Correspondence

Idiopathic angiokeratoma corporis diffusum

SIR Angiokeratomas can appear as solitary or localized lesions, as in the case of angiokeratoma circumscribed, angiokeratoma of Mibelli and scrotal angiokeratoma of Fordyce, or can involve large areas of the skin, in which case the term of angiokeratoma corporis diffusum (ACD) is used. This condition is usually regarded as a typical feature of Fabry’s disease (α-galactosidase deficiency), but several reports have shown that ACD can also occur in a variety of lysosomal storage diseases, such as sialidosis, fucosidosis, asparaglycosaminuria and β-mannosidase deficiency. In rare cases, however, ACD has been observed in the absence of any evident metabolic disease1–3 and as such was recently shown to be transmitted in a family as an autosomal dominant trait.4 We report here the case of a 34-year-old man who sought specialist advice about asymptomatic lesions which had first appeared eight years earlier and slowly spread to cover his entire body surface. Familial and personal history was negative for signs or symptoms of storage diseases. Clinical examination revealed many

Figure 1. Angiokeratomas on trunk and neck. Inset: close-up view of tiny angiokeratomas grouped in small plaques.

Figure 2. Histological appearance of lesion. Superficial enlarged vascular spaces lined with flat endothelium and filled with erythrocytes can be seen. Epidermis is moderately acanthotic and characteristically ‘embraces’ the dilated vascular structures in places. Modest orthokeratotic hyperkeratosis also seen (hematoxylin and eosin, original magnification ×200).
tiny, asymptomatic, dark red papules, grouped in small plaques, abundantly on the trunk but also on the face and legs (Fig. 1). A skin biopsy showed the features of angiokeratoma (Fig. 2). ACD was diagnosed and an attempt was made to determine the origin of the condition. Lysosomal enzyme activities (α- and β-galactosidase, β-hexosaminidase, β-glucuronidase, α- and β-mannosidase, α-fucosidase) in plasma and leukocytes were normal. Accurate electron microscopy examination of a biopsy specimen failed to reveal lysosomal inclusions in endothelial cells, pericytes or fibroblasts. Complete microscope examination of a biopsy specimen failed to reveal lysosomal (e.g. aspartyl glycosaminase deficiency), but we assume that these are all metabolic diseases so far associated with ACD are characterized by lysosomal inclusions, either in endothelial cells, fibroblasts or pericytes. Our case seems to support the opinion that ACD can be a purely cutaneous disease, unrelated to lysosomal storage diseases; however, molecular biology studies of Fabry’s disease have demonstrated more than 40 different mutations. Most of them were associated with the classical phenotype, but a few were found in atypical hemizygotic individuals who were asymptomatic or had cardiac involvement only. Hence we cannot exclude that ACD is linked to the genetic and phenotypic polymorphism of lysosomal storage diseases such as Fabry’s disease in otherwise apparently healthy patients. Molecular biology studies of apparently healthy ACD patients should clarify the possible relationship between genetic profile, enzyme alterations and the development of angiokeratomas. Until this relationship is proven, we propose that these cases be denoted by the term ‘idiopathic ACD.’

References

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Eczema craquelé and internal malignancy
Sir, I was interested to read the recent report by Guillet et al. of eczema craquelé as a pointer of internal malignancy.1 I have also seen a patient, a 73-year-old man, with a tumour of the gastrointestinal tract, who presented to a dermatologist with a 3-month history of weight loss and dry, itchy skin. Examination revealed eczema craquelé confined to the dorsa of the hands. In this case, however, rather than being a paraneoplastic phenomenon, the skin disorder was felt to be a direct consequence of the patient having to wash his hands more frequently as a result of his sudden increase in bowel motions (up to 13 times a day). Sigmoidoscopy revealed an annular carcinoma of the rectum, which at operation was found to be a Duke’s B adenocarcinoma. The patient underwent an abdominoperineal resection of his tumour and remains well 7 years later, with no recurrence of his skin problems.

Although this patient’s symptoms were a localized irritant contact dermatitis secondary to the excessive use of soap and water, the history revealed the true cause of the problem and highlighted the likelihood of an underlying malignancy of the bowel. Had the patient not been seen fortuitously by a dermatologist, his true diagnosis may have been even further delayed.

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E.M.Higgins

Hidradenitis suppurativa and monoarthrotis of the hip
Sir, Hidradenitis suppurativa (HS) is an indolent condition caused by apocrine gland blockage with secondary bacterial infection leading to recurrent abscesses, scarring and sinus formation. Arthritis is a rare association. We now describe a chronic monoarthritis and for the first time the associated synovial biopsy findings in a young Caucasian male with HS.

A 27-year-old man presented with a 2-year history of progressive pain in his right hip. There was no history of trauma. For 8 years he had suffered from inflammatory, discharging and scarring skin lesions in the axillae and groins diagnosed on this admission as HS. In 1988 he had been admitted with recurrent abdominal pain, a laparotomy on the third admission later showing adhesions, and biopsy demonstrating organized peritonitis with no evidence of granulomata. Every few weeks for the previous 2 years, he had passed intermittent loose motions with no blood or mucus four times daily for 2–3 days, but this had resolved 2 years before admission. In addition, he had had haematuria as a child, for which he had been investigated but the hospital notes concerning this were not available. There was also a history of depression, and both his mother and grandmother had suffered tuberculosis in the past, and his mother had psoriasis; the patient had not had a BCG vaccination. Examination revealed only global painful restriction of the right hip movements and axillary and groin HS. A provisional diagnosis of a sero-negative monoarthritis was made. On investigation, urinalysis was normal, as were full blood count, electrolytes, glucose, liver function tests and bone biochemistry, while rheumatoid factor, antinuclear antibodies and Brucella titres were negative. The erythrocyte sedimentation rate was 40–60 mm/h and the C-reactive protein concentration was 8–15 mg/ml (N < 5). Right hip X-ray showed a concentric loss of joint space and sclerosis of the acetabulum, while left hip, pelvis and lumbar spine X-rays were normal. A magnetic resonance imaging scan of the lumbar spine and pelvis revealed loss of bone density and erosion of the acetabulum. A Heaf test was strongly positive, but three early morning urine specimens revealed no growth on culture for acid and alcohol fast bacilli (AAFB), while chest X-ray was also normal. Thus, to exclude tuberculous monoarthritis, a right hip synovial biopsy was performed, which was negative on Ziehl-Nielson stain and culture and histology for AAFB, but otherwise showed features consistent with chronic non-specific synovitis with evidence of previous haemorrhage.

The patient was administered analgesia and in-patient physiotherapy, and on the advice of chest physicians given his grade IV Heaf test was also treated with a 6-month course of anti-tuberculous...
chemotherapy. He did not require any treatment for his HS during the admission.

The combination of arthritis and HS is rare, 1 previous reviews having mostly described arthritis occurring in combination with both acne conglobata and HS rather than HS alone.2 The arthritis described is seronegative, typically asymmetrical, and affects the knee, ankle and elbow joints most often, occurring between the ages of 22 and 46 with equal sex incidence; axial involvement, typically sacroiliitis, also occurs. Case reports have described the arthritis as occurring before the skin condition, 1 but most cases follow the skin lesions by 2–25 years. In all these respects our patient is typical, although most previous reported cases of HS and associated arthritis have been in Afro-Americans; however, this may reflect the racial distribution around the reporting centres.

It is possible that our patient’s sero-negative monoarthritis is associated with other causes, which include trauma, psoriatic arthropathy, inflammatory bowel disease-associated arthritis and tuberculosis monoarthrosis. Trauma is an unlikely explanation in the absence of a history of significant trauma, however, but the patient has a family history of psoriasis and it is thus just possible that he could have psoriatic monoarthropathy, particularly in that this may precede skin involvement by many years.3 HS has also been reported in association with Crohn’s disease,4 and it is therefore maybe possible that our patient has HS and monoarthritis associated with now quiescent inflammatory bowel disease. Tuberculous monoarthritis of the hip was also an important diagnosis to exclude, which was done.

In summary, the synovial biopsy findings from the affected hip of our patient were those of a chronic non-specific synovitis with an infiltrate of small lymphocytes, plasma cells and occasional mononuclear cells, although complicated by the finding of haemosiderin granules suggestive of previous haemorrhage and therefore possibly trauma. As far as we are aware, this is the first report of the synovial biopsy findings of arthritis in a patient with HS.

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References

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News and Notices

Cutaneous Microdialysis Club

The use of microdialysis for investigations in the skin is a rapidly expanding field of research. In order to facilitate contact between international study groups who use microdialysis to investigate the skin and the subcutaneous tissues, the Cutaneous Microdialysis Club was founded on April 22 1997.

The aim of this non-profit Society is to promote the knowledge of the technique, to promote standardisation and validation of cutaneous microdialysis and to promote investigations on and development of microdialysis instruments, dialysis analysis and data interpretation.

The Society covers a variety of fields of interest in microdialysis: pharmacokinetics, percutaneous absorption, allergology and inflammation, neurotransmitters, immunology, endocrinology and methodology. The Society has members from 12 countries and will hold scientific meetings on selected topics on a regular basis.

For further information, please contact: Eva Vejlstrup, Chairman, Department of Dermatology, University of Copenhagen, Gentofte Hospital, Niels Andersensvej 65, DK-2900 Hellerup, Denmark, Tel.: +45 39773213, Fax: +45 39777615 or Lotte Groth, Secretary, Department of Dermatological Research, Leo Pharmaceutical Products Ltd., Industriparken 55, DK-2750 Ballerup, Denmark, Fax: +45 44947488, e-mail: CMC@leo.dk.

Sixth International Conference on Perspectives in Percutaneous Penetration in collaboration with the Leiden/Amsterdam Centre for Drug Research, 22–26 September 1998, Leiden, The Netherlands

The Sixth International Perspectives in Percutaneous Penetration conference is to be held in Leiden, The Netherlands in September 1998. For further information contact the Conference Secretariat at: PPP Conference, Redwood Building, Cardiff CF1 3XH, U.K. Tel./Fax: +44 (0)1222 874932; e-mail: ppp@an-ex.co.uk.

VIIth Annual BEES Workshop, 16 January 1998, University of Nottingham, Nottingham, UK

The British Epidermo–Epidemiology Society (BEES) was founded in 1990 with the aim of promoting a high standard of epidemiological research into all forms of skin disease, with emphasis on an interdisciplinary approach. The VIIth BEES workshop will be held on 16 January 1998 at the University of Nottingham, UK.

Seating will be limited to the first 100 applicants. For further information please contact Mrs Melanie Bowesman-Jones, Secretary to Dr Hywel Williams, Department of Dermatology, C Floor, South Block, Queen’s Medical Centre, Nottingham NG7 2UH, Tel.: +44 (0)115 924 9924 ext. 44539; Fax: +44 (0)115 970 9003.

1998 UMDS Update in Dermopathology, Friday 20 February 1998, St. Thomas’ Hospital, London, UK

The theme of the 1998 UMDS Update in Dermopathology will be adnexal tumours. Seminars will be held on Sweat Gland Carcinomas (Dr D Santa Cruz), Microcystic Adnexal Carcinoma (Dr PH McKee), Sebaceous and Apocrine Tumours (Dr E Calcone) and Hair Follicle Tumours (speaker TBA). Further details can be obtained from Dr PH McKee, Department of Histopathology, St. Thomas’ Hospital, Lambeth Palace Road, London SE1 7EH, Tel.: +44 (0)171 928 9292 ext. 3493/2295; Fax: +44 (0)171 401 3661.
International Dermatopiediology Association Third Annual Scientific Meeting, 26 February 1998, Orlando, Florida, USA

This meeting – an international forum for the presentation and discussion of ongoing investigation in epidemiology and health services research in skin diseases – will take place on the evening of 26 February 1998, directly before the annual convention of the American Academy of Dermatology in Orlando, Florida. Abstracts will be selected by peer-review, based on scientific merit. Topics will include the epidemiology of cutaneous disorders (including descriptive, analytical, interventional and methodological investigations) and health services research (including studies of dermatological outcomes and the quality, costs and delivery of care). The deadline for the receipt of abstracts is 20 October 1997. Information and abstract forms are available from: Luigi Naldi, MD, Clinica Dermatologica, Ospedali Riuniti, L.go Barozzi 1, 24100 Bergamo, Italy. Tel.: +35 400625. Fax: +35 253070. E-mail: gised@uninetcom.it.

Second Professor Hans Storck Scientific Award
The Allergy Unit at the Department of Dermatology of the University Hospital Zurich will celebrate its fiftieth anniversary in 1998. The Allergy Unit was founded 1948 by Prof. Hans Storck after he returned from a visiting fellowship to various allergy centres in the USA. In recognition of this anniversary, on Friday and Saturday morning, 2–3 October 1998, an International Symposium dealing with “The Atopy Syndrome in the Third Millennium” is planned in collaboration with the Section Dermatology of the European Academy of Allergology and Clinical Immunology (EAACI). The organising committee announces the 2nd Professor Hans Storck Scientific Award of Sfr. 5000! This award, sponsored by an educational grant from Pharmacia & Upjohn Diagnostics Switzerland, is offered to a clinician or scientist, not older than 45 years, working in Europe in the field of Allergology and Clinical Immunology. The topic chosen for this award is “atopic dermatitis.” The contribution of the applicant should lead to a better understanding of the pathomechanisms involved in atopic skin disease or improve the management of patients. The paper should stress the contributions of Prof. Hans Storck and the Zurich School of Allergy concerning atopic dermatitis with particular emphasis on the so-called “intrinsic-type” and mention the paper of the winner of the 1st Award, U. Reinhold: “cell-mediated immunoregulation in atopic dermatitis. In: B. Wuthrich (ed.): Highlights in Allergy and Clinical Immunology, Hogrefe & Huber Publishers, Seattle, 1992, pp. 3–6. Five copies of an unpublished original article should be submitted together with a curriculum vitae and a list of publications and must be received not later than 30 June 1998.

The selection of the award winner will be made by the members of the organising committee and the Section of Dermatology of the EAACI. The article will be published in the a Proceedings Volume of Current Problems of Dermatology (ed. G. Burg), Karger, Basle. The application should be sent to: Prof. Dr B. Wuthrich, Allergy Unit, Department of Dermatology, University Hospital, Glierastrasse 31, CH-8091 Zurich, Switzerland.

Book review


Since patients with vulval disease may present in many different medical settings, there are several different groups of doctors who need to know about it. An atlas offers a useful way in which to serve this purpose, providing a sound didactic introduction to the subject. A multidisciplinary vulval clinic, such as that with which the authors have been associated, is a good source of expertise and of clinical material.

The collaboration of Miche`le Leibowitch, who was distinguished not only as a dermatologist but also as a pathologist, and who sadly died before this book became available, has led to description of the histological as well as of the clinical features for many of the conditions discussed; although this is essentially a book for reference in the clinic, the addition of such histological material may stimulate further interest in the reader.

When all vulval disease is to be encompassed, an entirely logical subdivision of the subject matter is difficult or impossible to attain. The authors have settled for a serviceable compromise between the ‘red lesions/white lesions’ method and the strictly aetiological or morphological approach. The contents range through anatomy, classification, examination, commonly observed non-pathological lesions, inflammatory diseases, ulcerating and blistering disorders, pilosebaceous inflammation, infections and infestations, tumours (meaning benign neoplasms, cysts and various other lumps), intraepithelial neoplasia, malignant disease, pigmented lesions, vulval diseases in childhood and psychological aspects of vulval disease. As is usual and useful in the books of this type currently available, rare conditions are noted as well as the common ones.

Tinted ‘box’ sections draw attention to essentials of management. The photographs are in general very good and the occasional line drawings clear. There are appendices which give brief details concerning plastic repair of the vulva, topical corticosteroids and practical treatment as well as of an information leaflet on lichen sclerosus and lichen planus for patients. There is also a list suggesting further reading.

It behaves a reviewer to consider areas of potential improvement. I think that in a further edition the authors might with advantage allow themselves more space for specifically psychological concerns. That section at present notes only dyssyestheath vulvodynia and cyclical vulvitis, neither of which are of certainly or entirely psychological origin; it would be nice to have some mention of, for example, the effects of surgery and of the role of psychosexual advice. More emphasis on the differential diagnosis, particularly of the infective conditions, would also be useful. Further clarity would be achieved by minor rearrangements of material; for instance to consider Paget’s disease with other forms of intraepithelial neoplasia. The appendices would benefit from expansion.

The authors are to be congratulated on producing a bigger and better second edition so quickly. It offers excellent value for money. I am sure that it will prove valuable in impressing upon those concerned the need for a multidisciplinary approach and, most importantly, in helping them to improve the everyday management of vulval disease.

C.M.Ridley

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