Organoid Nevus Phakomatosis

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Received May 5, 1980

Abstract. A case of organoid nevus phakomatosis with CT (computed tomography) findings resembling those seen in tuberous sclerosis.

Several names have been proposed for the condition called organoid nevus phakomatosis (ONP) by Hornstein & Knickenberg (4): Jadassohn's nevus phakomatosis (11), linear nevus sebaceous syndrome (5), epidermal nevus syndrome (10) and Schimmelpennig-Feuerstein-Mims syndrome (4).

Feuerstein & Mims in 1962 first noted the association of central nervous system involvement with linear nevus sebaceous (1). Subsequently over 40 cases of the ONP have been reported.

The involvement of the central nervous system in the ONP closely resembles tuberous sclerosis (11).

The extensive, often laterally localized and linear nevi are situated on the skin of the face and scalp. The skin of the trunk and the extremities is sometimes also involved (4). Histopathological descriptions of the nevi are not uniform. Nevus sebaceous Jadassohn (1), epidermal nevus (10) and intermediate forms between these and other structures of the skin (5) have been described. Other congenital malformations in the ONP have been reported, such as involving the eyes and skeletal or vascular systems (4, 10).

Recently we have had the opportunity to follow a 1-year-old boy clinically presenting with the ONP. The CT findings, not previously reported in the ONP, show a similarity to tuberous sclerosis.

CASE REPORT

This boy was born the second child of healthy parents. The gestation time was 41 weeks, the delivery was normal and the Apgar score at one and five minutes was nine. Birth weight was 4230 g, length 54 cm and the head circumference 36 cm.

At birth, multiple, raised, hairless flesh-coloured or red-dish patches 3–5 cm in diameter were noticed on the scalp. Similar large, soft, flesh-coloured patches were girdling the middle of the trunk (Fig. 1). Smaller, nodular nevi were diffusely distributed on the skin of the trunk and the extremities.

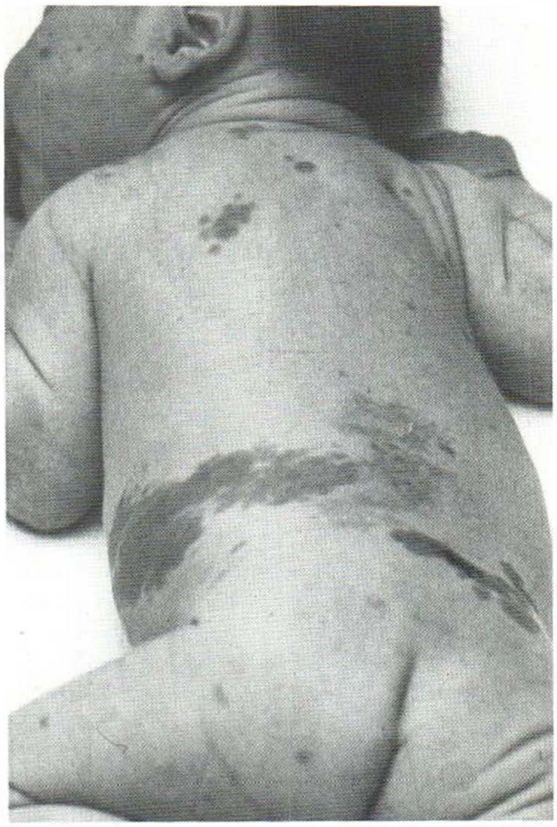

Fig. 1. Organoid nevus phakomatosis. A newborn boy with multiple soft, flesh-coloured patches on the skin of the trunk.

Numerous orange, somewhat verrucous nevi were also located on the facial skin (Fig. 2). On both earlobes there were several soft bulging nodules, almost blocking the meati.

The index finger of the right hand was thicker than normal and moved clumsily. Slightly hyperkeratotic, yellowish nevi involved the whole of its palmar aspect and extended linearly across the palm to the volar side of the wrist.

Several investigations were performed: skull X-ray was normal, the fundi were normal, as was the initial EEG. The thickened index finger showed thick bones in the X-ray (Fig. 3). At the age of 3 days brief seizures were seen with jerking in the extremities. Computed tomography (CT) was performed at the age of 3 months. Several nodular areas with increased density were seen in the periventricular regions (Fig. 4).

Skin biopsies were taken from three lesions. His-

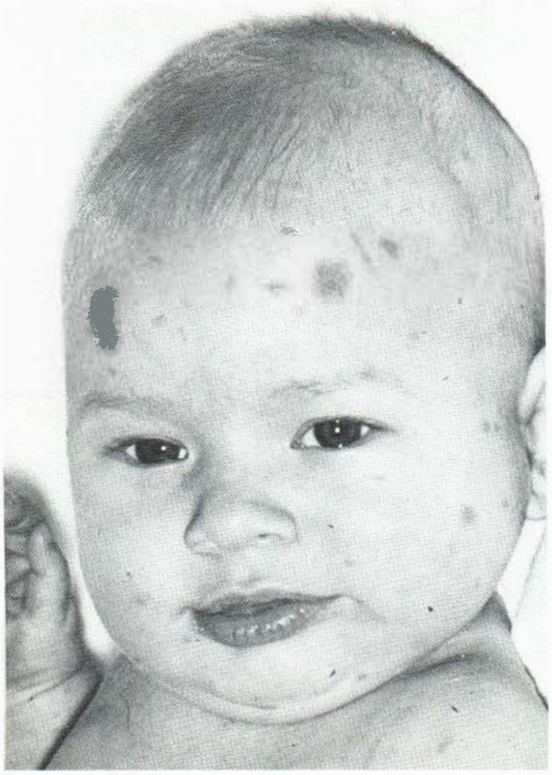

Fig. 2. Hairless patches on the scalp and multiple small, orange, verrucous nevi on the facial skin at the age of 2 months.

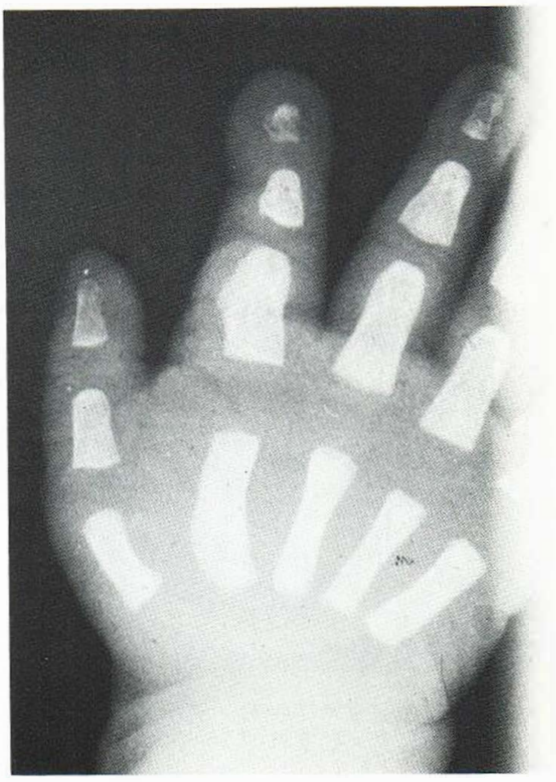

Fig. 3. The X-ray of the right index finger shows thicker bones than normal.

topathology of a lesion on the scalp showed (Fig. 5) hyperkeratosis and numerous epithelial buds resembling embryonic hair follicles. Some underdeveloped pilosebaceous complexes and hamartomatous dilated capillaries were seen in the dermis.

A specimen from the largest nevus on the forehead showed hyperkeratosis and acanthosis with elongation of the rete ridges. Some well-developed pilosebaceous complexes as well as epithelial buds resembling embryonic hair structures and an accentuated nevoid capillary component were also seen.

Histology of a lesion on the trunk showed hyperkeratosis, papillomatosis and acanthosis with some well-developed pilosebaceous complexes, but also epithelial cell buds resembling embryonic hair structures. Hamartomatous dilated capillaries surrounded by dense collagen were seen in the dermis.

During the first months the colour of the nevi become lighter, turning from red to flesh-coloured or yellow. Some of them seemed to reduce in size. Short, snow-white, kinky hair started to grow on some patches on the scalp. On oval white macule about 2 cm in diameter was observed on the right side of the middle of the trunk at the age of 6 months.

The seizures continued, phenobarbitone and diatsepam was given, but to no avail. At the age of 10 months calcifications emerged in the skull X-ray. The EEG showed only moderate excess of slow activity but no discharges, not even during a seizure. Nothing suggesting hypsarrhythmia was noted.

At the age of one year the boy can sit easily without support and is mentally alert. The head circumference has followed roughly the 50th percentile.

DISCUSSION

The clinical picture of the patient described above—low degree of mental retardation, infantile spasms and numerous large organoid nevi—agrees with the earlier descriptions of the ONP (11).

The lesions on the scalp were nevus sebaceous Jadassohn, while in the facial lesions there were features of epidermal nevi as also observed in the ONP by other investigators (4, 6). The lesions on the trunk in our case were epidermal nevi with a prominent nevoid capillary component. Histologically the nevus sebaceous Jadassohn demonstrated the infantile form with papillomatous epidermal

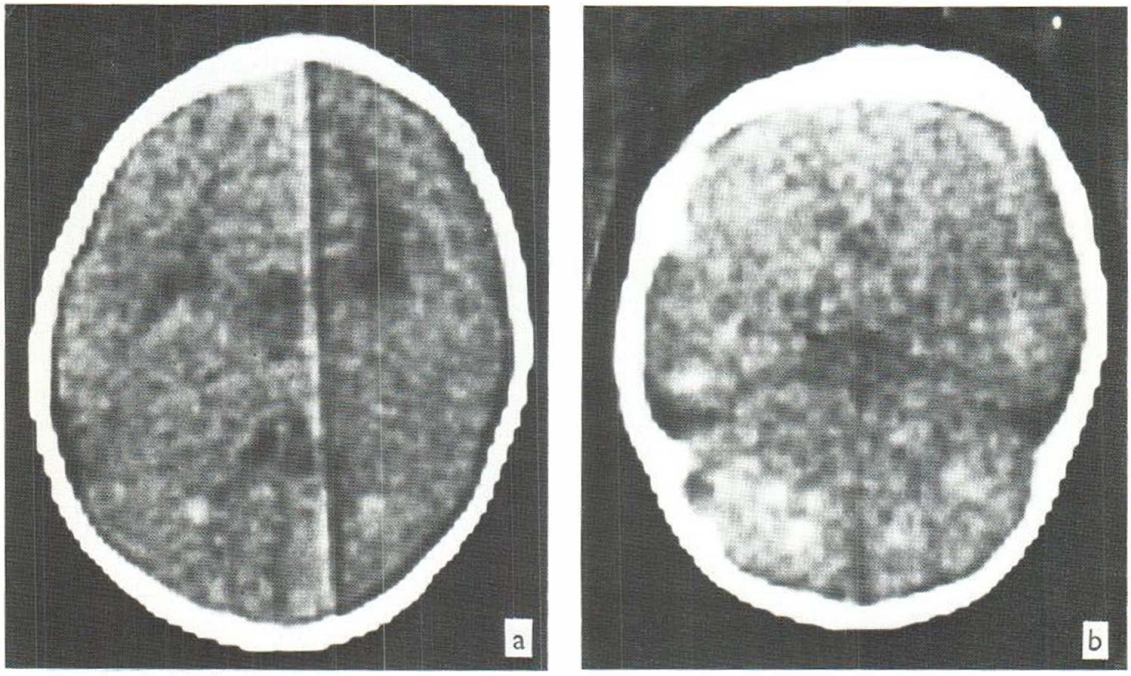

Fig. 4 a and *b*. The dense masses in preventricular tissue are clearly visible in computed tomography (*CT*) at the age of 3 months.

hyperplasia and underdeveloped pilosebaceous complexes (7). Pinkus & Mehregan (7) have used the name "organoid nevi" for all epidermal and adenexal nevi. Like Hornstein & Knickenberg (4) we also chose the name organoid nevus phakomatosis for our patient.

The only relevant disease in differential diagnosis is tuberous sclerosis. In the reports by Lovejoy &

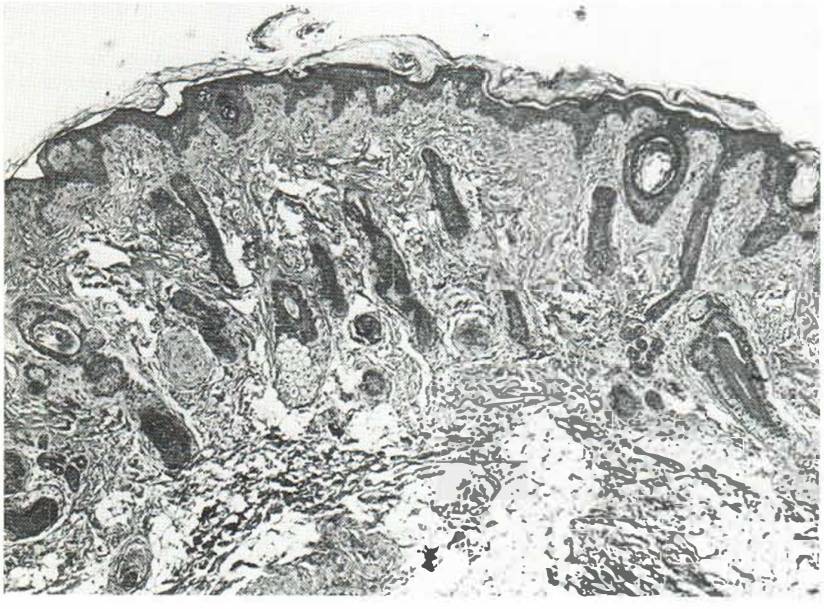

Fig. 5. Infantile form of Jadassohn's sebaceous nevus on the scalp with underdeveloped pilosebaceous complexes. Cords and buds consisting of germ-type cells are projecting from epidermis to dermis. (Herovici, ×25.)

Boyle (6) and Lansky et al. (5) the authors stress the possible confusion of the ONP with tuberous sclerosis. The present case gives rise to the question whether the ONP is a distinct entity as assumed by many authors (10, 11) or can perhaps be considered as a variation of tuberous sclerosis.

Skin lesions pathognomonic of tuberous sclerosis are "adenoma sebaceum" consisting oa angiofibromatous proliferation, and subungual fibromas. These lesions are practically never present at birth. Rather, leaf-shaped white maxules sometimes present at birth are the earliest diagnostically important cutaneous sign of tuberous sclerosis (2). Retinal phakomas are the most frequent ocular signs.

Computed tomography (CT) has become a useful investigative technique in the recognition of tuberous sclerosis also in cases without typical clinical signs (3).

Our patient did not show retinal phakomas. His neurological signs and CT findings might also lead to a diagnosis of tuberous sclerosis, especially when the bone defect, one leaf-shaped white macule and the radiologically confirmed calcification in the brain are considered together.

Can the present patient thus have simultaneously two phakomatoses? This possibility is not excluded, since case reports of the coexistence of tuberous sclerosis with Morbus Recklinghausen (8) and with Sturge-Weber syndrome (9) are to be found in the literature.

The picture of the ONP will be completed when CT findings in other patients are reported. Time will tell whether our patient will show further manifestations of the disease.

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Effects of Topical Erythromycin on Aerobic and Anaerobic Surface Flora

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Received March 12, 1980

Abstract. The contents of acne lesions from 30 men with inflammatory acne vulgaris, obtained before and at the conclusion of 4 weeks of topical 2% erythromycin therapy, were cultured on aerobic and anaerobic plates. Sensitivity testing of cultures was performed with discs containing 50 mg erythromycin, in order to ascertain the prevalence of erythromycin-resistant strains of *Propionibacterium acnes* and aerobic micrococci. The percentages of patients in whom erythromycin-resistant micrococci could be isolated increased from 3 to 60% during topical erythromycin therapy, but no resistant *P. acnes* strains were isolated either before or after therapy. Considering the widespread use of topical antibiotics, long-term studies on their ecological effects are desirable.

Key words: Surface flora; Topical erythromycin; Acne vulgaris

Ecological disturbances in surface microflora have been well documented with the use of systemic