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Case Report

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ADRENAL GANGLIONEUROBLASTOMA IN ADULTS: RARE CLINICAL CASE

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SUMMARY

We report the case of an adrenal ganglioneuroblastoma (GNB) in a 32-year-old patient, incidentally discovered, who underwent a complete surgical excision. The pathology study concluded that a nodular adrenal GNB which is a very rare tumor in adults with an unfavorable prognosis, only 19 cases of adrenal ganglioneuroblastoma in adults have been observed in the literature.

KEYWORDS: Ganglioneuroblastoma. adrenal. Adult.

INTRODUCTION

Ganglioneuroblastoma is a histological type of childhood neuroblastic tumor that grows at the expense of neuronal cells with a mixture of neuroblasts and ganglion cells.

Rare in adults, only 19 cases of adrenal ganglioneuroblastoma in adults have been observed in the literature.[1]

CLINICAL CASE

We report the case of a 32 year old patient, mother of 3 children with no personal or family medical history, who consults with a general practitioner for isolated vomiting, the clinical examination was without particularities, an abdominal ultrasound was performed objectifying a mass at the expense of the upper pole of the right kidney. An abdomino-pelvic CT confirmed the presence of a mass at the expense of the right adrenal gland with tissue density of 13 cm of long axis, which increases heterogeneously after PDC injection, site of multiple macro calcifications. The biological assessment has objectified blood levels = cortisol from 8h to 13µg / dl (6.40-21), aldosterone 192pg / ml (42-209), renin 17.30pg / ml (< 20) and urinary levels = Metadrenaline 25nmol / 24h, Normetadrenaline 297nmol / 24h and a high rate of 3 Ortho Methyldopamine 3794nmol / 24h (69-315) .The rest of the biological balance was without particularities.

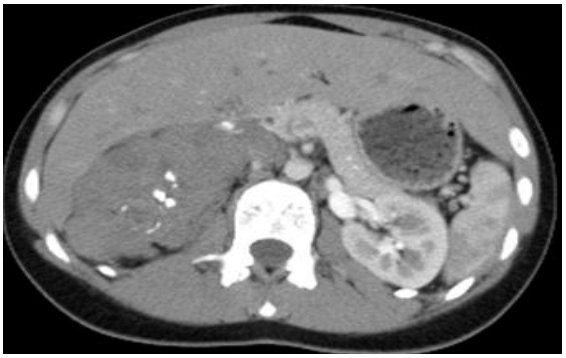

Figure 1: Abomino pelvic CT "cross section" objectifying a right adrenal mass of 13 * 10 * 6.6 cm, site of calcifications.

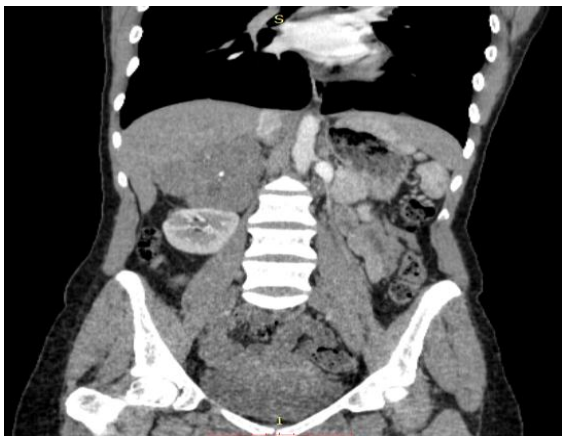

Figure 2: CT Abomino pelviennne "cut coronale" objectifying the right adrenal mass.

A right adrenalectomy was performed with total excision of the mass, the post-operative follow-ups were simple.

The histopathological examination relates to a multinodular tumor proliferation made of nodules of variable size containing hemorrhagic changes. It is made of neuroblastic cells at different stages of maturation. The stroma is an abundant fibrillar Schwanian type. Territories of necrosis are observed with the presence of vascular emboli. An additional immunohistochemical study led to the conclusion that a stromal ganglioneuroblastoma rich in nodular type classified as an unfavorable prognosis group according to the INPC classification.

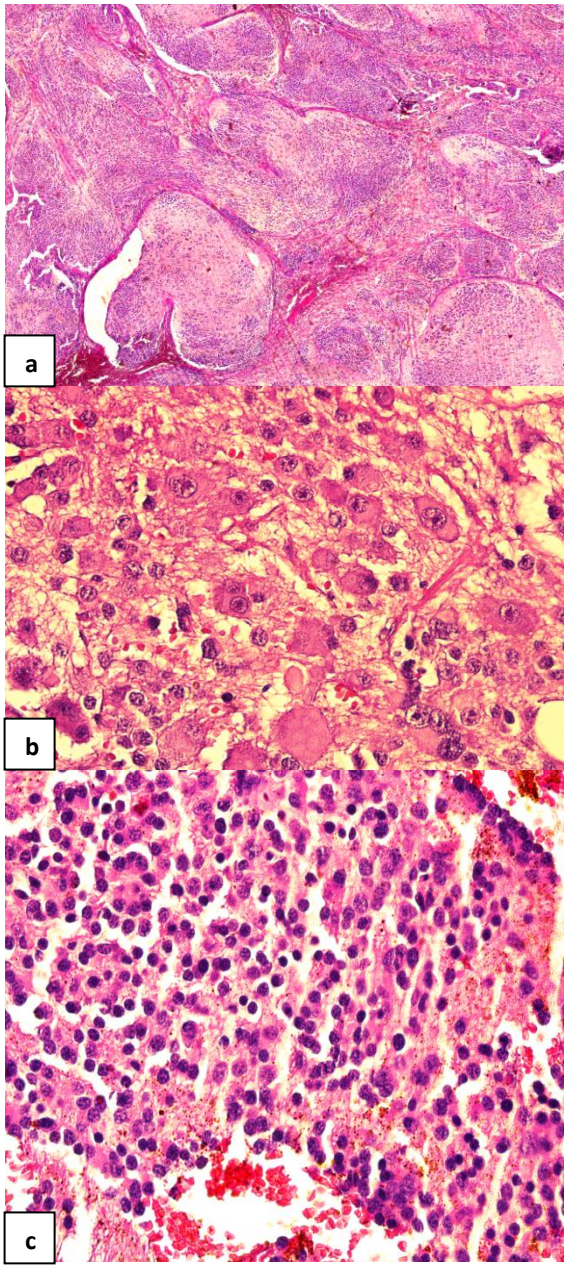

Figure 3: a) GNB with rich stroma in nodular type: HE x 40. b) GNB with rich stroma in nodular type (HEx400): predominant contingent of well differentiated tumor, with rich schwannian stroma. c) GNB with rich stroma under nodular type (HEx400): minor contingent of poorly differentiated tumor, with poor Schwannian stroma.

A postoperative extension assessment of a thoracoabdomino-pelvic CT scan did not show any macroscopic residue or secondary locations.

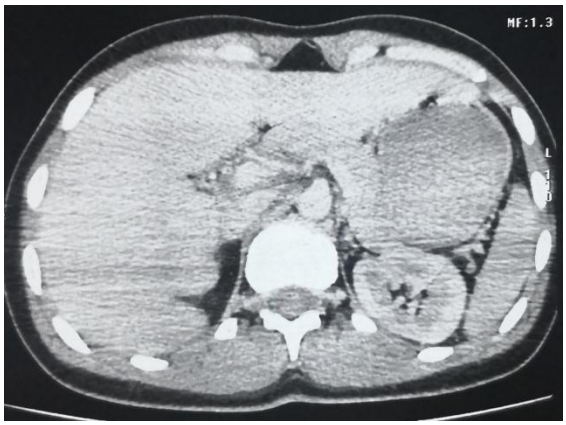

Figure 4: Post-operative TAP CT: Absence of residual mass or recurrence.

The case was discussed in CPR, and an additional extension assessment made of MRI and MIBG scintigraphy was requested before discussing possible adjuvant chemotherapy.

Table 1: Neuroblastoma prognosis according to classification (INPC / Shimada).[15]

Age	Pathology	Prognostic group
<1.5 years	Poorly differentiated or differentiating and low or intermediate MKI	Favourable
<1.5 years	Undifferentiated tumour or high MKI	Unfavourable
1.5–5 years	Undifferentiated or poorly differentiated and intermediate or high MKI	Unfavourable
>5 years	All tumours	Unfavourable

MKI, mitosis–karyorrhexis index.

DISCUSSION

Ganglioneuroblastoma is a tumor of the nervous tissue that occurs mainly in children, but their occurrence in adults is extremely rare.[1]

In a study of 1,111 adult patients with adrenal incidentalomas, GNB was diagnosed in only one case.[3]

In adults, GNB is characterized by a high potential for malignancy and distant metastases, especially in the liver and bone.[4]

The clinical picture remains atypical. Symptoms of gravity or pain may be related to the mass effect exerted by the tumor.[5] In some cases, the discovery is fortuitous during a routine examination such as the case in our patient.[6,7] While some are discovered at an advanced stage of distant metastasis.[8,9]

The contribution of urinary catecholamine dosage in the diagnosis of ganglioneuroblastomas remains limited. They cannot be used to differentiate ganglioneuroblastoma from pheochromocytoma.[1] In our case only 3 Ortho Methyldopamine was high.

For incidentalomas, imaging does not distinguish ganglioneuroblastoma from other adrenal incidentalomas.[10]

The confirmatory diagnosis is essentially based on the anatomopathological examination of the piece of excision or biopsy.

The International Classification of Neuroblastoma Pathologies (INPC), established in 1999 and revised in 2003, redefined the histological characteristics into 4 categories of tumors: Neuroblastoma, mixed GNB, Ganglioneuroma, nodular GNB. The four categories are divided into two distinct prognostic groups: favorable histology and unfavorable histology.[5,6] (Table 1)

The therapeutic protocols are derived from pediatric experience. The therapeutic arsenal includes surgery, radiotherapy and chemotherapy. If the tumor is considered to be resectable, surgery is the treatment of choice, if not, a diagnostic biopsy must be performed before chemotherapy or radiotherapy. Chemotherapy remains the treatment of choice in metastatic forms.[11]

Complete resection with negative margins is a good prognostic factor for GNB and constitutes the curative treatment of localized forms.[12] Some studies suggest that lymph nodeeuroblastomas in adults are slow growing.[13] And adjuvant therapy may not be necessary after surgery.[9]

The prognosis remains uncertain in the rare cases reported in the literature, but the risk of recurrence remains high during the first 2 years after surgery. Close monitoring with a MIBG scintigraphy every 6 months is recommended during these first 2 years.[14]

CONCLUSION

Adrenal GNB in adults remains a rare tumor, the diagnosis is essentially confirmed by the anatomo-pathological study, and the treatment protocol is derived from pediatric experiences hence the need to group the cases reported in the literature for a standardized protocol.

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