IRON AND CALCIUM IN STURGE-WEBER DISEASE

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THIS is a report of the chemical analysis of iron and calcium in the brains of three cases of Sturge-Weber disease. Although the clinical and pathological features of this syndrome have been well described, the chemistry of the characteristic deposits is less well understood, and there is a conflict of opinion as to whether iron is present in increased amount.

Owing to the kind co-operation of Mr. Alexander of the Neurosurgical Unit, Frenchay Hospital, it has been possible to re-investigate this problem using material derived from lobectomy specimens. The material had been stored in formol saline for some months before the analysis was carried out.

Microscopic examination showed the usual picture of meningeal angioma and massive mineral deposits in the outer part of the affected grey matter. There were also a few concretions lying more deeply in the white matter in all the specimens (Dr. R. M. Norman).

PRIOR LITERATURE

Chemical assessment of the iron and calcium in the cortex has been made in cases of this type by Eaves (1926) and by Wachsmuth and Löwenthal (1952). Eaves found a very large excess of total iron in a case preserved in formol; whereas Wachsmuth and Löwenthal, in fresh material, found no excess—the latter authors showed that preservation in formol raised the iron content but not to a level anything approaching that found by Eaves. All these authors found a large excess of total calcium. It should be noted that the iron estimated by these authors will include that due to the blood. Also that they did not assess the inorganic or readily available iron, which would have been increased in the case of Sturge-Weber disease if there had been a conversion of part of the organically bound iron to the inorganic form.

METHODS

"Inorganic" iron was estimated by the method of Tompsett (1935) and "Total" iron (i.e. all the iron except that due to the bound iron of the blood) by the method of Tingey (1937).

Total calcium was estimated by ashing and precipitation of the calcium from an HCl solution of the ash by saturated ammonium oxalate in the presence of a sodium acetate buffer at pH 5·0—the oxalate precipitate was re-dissolved in hot dilute H_2SO_4 and the liberated oxalic acid titrated against $KMnO_4$.

RESULTS

It will be seen from Table I that no excess of inorganic or total iron, over the control specimens preserved in formol, was found, either in the cortex or white matter.

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TABLE I
(Number of cases in brackets)

Cortex mg./100 m. Wet Tissue	"Inorganic" *Fe*	Total	Ca Total
Control fresh	1·28 (10)	2·99 (6)	6·60 (1)
Control formol	3·24 (5)	5·51 (5)	13·60 (5)
Case 1	2·27	4·30	10,824
Case 2	1·18	3·85	1,660
Case 3	3·37	5·75	490
White matter:			
Control fresh	1·23 (9)	2·59 (6)	8·00 (1)
Control formol	3·12 (3)	4·96 (5)	9·33 (5)
Case 1	1·20	1·80	156
Case 2	2·67	4·52	171
Case 3	3·35	5·80	110

TABLE II

Cortex mg./100 gm. Wet tissue	Ca	P	Ca in Excess of $Ca_3(Po_4)_2$
Calcified nodule hot acetic acid ..	10,500	3,800	8,600
Calcified nodule hot saline ..	220		
Homogeneous paste hot acetic acid	1,695	137	1,430
Homogeneous paste hot saline ..	149		

A large excess of total calcium as compared with the formal controls was found, both in the cortex and in the white matter, but especially in the cortex.

FORMS OF CALCIUM

A rough assessment of the way in which the calcium was combined was made by doing parallel calcium and phosphorus estimations on (1) a calcified nodule and (2) a homogeneous paste of the affected cortex. The material was boiled with 30 per cent. acetic acid in which calcium phosphate is soluble, and with dilute saline in which calcium oxide and sulphate are soluble.

It will be seen from Table II that there is more Ca than can be accounted for as $Ca_3(Po_4)_2$—this excess of Ca is probably largely $CaCo_3$, as the nodule evolved gas with N/10 HCl.

It would appear that in the calcified nodule about 220 mg. of the 10,500 mg. total Ca in 100 gm. cortex is water soluble, and therefore probably CaO or $CaSo_4$; and that at least 8,380 mg. is due to $CaCo_3$.

The Ca due to $Ca_3(Po_4)_2$ will be less than 1,900 mg. according to the amount of other P salts present.

A higher percentage of water soluble calcium salts was present in the homogeneous paste.

DISCUSSION

There would seem to be no doubt as to the enormous amount of calcium present in these cases, and that this element is present in excess in the white matter as well as in the cortex.

In these cases no excess of iron was found, thus confirming the findings of Wachsmuth and Löwenthal.

In the case reported by Eaves, there seems a possibility that she assessed some salts other than an iron salt. She did not estimate the iron as such, but weighed the insoluble residue remaining after extraction with hot concentrated acetic acid, and assumed that this was due to $Fe_3(Po_4)_2$.

It is curious that the mineral deposits give a positive Prussian Blue reactive histologically in these cases. The deposits are evidently not due to a conversion of organic to inorganic Fe in the tissue by pathological means. It would seem possible that the calcium deposits act as an adsorbent for readily available iron normally present in the tissue.

Summary

The cortex and white matter of the brain in three cases of Sturge-Weber disease contained no excess of inorganic or total iron, although iron was demonstrable by histological means.

These contained a large excess of calcium, mostly as phosphate and carbonate.

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