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TWO DIFFERENT TYPES OF EPIDEMIC ENCEPHALITIS: THE LETHARGIC AND THE MYOCLONIC FORM OF THE DISEASE

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The systematic and comparative study of many cases of encephalitis, which we are prosecuting at present at the Principal hospital in Milan, allows us to determine the clinical types of this interesting disease.

Besides the form which we will call the *classical form* of lethargic encephalitis, in which lethargy, fever and ocular paralysis are the cardinal symptoms, we must also consider other clinical varieties, less frequent but greatly different.

While in its usual form everything tends to torpor, sleep and paralysis, we can sometimes observe cases in which the symptoms are, on the contrary, excitement, delirium and myoclonia.

The two following cases are of this second type (*myoclonic type of Epidemic Encephalitis*).

CASE I. A girl, aged 19, entered the hospital on the 21st of January last. No hereditary antecedents. She had influenza with broncho-pneumonia in October, 1918, and typhoid fever in January, 1919.

On January 17 she began to feel great pain in the muscles of the neck and shoulders. A few days later she had fever and delirium. On entering the hospital her symptoms were: Light cyanosis of the face, irritation of the ocular conjunctives, irregular contractions of

the hands, delirium, temperature 38.2° C., pulse 104, respiration 26, arterial blood pressure 145 mm. Hg. (Riva Rocci). No paralysis of the eyes nor of the cranial nerves. Pupillary reflexes to light and to accommodation present and well maintained.¹

Cutaneous and tendonal reflexes normal. A lumbar puncture gave the following result: Liquid perfectly clear at almost normal pressure; albumen just noticeable; Nonne and Boveri's reaction negative; six lymphocytes per cu. mm. to the cell count (Cellule de Nageotte). Blood and spinal liquid cultures gave negative results.

The patient remained in this condition for about ten days. On the 4th of February *short and rhythmical contractions* appeared, which could be compared to *electric waves* in the flexing muscles of the arm and of the hand and fingers on both sides, but mostly on the right side.

These rhythmic movements are 66 to 68 per minute (P. 92, R. 22, T. 37° C.).

In the right thigh a contraction of the quadriceps was observed which produced attempts at flexion of the thigh. No sensory troubles. Two days later, to the above movements light contractions of the diaphragm were added which produced inspiratory contractions, synchronizing with the contractions of the arm. These movements remained even during sleep.

There was no lethargy. The pupillary reflexes to light and to accommodation were always regular; no visible paralysis of the cranial nerves.

This is the present condition of the patient who now has pain in the arms, temperature between 36.4° and 37° C.

CASE 2. A woman, aged 40, entered hospital on the 16th of February last. No hereditary antecedents, no influenza. On the 25th of January there had been great pain in the arm and shoulder on the left side, which passed on to the right side and to the wall of the abdomen. Shortly after, rapid and rhythmical muscular contractions appeared, which convulsed the patient. There was delirium, with slight fever, no lethargy, no affection of the ocular motion nor of the sight. On entering the hospital this woman, of average physical development and well nourished, offered a *most myoclonic symptomatology*. The *sterno-cleido-mastoid* and *trape-*

¹ See P. Boveri, "Abolition du Réflexe de l'Accommodation dans l'Encephalite Léthargique," Société de Neurologie de Paris, Séance du 4 mars, 1920.

In the above work Boveri states that in the lethargic form of encephalitis, one of the first and most important signs is the paralysis of pupillary accommodation, while the pupillary reflex to light may be maintained. This sign would not be present in the myoclonic form of epidemic encephalitis.

zoid muscles of the right side and the *diaphragm* presented *rhythmic contractions*, 48 per minute.

She perspired freely, was agitated and delirious, no paralysis of the ocular and cranial nerves. At the lumbar puncture, clear liquid with increased pressure, albumen 0.2 per cent. (Rachialbuminometre de Sicard). Nonne and Boveri's reactions negative, five elements per cu. mm. to the cell count (Cellule de Nageotte); T. 37.8°–38° C. No symptoms in the lower limbs. All reflexes normal.

Five days later the contractions of the neck disappeared, whilst in the wall of the *abdomen* strong *rhythmic convulsions* were observed, which never for a moment left the patient.

Synchronizing with the abdominal and diaphragmatic convulsions, the *right thigh* presented rhythmic contractions of the abducent muscles and sometimes of the sartorial muscle.

All these muscles are painful, as well as the back and the whole arm. Such is the patient's condition at present.

The new and striking symptoms of these two cases consist of myoclonic convulsions, both rhythmic and partial, as though produced by an electric current.

We can therefore speak of a *myoclonic type* different from the *lethargic type*, at least at a certain period of the disease.

Thus, waiting for the etiological moment to enlighten us, we ought to speak of *Epidemic Encephalitis* in general, to which might be added a lethargic or myoclonic type according to the different symptoms noted.

On studying this myoclonic form of encephalitis, we naturally ask whether we are in the presence of the disease described by Dubini and called by him "*Electric Chorea*."

This disease has not been mentioned since Dubini's time, 1840–1870. Dubini presented to his seventh meeting of Italian scientists (1846) a memorandum describing a new disease of which he had studied thirty-eight cases in a period of nine years. It is, Dubini writes, "nearly always a fatal disease," characterized "by muscular contractions occurring at different intervals, but always identical in each case, as if produced by a repeated electric current. These convulsions, affected at first a finger or limb (more frequently the upper limb on the right side) or a half of the face (the right half) and in a few days invaded the whole of that side of the body." Dubini adds that, as well as these rhythmic movements, two or three or even more times a day, one can observe convulsive attacks which repeat themselves every day, giving way afterwards to paresis and

paralysis. Dubini's description was followed by those of Frua (2), Morganti (3), Pignacca (4), Stefanini (5), Behrend (6) and, later, Grocco, 1884 (7).

But, however well these scientists had observed the symptoms of this disease, they were not able to understand them, on account of the limited knowledge of pathological anatomy at that time, for we must not forget that they lived about the middle of the last century.

Reading all these memoranda to-day, we see described under the name of Dubini's disease a great number of cases which could better be classed as typhoid or malarial fevers or Jacksonian epilepsies.

But, without entering into this question, it really seems to us that Dubini's disease might be connected with the myoclonic form of epidemic encephalitis, as we see it at present. It is also interesting to observe that in the two cases described above there were no pupillary affections to light or to accommodation tests, and that the ocular movements were also normal.

The different symptomatology between the lethargic and the myoclonic types suggests a different pathological localization of the virus. In the lethargic form the centers of encephalitis are localized, particularly on a level with the central peduncles, and the locus niger. It is a question whether, in the myoclonic type of the disease, the centers might not be localized in the opticus thalamus in different degrees in a transitory manner?

We are waiting for new discoveries in pathological anatomy to enlighten us on this point.

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3. Morganti. *Gazetta Med. Lomb.*, 1853-1854.
4. Pignacca. Della Corea Elettrica osservata in Pavia dall'anno 1848 al 1854; Pavia, Tipogr. Fusi, 1855.
5. Stefanini. Sulla Corea Elettrica, *Annali Universali di Medicina*, 1875, p. 201.
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6. Behrend. Ueber Spasmus Dubini oder Sogenante Chorea Electrica, *Journ. f. Kinderkr.*, XX, 34 (1854).
7. Grocco. Studi e Considerazioni sulla Corea Elettrica o Malattia del Dubini, *Annali Universali di Medicina*, Vol. 269, anno 1884.