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Case Report

Internal Medicine Section

Idiopathic Hypoparathyroidism Presenting with Erythrodermic Psoriasis and Fahr's Syndrome: A Case Report

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ABSTRACT

Idiopathic hypoparathyroidism may remain asymptomatic or present with hypocalcaemic neuromuscular manifestations. Dermatological manifestations like exfoliative dermatitis, pustular psoriasis and symmetrical brain calcification known as Fahr's syndrome are uncommon in clinical practice. Even in absence of neuromuscular symptoms, hypocalcaemia associated with exfoliative skin lesions and symmetrical brain calcification should prompt investigation for hypoparathyroidism. A 40-year-old male presented with diffuse erythematous pruritic plaques and fever of 10 days duration. Auspitz sign was positive on examination. Investigations showed leucocytosis with neutrophilic predominance. He was admitted with a diagnosis of erythrodermic psoriasis with secondary infection. On the second day of admission he developed an episode of generalised tonic clonic seizure for the first time. Investigations into the cause of seizure revealed low serum calcium while computerised tomography of head revealed symmetrical calcification of bilateral basal ganglia. Further, he was found to have a low level of intact Parathyroid Hormone (PTH) level and hyperphosphataemia. Based on clinical, biochemical and radiological findings, a diagnosis of idiopathic hypoparathyroidism, psoriasis and Fahr's syndrome was made. Serum calcium was corrected with infusions of calcium gluconate followed by oral calcium carbonate. The patient was kept under observation for seizure recurrence. Calcium and vitamin D supplements and hydrochlorothiazide were started and psoriasis treatment was continued. His skin lesions resolved over the next two weeks, serum calcium levels returned to normal level and no seizure episodes were reported over three months of follow-up.

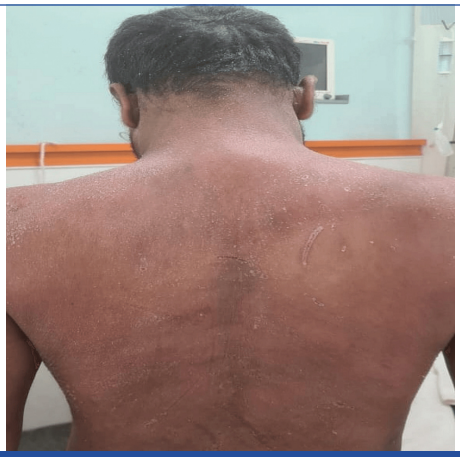
Keywords: Brain calcification, Dermatitis, Hypocalcaemia, Parathyroid hormone, Seizure

CASE REPORT

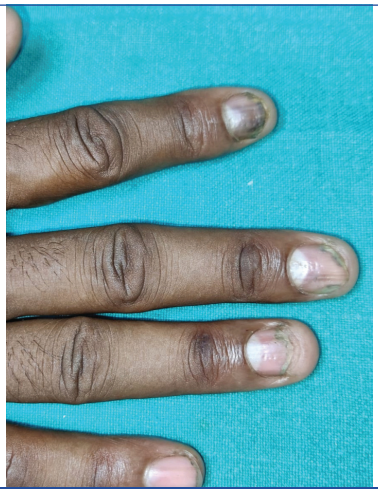
A 40-year-old male presented to the Dermatology Outpatient Department (OPD) with complaints of reddish, itchy eruptions over his entire body persisting for 10 days, and a fever for the past two days for which the patient had consumed over-the-counter anti-histaminic agents and applied topical medicines with no relief. The lesions started as reddish patches on the scalp and spread over the chest, abdomen, and all four limbs and were intensely itchy. Eight days later, the patient developed an intermittent fever of moderate grade which was not accompanied by chills. There was no history of drug intake, vaccination, trauma, diarrhoea, constipation, cough, or dysuria. A history of vague joint pain was present. The past and personal medical history was unremarkable. No similar family history was ascertained.

During the examination, the patient was conscious, oriented, and had normal behaviour. His pulse was 72 bpm, blood pressure was 118/70 mmHg, respiratory rate 18 bpm, and he was afebrile. The skin was dry with multiple erythematous, scaly plaques diffusely distributed over the chest, abdomen, scalp, back, and limbs, involving >90% of the body surface area {[Table/Fig-1] taken with consent}.

It was observed that the patient's fingernails were discoloured with horizontal grooves and onycholysis [Table/Fig-2]. The Auspitz sign was present with Psoriasis Area and Severity Index (PASI) of 36.3 [1]. Rest, general, and systemic examinations were normal. Complete Blood Count (CBC) in OPD showed Total Leucocyte Count (TLC) of 11600/cumm with 80% neutrophils. The patient was evaluated by the Dermatology Department and on the basis of diffuse erythema and papulosquamous eruption, onycholysis and Auspitz sign, he was admitted with a diagnosis of erythrodermic psoriasis and secondary infection. Treatment began with injections of Methotrexate 10 mg in 1 mL s/c weekly, oral Amoxycillin for secondary infection, cetirizine, ketoconazole and coal tar scalp wash, vitamin D and A supplements, coconut oil, folic acid, and acetaminophen. The



[Table/Fig-1]: Diffuse erythema, crusting, excoriation and pustules over the trunk.



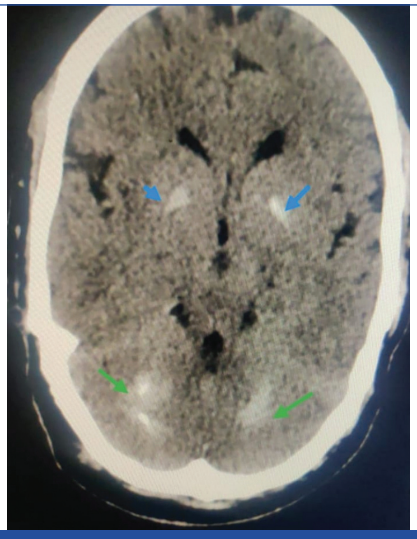
[Table/Fig-2]: Nail changes of psoriasis namely pitting, horizontal grooves and onycholysis.

patient became afebrile within 24 hours. On the second day after admission, the patient suddenly lost consciousness and developed tonic-clonic movements of all four limbs, accompanied by frothing from the mouth and urinary incontinence. The episode aborted spontaneously after about 2-3 minutes. Afterwards, the patient exhibited confusion. He was attended by the Emergency Medicine team. A diagnosis of Generalised Tonic-Clonic Seizure (GTCS) was made, and the patient was transferred to the medicine side. The patient had no history of similar episodes in the past. No other neurological deficits or signs of meningeal irritation were observed. On evaluation for the cause of GTCS, investigations were sent [Table/Fig-3].

Investigations	Results	Reference range
Haemoglobin	14.9 g/dL	Male 13-17 g/dL
Total Leucocyte Count (TLC)	8.9×1000/cumm	4-10×1000/cumm
Platelet count	130×1000/cumm	150-400×1000/cumm
Erythrocyte Sedimentation Rate (ESR)	35 mm/hr	0-20 mm/hr
Random Blood Sugar (RBS)	93 mg/dL	60-140 mg/dL
Blood urea	22 mg/dL	18-40 mg/dL
Serum creatinine	1.02 mg/dL	0.7-1.2 mg/dL
Serum sodium	140 mEq/L	135-145 mEq/L
Serum calcium	5.0 mg/dL	8.4-10.4 mg/dL
Serum magnesium	1.83 mg/dL	1.8-2.5 mg/dL
Serum phosphorous	5.1 mg/dL	2.4-4.6 mg/dL
Vitamin D	33.5 ng/mL	30-100 ng/mL
Anti-Nuclear Antibody (ANA)	0.97 u/mL	0-10 u/mL
Serum intact parathormone	3.32 pg/mL	12-88 pg/mL
24-hour urinary calcium	88.4 mg/day	0-300 mg/day
24-hour urinary phosphorus	255.0 mg/day	400-1300 mg/day

[Table/Fig-3]: Blood investigations of psoriasis patient with seizure.

In view of the severe hypocalcaemia, the patient was given i.v. infusion of calcium gluconate (10 mg) in 10 mL of 0.9% saline for 10 minutes, and his serum calcium was monitored for 12 hours a day. He was given 1g of Levetiracetam and 500 mg in 100 mL Normal Saline (NS) i.v. infusion every eight hours. An i.v. injection of 1g Ceftriaxone was given every 12 hours. Fever investigations were conducted and results were negative for malaria antigen and dengue screen and scrub Immunoglobulin M (IgM). Urine and blood cultures were reported sterile. The patient was seizure-free and non-febrile. The Non Contrast Computed Tomography (NCCT) brain scan revealed hyperdensities in the left basal ganglia, right thalamus, and bilateral cerebellar hemisphere, suggesting symmetrical calcification [Table/Fig-4]. Further, 24 hours of urinary calcium and phosphorus reports indicated that renal loss was not the cause of severe hypocalcaemia. The screening test for Human Immunodeficiency Virus (HIV) was negative.



[Table/Fig-4]: Axial sections of computed tomography of head showing calcification in bilateral basal ganglia-globus pallidus (blue arrows) and dentate nuclei (green arrows).

In view of typical skin lesions, hypocalcaemia with low PTH and symmetrical brain calcification, a diagnosis of idiopathic hypoparathyroidism associated with erythrodermic psoriasis and Fahr's syndrome was made. Antibiotic Ceftriaxone 1 g twice a day for seven days, calcium supplementation with injection calcium gluconate 10 mg under close monitoring was followed-up after two days with oral calcium carbonate, hydrochlorothiazide 12.5 mg/day, oral cholecalciferol 250 IU and supportive treatment were continued in addition to psoriasis management. The patient remained seizure-free. The psoriasis lesions and hypocalcaemia resolved over the next three weeks. He was discharged from the hospital with a treatment regimen of 1250 mg of calcium carbonate (500 mg of elemental calcium), and 250 IU of cholecalciferol daily, and weekly oral Methotrexate 7.5 mg and folic acid 5 mg. In follow-up of three months there was no recurrence of seizures and there was progressive improvement in skin lesions. A repeat NCCT of the brain was planned for follow-up to watch for regression of brain calcification but was declined by the patient due to financial constraints and asymptomatic status.

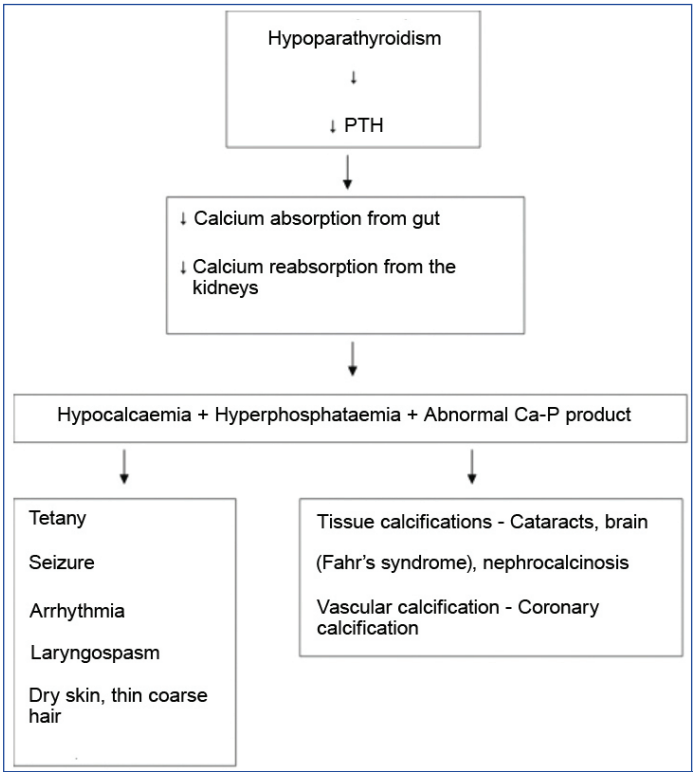
DISCUSSION

In adults, hypoparathyroidism has an incidence of 37/100,000 and is commonly caused by accidental surgical manipulation or removal during thyroid surgery or radical neck dissection. Rarely, it may occur due to autoimmune destruction secondary to polyglandular syndrome or activating antibodies to the calcium-sensing receptor, radiation, HIV, and infiltration. The absence of an identifiable cause results in a diagnosis of idiopathic hypoparathyroidism [2-4]. The clinical features of hypoparathyroidism result from low PTH causing low serum calcium, hyperphosphataemia and an increased calcium-phosphate product. Hypocalcaemia commonly presents with seizures, neuromuscular instability, laryngospasm, prolonged QT interval, refractory heart failure and skin changes like dryness and itching. hyperphosphataemia and increased calcium-phosphate product results in soft-tissue calcification. Cutaneous manifestations of hypoparathyroidism are more common in chronic hypoparathyroidism and include dryness, exfoliative dermatitis, itchiness, changes in the nails, and cataracts [4].

Psoriasis is a chronic inflammatory dermatosis which involves a genetic predisposition and autoimmune activation causing proliferation and dysfunctional differentiation of keratinocytes. Erythrodermic psoriasis is an acutely developing subtype characterised by erythematous inflammation of more than 90% of body surface area that may require emergency management. Proliferation and differentiation of keratinocytes is dependent on normal intracellular calcium levels and psoriasis lesions are known to flare up in presence of low serum calcium. Hypocalcaemia resulting from hypoparathyroidism plays a central role in the development of psoriasis. Intercellular junctional components in the dermis called cadherins are dependent on calcium, which is essential in keratinocyte proliferation and differentiation. Hypocalcaemia causes acantholysis, which recovers when calcium levels are restored. Treatment of hypocalcaemia with calcium replacements and vitamin D supplements improves manifestations in the skin [5-12].

In the present case, there was an acute development of psoriasis with diffuse erythema and was thus called erythrodermic psoriasis in contrast to previously reported pustular psoriasis or plaque psoriasis [6-13]. Brain calcification syndrome, or Fahr's syndrome, is a rare idiopathic neurodegenerative condition first described by Karl Theodor Fahr in 1930. It is characterised by the deposition of calcium phosphate and calcium carbonate symmetrically in vessel walls and perivascular space in the basal ganglia and cerebral cortex. It presents clinically with progressive neurological deficits like

Parkinsonism, seizures, and neuropsychiatric symptoms [4]. Fahr's syndrome or secondary brain calcification is known to be associated with hypoparathyroidism and pseudohypoparathyroidism. The incidence of symmetrical calcification of the basal ganglia is 0.49% in the general population, whereas it is reported to be as high as 12-74% in patients with hypoparathyroidism. In this condition, brain calcification is due to an abnormal calcium/phosphorus ratio, which results in the deposition of a calcium-phosphate complex in vessel walls and perivascular space and extends to the basal ganglia [Table/Fig-5] [14-18].



[Table/Fig-5]: Association of hypoparathyroidism with psoriasis and tissue calcification [14-18].

Despite the abovementioned cases of cutaneous manifestations and brain calcification described in the literature, there was no case of hypoparathyroidism with a clinical presentation of both psoriasis and Fahr's syndrome coexisting in the same patient, thereby making the presented case unique.

CONCLUSION(S)

The authors present a rare case of idiopathic hypoparathyroidism presenting with erythrodermic psoriasis, hypocalcaemic seizures and Fahr's syndrome which is the first such case to be reported. The authors concluded that serum calcium levels should be checked in cases of psoriasis and other exfoliative skin conditions for the timely diagnosis of hypoparathyroidism and its treatment for prevention of life threatening manifestations of hypocalcaemia.

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