THE HAND AS A SIGN-POST TO OCULAR DISEASE—III

Neurological Disease

Acromegaly (Fig. 1)

FIG. 1.—The spade-shaped hands of acromegaly demonstrating the scar from median nerve decompression.

Acromegaly results from an acidophilic adenoma of the pituitary occurring in an adult. The features are coarse, with prognathism, macroglossia, and enlarged extremities. Endocrine disorders are common and diabetes occurs in about 15 per cent. of cases.

The spade-shaped hands have a thick skin and an increase in subcutaneous tissue which may compress the median nerve in the carpal tunnel. Arthritis and peripheral neuropathy may also occur.

Optic atrophy with bitemporal field defects results from compression of the chiasm. Acute severe unilateral headache with oculomotor paralyses may indicate haemorrhage into the adenoma (pituitary apoplexy). Patients with acromegaly appear to have an increased incidence of simple glaucoma.

Myotonia (Fig. 2)

FIG. 2.—Myotonia demonstrating the hand 4 seconds after voluntary relaxation.

Myotonia is the inability to relax after muscular contraction and occurs in two main hereditary conditions: (1) Myotonia congenita (Thomsen's disease), which often appears soon after birth, is a generalized myotonia with a good prognosis. (2) Myotonia dystrophica develops in adolescence and involves mainly the hands and the tongue, though an associated muscular dystrophy is also present. Frontal baldness, gonadal atrophy, mental deterioration, and endocrine disorders are other features of this condition.

The hands demonstrate a delayed relaxation of the muscles with weakness and wasting.

Ptoxis, abnormalities of ocular movements, and sluggish pupillary responses may occur. In myotonia dystrophica degenerative changes may occur in the cornea, and the lens usually shows multiple punctate anterior and posterior subcapsular opacities.

THE HAND AS A SIGN-POST TO OCULAR DISEASE—III

Syringomyelia (Fig. 3)

This slowly progressive degenerative condition results from a cavity appearing in the central grey matter of the spinal cord. This may extend into the brain-stem (syringobulbia). A dissociated anaesthesia occurs from interruption of the spinothalamic tracts mediating pain and temperature, while other modalities are unaffected. As the condition progresses, weakness and wasting of the upper limbs occur and brain-stem signs appear.

The hands show trophic changes, often with indolent ulcers, and are enlarged and brawny with multiple scars from previous trauma (*main succulente*).

The main ocular defects are from brain-stem involvement and include Horner's syndrome, rotatory nystagmus, and paralyses of ocular muscles. Optic atrophy has been reported.

FIG. 3.—Syringomyelia.

Wasting of the Small Muscles of the Hand (Fig. 4)

This may result from neurological lesions in the spinal cord or the first thoracic nerve. In Pancoast's syndrome wasting occurs because of an apical pulmonary lesion which may be neoplastic, with an ipsilateral Horner's syndrome. In Refsum's syndrome a peripheral neuropathy, nerve deafness, and ichthyosis, occur with atypical retinitis pigmentosa. Many diseases may involve both peripheral nerves and optic or oculomotor nerves. Infective processes (leprosy, diphtheria, Guillain-Barré syndrome), deficiency diseases (vitamin B group), and toxins (apiol, isoniazid) may combine ocular abnormalities with wasting. Systemic diseases, such as sarcoidosis, amyloidosis, collagen diseases, and malignant processes, may involve peripheral nerves concurrently with their more characteristic ocular signs.

FIG. 4.—Pancoast's syndrome with wasting and sensory loss over the first thoracic distribution.

Illustrations:

INSTITUTE OF NEUROLOGY, LONDON, W.C.1. (Figs 1-3).
WESTMINSTER HOSPITAL, LONDON, S.W.1. (Fig. 4).

M. D. SANDERS,
MOORFIELDS EYE HOSPITAL,
LONDON, W.C.1.