ingested therefore equates to 25–30 mg of prednisolone, a reasonable adult dose and, for this child, 1 mg kg\(^{-1}\). There are no available data concerning the likely absorption of corticosteroid from a topical preparation such as Eumovate in a child of this age; however, it would be a coincidence if this event were not involved in the development of GPP in this case. While unlikely to be a common occurrence in everyday clinical practice it does appear that ingestion of topical steroid preparations may be a potential trigger for acute GPP.

References


Conflicts of interest: none declared.

A case of Bazex–Dupré–Christol syndrome associated with multiple genital trichoepitheliomas

DOI: 10.1111/j.1365-2133.2005.06819.x

Sir, Bazex–Dupré–Christol syndrome was first described by Bazex et al. in 1964.\(^1\) It is characterized by a triad of congenital hypotrichosis, follicular atrophoderma (affecting the dorsa of the hands and feet, the face, and extensor surfaces of the elbows or knees) and the development of basocellular neoplasms (including basal cell naevi and basal cell carcinomas) from the second decade onwards.\(^2\)–\(^4\) Other reported features include associated hair shaft abnormalities (pili torti and trichorhexis nodosa) admixed with hypotrichosis, prominent milia affecting the face, hypohidrosis, pinched nose with hypoplastic nasal alae and prominent columella, atopic diathesis with comedones, keratosis pilaris, joint hypermobility, lingua plicata and hyperpigmentation of the forehead.\(^5\)–\(^6\)

A 3-year-old girl presented at the age of 2 years with increasing numbers of multiple brown asymptomatic papules over the genital area and medial aspect of the thighs (Fig. 1a). She had hypotrichia since birth (Fig. 1b), and had prominent facial milia and follicular atrophoderma of the cheeks (Fig. 1c) consistent with a diagnosis of Bazex–Dupré–Christol syndrome. Multiple members of her family showed similar features (the patient’s grandfather was originally reported by Gould and Barker in 1978\(^7\)) (Fig. 2). Her mother had features of facial milia, follicular atrophoderma of the cheeks and dorsa of the hands, hypotrichia (since birth), hypohidrosis and axillary hidradenitis suppurativa. The patient’s newborn brother was born with hypotrichia, mild erythema and ichthyosis and has subsequently developed multiple facial milia, suggesting that he also has Bazex–Dupré–Christol syndrome. A biopsy from the labial area in our patient revealed multiple dermal nodules consisting of well-defined arrangements of rounded massess of basaloid cells in a loose lobular (or ‘organoid’) pattern (Fig. 1d). A loose myxoid stroma without retraction artefact was present surrounding the interconnecting strands of basaloid cells (Fig. 1e). Well-formed papillary mesenchymal bodies, mitotic figures and necrosis were not seen. A diagnosis of multiple benign trichoepitheliomas occurring in Bazex–Dupré–Christol syndrome was made.

Bazex–Dupré–Christol syndrome is thought to be transmitted by an X-linked dominant pattern of inheritance.\(^7\) The genetic defect has been reported to localize to Xq24–q27.\(^8\) Neither trichoepitheliomas nor hidradenitis suppurativa have been reported in association with Bazex–Dupré–Christol syndrome. Most of the reported clinical features of Bazex–Dupré–Christol syndrome demonstrate abnormalities in the development of follicular structures, suggesting that the defective gene codes for a protein intimately involved in follicular differentiation and development.

Trichoepitheliomas are benign tumours which arise from cells derived from the hair follicle.\(^7\) Lesions may occur singly as a papule or nodule (up to 2 cm in diameter) or as multiple 2–8 mm diameter skin-coloured papules on the face of children or young adults centred around the nasolabial folds and preauricular regions. Trichoepitheliomas rarely affect the vulval region.\(^9\) The major histological and clinical differential diagnosis of trichoepithelioma includes basal cell carcinoma and trichoblastoma.\(^9\),\(^11\) There is considerable controversy regarding the histological definition of these follicular tumours and differentiation between these entities can be difficult, although various features are reported to be helpful in differentiating trichoepitheliomas from basal cell carcinomas.\(^11\),\(^12\) Furthermore, it remains unclear whether basal cell carcinomas may arise from trichoepitheliomas.\(^7\)
Familial multiple trichoepithelioma is inherited as an autosomal dominant disorder. The association of multiple familial trichoepitheliomas with basal cell carcinoma, cylindroma and occasionally eccrine spiradenoma is denoted Brooke–Spiegler syndrome, an autosomal dominant disorder due to loss of heterozygosity at 9p21.14 In contrast, sporadic trichoepithelioma may be associated with loss of heterozygosity at 9p22.3, a common site of genetic defects found in basal cell carcinomas.15–17 Recent studies in Chinese families with multiple trichoepitheliomas (without cylindromatosis) have demonstrated genetic defects at 16q12-q13, the site of the recessive cylindromatosis oncogene CYLD, the tumour suppressor gene responsible for the development of cylindromatosis. This observation confirms that different genetic mutations may manifest with identical phenotypic expression.18–20 This suggests that the genetic mutation responsible for Bazex–Dupré–Christol syndrome may also have pleiotropic effects responsible for the multiple trichoepitheliomas seen in our case. It is unclear whether our patient will go on to develop basal cell carcinomas (or cylindromas and spiradenomas).

We report what we believe to be the first case of Bazex–Dupré–Christol syndrome associated with trichoepitheliomas and hidradenitis suppurativa. Bazex–Dupré–Christol syndrome is associated with the development of basal cell carcinomas; however, it is unknown whether they arise from previously undiagnosed trichoepitheliomas.

Department of Dermatology, Leeds General Infirmary, Leeds LS1 3EX, U.K.
*Department of Dermatology, St James’s University Hospital, Leeds LS9 7TF, U.K.
E-mail: Anthony.Yung@leedsth.nhs.uk; ants_ange@hotmail.com

A. Yung
J.A. Newton-Bishop*

References
Correspondence


Conflicts of interest: none declared.

Superficial granulomatous pyoderma associated with chronic osteomyelitis

DOI: 10.1111/j.1365-2133.2005.06822.x

Sir, Superficial granulomatous pyoderma (SGP), first described by Wilson-Jones and Winkelmann in 1988,1 is a vegetative form of pyoderma gangrenosum (PG). It is characterized by sterile abscesses that form sinuses to the skin surface, and vegetative ulcers, mainly at sites of trauma. It presents with a papule, nodule or plaque, which can discharge pus. The base of the ulcer is clean and granulating, and borders are not undermined. The most frequent site is the back, followed by the hip, arm and abdomen. Culture for bacteria, fungi and mycobacteria is negative. Healing takes between 3 months and some years, and spontaneous healing is infrequent.

Histologically, SGP is characterized by a three-layered granuloma: inner neutrophils and necrosis; a surrounding layer of histiocytes and giant cells; an outer layer of plasma cells and eosinophils.2 In contrast to PG, SGP is less likely to be associated with systemic diseases; however, it has been reported in association with rheumatoid arthritis, chronic lymphocytic leukemia, IgA paraproteinaemia,3 cystic acne4 and other diseases.5 The differential diagnosis includes deep chronic infections and classic PG.6 Pyoderma-like lesions can also be observed in deep fungal infections, mycobacterial disease, foreign body granuloma and other conditions.7 Treatment response in SGP is variable and unpredictable. Therapeutic options include oral tetracyclines, sulphapyridine, dapsone, clofazimine, immunosuppressants and topical steroids.8 Systemic steroids are usually unnecessary.

The aetiopathogenesis of SGP and PG is not clear. The association of PG with systemic diseases (ulcerative colitis, Crohn disease, haemorrhagic malignancies and others) suggests an underlying autoimmune condition. T helper–suppressor imbalance, defective neutrophil chemotaxis, impaired phagocytosis and deranged monocyte function have been postulated in the pathogenesis of PG. An acute vascular insufficiency to the skin, as a consequence of deposition of immunocomplexes and lymphocytotoxicity leading to infarcts, has also been proposed. The histological findings of vasculitis and the occasional demonstration of deposits of immunoglobulin, complement and fibrin by immunofluorescence supports this hypothesis. Even in SGP the histological findings, especially the three-layered granuloma with eosinophils, are suggestive of an immunological granuloma, that is a delayed type hypersensitivity to an otherwise non-pathogenic organism.9 Until now the antigen has not been recognized, but it could be either endogenous or exogenous.1

Brodie’s abscess is a localized form of chronic osteomyelitis. It is more frequent in long bones, before epiphyseal closure. Sphynykosaurus aura is cultured in only 50% of cases. We present a patient with Brodie’s abscess, with clinicopathological features supporting the diagnosis of SGP.

A 14-year-old girl presented to the orthopaedic surgeons with a painful right upper femur. X-ray, magnetic resonance imaging scan and bone biopsies were performed, showing Brodie’s abscess. On the bone biopsies fragments of bone were associated with intense inflammation, consistent with osteomyelitis. She was started on full-dose fusidic acid, dinitrophenyl and ciprofloxacin, with improvement of the bone infection; however, the cutaneous lesion due to the bone biopsy failed to heal. Cutaneous mycobacterial infection was suspected, but no acid-fast bacilli were shown on microbiology or culture, and Ziehl–Neelsen stain was negative. Swabs were also negative.

One month later, she developed a similar lesion on the upper left arm. When asked, she did not deny picking the