Silent corticotroph adenoma with adrenocortical choristoma in an 11-years old boy

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BACKGROUND

Silent corticotroph adenomas are adenomas composed of corticotrophs but are different from corticotroph adenomas because they are not associated with clinical and biochemical evidence of ACTH production or release in vivo. Despite being silent, they show more aggressive behavior than other clinically nonfunctional adenomas.
Adrenocortical choristomas in silent corticotroph adenomas (the presence of adrenocortical cells in the heterotopic location of the sella) were reported in three patients 16 years or older until now. Here we report, to our knowledge, the fourth and the youngest case of silent corticotroph adenoma with adrenocortical choristoma.

CASE REPORT

4 months old,
4.050 kg,TSH slightly high
TRH stimulation test:

	TSH(mIU/L)	PRL(ng/ml)
0’	3.6	25.3
20’	37.9	75.3
30’	38.0	72.6
60’	25.1	47.6

Started on 25 microgram L-thyroxine

10 years 4 months old,
Testicular volumes 5/5 ml, stretched penile length 7 cm,P1A1

11 years one month old,
56 kg,receiving on 5 week days 50 microgram and 2 week days 25 microgram L-thyroxine
Free T4 low,TSH low: Secondary hypothyroidism
ACTH :11.78 pg/ml,cortisol:4.82 microgram/dl.
Low dose ACTH stimulation test:

	Cortisol microgram/dl
0’	7.31
30’	15.27
60’	14.38

Started on hydrocortisone

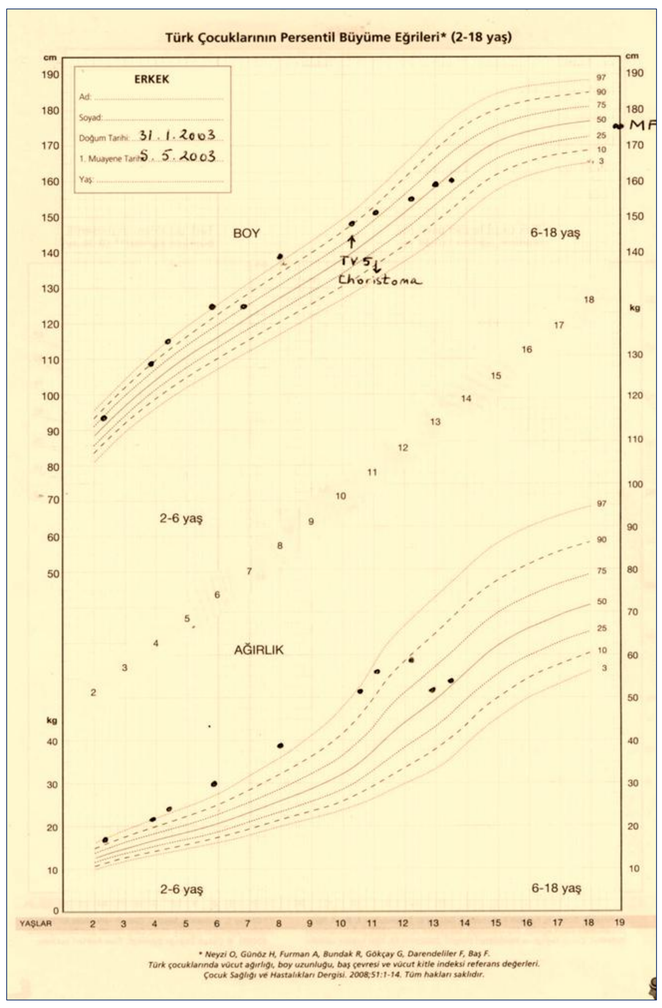

Figure -1: Growth chart of the patient

Radiological investigation:
On MRI, tumor was discovered
In the location of the hypophysis gland,a lesion of 11x11x10 mm which
- contrasts later than the hypophysis gland
- is located centrally and in the left paramedian area
- pressurizes the chiasma by proceeding to the suprasellar cisterna
- pushes the infundibulum to the right and anterior

Probable adenoma
Adenomectomy

Pathological diagnosis:
ACTH expressing hypophysis adenoma,adrenocortical choristoma

Histological sections of the tumour revealed a mixture of :
- small round cells with amphophilic or basophilic cytoplasm
- large spherical and oval cells with abundant, granular, partly vacuolated acidophilic cytoplasm

On PAS staining(Fig 2-3):
- small cells weakly to moderately positive
- large cells negative

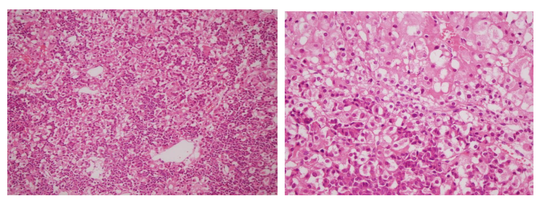

Figure-2-3: Mixture of the small round well-granulated cells with amphophilic or basophilic cytoplasm (Corticotroph cells) and the large spherical or oval cells with abundant, granular, partly vacuolated cytoplasm (Adrenocortical cells) form groups (H&EX100-400)

Small round cells(Fig 4,5):
- immunpositive for synaptophysin and ACTH
- negative for GH, PRL, FSH, LH, TSH and inhibin

Large cells(Fig 6,7):
- positive for inhibin
- mitochondrial protein.

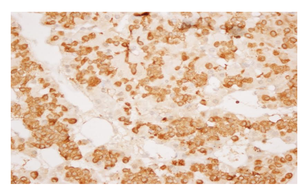

Figure-4: Small cells immunpositive for ACTH (ACTHX400)

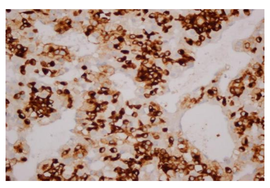

Figure-5: Adrenocortical cells are immunnegative and corticothop cells immunpositive for synaptophysin (Synaptophysin X400)

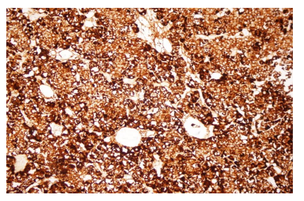

Figure-6: Large, vacuolated cytoplasm adrenocortical cells are dense immunpositive for mitochondrial antigen (Mitochondrial ag X100)

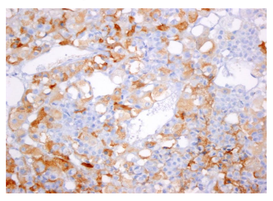

Figure-7: Adrenocortical cells are immunpositive for inhibin (InhibinX400)

Based on the findings above, pathological diagnosis was adrenocortical choristoma in endocrinologically silent corticotroph adenoma.

CONCLUSIONS

The lack of biochemical and clinical evidence of Cushing syndrome despite ACTH expressing cells in the adenoma indicated the presence of a silent adenoma. The presence of a second group of cells similar to adrenocortical cells in this heterotopic location is compatible with choristoma. The origin of adrenal cortical cells within a pituitary adenoma is unexplained. Previous reports suggested the possibilities of either an abnormal proliferation and differentiation of uncommitted mesenchymal stem cells within the sella or misplaced adrenal cortical cells derived during embryogenesis. The younger age of our patient than those of previously reported cases and clinical significance of silent corticotroph adenoma in general make this case of rare entity more remarkable.

References:
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