Some Unusual Syndromes

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SOMEx UNUSUAL MEDICAL SYNDROMES*
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The word "syndrome" has been in recognized use since 1541. Even after more than 400 years it continues to be one of the most misused terms in our medical vocabulary. It is most frequently used as a synonym for such terms as symptom complex, clinical entity, pathologic process or disease.

Dictionary definitions of a syndrome are vague. The term is defined as a con­currence or running together (togetherness) of abnormal signs or symptoms. In the Spanish language it is interpreted as any disturbance of physiology. As used in French medical literature, this convenient and elastic term seems to be an indispensable one. It is rarely used in German medical writing. More correct usage has broadened the concept to include as a composite group two, three, or even four totally unrelated conditions which occur in combination sufficiently often to symbolize a syndrome. The gradual evolution and final recognition of the component parts of a typical syndrome may require years or decades. A characteristic example is Van der Hoeve's syndrome, which consists of blue sclerae, brittle bones and otosclerosis with deafness. Innumerable other odd combinations could be cited. Unfortunately, the literature also abounds in such loosely used terms as "brassiere strap syndrome".

Categorically, syndromes may be classified as (1) eponymic, those which bear the name of one or more individuals who contributed to the clarification of the subject, and (2) descriptive. Most true syndromes are either congenital, familial or the result of genetic abnormality. Frequently only a partial anomalism is present. Syndromes occur in various fields of medicine, especially in disturbances of multiglandular physiology, dermatology, neurology, and ophthalmology. As a rule, they evoke more interest and are more challenging than a disease, because the relationships are obscure, and the etiology is usually unknown. Their variable characteristics will be more evident after a review of the small group herewith presented.

"Women Who Fall" syndrome. From this name it may be inferred, perhaps not incorrectly, that a permissible synonym is "fallen women." This is a narcissistic syndrome, occurring only in women and characterized by stumbling and falling without apparent reason. While talking or walking and without encountering the slightest obstacle, the patient may fall. After one or more such incidents, a neurologist is usually consulted. If the syndrome is promptly recognized, the patient may be referred to a psychiatrist or to a psychoanalyst. As an example, psychoanalytic study of one woman who suddenly fell to the street indicated that at the moment she fell she was thinking that the imminent betrayal of her husband was well justified because he was effeminate. An analytic interpretation of the emotional and personality make-up of such a patient indicates an ego that is weak, inconsistent, and, in fact, almost non-existent. Under certain circumstances, this timid ego may be submerged by anxiety and wholly or partly obscured. When the obscurations are total, the ego vanishes for a fraction of a second, falling occurs.¹

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“Alice in Wonderland” syndrome. It requires an elastic imagination to categorize as a medical syndrome the story of “Alice in Wonderland”, which has delighted so many readers. Through psychoanalytic interpretation, the illusionary dreams, feelings of levitation, and alteration in the sense of passage of time which Alice experienced have been made more understandable; in essence, Alice trod the paths and byways of a wonderland well known to her creator, Lewis Carroll. He suffered severely from migraine and many epileptic and migrainous patients experience bizarre disturbances of body image. Alice, as you will recall, dreamed that she was either remarkably tall or remarkably small, was a dual personality, and had illusions concerning the size, position, or distance of objects. In other words, according to the analysts, the record of her bizarre experiences was essentially a mirror of the experiences suffered by Carroll as a sequel to his migraine. According to neurophysiologists, the site of origin for such psychic alteration may perhaps be in the parietal lobe. Some degree of reproduction of similar symptoms has been reported to occur after electrical stimulation of this area.

Bogorad’s syndrome. This title is not a euphonious one. Its synonym, the syndrome of crocodile tears, evokes more interest. This latter title has been applied because it was formerly believed that the crocodile wept hypocritical tears while devouring its victims. Clinically, this syndrome is characterized by paroxysms of lacrimation which occur during eating. The exact etiology is obscure, but the condition has been observed after facial nerve palsies or in association with certain tics. The nerve fibers which normally cause salivary gland stimulation become misdirected to the lacrimal glands and produce tears. The condition is apparently due to a deviation of regenerating axons from their normal course. A similar overflow of nerve impulses into adjacent nerves has been seen in other neurologic conditions which will be mentioned.

Marcus-Gunn syndrome. This neurologic phenomenon is also known as the jaw-winking syndrome. Characterized by conspicuous increase in the width of the eye-slits, the syndrome occurs during mastication. In some instances, there is a rhythmic elevation of the upper eyelid when the mouth is opened and a drooping of the lid as the mouth is closed. Widening of the palpebral fissures is not limited in its occurrence to chewing motions. It has also been observed during loud speaking or shouting, while abducting the jaw to one side, and in children during episodes of crying. The condition is congenital, often occurs in successive generations, and one report described eighteen affected members in three generations. Neurologists and neurophysiologists suggest that the condition is best explained by a supranuclear lesion, a probable connection of the third and fifth cranial nuclei in the posterior longitudinal bundle or the corticopontine tract.

The two neurologic syndromes just described have their clinical counterpart in other synchronized neuromuscular motions. Observations show that cats raise their eyebrows as they open their mouths to eat. In addition, the writer has also observed and demonstrated in a posthemiplegic patient flexion of the affected hand and forearm and some elevation of the entire arm which invariably occurred during the act of yawning.

These several neurologic aberrations strongly suggest that under certain circumstances there may be a short-circuiting or misdirection of impulses from one
Durham cranial nucleus or nerve into another with consequent disturbance of effects upon the end organs.

Gilles de la Tourette syndrome. This interesting syndrome was first described in 1884 by a well-known French neurologist, whose eponym it bears. It is characterized by incoordinated motor movements, including facial twitchings or tics, blinking of the eyes, or jerking of the face, arms, or legs. These movements are often accompanied by verbal exclamations, which include profanities, swearing, and coprolalia, that is, the uttering of filthy words. The verbalization is often explosive, accompanied by imitative gestures. The onset of this strange behavior often occurs by the age of ten years, and the extent of the profane vocabulary at this age is sometimes astounding. The syndrome is not infrequently brought on by strong emotional stimuli or sudden noises. Affected patients find it difficult to concentrate and are easily distracted. With increasing age they tend to withdraw from social intercourse. The etiology is not known. Psychotherapy has been tried without noticeable success.

Stiff-man syndrome. This peculiar myopathy is characterized by generalized muscular spasm, rigidity, and myalgia. The onset may be rapid, developing within a few days, or more slowly, extending over several months. There is marked fluctuation in the intensity of the symptoms; the severity of the pain parallels the degree of muscular spasm. Muscles of the trunk are usually first affected. As the disorder progresses, other muscle groups become involved symmetrically until the effect of the over-all rigidity resembles a poker spine. The gait becomes stiff and laborious, and the patient justly becomes apprehensive about impending total rigidity.

Detailed studies of numerous patients have revealed no biochemical disturbance, except reducing substances occasionally found in the urine. No etiologic concept has been formulated. It has been postulated that the rigidity may occur reflexively by way of the spinal cord or that the basal ganglia are involved. Antispastics have not been helpful in treatment; in fact, no measure has been effectual.

Restless legs syndrome. Paresthesias and a sensation of restlessness felt in the legs, with inability to sit still, are characteristics of this syndrome. It occurs in mentally healthy patients, more often in women. Symptoms are felt only when the legs are at rest and consist of an unpleasant creeping sensation which seems deep in the tissues of the leg, more often below the knee, although the thigh is sometimes involved. The condition may occur while in the theater or at home, perhaps while entertaining certain friends. Many patients will admit that the symptoms are most likely to occur during a period of boredom. Movements of the legs or walking affords relief. Symptoms are usually periodic. In severe cases the feeling continues for several hours and may occur in rare instances night after night for years. The cause is not known, and no cure has been found.

Kleine-Levin syndrome. This unusual condition is characterized by hypersomnia and abnormal hunger. It apparently belongs to the group of narcoleptic disorders. The hypersomnia occurs in crises and is accompanied by slight agitation or mental confusion. In one recorded case observed in Coventry, England, the patient slept like a child during an eleven-hour period of intensive bombing and destruction of the city. Attacks occur at irregular intervals and may last two or three days. Males are more frequently
affected. Various studies have thrown no light whatsoever on the cause. Electroencephalograms have for the most part been normal. There is no evidence of a frontal lobe tumor which often causes an excess of hunger. Glucose tolerance tests are also normal, and there is no evidence to suggest a pancreatic tumor. Some observers believe that the syndrome is the result of a lesion in the area of the hypothalamus and third ventricle.

Anton's syndrome. This unusual eponymic syndrome was first described by Anton over half a century ago. Also known as amaurosis denial syndrome, it consists of blindness with confabulation and denial that the blindness exists. Besides stoutly denying their inability to see, afflicted patients describe objects which they believe they can see. Eventually, however, the patient may recognize that the blindness exists. The mechanism of the syndrome is not known, but it has been postulated that it is due to isolation of the diencephalon from the occipital lobe. In certain cases, the denial part of the syndrome is thought to relate to a lesion of the calcarine-thalamic connections.

Spasmodic laughter syndrome. Episodic outbursts of irrepresible laughter which occur without any idea of merriment are occasionally observed. The laughter may be spontaneous without any apparent psychic stimulation, or the paroxysms may be initiated by such simple movements as pointing a finger at the unfortunate subject. The episodes are usually recurrent, may be relatively brief, or may continue incessantly to the point of complete neuromuscular fatigue.

Pathologic laughter is usually a sequel to some type of organic brain lesion and may occur in association with epilepsy, postencephalitic states, more rarely in multiple sclerosis, or pseudobulbar palsy. It also occurs after prefrontal lobotomy. Cerebral pathology may usually be demonstrated by electroencephalographic study.

A more severe type of almost continuous episodic laughter has been described as a part of the symptom complex of Kuru, a puzzling, highly-fatal, epidemic, neurologic syndrome which was recently discovered in New Guinea. Because of the associated lesions, the prognosis of the spasmodic laughter syndrome is usually grave.

BIBLIOGRAPHY