Cushing´s disease:experience in a third level hospital from Zaragoza (Spain)

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Topic:Adrenal

Objectives

•Cushing´s disease (CD) is caused by pituitary corticotrophin (ACTH)-secreting tumors.
•Our aim is to show our experience in a third level hospital attending a 500000 based population area from 1990 to now.
•We want to compare our experience with data collected from other series
•We want to know the patient profile we are attending to elucidate the best way to manage them

Methods

•we rewiew retrospectively the medical reports of all patients diagnosed as CD from 1990 by a data collection protocol.

•We desing an algorithm protocol to rewiew in the same order the medical information of our patients

•We reject the reports we consider have not enough information or have inadequate information.

•We present results by using descriptive statistics

Results

29 patients were diagnosed as CD in the period study (stimate average incidence of 2,41 cases per million people per year).9 medical reports contain insufficient or misleading information including 2 from dead patients.The remaining 20 patients were included in our analysis.Main results are presented in tables 1-4 and in figure 1.

Table 1.Population caracteristics

Age at diagnosis	39 years (17-75)
Women	90%
Body mass index	28,45 kg/m² (19,55-44,6)
Follow up time	11 years (2-24)

Table 2.Clinical caracteristics

Most common reference reason		
	55%	hyperandrogenism
	25%	facial/troncal adiposity
Most common symptom	65%	hirsutism
	55%	obesity/weigth gain
	55%	HTA/DM worsening
	55%	muscle weakness,fatigue
	50%	ecchymosis

figure 1.Treatment

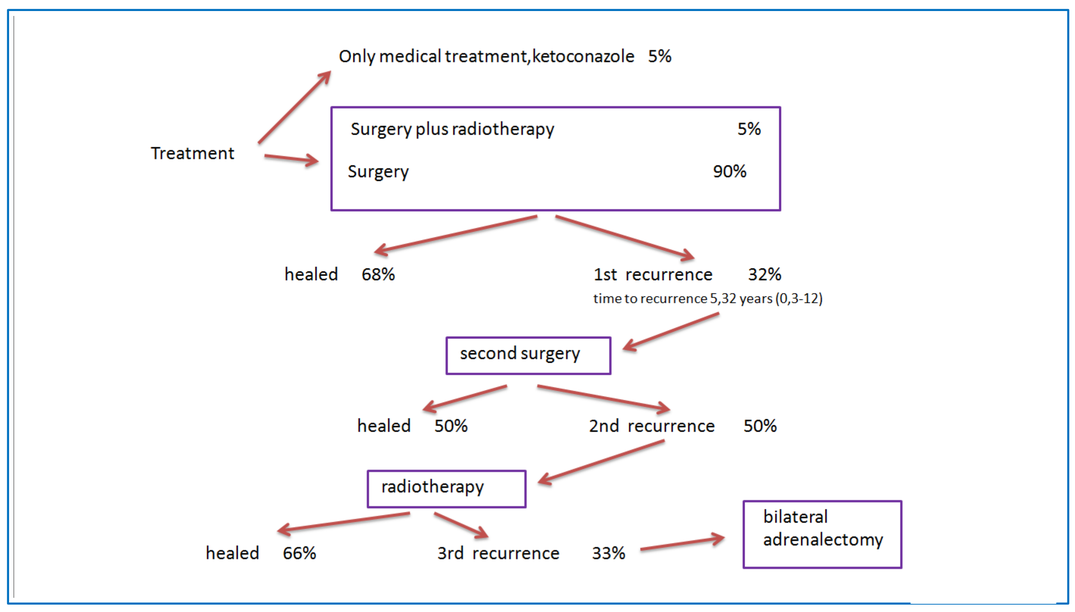

Table 3.Diagnostic findings

cortisol rhythm	with	12,5%
	without	87,5%
	inverted	31,25%
plasma basal cortisol		27,9 mcg/dl (18-51)
plasma basal ACTH		71,96 pg/ml (20-231)
plasma cortisol after 1 mg DXM (test A)		14,6 mcg/dl (3,38-34,9)
plasma cortisol after 4 mg DXM	14,53 mcg/dl(3,08-23,46)	
plasma cortisol after 8 mg DXM (test B)		7,96 mcg/dl(0,34-24,45)
difference betwen test A and test B		-5,6 mcg/dl(-17,7- +2)
plasma cortisol at midnight		13,63 mcg/dl (9,26-18,03)
24-h free urinary cortisol		840,7 mcg/24-h(97-5530)
MRI	any finding	35%
	microadenoma	55% (2 mm to 10 mm)
	macroadenoma	10% (12 mm and 45 mm)
inferior petrosal sinus sampling		done before treatment 70%
	undone	20%
	done after first line treatment 5%	
	done after second line treatment 5%	
	peak ACTH value	2560,45 pg/ml(182-11100)
	right lateralization	45%
	left lateralization	35%
	no lateralization	20%
pituitary hispopathology	no findings	20%
	multisecretor tumor	60%
	only ACTH-secreting tumor	20%

conclusions

•our patients reproduce well referenced data from other series epidemiologically,clinically and in their global outcomes.We have a ratio slighly worse for surgical cure than other recent series.
•hyperandrogenism was the main cause of consultation and the most common symptom
•12,5% of patients keep their diurnal physiological variation of cortisol secretion
•the suppression test with 4 mg of DXM contributed nothing to the diagnosis appart from the 1 mg DXM suppression test
•the minimum threshold cortisol value in the 1 mg DXM suppression test of our patients was 3,38 mcg/dl and the minimum midninght plasma cortisol value was 9,26 mcg/dl.
•plasma cortisol in the 8 mg DXM suppression test was highly variable and less helpful in our serie.
• 24-h free urinary cortisol treshold value of 200 mcg/24-h have a diagnostic sensitivity of 92% in our serie.
•70% of patients have required inferior petrosal sinus sampling to comfirm the pituitary ACTH-hypersecretion before entering the surgical procedure.
•worsening of vascular risk factors and loss of bone mass were the main complications of CD before and during diagnostic procedure.
•pituitary deficiencies and perisurgical diabetes insipidus and nasal liquorrhea are the main complications after treatment.
•It would be desirable to develop uniforms diagnostic protocols through different hospitals and countries

Table 4.Complications

osteoporosis		30%
osteopenia		45%
hypopituitarism	presurgery	5%
	postsurgery	15%
nasal liquorrhea		20%
diabetes insipidus		10%
need for cronic hydrocortisone supplementation		35%
meningitis		5%
derivation to intensive care unit		5%
death		0% (2/29,6,8%)

bibliography

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