

Benign persistent papular acantholytic and dyskeratotic eruption: a case report and review of the literature

TH. VAN JOOST, V.D. VUZEVSKI,* B. TANK AND H.E. MENKE†

Departments of Dermato-Venereology and *Clinical Pathology, Erasmus University, Rotterdam

†Department of Dermatology, St Franciscus Hospital, Rotterdam, The Netherlands

Accepted for publication 16 July 1990

Summary

We report a case of a 35-year-old female with a persistent pruritic acantholytic and dyskeratotic eruption on the chest and vulva. The light and electron microscopic studies showed suprabasal epidermal clefting with acantholysis and dyskeratotic cells. We suggest that the most appropriate term for this case is that of benign persistent papular acantholytic and dyskeratotic eruption.

Several different papular or papulo-vesicular conditions with acantholysis and accompanied by various degrees of dyskeratosis have been described.¹⁻¹⁰ Except for the condition of transient acantholytic dermatosis (Grover's disease),⁴ all the other dermatoses usually followed a more or less chronic course (Table 1).

There have been several reports of a papular eruption with acantholysis and dyskeratosis that is restricted to the vulvocruval areas^{10,11-17} (Table 2). These cases have been described as atypical vulval Hailey-Hailey disease¹¹⁻¹⁴ or vulval warty dyskeratosis.¹⁵ We report a patient with a persistent papular acantholytic and dyskeratotic eruption that involved the submammary areas and the vulva.

Case report

A 35-year-old female patient had a persistent history, of more than 3 years, of discrete and partly symmetrical papular lesions in the submammary region and, at the same time, more itchy and painful papules on the inner aspect of the labia majora. There was no family history of similar eruptions. The vulval lesions were clearly defined, flesh-coloured, partly grouped papules that measured 3-5 mm in diameter and some of them were eroded (Fig. 1). There were slightly keratotic discrete, light-brown skin lesions in the submammary region that clinically resembled Grover's disease. The more painful vulval lesions were excised. There were no lesions in the perianal region or in the mouth. Swabs taken from the patient and her sexual partner showed no evidence of *Candida* infections.

Histology

Light microscopy studies of biopsies from the submammary regions showed marked acantholysis and minimal



Figure 1. Grouped and solitary flesh-coloured papules on the right labium (arrows).

Table 1. Acantholytic eruptions with combined variations in dyskeratosis

Reference	Classification	Hereditary	Duration	Dyskeratosis (usual pattern)
1	Darier's disease	+	Persistent	Marked
2	Acantholytic dyskeratotic (zosteriform) naevus (Darier's disease?)	-	Persistent	Marked
3	Warty dyskeratosis	-	Persistent	Marked
4	Grover's disease (transient acantholytic dermatosis)	-	Transient	Moderate, Minimal or lacking
5	Hailey-Hailey disease	+	Persistent	Minimal
6	Acantholytic squamous cell carcinoma	-	Persistent	Minimal
7	Persistent acantholytic dermatosis*	-	Persistent	Moderate
8	Persistent acantholytic dermatosis related to actinic damage*	-	Persistent	Moderate or marked
9	Benign papular acantholytic dermatosis (BPAD)*	-	Persistent	Minimal or lacking
10	Acantholytic dermatosis* localized to the vulvocrural area	-	Persistent	Minimal or moderate

* Descriptive (provisional) diagnosis.

Table 2. Acantholytic and dyskeratotic lesions of the vulvocrural region

Reference	Number of cases	Age (years)	Provisional classification	Duration (months)	Other location
10	6	28, 37, 47, 53, 63, 83	(Distinct syndrome?) Acantholytic dermatosis of the vulvocrural area	12-36	No
11	1	22	Hailey-Hailey disease	12	No
12	1	29	Hailey-Hailey disease	7	No
13	1	39	Hailey-Hailey disease	12	No
14	1	38	Hailey-Hailey disease	?	No
15	3	49, 38, 52	Warty dyskeratosis	6-24	No
16	1	29	Papular acantholytic dyskeratosis of the vulva (Hailey-Hailey or Grover's disease or distinct syndrome?)	72	No
17	1	47	Papular acantholytic dyskeratosis	36	No
Present case	1	35	(Distinct syndrome?) Benign persistent papular acantholytic dyskeratotic eruption	36	Yes

dyskeratosis. Histology of the labia majora lesions showed a suprabasal epidermal cleft with typical acantholytic cells. Above the split, within the epidermis, there were dyskeratotic cells (Fig. 2). Small villous-like bodies

were observed consisting of dermal papillae covered with a single layer of cuboidal cells with pyknotic nuclei that projected into the cleft. There were a number of inflammatory cells surrounding the cleft.

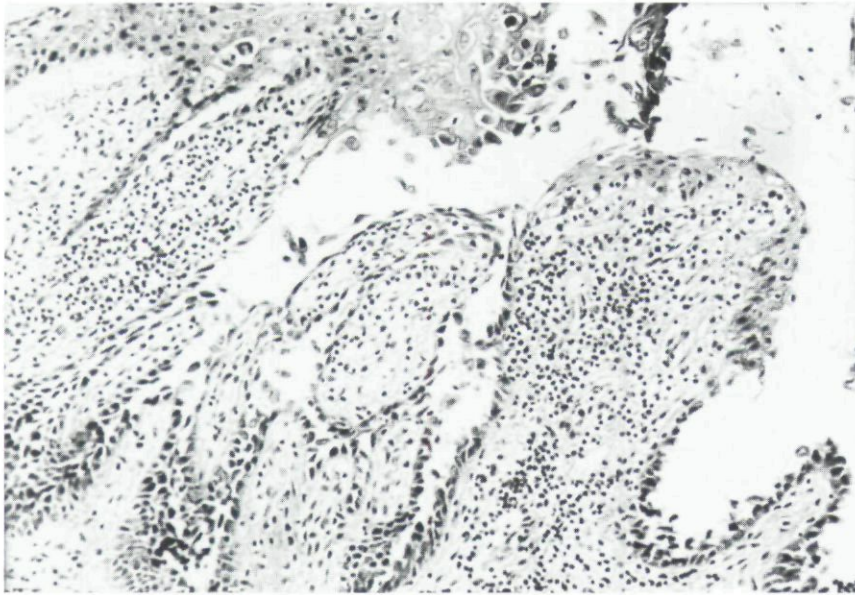


Figure 2. Histology of a papular lesion on the vulva showing suprabasal acantholysis and to a lesser extent dyskeratosis of the upper epidermis (haematoxylin and eosin, $\times 216$).

Immunofluorescence

Direct immunofluorescence was negative with no deposition of immunoglobulins or complement in the skin.

Electron microscopy

This was performed only on the vulval lesions and the ultrastructural changes were those of acantholysis and dyskeratosis. There was a total disappearance of intercellular connections and the number of desmosomes appeared to be reduced. The tonofilaments were detached from the desmosomal plate, often aggregated around the nucleus. The tonofilament aggregations were dense, wavy and band-like and were mostly observed in the cells of the Malpighian layer, but also in isolated cells within the cleft (Fig. 3).

Cytokeratin studies

These were carried out on the vulval lesions using commercially available monoclonal antibodies and using methods described previously¹⁸. Staining for anti-polykeratin was observed but there was no staining for anticytokeratin 7 (RRCK 105), anticytokeratin 18 and 19 (5D3). RKSE 60 (anticytokeratin 10) detects suprabasal cytokeratin filaments,¹⁹ but was absent in the vulval lesions in our patient.

Discussion

In 1972 Ackerman⁶ introduced the term 'focal acantho-

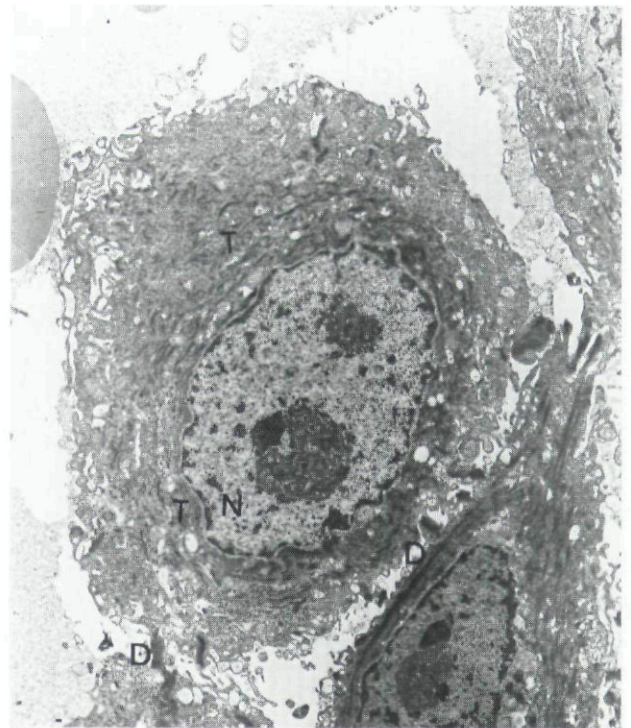


Figure 3. Electron micrograph of a vulval lesion. This shows acantholytic cells, tonofilaments (T) detached from the desmosomal plate (D) and aggregated around the nucleus (N). $\times 5200$.

lytic dyskeratosis' for clinical and histopathological conditions other than typical Darier's disease. A case was reported¹⁶ of a papular acantholytic and dyskeratotic dermatosis of the vulva and it was questioned as to

whether this may be a distinct entity. Coppola *et al.*¹⁷ reported a patient with vulval and perineal papular lesions that coalesced to form plaques and this case was described as being 'papular acantholytic dyskeratosis'. Six patients were later reported with an acantholytic dermatosis localized to the vulvocruval area.¹⁰ The histology in these cases showed acantholytic dyskeratosis that resembled Darier's disease or when there was minimal dyskeratosis, Hailey-Hailey disease (Tables 1 and 2). The presence of *Candida* in some of these cases could explain the apparent location of the vulval lesions. None of these patients had a family history and none had similar lesions elsewhere on the body as was observed in our case.

In Grover's disease and in the syndrome of benign papular acantholytic dermatosis described by Heaphy *et al.*,⁹ in which lesions occurred on the trunk and the neck, dyskeratosis can be minimal or absent. The histology of the submammary lesions in our case resembled that seen in Grover's disease or that described by Heaphy *et al.*⁹ The vulval lesions in our case ultrastructurally resembled the pattern described for transient Grover's disease.^{20,21} The combination of the clinical features, persistent nature and absence of family history with the histopathology of the submammary lesions, however, resemble more closely that of the acantholytic and dyskeratotic type of benign papular acantholytic dermatosis. Our patient had not only vulval but also submammary lesions. A further finding of interest in our case was that staining with antipolykeratin monoclonal antibody was observed, but that cytokeratin 10 was absent.

Although this is a report of a single case, we conclude from the clinical and histopathological findings that our patient is best classified as having a benign persistent papular acantholytic and dyskeratotic eruption.

References

- Gottlieb SK, Lutzner MA. Darier's disease. *Arch Dermatol* 1973; 107: 225-30.
- Starink TM, Woerdeman MJ. Unilateral systematized keratosis follicularis. A variant of Darier's disease or an epidermal naevus (acantholytic dyskeratotic epidermal naevus?) *Br J Dermatol* 1981; 105: 207-14.
- Gorlin RJ, Peterson WC. Warty dyskeratoma. *Arch Dermatol* 1967; 95: 292-3.
- Grover RW. Transient acantholytic dermatosis. *Arch Dermatol* 1970; 101: 426-34.
- Hailey H, Hailey H. Familial benign chronic pemphigus. *Arch Dermatol* 1939; 39: 679-85.
- Ackerman AB. Focal acantholytic dyskeratosis. *Arch Dermatol* 1972; 106: 702-6.
- Simon RS, Bloom D, Ackerman AB. Persistent acantholytic dermatosis. *Arch Dermatol* 1976; 112: 1429-31.
- Fawcett HA, Miller JA. Persistent acantholytic dermatosis related to actinic damage. *Br J Dermatol* 1983; 109: 349-54.
- Heaphy MR, Tucker SB, Winkelmann RK. Benign papular acantholytic dermatosis. *Arch Dermatol* 1976; 112: 814-21.
- Cooper PH. Acantholytic dermatosis localized to the vulvocruval area. *J Cutan Pathol* 1989; 16: 81-4.
- Lyles TW, Knox JM, Richardson JB. Atypical features in familial benign chronic pemphigus. *Arch Dermatol* 1958; 78: 446-53.
- Hazelrigg DE, Stoller LJ. Isolated familial benign chronic pemphigus. *Arch Dermatol* 1977; 113: 1302.
- King DT, Hirose FM, King LA. Simultaneous occurrence of familial benign chronic pemphigus (Hailey's-Hailey's disease) and syringoma of the vulva. *Arch Dermatol* 1978; 114: 801.
- Evron S, Leviatan A, Okon E. Familial benign chronic pemphigus appearing as leukoplakia of the vulva. *Int J Dermatol* 1984; 23: 556-557.
- Duray PH, Merino MJ, Asiotis G. Warty dyskeratosis of the vulva. *Int J Gynecol Pathol* 1983; 2: 286.
- Chorzelski TP, Kudejko J, Jablonska S. Is papular acantholytic dyskeratosis of the vulva a new entity? *Am J Dermatopathol* 1984; 6: 557-9.
- Coppola G, Muscardin LM, Piazza P. Papular acantholytic dyskeratosis. *Am J Dermatopathol* 1986; 8: 364-6.
- Habets JMW, Tank B, Vuzevski VD *et al.* Absence of Cytokeratin 8 and inconsistent expression of cytokeratins 7 and 19 in human basal cell carcinoma. *Anticancer Research* 1988; 8: 611-16.
- Leigh IM, Pulford KA, Ramaekers FCS, Lane EB. Psoriasis: maintenance of an intact monolayer basal cell differentiation compartment in spite of proliferation. *Br J Dermatol* 1985; 113: 53-64.
- Kanzaki T, Hashimoto K. Transient acantholytic dermatosis of oral mucosa. *J Cutan Pathol* 1978; 5: 23-30.
- Grover RW, Duffy LW. Transient acantholytic dermatosis: Electron microscopic study of the Darier type. *J Cutan Pathol* 1975; 2: 111-27.

This document is a scanned copy of a printed document. No warranty is given about the accuracy of the copy. Users should refer to the original published version of the material.