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Characteristic Striation and Ridge Pattern of the Cervical Skin in Lymphogenic Encephalopathy

By

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Földi *et al.* [1] observed a phenomenon similar to pseudoxanthoma elasticum on the skin of the neck of patients suffering from lymphogenic encephalopathy due to a failure of cervical lymph flow.

These patients displayed a peculiar pattern on both sides of the neck which, sometimes symmetrically, extended to the subclavicular area. It consisted of parallel, undulating or converging, slightly elevated ridges 1—2 mm to several cm long, which ran an oblique course above the clavicles. Inspection under a magnifying glass showed them to be composed of linearly arranged rows of densely packed or sometimes discrete pinhead-sized or smaller acuminate follicles from bright yellow to a pale bluish red in colour. The course of the ridges did not conform to the plication of the skin nor did they run in the direction in which the skin splits; their configuration was rather like that of the rows of elevated hair follicles which run obliquely downward on the neck [2]. On rubbing, the ridges become darker and assume a bluish red or red colour, while the intact skin between them becomes lighter. The contrast between the reddened ridges and the interlying paler areas make the skin of the neck appear to be striped, although there are neither atrophic nor scarred striae in this area. The cyanotic ridges become paler under pressure by a glass plate (Fig. 1).

Histological examination in some cases [1] revealed a destruction of the elastic fibres in the corium.

The phenomenon in question has been encountered in 85% of the examined cases of lymphogenic encephalopathy.

Schneider *et al.* [3] have shown that after ligature of cervical lymph vessels the cervical skin of dogs assumed a flagstone-like pattern. Histology revealed extensive fragmentation and destruction of the elastic fibres in the corium and the appearance of intraadventitial gaps in the vascular walls (Figs 2 to 5).

FIG. 2. Cervical skin of dog before ligature of cervical lymph vessels

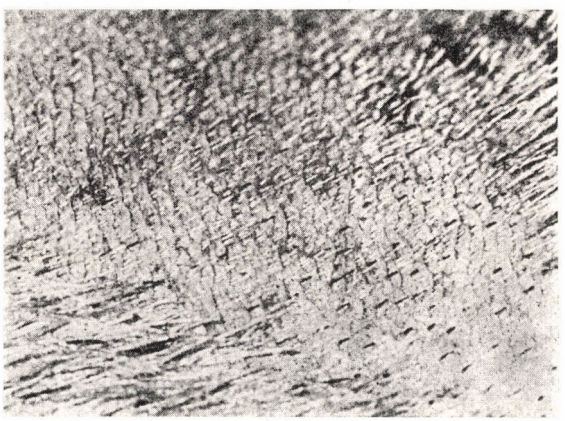


FIG. 3. Cervical skin of dog after ligature of cervical lymph vessels. Note flagstone-like pattern

FIG. 4. Extensive destruction of elastic fibres in the cervical skin of dog after blocking of cervical lymph flow

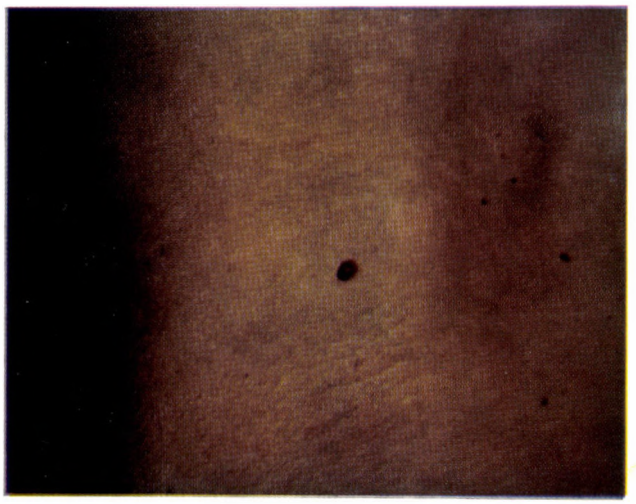


FIG. 1. Characteristic striation and folding of the cervical skin in lymphogenic encephalopathy

The present study had the aim to determine on a large material the extent to which the destruction of elastic fibres was characteristic of the said phenomenon.

Material and Method

Ten patients suffering from lymphogenic encephalopathy and displaying the pseudoxanthoma elasticum-like phenomenon and ten controls were studied. From their cervical skin a specimen was excised, stained with resorcin-fuchsin for elastic fibres, and examined.

Results

The corium showed a fragmentation of elastic fibres in eight of the ten patients and in five out of ten control cases (Fig. 6). The chi-square test as modified by Yates showed the difference to be significant statistically ($p < 5\%$). While the formation of intraadventitial gaps was observed in three cases of lymphogenic encephalopathy, no such phenomenon occurred in the controls (Fig. 7).

Discussion

Striation and specific ridge pattern on the neck are characteristic concomitants of lymphogenic encephalopathy. They have to be distinguished from the initial form of pseudoxanthoma elasticum, a distinction difficult even for skilled dermatologists. Pseudoxanthoma elasticum commonly affects both sides of the neck and often begins with hardly perceptible manifestations. Small yellowish white papules may develop which slightly emerge from the skin; if discrete, the papules may resemble localized cutaneous lesions as found in lymphogenic encephalopathy. At a later stage, the diffuse infiltration, atrophy and eventual haemorrhages already ensure the diagnosis.

What the above described skin pattern resembles perhaps most closely is the condition called cutis punctata linearis colli or stippled skin [4]. In this disease, pinhead-sized or larger yellowish white, hypertrophic sebaceous glands in parallel, sometimes irregular or undulatory, rows, appear mostly on the neck, the nape, the upper part of the chest or on the shoulders. The skin around the glands is bright red or brownish, cyanosed. If these phenomena occur after prolonged corticosteroid or testosterone treatment, they may be accompanied by the so-called steroid acne. The histological picture is characterized by atrophy of the epithelium and connective tissue and by enlarged sebaceous glands. Interruption of testosterone or cotricosteroid treatment is usually followed by a disappearance of the change. The condition may be elicited also by intensive sunbathing; in this case the picture is termed erythrosis interfollicularis colli (Leder).

The cervical lymph nodes collect lymph from the cervical skin, from the eyes and the brain, and it is this anatomical arrangement which explains why striation and the characteristic ridges of the cervical skin are such a frequent symptom of lympho-

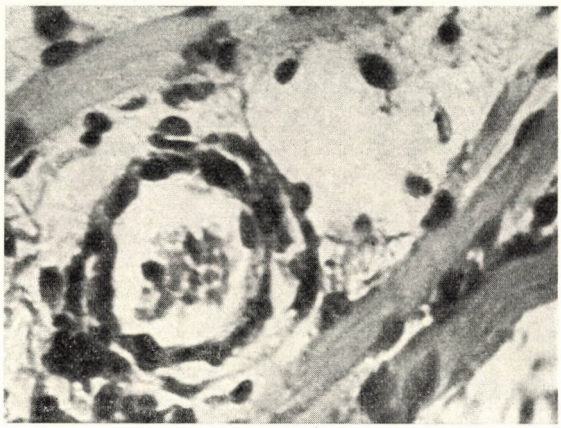


FIG. 5. Formation of intraadventitial gaps in blood vessels of the cervical skin of dog after blocking of cervical lymph flow

FIG. 6. Fragmentation of elastic fibres in middle layer of the corium. Cervical skin of patient suffering from lymphogenic encephalopathy

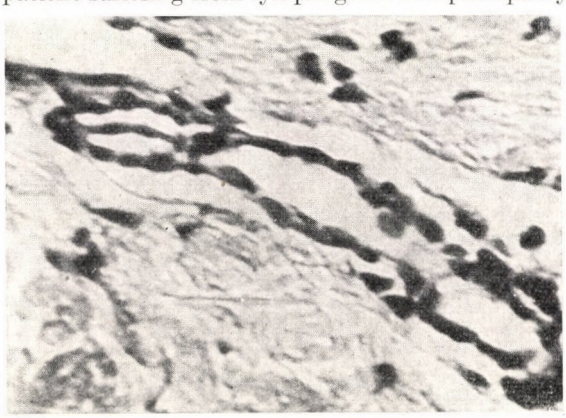


FIG. 7. Intraadventitial gap in blood vessels of cervical skin of patient suffering from lymphogenic encephalopathy

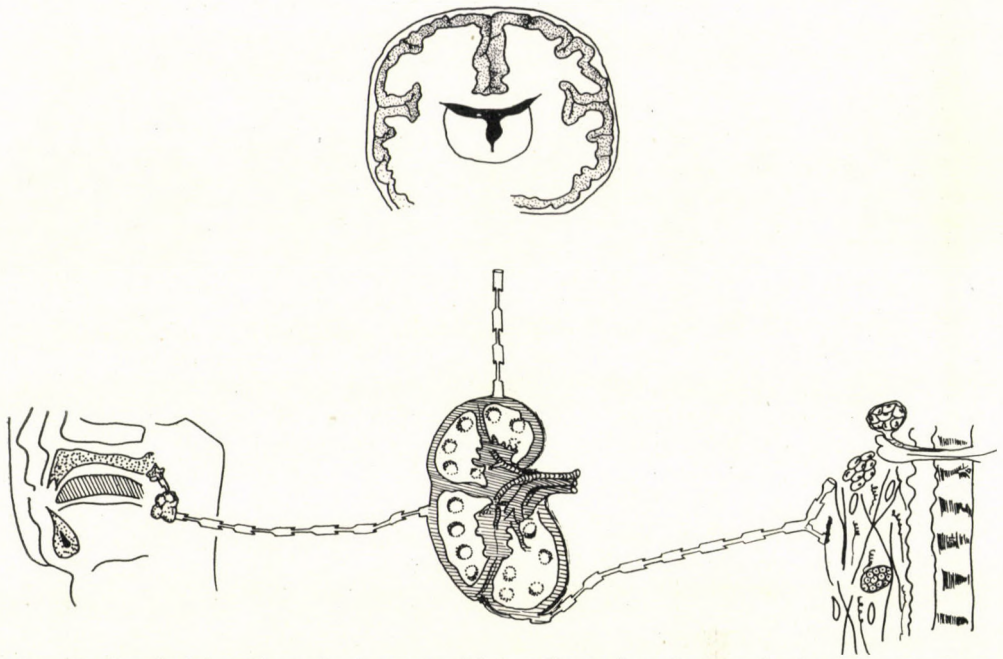


FIG. 8. Cervical lymph node (centre) which collects lymph from brain (above), from Waldeyer's tonsillar ring (left) and from skin (right)

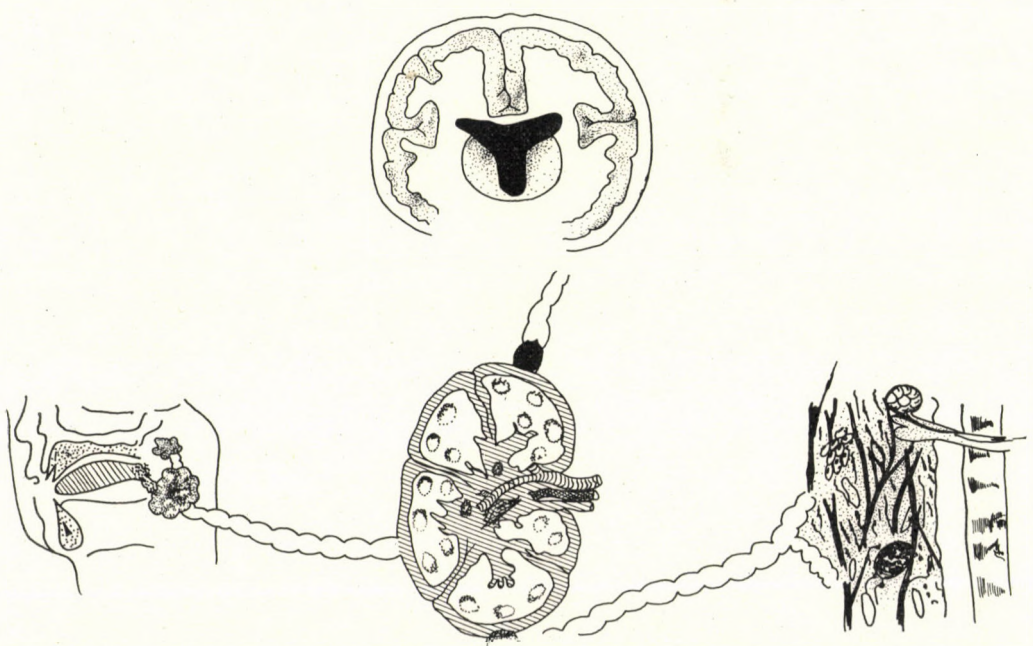
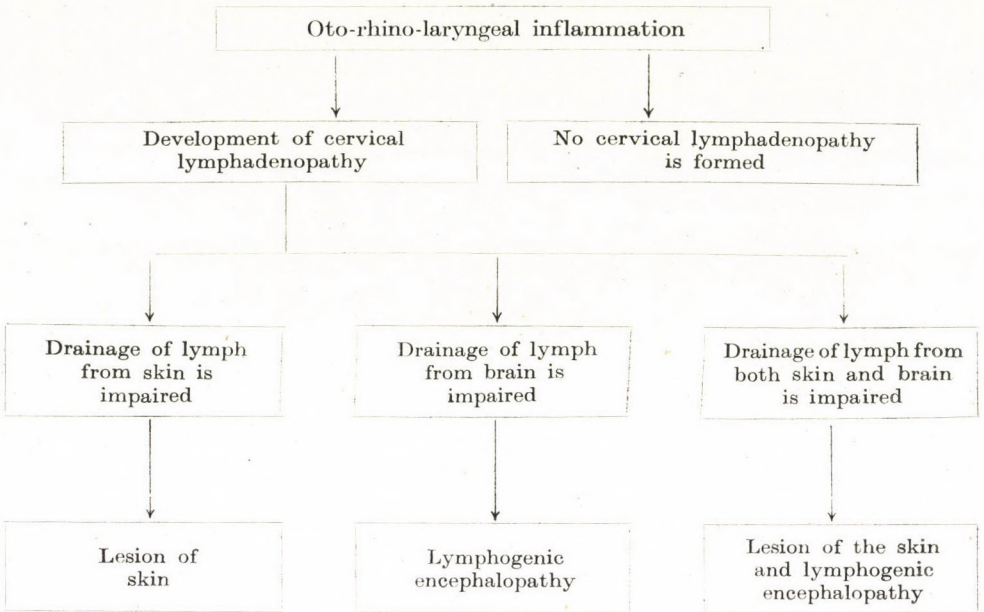


FIG. 9. Inflammation in the area of Waldeyer's tonsillar ring had led to cervical lymphadenopathy. After blocking of cerebral lymph flow by a thrombus in the lymph vessel at its opening into the lymph node, cerebral oedema and hydrocephalus have developed. Note oedema of skin

TABLE I



genic encephalopathy (Figs 8, 9, Table I).

SUMMARY

Striation and a peculiar ridge pattern of the skin of the neck have been observed as a characteristic feature of lymphogenic encephalopathy. Histological examination revealed destruction of the elastic fibres and the occasional appearance of intraadventitial gaps.

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