



UNFAMILIAR PRESENTATION OF ACROMEGALY WITH NEUROFIBROMATOSIS AND HYPER PROLACTONEMIA

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ABSTRACT Acromegaly results from anterior pituitary adenoma with hypersecretion of growth hormone. Acidophilic adenoma secrete both growth hormone and prolactin (in our case), we report a case of increase in growth hormone and prolactin with associated neurofibromatosis and hirsutism.

KEYWORDS : Acromegaly-Hyperprolactonemia-Hirsutism-Neurofibromatosis

Case history:

40 yrs old female presented since the age of 30 yrs with acral enlargements (hands and feet) Fig.1, bony prominences of face (frontal bossing) with wide gap between the teeth of lower jaw, oily skin, decreased distant vision in both eyes, irregular menses and cutaneous nodules on left forearm, lower back (2x2 cm) and OA changes in both knees (Fig. 1), dorsal kyphosis, increased size of fingers and toes, coarse facial features with history of galactorrhoea and snoring. No past history of diabetes, hypertension, asthma, tuberculosis, seizures. Physical findings include pitting edema of feet, wrinkles on forehead, 2 x 2 cm mobile subcutaneous nodules over left forearm and lower back (circled ones Fig. 1). Pulse rate 88 per minute, blood pressure 190/110 mm.Hg. Cardiac examination revealed forcible apex beat., Respiratory system and abdomen examinations were normal. Genu valgum deformity of both knees (Fig. 1) present. Hirsutism score 18/36. Clinical diagnosis of acromegaly, galactorrhoea, hirsutism, hypertension and diabetes made.

Investigations:

Ophthalmological exam – showed bitemporal hemianopia with more involvement of right eye (Fig.2), X-ray of skull lat view (Fig. 3) showed increased diploe, erosion of posterior clinoid process, enlarged and widened pituitary fossa (sella turcica). MRI of brain in T1 and T2 images (Fig. 3) showed heterogenous cystic component compressing optic chiasma.

Biochemistry showed raised blood sugars, increased prolactin and growth hormones (table 1). Biopsy of subcutaneous nodules confirmed neurofibroma histology (spindle shaped cells, irregular nuclei, lying against myxoid background).

Management:

Treated with metformin, dopamine agonists bromocriptine and cabergoline., SRIF receptor ligands octreotide and anti hypertensives. Increased nocturnal prolactin levels treated with cyproheptadine. These measures helped in shrinkage of tumour size. Later submitted to transsphenoidal pituitary surgery. Within 4 hrs after surgery growth hormone level decreased, prolactin level remained normal, vision improved but patient developed SIAADH postoperatively which was managed conservatively. Neurofibromas excised.

Followup:

Periodically with T1 and T2 weighed MRIs, growth hormone, prolactin, cortisol and perimetry showed no recurrence of pituitary tumour

DISCUSSION:

We presented this case of acromegaly for its unfamiliar association with neurofibromatosis and hirsutism and hyperprolactonemia. Literature shows 50% of patients with acromegaly have elevated prolactin levels. Postoperatively measurement of growth hormone showed fall in growth hormone to less than 1ug/L with significant decrease in blood pressure, LV mass and heart rate.

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Fig. 1 genu valgum, acral enlargements and neurofibromatosis

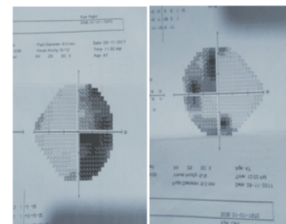


Fig : 2 Rt. eye & Lt. eye



Fig: 3 Lat view skull X ray & MRIs

Table 1 Biochemistry

Fasting blood glucose 149 mg/dl
Serum growth hormone – 1hr 42.13 ng/ml
Postprandial blood glucose 277 mg/dl
Serum growth hormone – 2hr >50 ng/ml
Serum prolactin >200 ng/ml
Serum FSH 4.29 mIU/ml., LH 0.62mIU/ml
Serum cortisol (8am) 5.41ug/dL
Anti HCV, HIV 1 & 2, HBsAg non reactive

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