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Case report

Soft-tissue necrosis complicating tibial osteotomy in a child with Proteus syndrome

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ABSTRACT

Introduction: Proteus syndrome is a rare congenital hamartomatous disease frequently responsible for musculoskeletal deformities. The results and complications of surgical treatment are not well documented owing to the scarcity of reported cases.
Case report: The authors report a case of poor evolution of valgus proximal tibial osteotomy in a 6-year-old girl with Proteus syndrome. The surgery was complicated by extensive deep wound necrosis exposing the tibial bone, necessitating surgical excision, antibiotherapy and controlled wound healing. At 1 year postoperatively, the deformity recurred.
Discussion: The possibility of serious wound complications and of recurrence must be kept in mind when operating on a limb deformity in patients with Proteus syndrome. Potential complications should be taken into account in selecting the surgical correction technique: epiphysiodesis may be preferable to osteotomy.

1. Introduction

Proteus syndrome (PS) is a very rare congenital hamartomatous pathology featuring continuing excessive development of various body parts [1], including severe musculoskeletal deformities and progressive deterioration in the limbs and axial skeleton [2,3]. Surgery is frequently recommended to correct musculoskeletal deformities, and may consist in epiphysiodesis or osteotomy [4]; however, due to the rarity of PS, results and complications are poorly known.

We report a case of poor evolution of valgus tibial osteotomy in a 6-year-old girl with Proteus syndrome.

2. Case report

A 6-year-old girl with no particular familial history was referred with progressively worsening left genu varum. Interview revealed that a small left fronto-orbital protuberance and a skin blemish on the back had been observed at birth. Her parents further reported onset, at around 1 year of age, of a deformity of the left knee, an increase in the volume of the facial protuberance, onset of truncal deformity and digital deformity of slow evolution.

Clinical examination found craniofacial dysmorphism with dolichocephaly, lengthened neck, protruding ears and bilateral supraorbital protuberance (Fig. 1). Orthopedically, there was truncal imbalance toward the left. There was also macrodactyly of the second and third fingers of the left hand and clinodactyly inducing a forked aspect (Fig. 2). The second and third fingers of the right hand also showed slight macrodactyly.

Examination of the knees found left genu varum and moderate genu recurvatum (Fig. 3), without soft-tissue trophic disorder or vascular abnormality.

There were 3 angiomatous blemishes with long axes of a few centimeters on the back and abdomen.

Plain spinal X-ray found asymmetric enlargement of the vertebral bodies and pedicles (Fig. 4), predominating on the 12th lumbar vertebra and inducing dorsolumbar scoliosis with severe thoracic left-convexity curvature (Fig. 4). There was also hypertrophy of the cervical vertebral spinous processes.

Lower-limb X-ray showed a dystrophic left proximal tibial growth plate (Fig. 5).

Exploration was completed by CT, which found frontal bone hypertrophy corresponding to the facial protuberances. Given the association of general and specific criteria as defined by Bieseker et al. [5], PS was diagnosed.

Left proximal tibial valgus osteotomy was indicated and performed under pneumatic tourniquet. The superior tibial approach was anterolateral. While performing the approach, we were astonished by the poorly vascularized aspect of the soft tissue and bone. The lateral closing-wedge valgus osteotomy was stabilized using 2 K-wires. Soft-tissue closure was performed without tension and the lower limb was immobilized in a long-leg cast.

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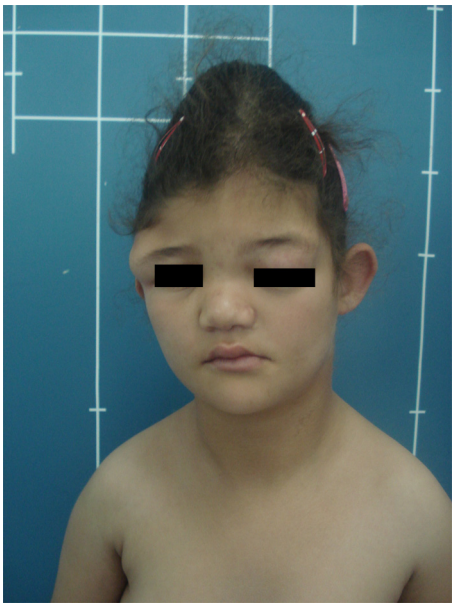

Fig. 1. Frontal photograph showing supraorbital osseous protuberances, more pronounced on the left side (hunched skull), and dolichocephaly.

Postoperative anticoagulation therapy was initiated due to the risk of thromboembolitic complications in PS [6]. Immediate postoperative course was simple and the patient was discharged at 2 days. Three weeks later, she came to emergency with sharp pain in the operative site and a sweating aspect in the cast. The cast was removed, to reveal extensive soft-tissue necrosis in the anterior side of the limb, exposing the bone (Fig. 6). Emergency surgical resection of the necrotic tissue and infected bone was associated to stabilization of the osteotomy site by a circular external fixator with general route antibiotics. The wound received local treatment without soft-tissue closure. Healing was controlled by iterative flat dressing. Evolution toward osteotomy site cover was very gradual, over a period of 3 months, leaving stiff, adherent, umbilicated skin and persistent cutaneous fistula. The tibial osteotomy consolidated, with recurrence of tibial deformity at 1 year.

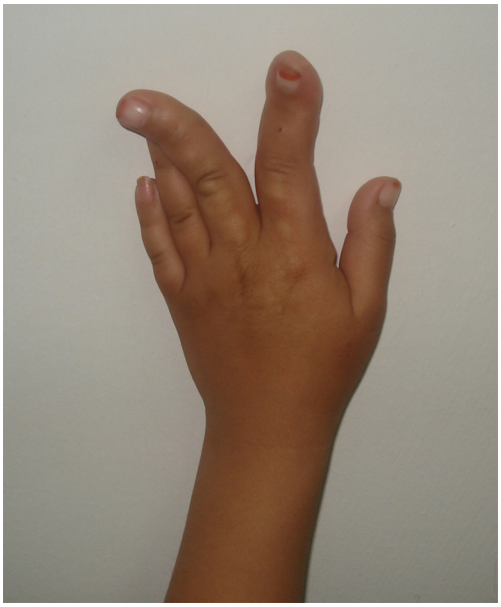

Fig. 2. Macrodactyly of the forefinger and the middle finger (forked aspect).

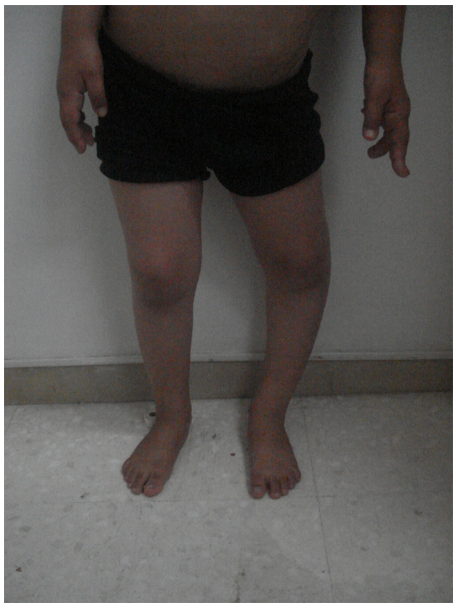

Fig. 3. Left genu varum.

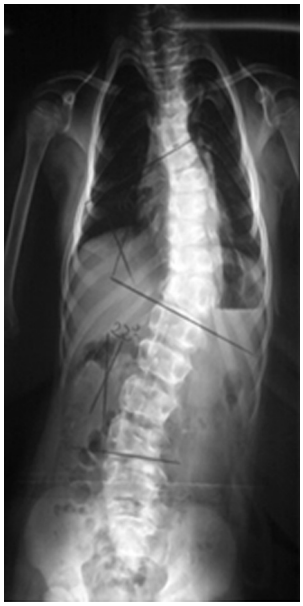

Fig. 4. Asymmetric aspect of pedicles and vertebral bodies, especially on the twelfth dorsal vertebra, causing thoracolumbar scoliosis with leftward convexity.

3. Discussion

PS involves deformity by excessive asymmetric growth of various tissues: osseous, conjunctive and adipose [1]. The most famous case of PS was John Merrick, also known as the Elephant Man, who was long mistakenly thought to have had neurofibromatosis [7]. Temtamy and Rogers first described PS in 1976 [8], and Wiedman et al. [9] coined the name “Proteus syndrome”, for the Greek god able to change shape to escape his enemies.

PS is due to a somatic mosaic induced by mutation of the oncogene AKT1 that controls cell proliferation [10], activating growth in cells including the mutation, resulting in tissue proliferation.

It is a rare condition. Turner et al. [1], in a systematic review in 2004, retrieved 205 cases in the international literature, only 97 of which were true PS on the diagnostic criteria of Bieseker. This highlights the rarity and diagnostic difficulty of PS. Diagnosis