The immunohistochemistry was done and was positive for α-smooth muscle actin and S-100 was negative.

Kloepfer *et al.*[3] first described an Italian family with cutaneous leiomyomatosis, and autosomal dominant inheritance pattern with incomplete penetrance. Reed *et al.*[4] emphasized the association with leiomyoma of uterus and, therefore, this syndrome was known as Reed's syndrome. Launonen *et al.*[5] reported that a small proportion of families with MCUL also cluster renal cell cancer (RCC). Tomlinson *et al.*[6] showed that germline mutations in the fumarate hydratase (FH) gene underlie hereditary cutaneous leiomyomatosis.

The largest reported genital leiomyoma till date is of the size 15 cms in largest diameter.[7] Thus, we speculate them to be the biggest reported size in the world to the authors' best of knowledge.

Recent studies have demonstrated the involvement of a classical tumour suppressor gene encoding fumarate hydratase (FH). Early studies of two dominantly transmitted Mandelian syndromes, multiple cutaneous and uterine leiomyomatosis (MCUL1; OMIM 150800) and hereditary leiomyomatosis and renal cell cancer (HLRCC; OMIM 605839) have implicated a subregion on chromosome 1q43 that contains FH, a gene encoding the tricarboxylic acid cycle (Krebs cycle) fumarate hydratase enzyme.[8]

Kubinova *et al.*[9] suggested fumarate hydratase activity or FH mutation screening in women with onset of uterine fibroids in their 20s and family history of uterine fibroids or other HLRCC-associated malignancies. In our patients, the genetic and fumarate hydratase testing were declined by the family.

The differential diagnoses are eruptive syringomas, neurofibromas, neurinoma or eccrine spiradenomas. Being benign condition, the avoidance of the precipitating factors such as pain, cold and emotional stress may be the only treatment required.[10]

Amlodipine, nifedipine, hyoscine butyl bromide, gabapentin and doxazosin and CO_2 lasers, Cryosurgery are other modalities of treatment.

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Paediatric anogenital Crohn's disease 11 years after Lichen planus – same background for two different entities?

Editor

Inflammatory bowel disease has a multiplicity of cutaneous manifestations, which are frequent and can be present in about 40% of patients. Cutaneous Crohn's disease (CCD) also called metastatic CD refers to non-caseating granulomatous lesions that are not contiguous with intestinal CD.

There are less than 30 cases described in children.

Lichen planus is an inflammatory papulosquamous disease, and while there are some well-known associations such as Hepatitis C, metal allergy and vaccination induction its aetiology remains largely unknown. It is rare in the paediatric setting with an estimate prevalence of 1–4%[1] and although it has been associated with personal and familial autoimmune history, this finding is not constant through the series.

We report the case of a 12-year-old female patient observed eleven years before for a lichen planus (LP) lesions on the hands, feet and oral mucosa confirmed by histopathological features (Fig. 1). All the lesions were successfully treated with the exception of the lesion of the hard palate that remained not completely healed.

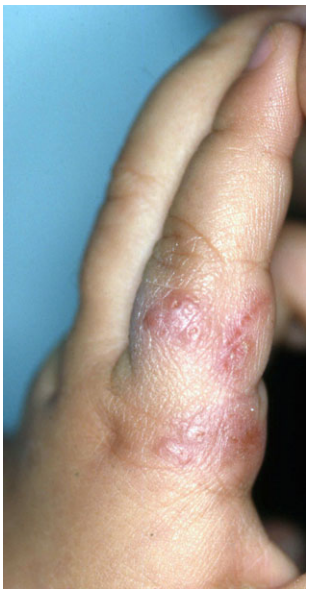

Figure 1 Poliedric erythemato-violaceous papules on the side of the finger.

This patient was again referred to our consultation due to a postexcision surgical wound of pilonidal cyst of difficult healing, denying any bowel symptoms. She had a 16 year-old sister with type 1 diabetes mellitus.

On physical examination, an ulcer was observed in the intergluteal sulcus, with 4,5 cm of length and pediculated lesions around the anus (Fig. 2). In the vulvar area, a whitish exudate was present and multiple small fissures were observed around the clitoris. In the hard palate a violaceous patch was present. Multiple biopsies were taken and the surgical specimen was reviewed, the features being unspecific. In the perianal lesions inflammatory tissue with fistulous and epithelioid granulomas was compatible with inflammatory bowel disease. Colonoscopy and ileoscopy were normal.

Immunoglobulins G,A and M were raised. C-Anca was negative. HLA typing was DRB1*04*15; DQB1*03*06.

Treatment was initiated with prednisolone, azathioprine, ciprofloxacin and metronidazole with an initial good response, but due to the relapses associated with the corticosteroid tapering, infliximab was started. After 10 months of follow-up, the patient remained stable with the exception of an anal abscess that has led to surgical drainage.

Although Crohn's disease is frequent in children, cutaneous manifestations are the first manifestation in only 20% of patients. It may rarely precede bowel disease, by 3 months to 8 years.[2] Two thirds of children with CCD had genital involvement and 25% of patients with genital CCD presented with the skin disease first[2]. Perianal disease was present in 81% of CCD patients in the paediatric setting[3].

In the paediatric population, it can be an important differential diagnosis with genital venereal disease such as syphilis,

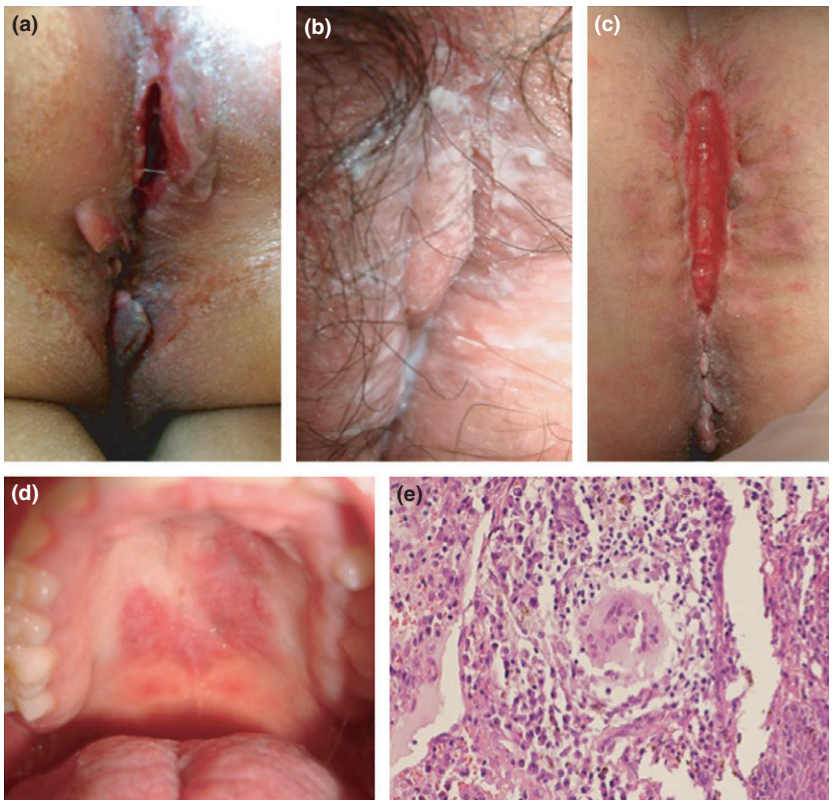

Figure 2 Ulcer in the intergluteal sulcus, with 4, 5 cm of length and ill-defined borders; pediculated lesions of variable dimensions with elastic consistency (a) Whitish exudate and multiple small fissures between the major and minor labia, around clitoris (b) After 3 months of treatment, intergluteal cleft with *coup de couteau* (c). Violaceous patch of circinate borders on the hard palate (d). Haematoxylin-eosin, 100× – Inflammatory tissue with fistulous and epithelioid granulomas (e).

lymphogranuloma venereum, herpes simplex, and non-veneral genital disease such as Behçet's disease[4].

To our knowledge, there are only 3 cases described of patients with Crohn 's disease and LP[5], none in the paediatric setting. Although the findings can be coincidental, in our case, the family history of type 1 DM, raised immunoglobulin levels, the presence of LP in a precocious age contributes for what could be an autoimmunity background favouring the development of CCD.

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Surgical management of extranasal nasal glioma

Editor

Nasal glioma is characterized by the appearance of heterotopic brain tissue in the frontonasal region. Despite its name it is a benign, slowly growing process. Several developmental theories exist, the most prevalent is that of a possible sequestered encephalocele. An extranasal form (60%) is divided from an intranasal form (30%). Mixed forms occur in 10%. Nasal glioma may cause functional impairment such as nasal deformation, airway obstruction, visual impairment or failure to thrive. Differential diagnoses include encephaloceles, (epi-) dermoids, infantile hemangiomas (IH), teratomas, lipomas and rhabdomyosarcomas.[1] Imaging is crucial to exclude intracranial anastomosis, the preferred method is magnetic resonance imaging (MRI).[2] With regard to the usually young age of affected patients, diagnostics and adequate treatment can be challenging.

Our first patient, an 11-day-old girl, presented with a congenital 16 × 14 × 12 mm swelling on the left side of the nasal root (Fig. 1a). The firm mass was non-dislocatable and non-pulsatile with small teleangiectasias, impairing the vision field of the left eye. MRI showed a local supranasal mass without intracranial anastomosis. At the age of 5 weeks, complete tumour excision including the overlaying skin with primary plastic reconstruction by a local transposition flap was done. Histopathology showed glial tissue without any signs of malignancy, also involving the dermis. Immunohistochemistry stained strongly positive for

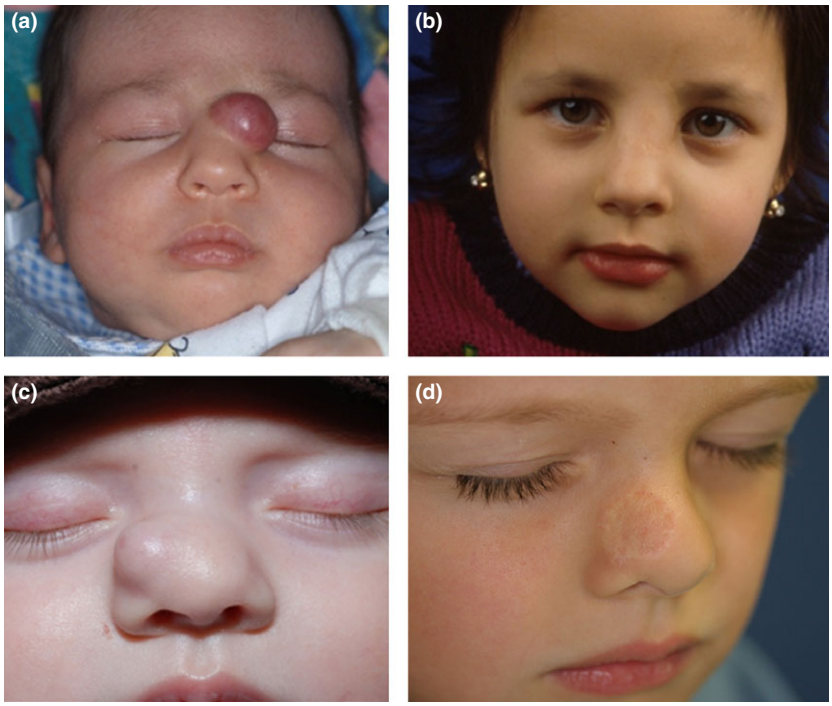

Figure 1 Initial presentation of patient 1 (a) and patient 2 (c). Cosmetic results after 5 years post-surgery in patient 1 (b) and after 4.75 years in patient 2 (d).