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Retroperitoneallocalized Neuroblastoma in the Adult: Case Report and Literature Review

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ABSTRACT

Retroperitoneal neuroblastoma is an embryonic tumor of the peripherical 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 pediatrics staging systems and guidelines. She is now 20-years-old, and her CT scans and MIBG scintigraphys 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.

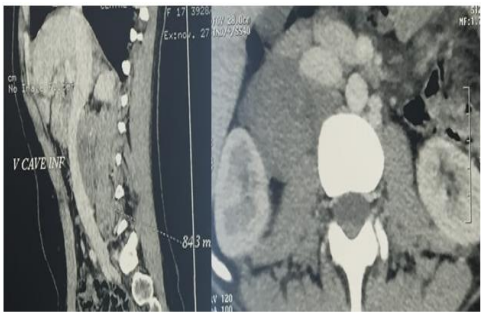

Figure 1. Abdominal CT scan showing right retroperitoneal mass.

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.

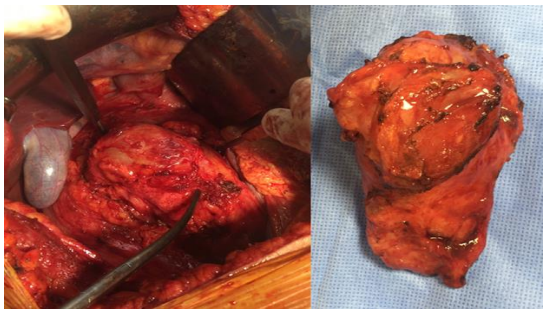

Figure 2. Overview of the mass and “en bloc” excised tissue (tumor and lymphadenectomy).

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).

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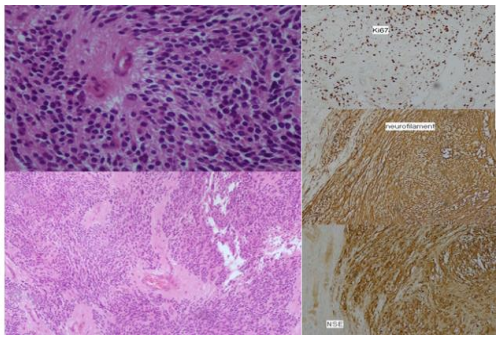

Figure 3. Morphological and immunohistochemical features of the excised tissue.

Given the resection was complete, adjuvant treatment was not indicated. Twenty-two months after the intervention, there was no sign of recurrence neither in the CT scans nor in MIBG (Meta-iodo-benzyl-guanidine) scintigraphys.

Discussion

Neuroblastoma tumors are a very rare etiology of retroperitoneal tumors in the adult [3].

While in children, primary tumors develop in 79 % of cases from the adrenal gland, the sympathetic ganglia of the abdomen or chest, in adults, 81 % of primary tumors are located at head and neck [4]. In cases where the location is abdominal, the main clinical signs are the perception of an abdominal mass, abdominal pain and digestive disorders such as diarrhea [5]. Our patient complained of an isolated right lumbar pain.

Concerning the imaging techniques, abdominopelvic CT scan or MRI are necessary to characterize the mass (localization, tumor size, density, contrast enhancement and extension to adjacent organs or vessels). MIBG scintigraphy can help searching for distant metastases. Chest CT or Brain CT should be performed if there are suggesting clinical symptoms or suspicion of lymph node disease in abdominopelvic CT scan [6]. In the adult, retroperitoneal soft-tissues masses are essentially represented by sarcoma, histiocytoma, ganglioneuroma, schwannoma and neurofibroma. Besides the fact that retroperitoneal soft-tissues masses represent less than 1 % of all localizations in the adult, neuroblastoma is a particularly rare etiology [7]. In our case, given we did not suspect a neuroblastoma, we didn't perform MIBG scintigraphy before surgery. Regarding to CT scan, we suspected a paraganglioma or adrenocortical carcinoma.

Meanwhile in children, measurement of urinary catecholamines can be useful in diagnosis given that 95 % of neuroblastoma are secreting, in the adult, only 40 to 57 % have elevated urinary catecholamines. Therefore, it has a low negative predictive value in adults [8].

Because neuroblastoma is a rare entity in adult, staging systems and treatment protocols are inspired by pediatric classifications and guidelines [1-6]. A recent study compared children and adult outcomes based on pediatrics staging systems and concluded that it can be used to stratify adult neuroblastoma patients and help select treatment [4].

In our case, according to INRG (International Neuroblastoma Risk Group) staging system, a preoperative imaging-based classification of predictive risky surgery and disease extension, our patient was stage L1 (localized tumor with no vital structures involved) [9]. Indeed, the surgical exploration found a non-adherent isolated tumor without adjacent organs infiltration.

In localized neuroblastoma with complete excision, the histological and immunohistochemical study determine treatment strategy. Indeed, according to INRG Pretreatment Classification Schema, an amplification of MYCN upgrades the pretreatment risk group from very low to high [10]. In children, MYCN amplification is associated with advanced stages and a poor outcome, and even for lower stages, with a higher progression [11]. Whereas, in adults, incidence of MYCN amplification is lower, which correlates with a better outcome, studies reported a higher incidence of unfavorable histologies in comparison with children [4,12].

According to the COG guidelines of Neuroblastoma Low-Risk Group, the standard is surgery followed by observation, which is curative for most patients, even without a complete resection. Surgery allows establishing diagnosis, resect tumor as much as possible and histological study for staging. Chemotherapy is not indicated [13,14]. Conter compared surgical resection alone versus surgical resection and radiotherapy in adult and concludes that adding radiotherapy brings a better PFS (progression-free survival) and OS (overall survival) than surgery alone [4]. On the other side, for L1 patients with MYCN amplification, whom are upgraded to high risk, the treatment is more demanding and is divided into three phases. The first is induction with chemotherapy and surgery after response. Then a consolidation phase with myeloablative therapy, stem cell transplant and radiation therapy to the site of the primary tumor. The last one is postconsolidation phase with immunotherapy and isotretinoin [14].

The only report studying outcome of neuroblastoma in adults and is statistically significant is Conter report. The median OS of adult patients was 18.1 years for localize stage and OS of 94%, 90%, and 69% at years 3, 5, and 10, respectively [4]. The prognosis of neuroblastoma in adult was not statistically different compared to pediatric neuroblastoma patients. The limit of the study was the small incidence of early stage in pediatric patients (57 L1 stage adults versus 24 L1 stage children).

Conclusion

Neuroblastoma is rare in adults, retroperitoneal localization is even rarer, and its therapeutic strategy is not well codified. Localized tumors are more frequent in adults than in children, but adults have more histological poor prognostic factors. There are few cases in literature and this one strengthens that pediatrics guidelines and staging systems can be used for the management of neuroblastoma in adults.

Competing interests

The authors declare no competing interest.

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