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INTRAORAL WARTY DYSKERATOMA

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Warty dysteratoma (isolated dyskeratosis follicularis or focal Darier's disease) is a benign cutaneous tumor which occurs commonly on the scalp, face or neck. In several instances it has been reported to occur in areas not customarily exposed to the sun, including the mouth. Only four cases have been reported on the oral mucosa. The purpose of this paper is to review these reports and describe three additional oral cases.

Introduction

On 1954 Helwig[1] described a lesion which he called "isolated Darier's disease" because of its histomorphologic similarity to Darier's disease. In the same year Allen[2] referred to this lesion as "isolated or solitary Darier's disease," and in 1967[3] he suggested that it might

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be labeled "solitary verrucal dyskeratoma (pseudo-Darier-'s disease). Szymanski[4-5-6] suggested the term "Warty dyskeratoma" for this lesion.

In 1958 Graham and Helwig[7] described a lesion which they called "isolated dyskeratosis follicularis." 50% of the patients having this lesion were over fifty years of age. Ninety per cent of them were male. The majority of the lesions (88 per cent) were from the head and neck area, anatomic sites with prominent pilosebaceous elements.

The first intraoral warty dyskeratoma was reported in 1967 by Gorlin and Peterson.[8] It was observed on palatal mucosa of a forty-five year old white male. In 1971 three cases were reported by Tomich and et al,[9] all of these patients were males between forty-nine and sixty-one years of age. (Table I) Below are described three additional cases, reported here for the first time.

Case Reports

Case 1: A forty-two year old Caucasian male visited his dentist for the examination of a generalized white patch. Generalized hyperkeratotic areas were noted over the lower ridges, and a discrete 2mm lesion was noted on the buccal surface of the lower left second bicuspid area. The patient was a heavy smoker. A physical examination disclosed no evidence of other cutaneous or mucosal lesions. A family history was obtained.

Gross findings: Received in formalin was an elliptical excision of tissue measuring. 3x.3x.3 cm. The apparent epithelial surface was gray-white and near one pole of the ellipse, two similar-appearing, fairly wellcircumscribed arease were noted. The central portion of the areas contained some yellow-tan tissue surrounded by a slightly elevated, ovoid ring of gray-white tis-

Summary of Seven Patients
with Intraoral Dyskeratoma

Table I

Authors	Age	Sex	Location	Clinical	History
Gorlin & Perterson	45	M	Palatal mucosa	.5x.3x.3mm verrucose	Normal
Tomich & Burkes	49	M	Left man. ridge	Small depression	Used snuff No skin lesion
"	61	M	Mandibular ridge	5mm white with de-pressed center	--
"	61	M	Palatal mucosa	White lesion 5 mm.	No skin lesion
Forouzan-deh	42	M	Lower buccal mucosa	.2 mm diameter	No skin lesion Heavy Smoker
"	60	M	Left pos-terior Hard Palate	.5x.6x.1 mm	No skin lesion
"	58	F	Hard Palated	.9x.6x.3 mm	Heavy smoker No skin lesion

Table I

sue.

Microscopic examination: The section revealed multiple strips of mucosa covered by intact squamous epithelium. In many areas, the epithelium was somewhat acanthotic and was covered by a moderately thick layer of keratin alternating with foci of parakeratin. No dyskeratosis was seen. One segment presented a central zone of epithelial proliferation characterized by an extension of narrow cords of basal cells into the underlying connective tissue. There was also some evidence of a papillary growth in association with this, since numerous clefts appear to be formed between the rows of columnar basal cells. In the superfical central portion there appeared to be a plug of more typical squamous cells. The epithelial cells covering these villi were somewhat dyskeratotic and showed enlarged prominent nucleoli. One small whorl of squamous epithelium was noted in the deeper aspect of these epithilial cords. Only minimal chronic inflamation was associated with this fairly well-demarcated vasculature, and the epithelial cells covering some had taken on a flattened appearance.

Case 2: A sixty year old Caucasian male visited his dentist for the examination of lesions on the soft and hard palate. Excisional biopsies were performed. A subsequent physical examination disclosed no evidence of other cutaneous or mucosal lesions and a normal family history was obtained.

Gross findings: The formalin-fixed specimen consisted of a light to dark tan irregular mass which measured 6x.5x. 1 cm.

Microscopic examination: The section revealed mucos-

al segments covered by well-differentiated hyperorthokeratosis, and stratified squamous epithelium. In one area there was a fairly large, cup-shaped invagination filled with parakeratin and hypergranulosis of the superficial epithelial cells at the orifice. The base of the invagination shows a moderate acantholysis with formation of a few pseudovillous processes lined by a single layer of epithelium. Adjacent connective tissue contained a mild infiltrate of chronic inflammatory cells.

Case 3: A fifty-eight year old female visited her dentist for the examination of a lesion on the left palatal mucosa. Examination revealed an irregular, slightly raised white lesion. The patient had smoked more than one pack of cigarettes per day for many years.Surgery was performed.

Gross findings: The formalin-fixed specimen consisted of a gray-white segment of curved tissue measuring. 9x.3x.6 cm, with a roughened surface. Selected transverse sections were taken and the cut surfaces embedded for sectioning.

Microscopic examination: The section revealed mucosal segments which were covered by stratified equamous epithelium showing marked hyperorthokeratosis with hypergranulosis at the orifice of a cup-shaped epithelial invagination partially filled with parakeratin. The base of the invagination was characterized by numerous "villi" lined by a single-to-double layer of squamous epithelial

ses, including the three cases reported here. From this data, it appears that warty dyskeratoma occurs as a solitary lesion ranging from two to ten mm in diameter. It apears as an elevated nodule with a depressed center and varying color. In all of the reported patients there was no evidence of other cutaneous lesions; each had a normal family history.

ETIOLOGY: Graham and Helwig[7] believe that a viral etiology is most logical. They speculate that a virus with an affinity for pilosebaceous structures can enter the hair follicle, disrupt, and finally destroy the hair and the sebaceous gland.[7-10]

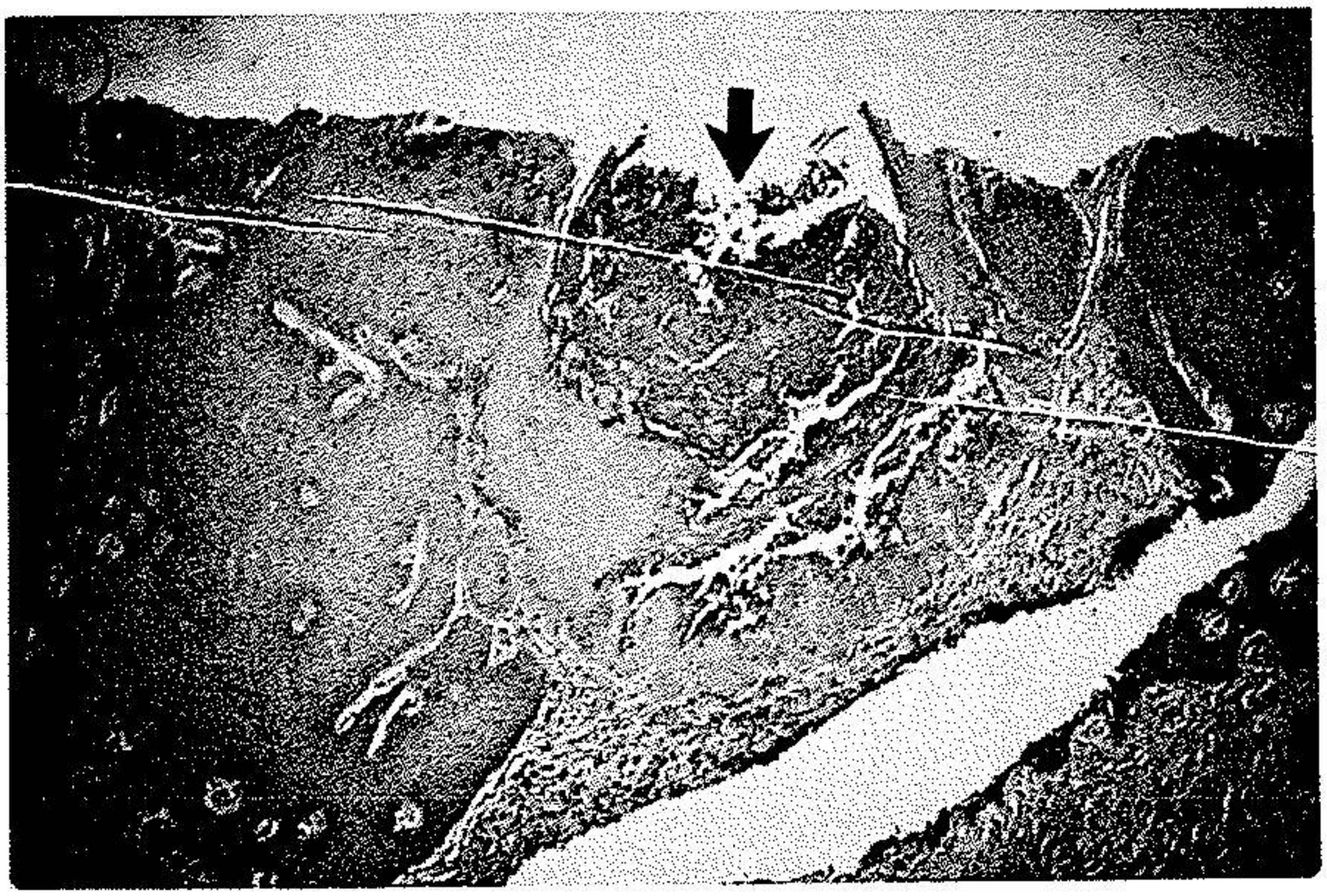
Szymanski[4] suggests that the cutaneous lesion is induced by virus and closely related to a viral wart(verruca). He reports what he considers to be histochemical evidence to support his theory-histochemical demonstration of DNA and RNA in the nuclei and cytoplasm. On the other hand the specific site for occurrence of warty dyskeratoma and recurrence of intraoral herpes simplex are the same, and both of these lesions occur in attached mucoşa overlining bone; this fact can support a viral theory for this lesion.

HISTOMORPHOLOGY FEATURES: The center of the lesion is occupied by a large, cup-shaped invagination containing keratinous material much as orthokeratin or para keratin, and numerous acantholitic, dystkeratotic cells in its lower portion (Fig.1,2,3). Numerous villi lined often with only a single layer of basal cells project upward from the base of the cup-shaped invagination (Fig.3). These papilla or villi are lined with either cupoidal,or columnar epithelial cells which are usually in a single

row (Fig. 4). The supporting connective tissue invariably contains a mild chronic inflammatory cell infiltrate.

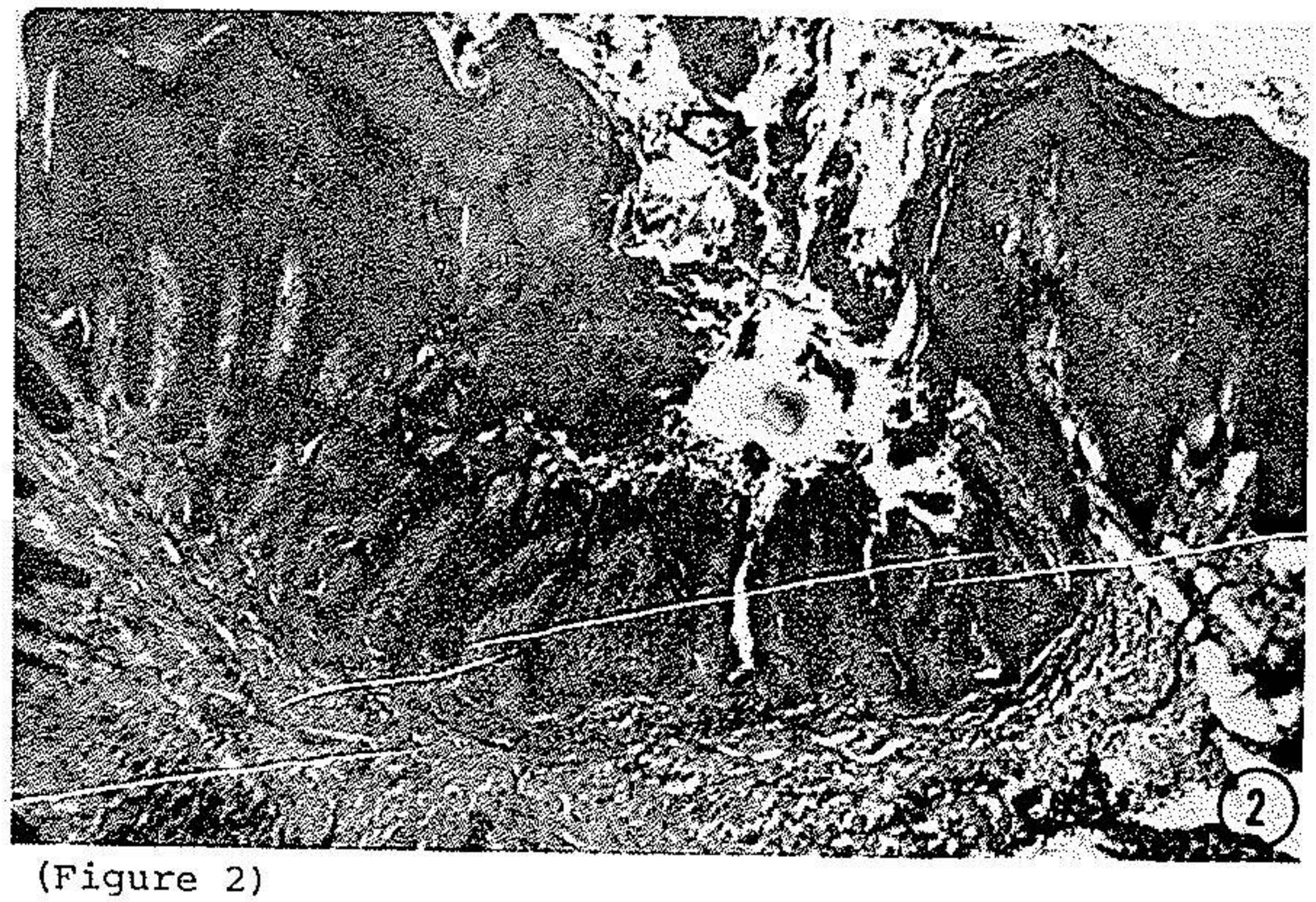
Discussion of Clinical Features

Warty dyskeratoma or isolated dyskeratosis follicu-



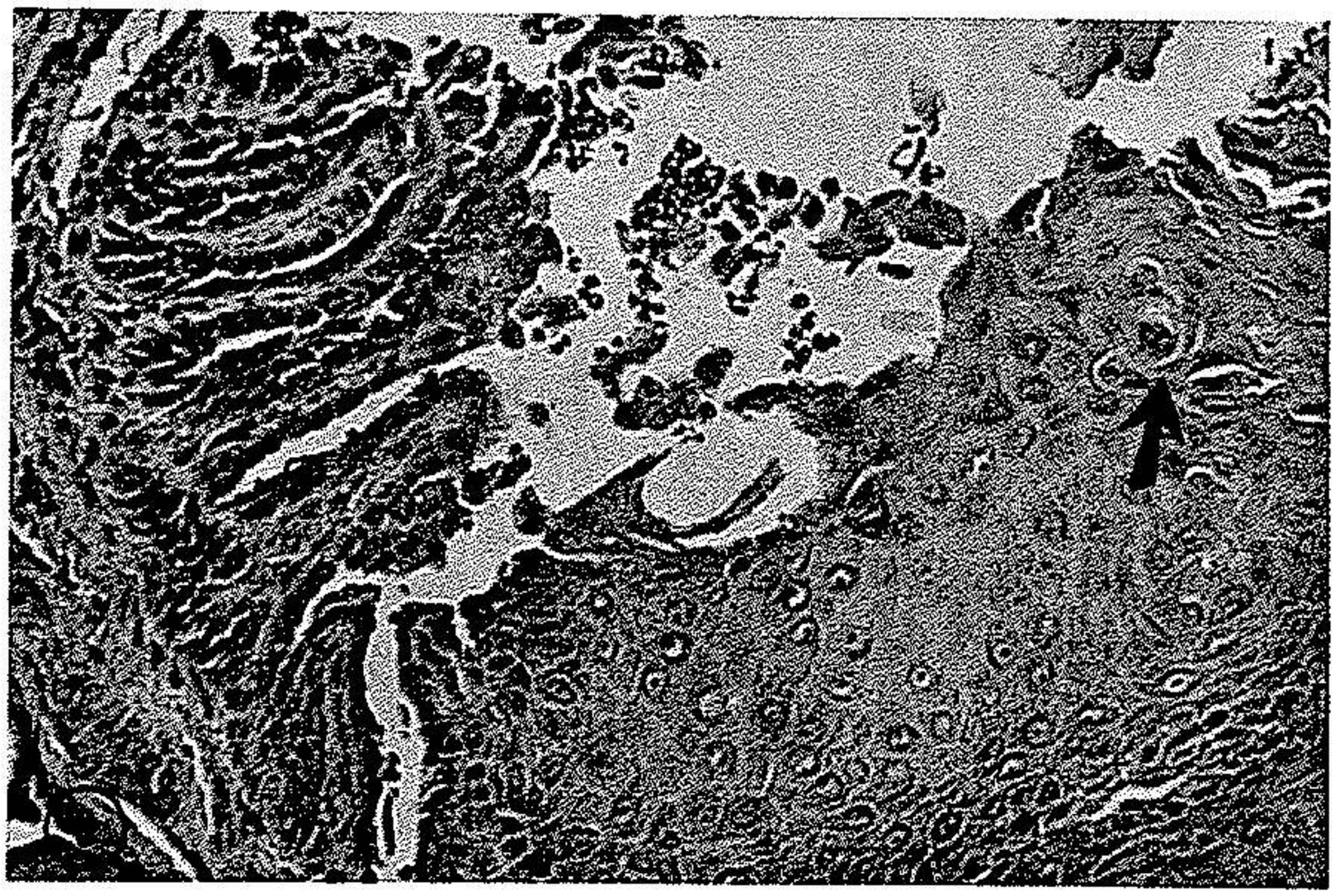
(Figure 1)

Photomicrograph of an intraoral warty dyskeratoma(case 2) showing the keratin core, superabasilar cleft and villi. (Hematoxylin and eosin stain, magnification, x40)

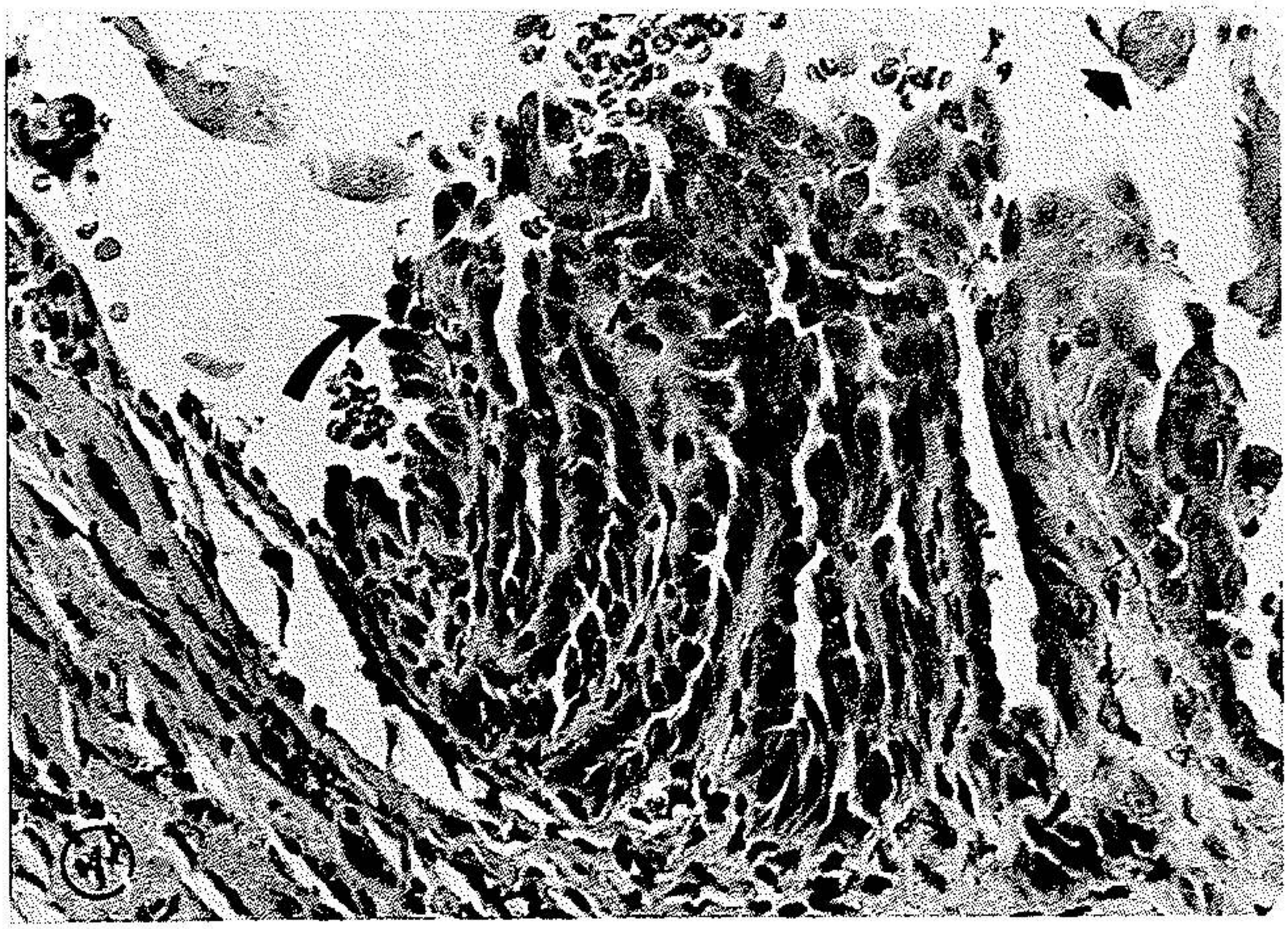


(Figure 2)

Photomicrograph of an intraoral warty dyskeratoma(case 3). Hematoxylin and eosin stain, magnification x40).



Photomicrograph of an intraoral warty dyskeratoma (case 1). Suprabasilar cleft, villi, acantholytic cells and dyskeratosis (arrow). (Hematoxylin and eosin stain, magnification, x 100.)



(Figure 4)

Higher power photomicrograph of a warty dyskeratoma shown in figure 3. Shown is the connective tissue villi with a lininq row of epithelial cells (arrow). Note the acantholitic cluster of cells (arrow). (Hematoxylin and eosin stain, magnification x 450).

laris occurs always as a solitary lesion[6-12-13] most common on the scalp, face or neck. The occurence of warty dyskeratoma in the oral cavity is extremely uncommon. Only four intraoral cases of warty dyskeratoma have been previously reported in English-languqge journals.[8-9-12] Intraoral warty dyskeratoma seems to occur most commonly in men at middle and old age. All reported lesions have occurred in mucosa overlying bone palatal or alveolar mucosa. The clinical appearance of this lesion is usually a slightly elevated papule or nodule with a keratotic umbilicated center.[8-9-14] The lesion, after having reached a certain size, seems to persist indefinitely. Clinically, all of the lesions reported have behaved like benign tumors. Metastases or invasion have not been reported. However, if the tumors are not excised completely,there is a possibility of recurrence.[5-12-13,]

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face, hands and all over the body, In 1973 he noticed an ulcer of 0.5 cm. on the occiput. This was not improved by any topical treatment. In 1974 the first excisional surgical intervension was performed on the occiputal ulcer; but the ulcer recurred after three months. This was treated by wedge excision in July 1974. The histological report of the excised ulcer showed a squamous cell carcinoma.

Shortly after, the ulcer recurred again and this time it was treated by radiotherapy with 6000 rads; the ulcer healed with a remaining scar.

At this time a biopsy of one of the papules of the anterior aspect of the arm was performed. The histological report confirmed the suggested clinical diagnosis of epidermodysplasia verruciformis. Six months later redness and ulcers were found around the scar. A repeated biopsy showed a squamous cell carcinoma.

The patient was hospitalized and a part of the scalp and acciputal bone together with a small tumour on the sagital sinus were removed under the supervision of neurosurgeon. In 1975, at the site of the previous operation a new ulcer developed. For further treatment the patient was referred to London wherby a surgical treatment on the occiput using skin grafts (direct pedicle flaps)was performed. Two months later a fistula appeared at the site of operation and radiotherapy was administered.

Shortly after a lymph node enlargement developed on the right side of the neck, This was treated by radical neck dissection. In 1976, the patients was hospitalized in Israel, where he was treated by chemiotherapy.

The patient's parents show no evidence of the disease. Of three brothers and one sister of the patients,a

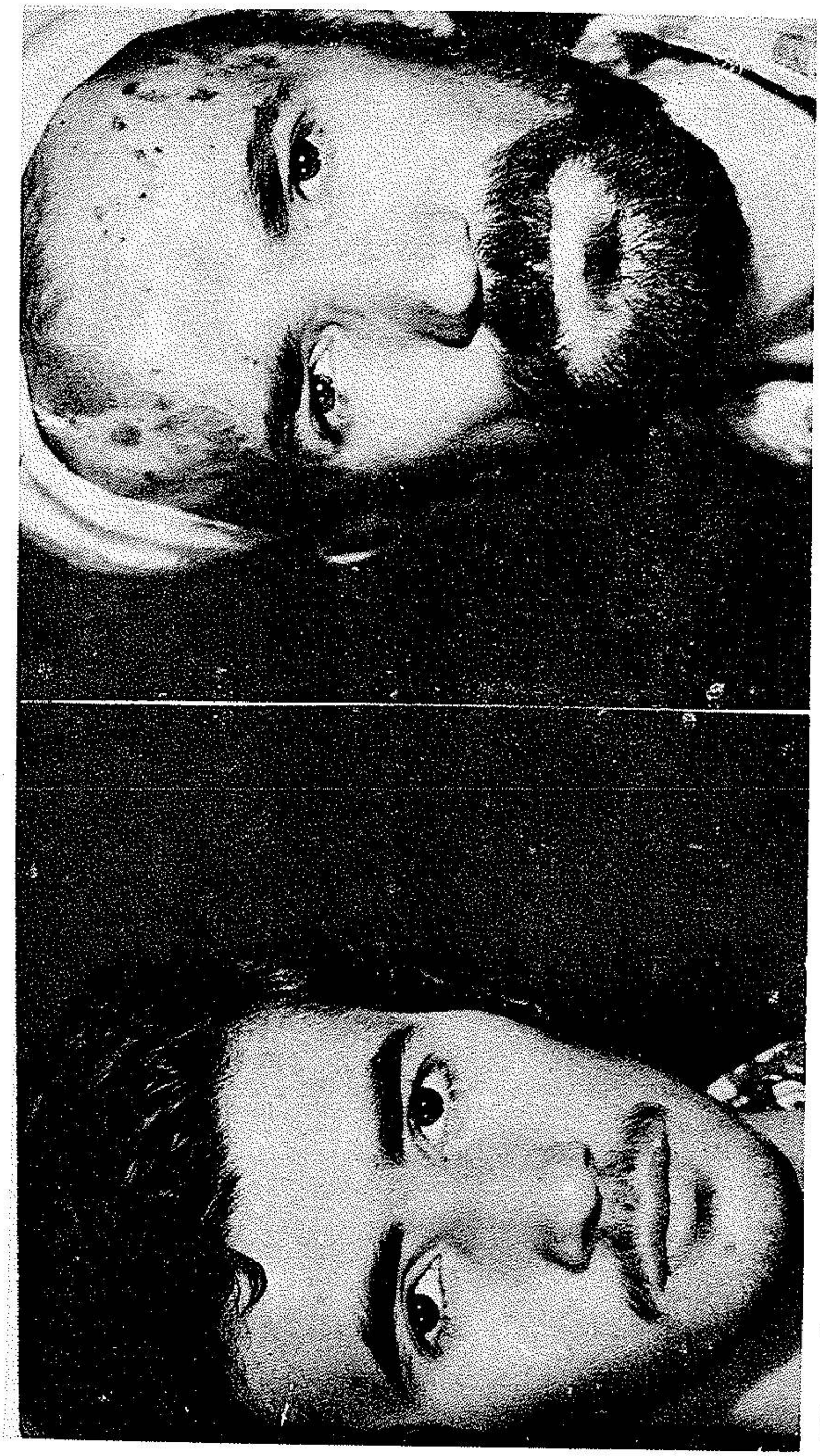

FIG. 1: Both brothersshowing similar papular lesions on foreheads.

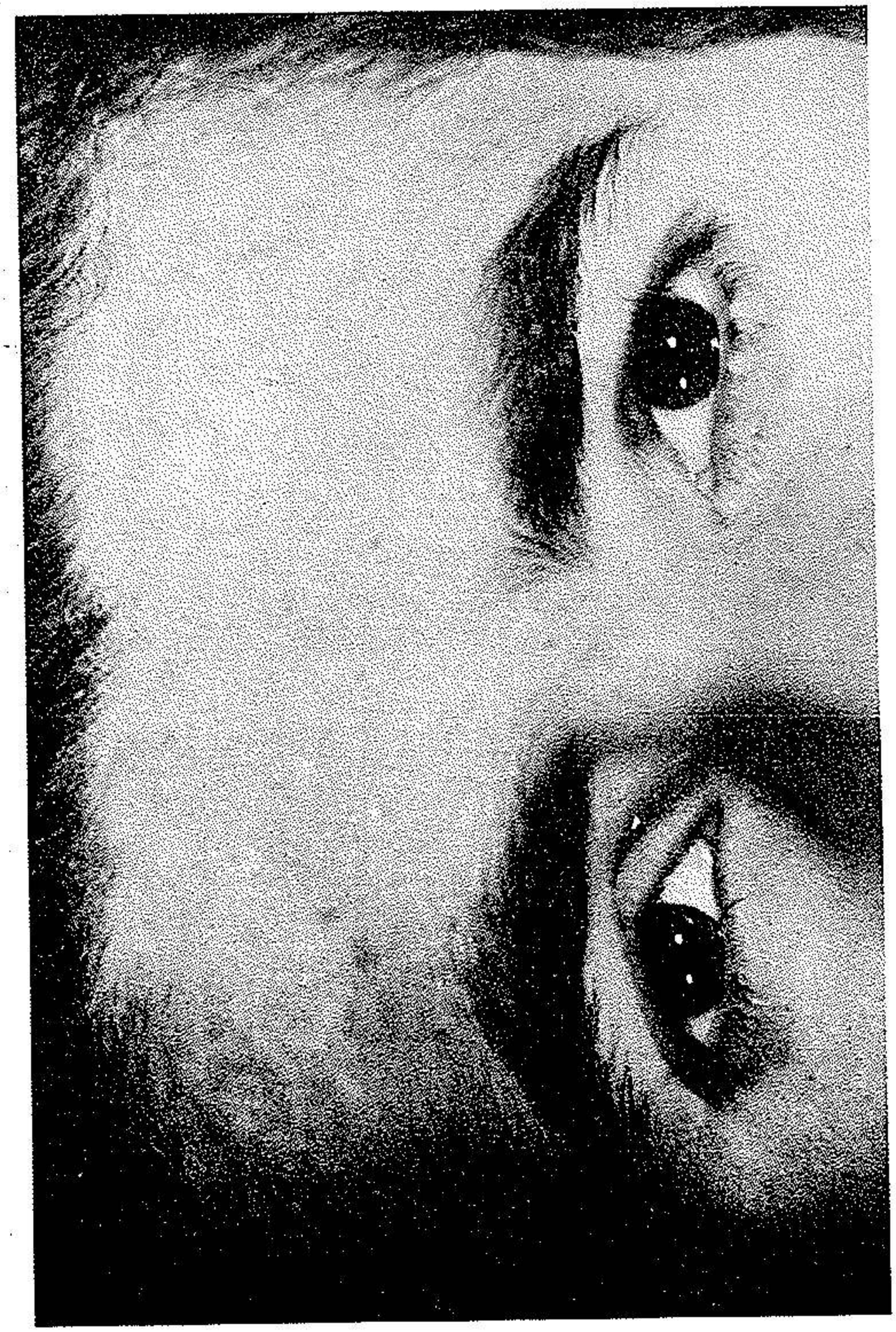

FIG. 2: Close-up picture of the forehead showing the papular lesions of younger brother.

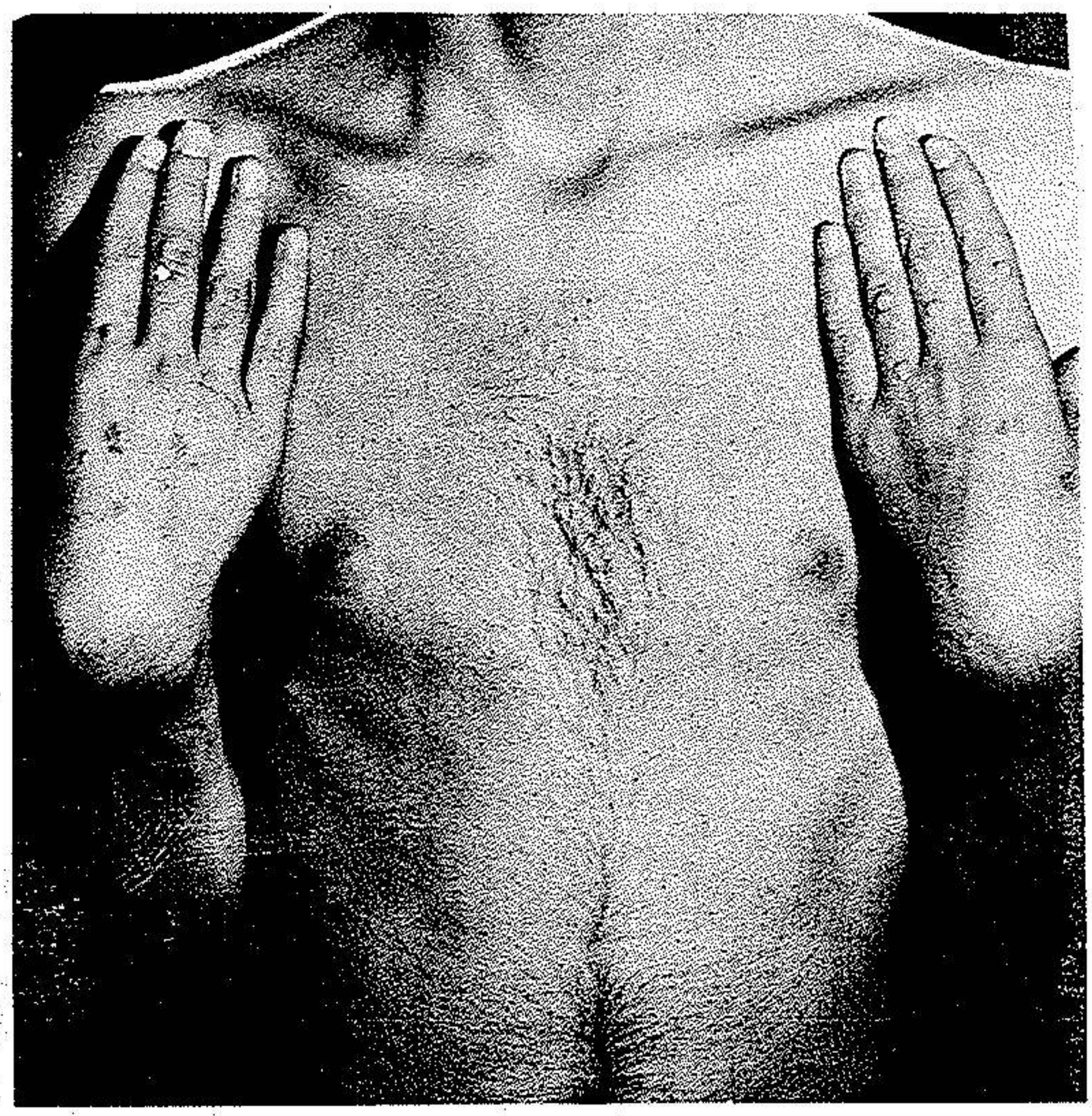

FIG. 3: Papular lesions on back of the hands of younger brother.

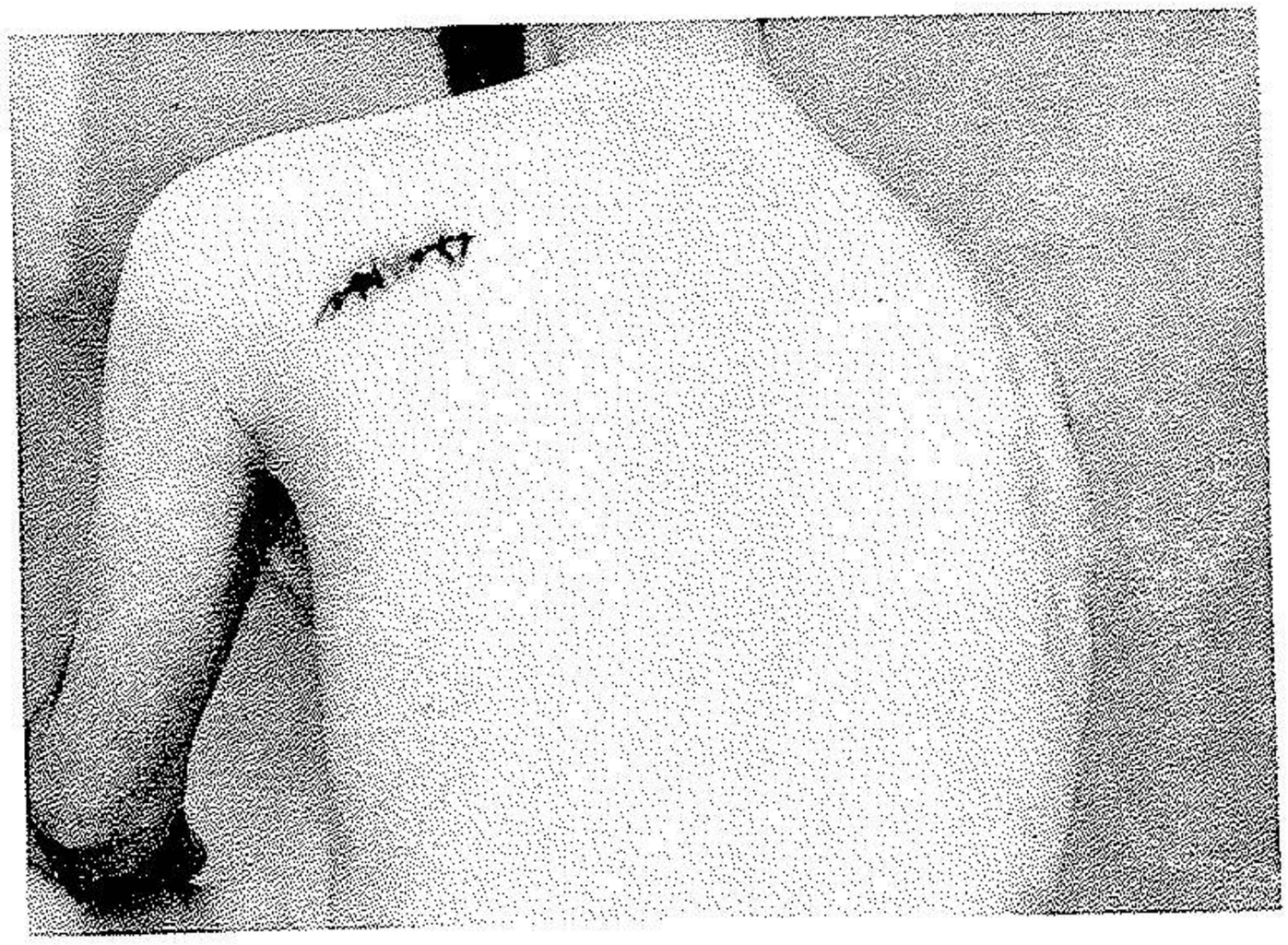

FIG. 4: Back of the patient with diffuse papular lesions.

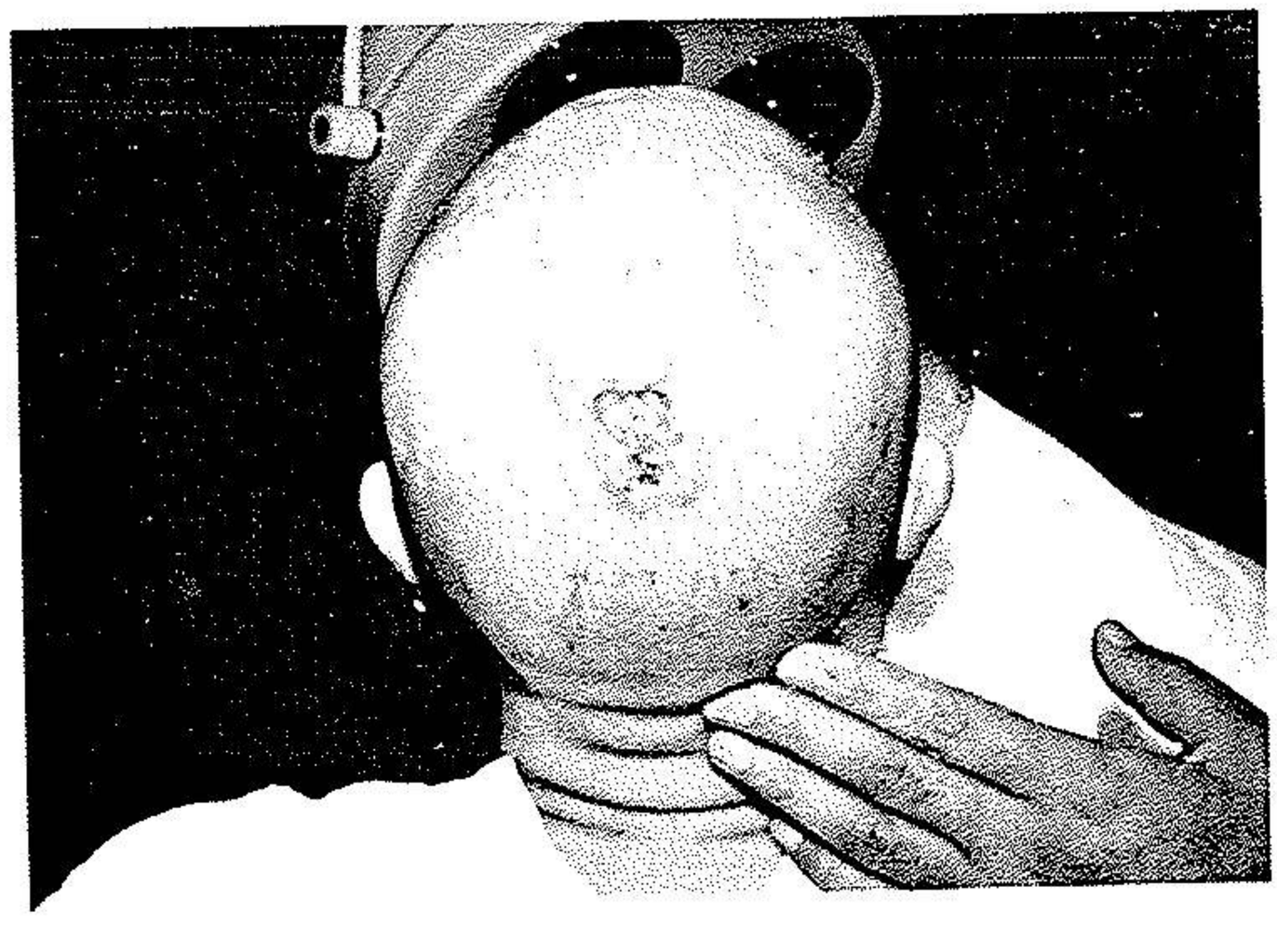

FIG. 5: Ulcer on the occiput of the patients. Repeated biopsies of this showed to be squamous cell carcinoma.

FIG. 6: Microscopic appearence of the papular lesion.Note hyperkeratosis with basket-weave appearence and acanthosis with vacuolated cells.

16-year-old brother has similar papules on the hands,posterior part of the chest and forehead since childhood the biopsy one of these papules has confirmed the suggested clinical diagnosis of erythrodysplasia verruciformis. However, no malignant change has yet been observed.

Commentary:

A case of E.V. with malignant degeneration has been reported in a 26-year old male and the literature is reviewed with the following conclusion:

(1) E.V. represents an unusual response to the wart virus. This explanation has been confirmed on histological grounds, successful inocu-

lation experiments and the demonstration of intracellular virus electronmicroscopically.

(2) A familial incidence is often reported suggesting that a predesposition to this exceptional reaction to the virus may be inherited.

(3) Malignancies of basal cell, squamous cell carcinomas, Paget's disease and other intermediated forms are known to occur in association with E.V.

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