Retroperitoneal localized Neuroblastoma in the Adult: Case Report and Literature Review

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ABSTRACT
Retroperitoneal neuroblastoma is an embryonic tumor of the peripheral sympathetic nervous system. In this study, we report the case of an 18-years-old woman who underwent a subcostal laparotomy for a localized retroperitoneal tumor. The histological findings after resection showed it to be a neuroblastoma. Immunohistochemical staining showed no MYCN amplification. We managed this case using pediatric staging systems and guidelines. She is now 20-years-old, and her CT scans and MIBG scintigraphies are showing neither recurrence nor progression.

Introduction
Neuroblastoma is derived from peripheral sympathetic nervous system. The most frequent localization in children is the adrenal gland, sympathetic ganglia of retroperitoneum or chest [1]. It is the most common solid tumor in children aged less than one year. 90 % of patients are diagnosed before age of five, and only 6.1 % are diagnosed after 20 [2]. In adult, the immunohistochemical study is precious to set the diagnostic given this rare entity [1]. Currently, there are no recommendations for staging or treatment of adult neuroblastoma. The support is mostly based on children guidelines [3]. We report here a case of localized retroperitoneal neuroblastoma in an 18-years-old female.

Patient and observation
An 18-year-old woman with no medical history consulted for right lumbar pain for 6 months. Her physical examination was unremarkable.

Abdominal ultrasound revealed a subhepatic mass. The computer tomography (CT) scan shows a large tissular, hypodense and heterogeneous retroperitoneal mass of 37 H.U. (Hounsfield units), slightly enhanced after contrast injection, measuring 83 mm x 52 mm x 46 mm, pushing forward the inferior vena cava, without evidence of adjacent structures infiltration, evoking a paraganglioma or adrenocortical carcinoma (FIGURE 1). Blood analysis and urine catecholamines were normal. No biopsy was required.

Surgical exploration with right subcostal incision found retrocaval mass isolated from adrenal gland and kidney. The patient underwent an “en bloc”, macroscopically complete, resection of the mass, with laterocaval and retrocaval lymphadenectomy (FIGURE 2). The postoperative course was simple.

Figure 1. Abdominal CT scan showing right retroperitoneal mass.

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Figure 2. Overview of the mass and “en bloc” excised tissue (tumor and lymphadenectomy).

Pathologic examination revealed an undifferentiated proliferation of tumoral cells with nodular architecture, forming Homer-Wright rosettes. The mitosis-karyorrhexis index (MKI) was elevated. The Schwannian stroma was represented in 30 to 40 %. Immunohistochemical (IHC) staining showed tumor cells strongly positive for neurofilament 1 and neuron-specific enolase (NSE), confirming the diagnosis of neuroblastoma, and an absence of MYCN amplification (FIGURE 3).
Neuroblastoma is a rare type of cancer that occurs in children. It typically starts in the nerves near the adrenal glands, which are small glands located on top of each kidney. Neuroblastoma can also develop in other parts of the body, such as the bone marrow, liver, and lungs.

Most cases of neuroblastoma occur before age 5, and the disease is very rare in adults. However, when it does occur in adults, it can be challenging to diagnose and treat.

The symptoms of neuroblastoma in adults can vary widely and may include:

- Abdominal pain or swelling
- Back pain
- Bone pain
- Night sweats
- Unexplained weight loss
- Fatigue

Diagnosis of neuroblastoma in adults can be difficult because the symptoms are not specific to the disease. The diagnosis usually involves a combination of imaging studies, such as CT scans or MRI, and tests to check for the presence of a tumor.

Treatment for neuroblastoma in adults depends on the stage and type of cancer, as well as the overall health of the individual. The treatments may include:

- Surgery: removing the tumor
- Chemotherapy: using drugs to kill cancer cells
- Radiation therapy: using high-energy radiation to kill cancer cells
- Immunotherapy: using the body's immune system to fight cancer
- Targeted therapy: using drugs that target specific molecules involved in cancer growth

The choice of treatment will be based on a number of factors, including the size and location of the tumor, the presence of metastases, and the overall health of the patient.

Prognosis for neuroblastoma in adults is generally worse than in children due to the advanced stage of disease at diagnosis. However, recent advances in treatment, such as the use of targeted therapies and immunotherapy, have improved outcomes in some cases.

In conclusion, neuroblastoma in adults is a rare and complex disease that requires a multidisciplinary approach to treatment. Early recognition and prompt treatment are crucial for improving outcomes.