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Case Report

Adrenal neuroblastoma in adults: A case report and review of the literature

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Abstract

the clinical manifestations of Neuroblastoma are very variable. They depend on the site of the primary tumor, most often in the abdomen, along the spine or in the adrenal gland Very often, it is the existence of metastases which reveals the disease, with bone pain, and / or difficulty in walking or urinary problems. Management must be carried out in a specialized multidisciplinary team. This will confirm, by imaging, the existence of a neuroblastoma, perform a biological assessment, search for metastases from a distance, and define prognostic factors and the appropriate therapeutic strategy.

Key Words: adrenal tumor; neuroblastoma adult

Introduction

Neuroblastoma is the most common extra-cranial solid malignant tumor in young children. It is a malignant tumor derived from cells originating in the tissues that give rise to the sympathetic nervous system. It can be found in front of the spine and in the internal part of the adrenal gland.

It most often sits behind the peritoneum.

Neuroblastoma accounts for about 10% of solid tumors in children. It is the most common malignant neoplasm of infants: 50% of affected children are less than 2 years old. In a third of cases, the tumor occurs in a child under one year of age and in 95% of cases before the age of 10. For the majority of neuroblastomas, there is no known cause and they remain exceptional in adults.

Predisposition syndromes have been described: Neurofibromatosis type 1, Hirschsprung disease, Ondine syndrome. Very exceptionally, neuroblastoma has been observed in association with Beckwith-Wiedemann syndrome or Di-George syndrome.

Observation

Patient C.N female; 22 year old who has no particular pathological history; who consults for chronic constipation associated with dysmenorrhea; the patient shows no signs of hypercorticism; no signs of hyper androgenism or hypokalemia.

Also the absence of Menard's triad. Biological examinations:

- Urinary metanephrines = 0.12 mg / 24h (0-1).
- Cortisol = 284.4nmol / l. Cortisol after minute braking = 22.3nmol / l (+).
- Testosterone less than 0.45nmol / l. - SDHEA = 116.7ug / dl.

Imaging abdominal pelvic MRI: fig. 1
Fig. 1. Right retroperitoneal supra-renal mass of 124/75/86 mm enhancing heterogeneously and delimiting necrosis areas of malignant appearance compressing the inferior vena cava.

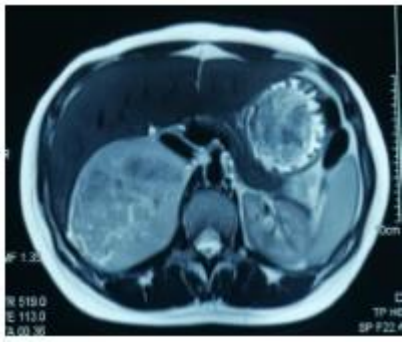
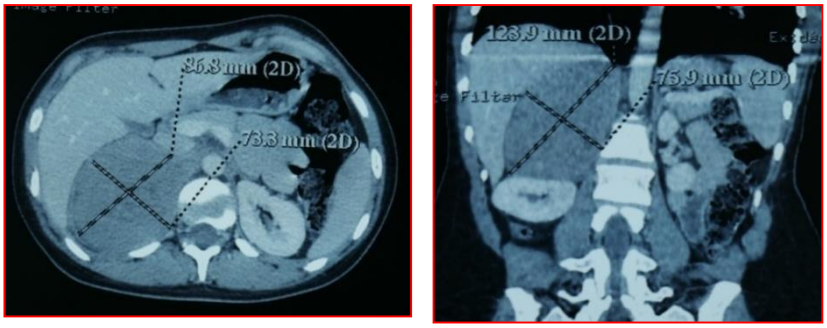


Fig. 1 MRI Axial Section

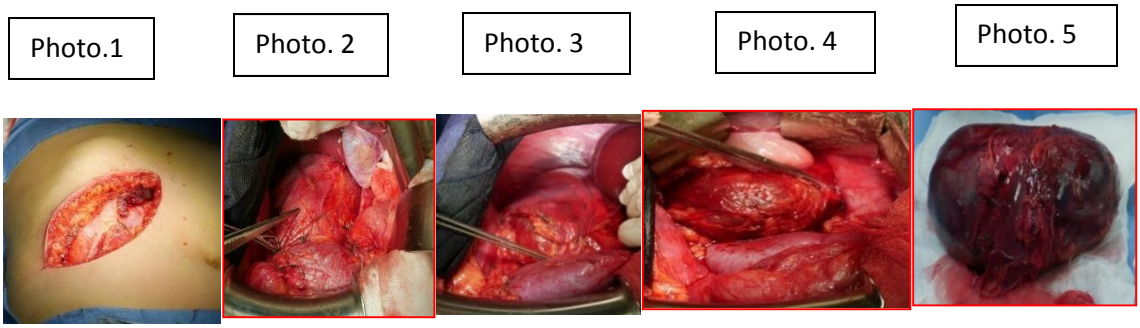
Abdominal Doppler ultrasound: Lower vena cava compressed by a right adrenal mass without thrombosis and without downstream impact.

Abdominal CT: Fig. 2 (a; b): Probable right adrenal mass looking suspicious.



Abdominal CT: Fig2 (a; b) axial and coronal sections Probable right adrenal mass looking suspicious.

Surgery



- Photo 1 Right costal incision.
- Photo 2 Adrenal tumor driving the kidney down; the VCI on the right.
- Photo3 Dissection of the mass compared to the IVC after dislocation of the liver.
- Photo 4: release of the tumor from its vascular attachments.
- Photo5 Neuroblastoma undergoing differentiation: (anatomo pathology study).

Post-operative follow-up:

The extension assessment including a bone scan and a chest CT shows a suspicious thymic retro-sternal mass. The patient was operated 03 months after the first intervention or a thymic resection was carried out and the anatomo-pathological study returns in favor of a benign thymic lesion.

Discussion

Neuroblastoma is a tumor originating from ganglion cells derived from the neural crest. It is the most common solid extra-cerebral tumor in children, responsible for 15% of deaths from neoplasia. 36% of cases are diagnosed before the age of one, 75% before five, and more than 97% before the age of ten. The tumor biopsy is used to look for abnormalities in the tumor chromosomes, which are used to classify the level of aggressiveness of the tumor: abnormalities in number of best prognosis.

The evolution depends on the age of the child at the time of diagnosis, the tumor extension. The therapeutic strategy is built taking into account these three factors: for tumors that can be immediately treated, surgery alone will most often be sufficient; for tumors that are not immediately operable, pre-surgical chemotherapy is necessary before surgery. The evolution is very variable. Indeed, in infants, certain localized or metastatic tumors can regress spontaneously; some others, in older children, escape treatment despite the use of very aggressive combinations.

In metastatic forms and / or in case of amplification of MYCN, consolidation by high-dose chemotherapy with hematopoietic stem cell transplantation is necessary, as well as radiotherapy of the tumor and maintenance treatment comprising immunotherapy. Survival varies according to the identified risk, between more than 90% of recovery for localized forms and young children, to 40 to 50% in the most serious forms.

The search for a genetic anomaly, initial or at relapse, makes it possible to develop treatments targeting these anomalies in the event of non-control of the disease by conventional treatments.

One of the reasons for its rarity in adulthood and adolescence could be attributed to the tendency of these tumors to spontaneous regression or down staging without any treatment.

In adults, the anatomical localizations are most often abdominal in 60%, (of which 32 - 35% adrenal), followed by the medial localizations 20% and pelvic. To date, no environmental factors or parental exposure have been found to explain the occurrence of the disease.

Risk groups are defined according to the anatomical stage, age at diagnosis, histopathology classification and the amplified or non-amplified status of the N-MYC oncogene, the whole defining the criteria of the International Neuroblastoma Staging System (INSS) Table1.

Table 1. Neuroblastoma Clinical Stages

International Neuroblastoma Staging System (INSS):

Stage 1: Localized tumor confined to the area of origin.
Stage 2A: Unilateral tumor with incomplete gross resection; identifiable ipsilateral and contralateral lymph node negative for tumor.
Stage 2B: Unilateral tumor with complete or incomplete gross resection; with ipsilateral lymph node positive for tumor; identifiable contralateral lymph node negative for tumor.
Stage 3: Tumor infiltrating across midline with or without regional lymph node involvement; or unilateral tumor with contralateral lymph node involvement; or midline tumor with bilateral lymph node involvement.
Stage 4: Dissemination of tumor to distant lymph nodes, bone marrow, bone, liver, or other organs except as defined by Stage 4S.
Stage 4S: Age <1 year old with localized primary tumor as defined in Stage 1 or 2, with dissemination limited to liver, skin, or bone marrow (less than 10 percent of nucleated bone marrow cells are tumors).

International Neuroblastoma Risk Group Staging System (INRGSS):

Stage L1: Localized disease without image-defined risk factors.
Stage L2: Localized disease with image-defined risk factors.
Stage M: Metastatic disease.
Stage MS: Metastatic disease "special" where MS is equivalent to stage 4S.

Surgical resection is the standard curative treatment. Chemotherapy is indicated in locally advanced or metastatic forms. The most used molecules: Cyclophosphamide, Cisplatin, and Doxorubicin…Radiotherapy is an important complement to chemotherapy.

Adult forms, however, are exceptional. The few publications on this subject seem to show significant differences from pediatric forms, in particular the very indolent nature of adult forms and their most often fatal prognosis.

Conclusion

It should be noted that Adrenal Neuroblastomas are very rare in adulthood, but it should be borne in mind that each adrenal tumor without specific characteristics has a high probability of being identified as Neuroblastoma.

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