

## Localized Scleroderma 'en coup de sabre' and Iridopalpebral Atrophy at the Same Line

Jørgen Serup and Poul Helge Alsbirk

*Departments of Dermatology and Ophthalmology,  
University of Copenhagen, Rigshospitalet,  
Copenhagen, Denmark*

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**Abstract.** A case is reported of scleroderma 'en coup de sabre' in a 13-year-old girl with atrophy of the nasal part of the iris and loss of cilia on the upper eyelid. The lesions of the front and eye were located on precisely the same line. The line did not follow the innervation fields of the cranial, peripheral or autonomous nerves. Nor did it follow the tension lines of the skin (Langer), or underlying anatomical structures. It is discussed that the predisposition to the 'coup de sabre' line seen in localized scleroderma was laid down in the mesenchyme in early foetal life before the differentiation of the anatomical structures.

In the 'coup de sabre' form of localized scleroderma with sclerosis at the classical paramedian line of the front, involvement of parts of the internal eye is uncommon, with only a few cases of sectorial atrophy and heterochromia of the iris reported (2, 9, 10). When the scleroderma is associated with atrophic changes of the lower part of the face, changes of the external eye and uveal tract are often seen (3, 7).

In the present article a case of scleroderma 'en coup de sabre' is reported, with associated segmental atrophy of the nasal part of the iris and upper eyelid, but no changes of the lower part of the face.

### CASE REPORT

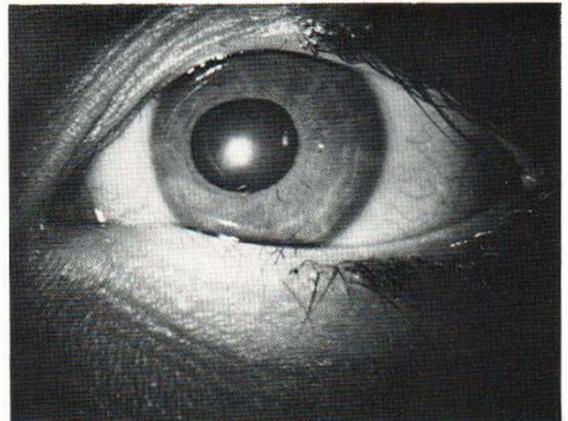
The patient, a girl born in 1966, was originally seen in the eye clinic at the age of seven years for symptoms of iritis persisting for one month. She was referred by her private ophthalmologist, by whom she had been followed from the age of four because of an oval and enlarged left pupil and loss of cilia in the nasal part of her left upper eyelid. The changes of the lid and pupil were confirmed at the eye clinic. Moreover, the left eye was pale, with fatty precipitates on the cornea endothelium, and there was a slight flare in the aqueous humor of the anterior chamber. Topical treatment with atropine 1% and prednisolone 0.5% as eye drops was started, and improvement was noticed after a few weeks. An examina-

tion at the paediatric outpatient clinic showed a normal child with good health. No cranial or dermal abnormalities were recorded.

At the age of 13 she was referred to the paediatric outpatient clinic, this time with an indolent nodule on her third right toe. For 2-3 years she had experienced monthly attacks of headache of increasing strength. The headache was hemifrontal, and the attacks were preceded by nausea, vomiting and photophobia. The nodule on the toe regressed spontaneously. No rheumatological diseases were found. She was referred to the eye clinic again, and examination of visual activity of the right eye showed 6/6+0.75, and of the left eye 6/9+1.5. On the right side the eyelids, the anterior chamber and fundus were normal. On the left side there was loss of cilia at the nasal part of the upper lid and atrophy of the nasal part of the iris (Fig. 1). Biomicroscopy revealed chronic precipitates on the corneal endothelium with a glossy appearance, in a pale eye with no aqueous flare. The iris was oval with moderate stromal atrophy in the nasal half, and the pigment epithelium was intact with no retrograde translucency. The pupillar ovalness was followed by non-reactivity to light and to convergence of the nasal part of the iris. The lens lacked synechiae. The chamber angle, the fundus, the intraocular pressure, and the electroretinogram were all normal. Central and nasal corneal thickness by Haag Streit pachymetry, and sensitivity by aesthesiometry gave normal and symmetric values. There was no involvement of eye motility, and no ptosis.

The second visit, 2 weeks later, added an essential finding to the ophthalmological status. As shown in Fig. 2, the changes of the iris and upper lid proved consistent with a paramedian cutaneous lesion in the left part of the front.

A retrospective survey of old portraits showed fully symmetrical and normal frontal regions as well as upper eyelids, cilia, and irides up to the age of four. Apart from



*Fig. 1.* Irido-palpebral atrophy of the left eye. The nasal half of the iris show paretic muscles and stromal thinning, while the temporal muscles and the stroma are normal. There is tarsal thinning and loss of cilia in the nasal half of the upper lid.



Fig. 2. Bilateral linear scleroderma 'en coup de sabre', primarily in left frontal region, with underlying atrophy including depression of the nasal part of the left supra-orbital margo.

the iritis episode, no inflammatory reactions had been noticed in the upper face and front.

Examination at the clinic of dermatology established the following status: On the left side of the front at a paramedian line the skin was slightly brownish pigmented, and on the anterior scalp, on the same line, there was an alopecic area, white, with increased dermal thickness and induration indicating dermal sclerosis (Fig. 2). There was underlying subcutaneous and pericranial atrophy, and a visible and palpable depression in the frontal bone and nasal part of the supraorbital margo.

In the right side of the front there were analogous linear changes, but the changes on the right side were only minimal. The face and skull exhibited no other changes of the skin, subcutaneous tissue, muscles or bone. The neurological status of the cranial nerves—including sensitivity of the lesions—was normal. No skin diseases were recorded on the trunk or extremities.

X-ray examination of the orbit and cranium showed no osseous disease. CT-scan showed paramedian depression and thinning of the frontal bone and supraorbital margo, but no underlying atrophy of the cerebrum or other pathological changes.

Light microscopy of a skin biopsy from the clinically sclerotic area on the anterior scalp showed an increase in dermal collagen indicating localized scleroderma.

## DISCUSSION

We find it remarkable that in this case of scleroderma 'en coup de sabre' the changes of the scalp, the front, and the external and internal eye were located on precisely the same line.

This paramedian line

1) did not follow the cranial nerve innervation

of the face and front, which is innervated by the trigeminal nerve and Gasserian ganglion (4);

2) did not follow the innervation fields of the peripheral nerves or the borderline between innervated fields. The borderline between the first and the second branch of the trigeminal nerve runs temporally to the eye and front. The supratrochlear and supraorbital branches do not innervate the internal eye;

3) did not follow the sympathetic and parasympathetic fibres, which have an irregular course in the head with different branches to the face and front, and to the internal eye;

4) did not follow the tension lines of the skin (lines of Langer), which are curved in the front with the convexity to the glabella;

5) did not follow other underlying anatomical structures such as muscles, cranial sutures and cerebral fissures.

The principal changes in this case were observed in mesodermal structures: dermis, subcutaneous tissue, muscle (including the iris) and bone. It is therefore conceivable that a common predisposition was laid down in the mesenchyme in early foetal life before the differentiation of the eye and other anatomical structures of the front and face, i.e. before 5–6 weeks of gestation. In this period the skull and upper face is expanding rapidly, and the eye and the maxillary process is progressing medially. At the same time neural crest-cells proliferate and migrate from behind to the face, on each side of the eye, in a disproportionate way to form parts of the soft tissue structures of the forehead (5). A congenital condition, median facial clefting, with hypertelorism and sometimes paramedian lines in the front can be seen as a probable result of defects in crest-cell migration (1, 5, 8). In another congenital condition with aberrant tissue bands, gross deformity of the midfrontal region medial to paramedian lines is often seen (6). These congenital conditions give relative support for the opinion that the predisposition to the 'coup de sabre' line seen in localized scleroderma is laid down in the mesenchyme in early foetal life before the differentiation of the anatomical structures.

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## Bilateral Follicular Basal Cell Nevus with Comedo-like Lesions

E. Crivellato,<sup>1</sup> G. Trevisan,<sup>1</sup> G. Grandi<sup>2</sup>  
and L. Lepore<sup>3</sup>

<sup>1</sup>Dermatological Clinic, <sup>2</sup>Institute of Anatomy and Histopathology and <sup>3</sup>Clinic of Pediatrics, University of Trieste, Italy

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**Abstract.** A 12-year-old white boy presented with symmetrically appearing papular, nodular, comedo-like, highly itching lesions involving the skin on his ankles. They had first appeared 3 years earlier. Numerous cutaneous biopsies showed tumoral buds of basalioma cells associated with follicular structures, infundibular cysts and also hypertrophy and hyperplasia of sebaceous glands. To the best of our knowledge, there are no reported cases with the same clinical and histopathological findings as in

those reported here. We believe the lesions represent a "bilateral follicular basal cell nevus with comedones".

**Key words:** Bilateral follicular basal cell nevus; Linear unilateral basal cell nevus; Basal cell hamartoma with follicular differentiation; Comedo-like lesions; Infundibular follicular cysts

The term "follicular basal cell nevus" denotes a heterogeneous group of histopathological features which are difficult to classify nosographically. In fact they are both related to tumours of follicular origin (3, 6, 7) and to multiple basal cell epithelioma (1) and basal cell nevus syndrome (4).

The case we present seems to belong to the follicular basal cell nevus group, although it differs from the other previously reported cases due to some clinical and histopathological findings.

### CLINICAL CASE

A 12-year-old white male came to our attention because of the presence of papular, nodular, crusted and comedo-like lesions localized on the internal and external aspects of the left ankle and the anterior aspect of the right ankle (Fig. 1). These lesions had appeared 3 years before.

The individual lesions consisted of numerous papular, nodular, pink-whitish, dome-shaped or flattened, solitary or bridged, slightly scaling elements, varying in size from 3 mm to 2 cm. Many of them were crusted; furthermore there was a large tumorous plaque on the right leg. Multiple clusters of comedones were present bilaterally. There were, moreover, some areas of slightly scaling "cigarette paper" atrophy.

Subjectively the patient felt intense itching on the affected areas.



Fig. 1. Clinical picture.