PP362 - ADRENAL GANGLIONEUROBLASTOMA PRESENTING AS INCIDENTALOMA IN A YOUNG WOMAN

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Peripheral neuroblastic tumors (PNTs) are a group of heterogeneous tumours (including neuroblastoma, ganglioneuroblastoma and ganglioneuroma) arising from sympathetic ganglion cells and frequently localized in the adrenal gland. Prognosis is variable and depends on combination of several factors: age at diagnosis, stage, pathology and genetic abnormalities. PNTs are one of the most frequent solid tumour in children, while the occurrence in older patients is rare. PNTs represent an exceptional cause of adrenal incidentaloma in adults: 15 cases of adrenal ganglioneuroblastoma (GNB) in adults have been described in literature so far.

We report the case of a 20-years old woman presenting to the emergency department because of left flank pain. Her clinical history was unremarkable. Abdominal ultrasound examination revealed a contralateral adrenal mass. Computed tomography confirmed a solid adrenal lesion measuring 11x10x7 cm, heterogeneous in density and with scattered calcifications. She had no Cushing phenotype or hyrsutism, and hormonal hypersecretion was excluded. Giving the size and the undetermined nature of the mass, according to the most recent guidelines, she underwent surgical removal. The tumour bulk included the aorta, superior mesenteric arteria, the renal hilum and vessels therefore additional nephrectomy was performed. The histological report described a spindle cell stroma in a fibrillary matrix interspersed with scattered nests of primitive neuroblasts and high proportion of ganglion differentiating cells. These findings were consistent with intermixed stroma-rich GNB arising from the adrenal gland with metastatic extension to ipsilateral lymphnodes.

Abdominal pain or compressive symptoms represent the most common clinical presentation of adrenal GNB in adult patients. These tumors grow silently measuring at diagnosis up to 18 cm (mean size 10.44 cm) and showing metastatic invasion in 50% of cases. In locally advanced PNTs with potentially associated surgery-related complications, pre-surgical chemotherapy should be administered in order to shrink the tumor and enable safe resection saving other abdominal viscera involved. Nevertheless radiological features of tumors are often misleading and neuroblastic origin of the mass was hypothesized only in a few cases. Consequently patients were addressed straight to surgery and the definitive etiology was histologically defined. Overall survival from PNTs in infant is >90% at 5 years and progressively decline parallel to the increased age at diagnoses. Our patient is alive 1 year after operation. Long term prognosis of adrenal GNBs in adults is uncertain, giving the short follow-up duration and small number of cases reported.

PP363 - CIRCULATING ADIPOKINE LEVELS IN DIABETIC PATIENTS WITH CUSHING DISEASE ON PASIREOTIDE TREATMENT COMPARED WITH PATIENTS WITH TYPE 2 DIABETES MELLITUS

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Background: Pasireotide treatment is associated with a good control of hypercortisolism and body weight decrease in patients with Cushing's disease (CD). Aim of the study was to evaluate the effects of pasireotide on circulating adipokine levels in diabetic patients with CD compared to patients with type 2 diabetes mellitus (T2DM).

Methods: Clinical and hormonal parameters and circulating adipokines were evaluated in 12 diabetic patients with active CD before and after 12 months of pasireotide treatment and in 12
patients with T2DM as controls.

Results: At baseline, patients with CD showed significantly higher levels of leptin (Ob) (p 0.008), adipocyte fatty acid binding protein (AFABP) (p 0.045) and non-esterified fatty acids (NEFAs) (p 0.022), with higher leptin/Ob receptor (Ob-R) ratio (p 0.024) than diabetic controls.

After 12 months of treatment, patients with CD showed a significant decrease in BMI (p 0.008), waist circumference (WC) (p 0.009), ACTH (p 0.049) and urinary free cortisol (p 0.007), in concomitant with a significant decrease in Ob (p 0.039), Ob/Ob-R ratio (p 0.017) and AFABP (p 0.036) and a decrease in Ob-R (p 0.028) than baseline, while no differences in all parameters were observed comparing with controls, except for leptin (p 0.037).

Conclusions: Treatment with Pasireotide is associated with an improvement of adipose function in diabetic patients with CD. However, despite the good control of hypercortisolism, these patients maintain a degree of adipose tissue dysfunction compared to T2DM patients.

PP364 - COMPARISON OF CLINICAL AND METABOLIC EFFECTS OF PASIREOTIDE AND PITUITARY SURGERY IN CUSHING DISEASE

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Background: Pituitary surgery is the first-line therapy for patients with Cushing's disease (CD). However, pasireotide is showing favourable results in the treatment of patients with CD. Aim of the study was to compare the surgery and pasireotide effects on hormonal, metabolic and clinical parameters in patients with active CD.

Materials and methods: Body mass index (BMI) and waist circumference (WC), ACTH, mean urinary free cortisol (mUFC) and metabolic parameters were evaluated in 8 patients with active CD treated with pasireotide and 16 patients with active CD treated by pituitary surgery.

Results: A significant reduction in BMI (p <0.001), WC (p 0.007), mUFC (p 0.003), ACTH (p 0.020) and total cholesterol (p 0.012), in concomitance with an increase in fasting glycaemia (p 0.007), HbA1c (p 0.015) and AUC2h glycaemia (p 0.007) was observed after 12 months of pasireotide. By contrast, surgery resulted in a significant decrease in BMI (p <0.001), WC (p <0.001), mUFC (p 0.001), ACTH (p 0.001), total cholesterol (p 0.020), LDL-cholesterol (p 0.045) and fibrinogen (p 0.025). Comparing the changes (Δ) in the hormonal parameters after 12 months of treatment, surgery resulted in a higher Δ_ACTH (p= 0.015), than pasireotide, notably in patients with moderate (p= 0.039) and severe (p=0.019) CD, and Δ_mUFC (p 0.019) in patients with severe (p 0.035) CD.

Conclusions: These preliminary data suggest that pasireotide and surgery are effective in the control of CD. However, surgery reduces the hormonal parameters more markedly in patients with moderate and severe CD.