

Short fourth and fifth metacarpals in a case of idiopathic primary hypoparathyroidism

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ABSTRACT

Shortening of metacarpals is a useful diagnostic marker in patients with pseudohypoparathyroidism type Ia (PHP-Ia) with Albright's hereditary osteodystrophy (AHO) phenotype or pseudopseudohypoparathyroidism (PPHP). There are very rare reports of metacarpals shortening in idiopathic primary hypoparathyroidism (IPH) cases in the literature. Here we described a young woman with IPH who presented with hypocalcaemia and generalized tonic-clonic seizure. She had shortening of fourth and fifth metacarpals which was prominent in her right hand. Based on our finding and other previous case reports we conclude that metacarpals shortening is not a specific finding of PHP-Ia or PPHP and it may be found in IPH cases.

Key words: Albright's hereditary osteodystrophy, hypocalcaemia, idiopathic primary hypoparathyroidism, pseudohypoparathyroidism, pseudopseudohypoparathyroidism

INTRODUCTION

Shortening of fourth and/or fifth metacarpal in a patient with hypocalcaemia and elevated parathyroid hormone (PTH) levels is a useful diagnostic clue for pseudohypoparathyroidism type Ia (PHP Ia) with Albright's hereditary osteodystrophy (AHO) phenotype.^[1,2] AHO patients have other diagnostic clinical signs including short stature, obesity, round face and mental retardation.^[3] IPH patients have similar biochemical findings as those PHP patients including hypocalcaemia and hyperphosphatemia, but they have PTH deficiency which is helpful in the making of definite diagnosis.^[4,5] As far as we are aware there are limited reports of idiopathic primary hypoparathyroidism (IPH) cases with short metacarpals in the literature.^[4] Here we report a rare case of idiopathic primary hypoparathyroidism who had short 4th and 5th

metacarpals bone and presented with tonic –clonic seizure in conjunction of hypocalcaemia and hyperphosphatemia.

CASE REPORT

A 22-year old woman was referred to endocrine clinic because of three episodes of generalized tonic-clonic seizure from one month ago. She was the product of consanguineous marriage. She was newly married and had no children. As far as she knew, family history was negative for similar disorder or other endocrine problems. She had normal regular menses. She was an oriented female with normal face appearance and short body stature (her height was 153 cm; her weight and BMI were 60 kg and 25.6 kg/m² respectively).

She had normal vital signs:

BP: 100/70 mm Hg, Temperature: 37°C, Pulse rate: 80/min and regular, Respiratory rate: 16/min.

Physical exam revealed shortening of fourth and fifth metacarpal in both hands with greater degree of fourth metacarpal involvement in right hand. The knuckles of her ring and little fingers looked depressed during clinching fist position [Figures 1a and b]. Hands

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DOI:
10.4103/2230-8210.117221

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Figure 1: (a) Photograph showing shortening of ring and little fingers (b) Depressed 4th and 5th fingers knuckles on the clenching fist position

movements and function were normal. Radiograph study of hands showed short fourth and fifth metacarpal in both hands [Figure 2]. Deep tendon reflexes were normal and plantar reflexes were flexor. Chvostek's and Trousseau's signs were negative at the time of admission to endocrine ward.

Laboratory results showing the following results: WBC: 7700/ μ l, hemoglobin (Hb):11.7g/dl, mean cell volume (MCV): 84 (fl), platelet (Plt):255,000/ μ l, blood urea: 19mg/dl, Cr: 0.8 mg/dl, blood sugar: 73 mg/dl, Potassium: 3.6 mEq/L, Sodium: 141 mEq/L, calcium:5.3-7.1 mg/dl (8.5-10.4), Ionized calcium: 3.88 mg/dl (4.6-5.3), serum phosphorus:6.4-7.3 mg/dl (2.8-4.5), serum Mg (magnesium): 2.20 mg/dl (1.8-2.6), serum ALP (serum Alkaline phosphatase):162 IU/L (64-306), Intact PTH: 17.91pg/ml (15-65), serum Albumin: 4.2 gr/dl (3.5-5.2). Liver function, urine analysis and thyroid function tests were all normal. Erythrocyte sedimentation rate (ESR) was 17 mm/hour and RF (Rheumatoid factor) was negative.

There were transient sharp epileptiform discharges in both frontocentral regions predominantly in right side in Electroencephalography (EEG) study.

CXR and ECG results were normal.

Brain magnetic resonance imaging (MRI) revealed multiple abnormal signal foci at subcortical white matter and there was no space occupying lesion.

There were nonspecific nonenhancing T2 and FLAIR high signal foci at left forceps major area in brain magnetic resonance imaging (MRI) with and without contrast.



Figure 2: Radiograph of hands showing short fourth and fifth metacarpals

We made the diagnosis of IPH and she was initiated on intravenous calcium infusion along with oral calcium tablets, calcitriol and also sodium valproate and after controlling of her hypocalcemia she was discharged to home and she was advised to return to the clinic for periodic monitoring with serum calcium and phosphorous levels and 24 hours urinary calcium collection.

DISCUSSION

Short metacarpals sign is a common finding in patients with pseudohypoparathyroidism type Ia or pseudopseudohypoparathyroidism (PPHP); but it had been rarely reported in patients with idiopathic primary hypoparathyroidism.^[1,2,4]

Shortening of metacarpals in conjunction of hypocalcaemic and increased serum PTH concentration is a useful clinical marker that suggests pseudohypoparathyroidism type Ia with phenotype of AHO diagnosis.^[3] Bilateral involvement of 4th and 5th metacarpals is the most usual form of disorder and these results from premature epiphyseal plates fusion of ring and little finger metacarpals.^[1] In our

patient with hypocalcaemia, hyperphosphatemia and low serum PTH levels, the diagnosis of idiopathic primary hypoparathyroidism was considered. We found short 4th and 5th metacarpals in this case. It was an interesting and unusual finding in an IPH case. Previously Isozaki and his colleagues reported a similar case of IPH with short metacarpals, round face and short stature.^[4] Based on our report and previous similar cases, we conclude that metacarpal shortening is not a specific sign for pseudohypoparathyroidism and its presence in a patient with low calcium levels could not differentiate between PHP Ia (AHO phenotypic variant) and IPH in the absence of biochemical test results.

CONCLUSION

Metacarpal sign is not a specific clinical marker of PHP Ia. It is not sufficient for making the diagnosis of PHP in a hypocalcaemic patient. Further clinical and biochemical

markers including serum PTH assessment is necessary for differentiating between PHP Ia and IPH.

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Cite this article as: Valizadeh N, Mehdizadeh A, Nazarbaghi S. Short fourth and fifth metacarpals in a case of idiopathic primary hypoparathyroidism. *Indian J Endocr Metab* 2013;17:924-6.

Source of Support: Nil, **Conflict of Interest:** None declared.