

Recurrent Panniculitis in an Adolescent Boy with Prader-Willi Syndrome

Alexander K.C. Leung, MBBS, FRCPC, FRCP (UK & Irel), FRCPCH, FAAP and
William Lane M. Robson, MD, FRCPC, FRCP (Glasg)
Calgary, Alberta, Canada

We describe recurrent panniculitis in a 15-year-old boy with Prader-Willi syndrome. To our knowledge, this association has not been previously reported.

Key words: Prader-Willi syndrome ■ panniculitis

© 2006. From the University of Calgary (Leung, clinical associate professor of pediatrics) and The Childrens' Clinic (Robson, medical director), Calgary, Alberta, Canada. Send correspondence and reprint requests for *J Natl Med Assoc.* 2006;98:1700-1701 to: Dr. Alexander K.C. Leung, #200, 233 16th Ave. NW, Calgary, Alberta T2M 0H5; fax: (403) 230-3322; e-mail: aleung@ucalgary.ca

INTRODUCTION

Panniculitis refers to inflammation of the subcutaneous fat. The disorder usually manifests as a tender, subcutaneous nodule covered by erythematous skin.¹ We report a case of recurrent panniculitis in a 15-year-old boy with Prader-Willi syndrome. This association has not been previously reported.

CASE REPORT

A 15-year-old Chinese boy presented with a two-day history of four painful red bumps on his lower back. Four and 12 months previously, similar skin lesions had appeared on the legs and on the back, respectively. The prior lesions were tender to touch and resolved spontaneously in 2-3 weeks. He received a monthly injection of intramuscular testosterone, but the sites of the injections did not correspond with the locations of the lesions. There was no history of preceding sore throat, fever or trauma.

There was concern about decreased fetal movement in the third trimester of pregnancy, but the pregnancy was otherwise uncomplicated. He was born at 39 weeks' gestation with a birthweight of 2.8 kg and a length of 49 cm. In the neonatal period, he was observed to have a weak cry, hypotonia, micropenis, hypoplastic scrotum and undescended testicles. Gross motor milestones for sitting, crawling, walking and running were delayed, as was speech development. A left orchidopexy was performed at 2 years of age. The right testicle was found to be atrophic and was removed. A voracious

appetite developed as a toddler. Chromosomal analysis showed a classical deletion of 15q11, and a diagnosis of Prader-Willi syndrome was established.

On examination, his height was 151 cm (4 cm below the fifth percentile) and weight 86 kg (6 kg above the 95th percentile). His pulse was 76/min and blood pressure 110/70 mmHg. Four subcutaneous nodules were noted on the lower back. The nodules measured 1-2 cm in diameter and were covered by erythematous skin (Figure 1). The lesions were firm and tender. He also had acanthosis nigricans that involved the nape of the neck and the axillae. He had thoracic kyphoscoliosis, microdontia, enamel hypoplasia, high-arched palate, keratosis pilaris, and small hands and feet. He had Tanner stage-3 genitalia and pubic hair. The rest of the examination was normal.

His hemoglobin was 142 g/L, white blood count 69 x 10⁹/L with a normal differential count, and platelet count 287 x 10⁹/L. His serum electrolytes, glucose, creatinine, cholesterol and triglycerides were normal. A clinical diagnosis of panniculitis was established. The lesions resolved spontaneously in 10 days without any specific treatment.

DISCUSSION

Panniculitis might be primary or secondary and associated with a variety of clinical conditions. Panniculitis has been reported with subcutaneous fat necrosis of the newborn, streptococcal infection, tuberculosis, connective tissue diseases, pancreatic diseases, gout, α_1 -antitrypsin deficiency, immunodeficiency, malignancy, prolonged cold exposure, physical trauma, subcutaneous injection of an oily material and systemic high-dose corticosteroid administration.¹⁻⁵

Bukhari reported an eight-month-old Egyptian girl with Niemann-Pick disease type C who had recurrent panniculitis.⁶ The cause of the panniculitis was not known, and the author suggested that panniculitis should be considered a cutaneous manifestation of Niemann-Pick disease type C.

Prader-Willi syndrome is associated with an abnormality or deletion of the paternal chromosome 15q11-13.^{7,8}

Figure 1. An erythematous subcutaneous nodule on the lower back



Although Prader-Willi syndrome is an autosomal dominant disorder, most cases are sporadic and arise from a de novo mutation.^{7,9} The syndrome is characterized by hypotonia, hypomentality, hypogonadism and obesity, sometimes referred to as the H₃O syndrome.⁹ Other manifestations include reduced fetal activity, infantile hypotonia, short stature, microdentia, enamel hypoplasia, high-arched palate, small hands and feet, scoliosis, kyphosis, and compulsive snacking and skin-picking.^{7,8,10} Many of the manifestations of Prader-Willi syndrome can be attributed to hypothalamic dysfunction. Potential complications in patients with Prader-Willi syndrome include behavioral problems, dental caries, diabetes mellitus, hypertension, atherosclerosis, joint contracture, osteoporosis, glomerulosclerosis, and development of a Pickwickian or obesity-hypoventilation syndrome.^{8,9} Physical trauma to the fatty tissue might lead to panniculitis.^{1,11} Patients with Prader-Willi syndrome are morbidly obese and are compulsive skin-pickers, which might increase their risk for panniculitis. With report of this case, it is hoped that more confirmatory case reports would be forthcoming.

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The University of Chicago

Section of General Pediatrics

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