Acromegaloidism Associated with Pituitary Incidentaloma

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Abstract

Acromegaloidism with pituitary microadenoma has not been previously reported. We present a case of a 28-year old male with typical features of acromegaly for 11 years with a pituitary tumor. He had characteristic acromegaloid facial features, clubbing of hands and feet, enlargement of fingers and toes. The natural history of the disease is reviewed and the differential diagnosis is discussed.

Introduction

Acromegaloidism is a syndrome consisting of clinical features similar to acromegaly but without excessive growth hormone (GH) production, and it is difficult to distinguish between these disorders on the basis of their clinical manifestations. Acromegaloidism has no dysfunction of pituitary or hypothalamus and these patients have normal levels of GH and insulin-like growth factor (IGF-1) both at baseline and dynamic testing.¹ GH-producing pituitary adenomas are associated with overproduction of GH, which may lead to acromegaly. Acromegaly is often confused with pachydermoperiostosis or primary hypertropic osteoarthropathy, which is a rare hereditary disorder. Acromegaloidism also closely resembles pachydermoperiostosis, which however has no demonstrable causative factors such as pulmonary, cardiac or hepatic disorders.² Pachydermoperiostosis is characterized by digital clubbing, thickening of the facial skin and/or scalp, and swelling of periarticular tissue and sub-periosteal new bone formation. It is associated with joint pain, polyarthritis, cutis verticis gyrata, seborrhea, eyelid ptosis,³ and hyperhidrosis. In the current case, the patient presented with classic dermal symptomatology of pachydermoperiostosis along with a pituitary microadenoma, thus making differential diagnosis of acromegaloidism a challenging task.

Case Report

A 28 year old male presented with complaints of enlargement of fingers and toes, and prominent skin folds on his forehead and cheeks since 17 years of age (Figure 1). He had a history of hyperhidrosis with thick upper and lower eyelids. On examination, his height and weight were found to be 174 cm and 68 kg respectively and he was normotensive. His skin was coarse and greasy, with hyperhidrosis and grade 4 digital clubbing. Patient had enlarged hands and feet (Figure 2) but there was no history of change in ring or footwear size. There was ptosis with thickening of upper and lower eyelids and mild prognathism, with blubbery lips. Oral examination showed a normally sized tongue. His examination of thyroid, heart and abdomen were normal with no evidence of goiter, cardiac dysfunction, or organomegaly. None of his family members had similar features.

On biochemical evaluation for GH suppression test with 75 grams of anhydrous glucose showed a 0 minute human growth hormone (HGH) level of 0.22 ng/mL, insulin-like growth factor-1 (IGF-1) level of 106.0 ng/ml (ref. range: 117-329) and HGH of 0.1 ng/mL at 30 minute and 60 minute with corresponding fasting blood glucose of 92 mg/dL and 2 hour blood glucose of 105 mg/dL. The other hormonal evaluation and serum calcium, phosphorous, potassium and sodium were in the normal range (Table 1). The hemoglobin level was 12.63 g/dL and the total count was 11780 cells/cumm. However, a higher level of alkaline phosphatase and gamma-glutamyl transferase was observed while the other liver function tests were normal.

Discussion

Since the description of pachydermoperiostosis in 1868 by Friedrich in two young brothers, the disease has been better understood but is still uncommon. In 1907, Unna described marked thickening of the skin of the forehead and its resemblance to the sulci and gyri of the brain and called it ‘cutis verticis gyrata’.⁴ The association of cutis verticis gyrata and pachydermoperiostosis is sometimes referred to as Tourane-Solente-Gole syndrome.⁵ While the disorder is inherited as an autosomal dominant trait with
variable expression (1/3rd of patients have a positive family history), there was no family history in our patient.

Pachydermoperiostosis has variable clinical presentation with respect to skin and bone changes. The various clinical expressions include (1) the complete form (pachydermia, periostitis, cutis verticis gyrata), (2) the incomplete form (absence of cutis verticis gyrata) and (3) the forme fruste (pachydermia with minimal or absent periostitis)\(^5\). Our patient presented with thickening of the skin over the face and forehead, hyperhidrosis, clubbing of fingers and toes, which are classically described in pachydermoperiostosis. However, there were no associated symptoms of joint pain/polyarthritis. On investigation there was subperiosteal new bone formation of the radius, ulna, tibia, all metacarpals and metatarsals radiologically. No changes were seen in the articular cartilage.

*Brain magnetic resonance imaging (MRI) showed a pituitary microadenoma (Figure 3). However, on hormonal evaluation for growth hormone suppression test with 75 grams of anhydrous glucose given orally, human growth hormone (HGH) was suppressed to a level of less than 1 ng/mL, and insulin like growth factor-1 (IGF-1) was within normal range for the patient’s age and sex. Evaluation of gonadotropins, prolactin, thyroid hormone and cortisol hormone levels was normal. Thus, the finding of pituitary microadenoma in MRI was incidental. A primary hormonal evaluation of a patient with pachydermoperiostosis is perhaps necessary before performing a brain MRI, which can lead to diagnosis of acromegaly due to the presence of incidental pituitary abnormalities.*

*Acromegaloidism is characterized by an acromegaly appearance without any abnormal growth hormone function. Its association with a pituitary microadenoma is a rare disorder and should prompt the treating physician to consider acromegaloidism as a differential diagnosis with suppressed growth hormone following an oral glucose challenge and normal serum IGF-1 levels. Additionally, other causes like severe insulin resistance, pachydermoperiostosis, hypothroidism, Ascher’s syndrome, drug intake (e.g., minoxidil, phenytoin) must be...
ruled out before confirming the diagnosis.\textsuperscript{5,6}

The normal growth hormone levels reported by hormonal investigations ruled out acromegaly. The absence of osteal changes specific to pachydermoperiostosis pointed acromegaloidism as the most probable diagnosis. Our patient had some features of pachydermoperiostosis, acromegaly or thyroid acropachy.

In conclusion, though acromegaloidism is rare it has to be considered in the differential diagnosis of patients presenting with clinical features of acromegaly. The presence of a pituitary tumor may lead to a misdiagnosis of acromegaly, so in such cases biochemical evaluation is the key. Further pachydermoperiostosis-like dermatological presentation is also a likely symptom, which confounds diagnosis. Thus, this troika of closely related, yet distinct disorders must be carefully assessed for accurate diagnosis.

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References