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Dactylyosis espontânea (ainhum)

Dear Editor,

A 76-year-old, white woman presenting with bone resorption in the fifth toes. For three years, the patient had experienced severe pain and local edema. Conventional radiography (Figure 1) demonstrated narrowing and osteolysis of the middle and distal phalanges of the fifth toes, most noticeable at left, in association with focal and concentric decrease in the soft parts thickness at the roots of those toes. Because of the intense local pain, the patient underwent surgical amputation of the fifth toes, and the symptoms disappeared.

Several spontaneously painful conditions of lower and upper limbs, particularly in their extremities, have been observed and reported in Brazil(1–5).

Dactylyosis spontanea (DS), that is also known as ‘ainhum’, is a rare disease occurring principally in Afrod descendant male individuals (2:1) aged between 30 and 50(6). “Ainhum” is an Angolan word meaning “to saw”. The first case report of DS in Brazil refers to a “quilombola” in Bahia state and was described by Silva Lima in 1867(7). The prevalence of DS ranges from 0.015 to 2% of the population in African countries. In Brazil, its prevalence is still to be studied.

Few reports of cases of DS in white individuals are found in the literature(8). In Brazil, due to ethnic miscegenation, a higher incidence of this disease may be observed in white individuals with some African ancestry that is not always noticeable in their phenotype.

The main DS feature of this condition is the development of a fibrotic constriction ring involving the base of one or more toes, conditioning eversion and absorption of distal structures, possibly progressing to spontaneous amputation(9). Recently a case of DS involving toes and fingers was reported(9).

The radiographic findings are typical and may be classified into four phases, as follows: the first one is characterized by the development of a deep sulcus along the medial aspect of the distal portion of the proximal phalanx, sometimes resembling the shape of a sand glass. The second phase progresses with increase in the volume distally to the constriction ring, secondary to lymphedema. The third phase is characterized by progressive bone absorption, and the fourth phase, by spontaneous amputation averagely occurring within four to six years after the disease onset(9).

The differential diagnosis should be made with other conditions involving the development of fibrotic constriction rings such as porokeratosis of Mibelli; erythropoietic protoporphyria; scleroderma; psoriasis; neuropathic plica; hanseniasis; syphilis; Raynaud’s disease; diabetes mellitus and syringomyelia. Also, factitious pseudoainhum, caused by hair-thread tourniquet syn drome should be considered(9).

No well established treatment for DS is available yet. Resection of the sulcus followed by z-plasty may alleviate the pain and avoid spontaneous amputation at early stages of the disease(6).

Surgical amputation may be recommended to alleviate the symptoms(10).

DS is a rare disease whose diagnosis is hampered by its low prevalence and variable clinical presentation. The radiological evaluation allows for an early diagnosis, preventing spontaneous amputation.

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Uncommon presentation of perforated Meckel’s diverticulum in preterm newborn

Divertículo de Meckel perfurado como causa incomum de pneumoperitônio em recém-nascido pré-termo

Dear Editor,

A male neonate with gestational age of 30 weeks, weighing 940 g at birth, with respiratory failure right after birth, and radiological signs compatible with hyaline membrane disease. At his tenth day of life, the patient presented vomiting and abdominal distention, presenting with radiological signs of pneumoperitoneum (Figure 1).

Initially, the neonate was submitted to peritoneal drainage, due to the lack of surgical conditions, and at the 19th day, after gaining weight and present with hemodynamically stable conditions, was submitted to exploratory laparotomy. During the surgery, a Meckel’s diverticulum (MD) was found, with jejunal perforation, hepatic blockage and obstruction distal to the blockage due to the development of adherence. Resection of about 6 cm of the jejunal loop including the perforated area was performed, with later termino-terminal anastomosis. The anatomopathological result was subacute diverticulitis with ulcer and severe peridiverticulitis. The neonate presented a favorable evolution and was discharged at his 82nd day of life.

Meckel’s diverticulum represents the most common congenital malformation of the digestive tube, and is asymptomatic in most cases (1–3). Symptomatic cases of MD are rarely found, affecting less than 20% of all pediatric cases (1).

Figure 1. A: Chest and abdominal radiography – Image acquired with the patients in supine position, with vertical x-rays, demonstrating hypertransparent abdominal cavity due to accumulation of free air. B: Chest and abdominal radiography – Image acquired with the patient in supine position with horizontal x-rays, demonstrating the free air collection located between the anterior abdominal wall and the bowel loops.