaluate his diffuse osseous metastasis which showed crush artifact, with positive chromogranin and synaptophysin staining.
- Right posterior pelvic crest lesion excision biopsy** showed primitive small round cell neoplasm with neuroendocrine features, favoring an adult type neuroblastoma confirmed by immunohistochemical staining
- N-myc: negative.**
- Immunohistochemical staining** showed positivity for synaptophysin, CD56, chromogranin and focal nonspecific nuclear reactivity for neurofilament SM 131.
- Treatment:**
- We started doxazosin 1 mg daily. The patient subsequently received chemotherapy and radiation therapy then underwent debulking surgery which revealed a 6 cm neuroblastoma with extraadrenal extension.
- Post surgery:**
- (I-123 MIBG):** Extensive diffuse osseous metastatic disease.
- There is no evidence of extraosseous metastatic disease.



## Follow up

- Bone marrow (BM) involvement was noted; therefore, the patient underwent bone marrow transplantation.
- He is doing well six months post-transplant.
- Catecholamine levels trended down, follow up BM biopsy is negative.

## Discussion

- PNTs are made up of two components: neuroblastic cells and Schwannian cells.
- The International Neuroblastoma Pathology Classification (INPC)** distinguishes four pathological groups according to the proportion of the two components.

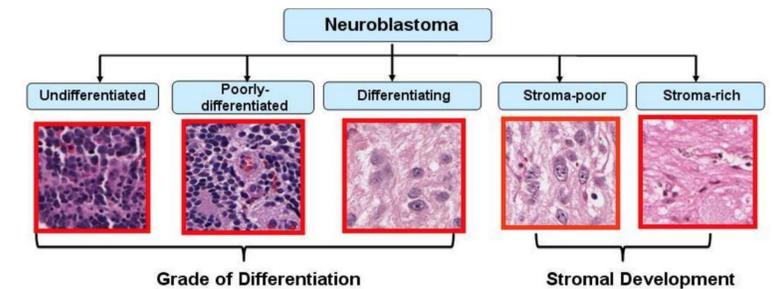


Figure 3: components of neuroblastoma

- It defines prognostic impact in association with age and two mitotic features: Mitosis Karyorrhexis Index (MKI) and grade of differentiation.
- N-myc oncogene:** A member of transcription factor family and is amplified in at least 20% of neuroblastomas in childhood. It promotes tumor growth and tumor progression. It is associated with aggressiveness of disease and is more prevalent in stage 4 disease and therefore is correlated with a poor prognosis. However, it is not as important in the adult population compared to pediatrics <sup>(3)</sup>.
- Surgical resection is the modality of choice for both diagnosis and treatment.
- Other treatment consideration is chemotherapy, radiotherapy and biotherapy.
- Metastatic dissemination occurs in 40% of adults, mainly to the bone and bone marrow. Therefore, adults should have a BM assessment upon diagnosis and throughout treatment <sup>(3)</sup>.
- I 123- MIBG** is performed to evaluate the site of the primary tumor and to detect metastases <sup>(3)</sup>.
- Our case highlights the challenge in diagnosing adrenal mass with high Hounsfield units. Consideration should be given for adrenocortical carcinoma, neuroblastoma and pheochromocytoma.
- Neuroblastoma and pheochromocytoma both secrete catecholamines and therefore, we cannot rely on the levels alone to make the diagnosis. In addition, imaging characteristics are variable. Diagnosis is likely to be made on histopathologic examination. Though rare, one should consider the possibility of neuroblastoma in adults in the differential diagnosis of an adrenal mass. Patients need to be on alpha blockade prior to surgery.
- Catecholamine and chromogranin levels can be used as tumor markers for monitoring.

## References

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