Pamidronate in Treatment of Calcinosis in Juvenile Dermatomyositis

Juvenile dermatomyositis is a rare systemic autoimmune disease wth calcinosis as its hallmark sequelae. We report three patients with juvenile dermatomyositis with calcinosis, who were treated with pamidronate. There was complete clearance of calcinosis in one child.

Keywords: *Management, Sequalae.*

Juvenile dermatomyositis (JDM) is an inflammatory disease of the muscle, skin and blood vessels with peak age of onset 5-14 years and female:male ratio 1.7:1 [1,2]. Calcinosis is a hallmark sequelae [3]. Pamidronate has been used earlier for this indication [4] but there is lack of reported experience in Indian set-up.

A 9-year-old boy presented to us with complaints of fever, pain in all limbs and difficulty in walking for one and half months. Child was initially treated with non-steroidal anti-inflammatory agents (NSAIDS) after which pain had initially subsided to recur again. On examination, there was peripheral myopathy, heliotrope rash and Gottron papules. Laboratory investigations showed hemoglobin 7.7 g/dL, elevated lactate dehydrogenase (399 U/L) and creatinine phosphokinase (321 ug/L), and normal electromyogram. A muscle biopsy from Vastus lateralis showed features of immune-mediated inflammatory myopathy. Child was diagnosed as having JDM, and was started on steroids (20 mg/day), hydroxychloroquine (100 mg/day) and methotrexate (7.5 mg once a week). The child improved over the next 3 years with good compliance and regular follow-up, and gradually steroids were tapered to 5 mg/day. He presented after fall from a bicycle with swelling of the little finger with chalky white discharge for 3 days (***Fig.* 1a)**. *X*-ray revealed calcium deposits in soft tissue of little finger (***Fig.* 1b)**. Pamidronate was infused at 1 mg/kg/day for 3 consecutive days every 3 months. On follow-up after one year, there was complete clearance of calcinosis of fingers without any new focus and good disease control without signs of myositis (***Fig.* 1c** and **d**).

An 11-year-old girl presented with complaints of swelling over right elbow and bilateral buttocks. She was diagnosed as having JDM at 8 years of age. She was initially treated with steroids, hydroxychloroquine and NSAIDS but had poor compliance to medicines. Following a fall from the stairs, she started developing calcinosis of buttocks followed by calcinosis of right elbow. There was presence of heliotrope rash and Gower sign. *X*-ray showed calcium deposits on affected areas. Child was started on 3-monthly pamidronate infusion after which there was significant decrease in calcinosis with no fresh foci. Complete resolution of disease process was not observed (***Web Fig.* 1a)**, but the compliance to drugs was also not optimal.

A 7-year-old girl presented with complaints of multiple swellings over the body and difficulty in walking for one year. The first swelling appeared in the waist region, followed by swellings in bilateral chest walls and scalp (***Web Fig.* 2 a)**. The swelling on the chest was excised by a local physician mistaking it to be an abscess. Following this, the child developed more swellings in the lateral chest near the site of excision (***Web Fig.* 2 b)**. On examination, there were nodules on left anterior chest wall with scar marks in bilateral infra-axillary area, Gottron papules and proximal myopathy. Laboratory investigations showed anemia with raised LDH and CPK. Hip *X*-Ray showed white nodular opacity around hip joint, suggestive of calcinosis (***Web Fig.* 2 c** and **d)**. Electromyogram showed membrane instability and fiber destruction. Child was diagnosed as a case of JDM with calcinosis and was started on steroids, hydroxychloroquine, methotrexate, folic acid and 3-monthly pamidronate infusion. Follow-up after 1 year showed significant decrease in scalp swelling with complete disappearance of swelling over chest wall and the waist region with no new calcinosis**.**

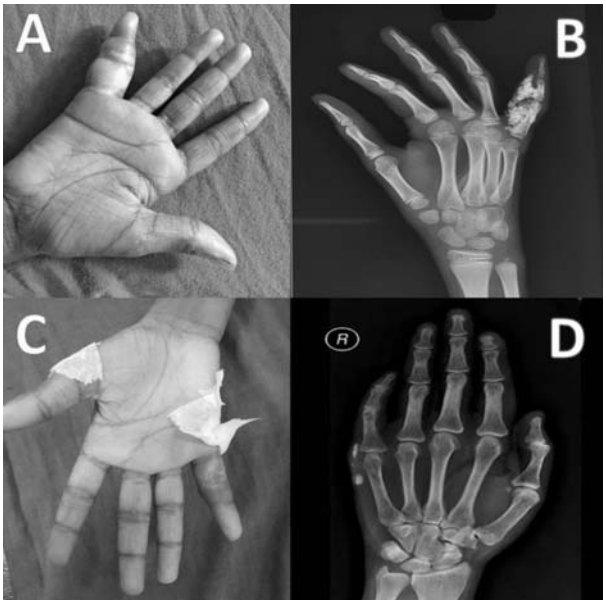

FIG. 1 *(a) Calcinosis in the little finger before treatment; (b) X-ray hand showing calcinosis before treatment; (c) clinical resolution of calcinosis after pamidronate; and (d) radiological resolution after pamidronate*

Calcinosis is a hallmark sequelae of JDM [1]. Alum, alendronate, diltiazem and rituximab are few drugs used for treatment of calcinosis [3]. Pamidronate is a nitrogen-containing bisphosphonate which inhibits bone resorption used to treat osteoporosis [4]. Although the mechanism of action of pamidronate is unclear, it was chosen based on available adult studies [1,4,6]. A significant decrease in calcinosis was found in two cases whereas there was complete clearance in one case. Aggressive treatment with disease modifying anti-inflammatory agents (DMARDs) early in the course of disease seem to be effective in good disease control as was evident from case 1 and 3. Prompt diagnosis and early intervention prevents further calcinosis. Our results suggest that treatment with pamidronate infusion may achieve good disease control in prevention of further calcinosis in JDM.

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Electrical Injury Causing Facial Nerve Palsy in a Toddler

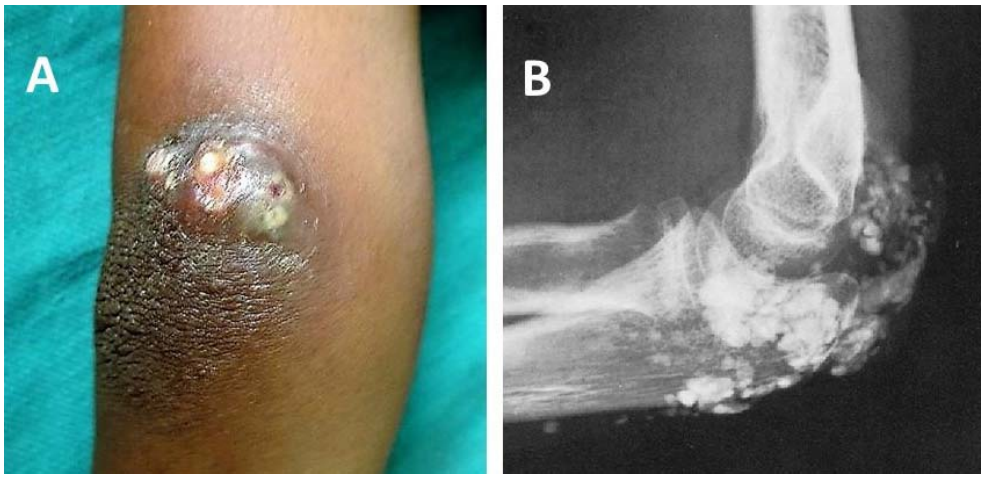
Although electrical injuries are one of the common injuries encountered in clinical practice, low voltage electrical injuries presenting as focal neurological deficits are rare. We report the case of a 3-year-old boy who presented with right facial palsy and hemotympanum after electrical injury.

Keywords: *Childhood injuries, Electric shock, Paralysis.*

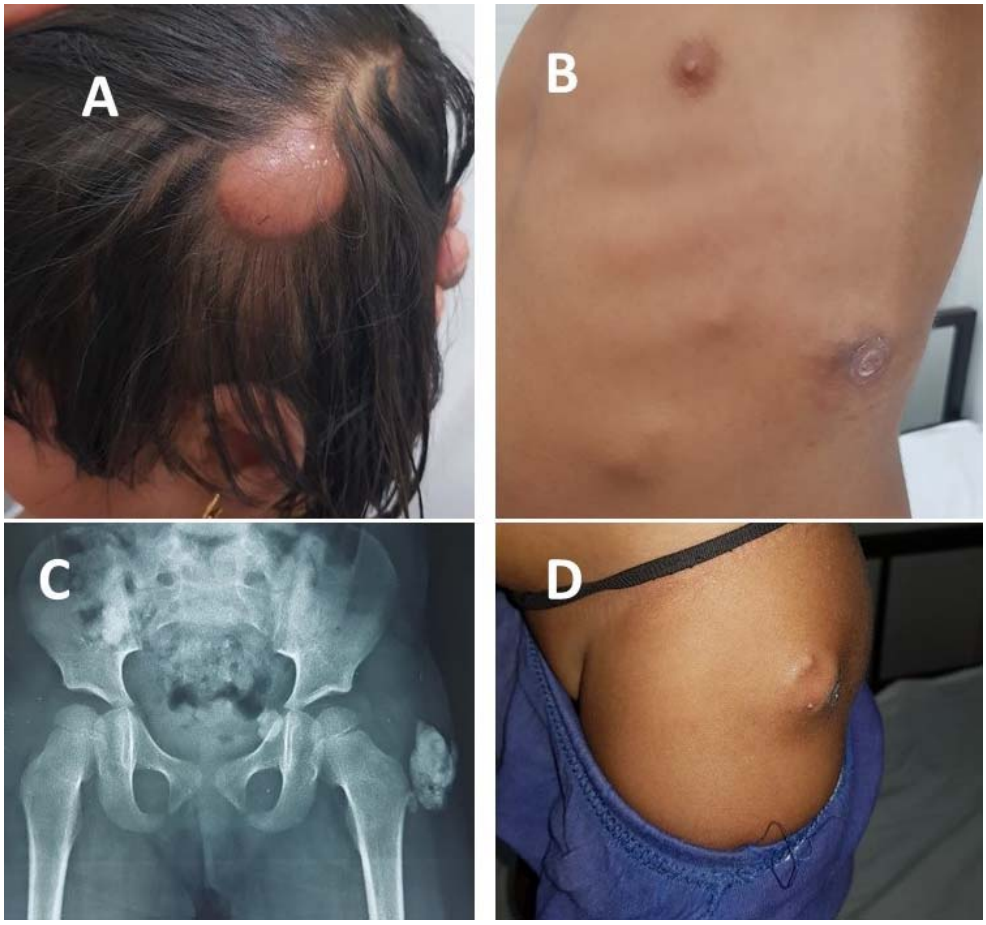
The curiosity of children to explore new things increases their risk of childhood accidents [1,2]. The most common sources for electrical injuries in children are electric sockets, faulty appliances, and live wires [3].

A 3-year-old boy was brought to us for consultation with deviation of angle of the mouth and inability to close his right eye for a day. Neurological examination showed deviation of angle of mouth to left side, absence of wrinkling of forehead on the right side, and incomplete closure of the right eye, with rest of the central nerve system and systemic examination was normal. Initially, idiopathic Bell palsy was considered as the diagnosis, but when a detailed history was elicited, a history of electric injury 1-day back was revealed which was thought to be irrelevant by the parents and hence was not initially revealed by them. The child did not have any history of trauma to the right ear or face, and there were no symptoms suggesting infection of right ear. Otoscopic examination revealed the presence of reddish-blue ear drum suggesting hemotympanum. Blood cell counts, creatinine kinase, urine analysis, renal and liver function tests, and electrocardiogram were within normal limits. Prothrombin time, activated partial thromboplastin time, bleeding time and clotting time were also found to be within normal limits.

The child was diagnosed as having Grade IV of House Brackmann lower motor neuron (LMN) type facial nerve palsy of right side with hemotympanum due to low-voltage electric current injury. Patient was started on low-dose oral steroids, eye lubricant and eye bandage to prevent exposure keratitis. The child was discharged after 3 days on low-dose oral steroids. On reviewing after one week, there was improvement of the facial palsy (grade 3). At follow-up, almost three months following the incident, the child had fully recovered with no residual facial nerve palsy or hemotympanum.



WEB FIG. 1 *(a) Calcinosis in the right elbow; and (b) X-ray elbow showing calcinosis before treatment.*



WEB FIG. 2 *(a) Scalp calcinosis; (b) multiple nodular swelling in lateral chest with scar mark; (c) X-ray pelvis with calcinosis; and (d) calcinosis in lateral aspect of the hip.*