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Case Report

Ultrastructure of Calcification in Sturge-Weber Disease

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Summary. The ultrastructure of calcareous deposits in a case of Sturge-Weber disease is described. Concrements were found mainly extracellularly, outside of blood vessels, but there were some also within the vessel walls. The laminated concrements are built up of fine filaments, similar to those seen in the Fahr's disease. Needle-like cristals (calcium apatit) were observed within the concrements. Increased permeability of the altered vessel walls may be responsible for these alterations.

Key words: Sturge-Weber Disease — Electron Microscopy.

Introduction

Calcification of blood vessels is a common but not a constant feature of Sturge-Weber's disease (SWD). Light microscopical description of calcium deposits (Frigyér *et al.*, 1953) and their localization are well known, however search of the literature failed to reveal any information concerning the ultrastructure of calcareous deposits in this disease. In striatodental calcification (Fahr's disease; FD) calcium deposits have been shown by Cervós-Navarro and Matakas (1974) in the vessel wall between the basement membrane lamellae and to a lesser extent also outside of vessels (Guseo *et al.*, 1975), well preserved after formalin fixation, too. Calcium deposits in the brain contain calcium apatit and calcium hydroxyapatit shown by the roentgen and electron diffraction (Earle, 1965; Brauman and Verbanck, 1973), but we failed to demonstrate unambiguously the typical needle-shaped cristals of calcium apatit in the FD (Guseo *et al.*).

The present study was designed to describe the ultrastructure of calcareous deposits in the SWD and to compare them with the ultrastructural findings in Fahr's disease, as means of better understanding the nature of pathologic calcifications in human brains.

Material and Methods

The brain of a girl aged 2.5 month suffering from SWD was stored in formaldehyd for 20 years. Small pieces were washed in cacodylate buffer for 2 days, than treated with OsO_4, dehydrated and embedded in epoxy resin. Double contrasted ultrathin sections were investigated with a Zeiss EM 9 electron microscope.

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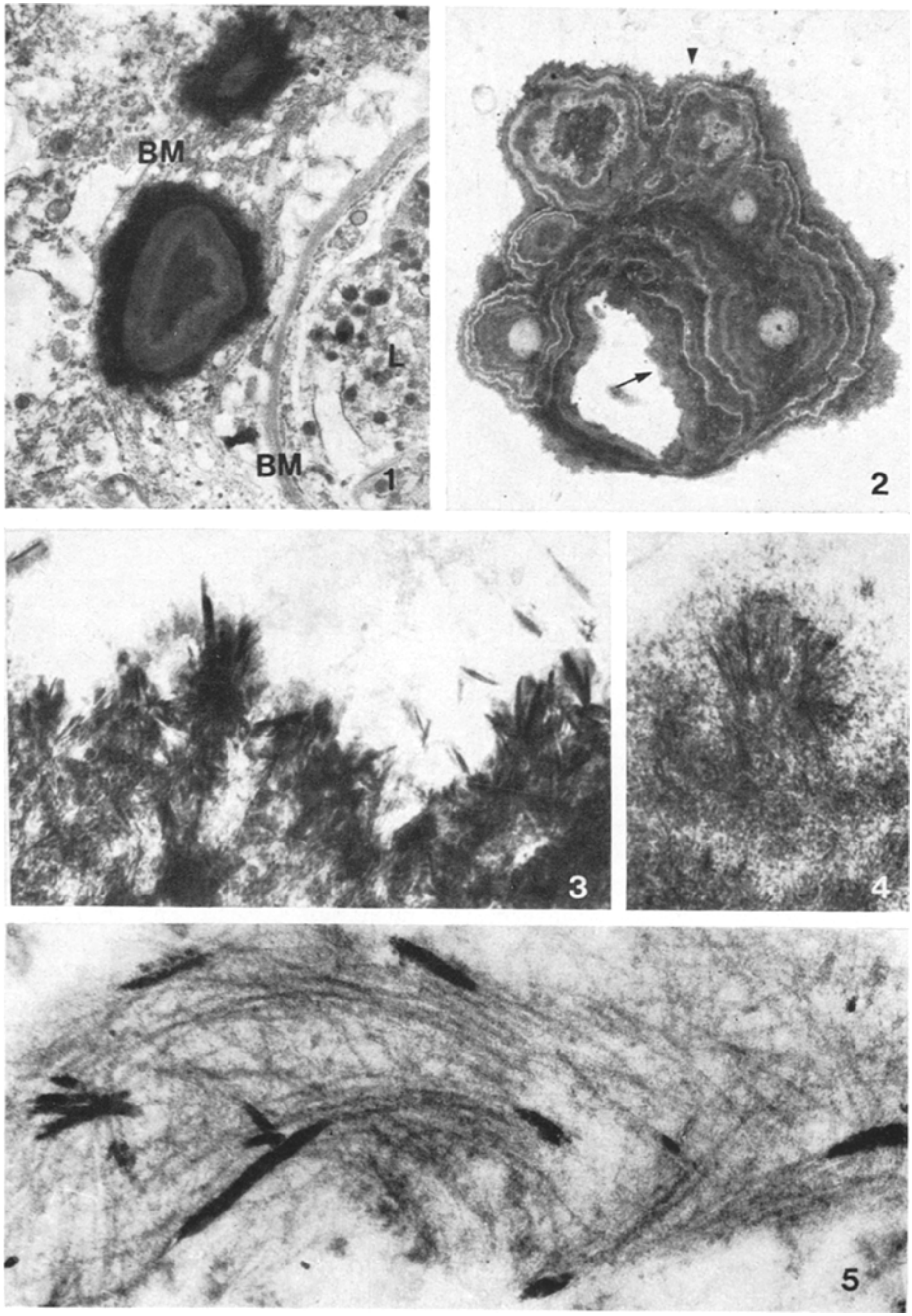


Figs. 1—5. Sturge-Weber disease

Fig. 1. Calcareous deposits in the vessel wall. (*BM* basement membrane, *L* Lumen) ×7400

Fig. 2. Concentric lamination of a great calcareous deposit. In the centre (→) and at the surface (►) needle-shaped calcium apatit cristals can be seen. ×3000

Results

Proliferation of altered cortical vessels were found in semithin sections. Great masses of mulberry-shaped laminated calcareous deposits were situated around the blood vessels but they occurred also within the vessel walls.

At the ultrastructural level the widened media was rich in collagen fibers, among which many small and some large round or amorphous particles of electron dense granulo-filamentous matter was found (Fig. 1). In many collagen fibers needle-shaped electron dark segments were seen which were thicker than the collagen fibers (Fig. 5). Collagen fibers were often found to be included into the calcareous deposits. Large calcium deposits were situated extracellularly, around blood vessels with or without any contact to the outer basement membrane. The large mulberry-shaped deposits originated by fusion of small ones. Almost all, even the smallest deposits had a concentric lamination like annual rings (Fig. 2).

At higher magnification calcareous deposits had a filamentous structure. There were irregular electron dark spots resulting from conglomerating of needle-like cristals (Figs. 3 and 4), more often in the central parts but also in the periphery of the concrements.

For comparison in a case of Fahr's disease we have shown that calcareous deposits in the vessel wall [Pseudokalk-Spatz, 1922] (Fig. 6) have a characteristic filamentous ultrastructure (acid mucopolysaccharides ?) in which 40–80 Å electron dense globules (calcium phosphate ?) were precipitated (Fig. 7). Calcium apatit cristals were not found by transmission electron microscopy.

Discussion

Roentgen and electron diffraction studies of calcareous deposits in human pineal gland and spinal arachnoid (Earle, 1965; Hassler, 1967) have shown that calcium deposits are built up of calcium apatit and hydroxyapatit cristals.

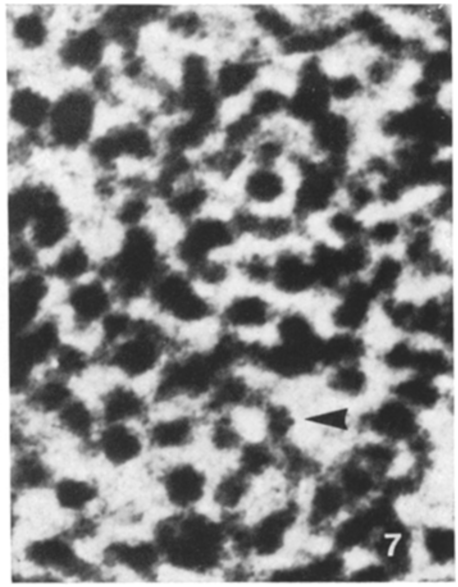
Calcareous deposits in the SWD have a filamentous ultrastructure similar to that, which have been found in the FD. The concentric lamination is a reflection of differing density of aggregated filaments. Cristallization, similar to that observed in bone calcification (Cameron, 1972) takes place most often in the deeper parts of the deposits, but occasionally it can be found at the outer surface of the concrements too.

In the SWD there is an increased permeability of cerebral blood vessels for protein and calcium phosphate or carbonate. These are precipitated within or outside the vessel wall in which later, depending on various factors (concentration of calcium and phosphor as well as carbonate ions, metal ions, acidotic sorroundings etc.) cristallization occurs. There were no essential differences in the substructure of calcareous deposits in the two cases investigated, except for an extensive cristal formation in the SWD.

Fig. 3. Calcium apatit cristals at the inner surface of Fig. 2. × 50000

Fig. 4. Calcium apatit cristals at the outer surface of Fig. 2. × 50000

Fig. 5. Calcification of collagen fibers. × 50000



Figs. 6 and 7. Fahr's disease

Fig. 6. Spherical calcareous deposits (Pseudokalk) between the basement membrane lamellae. × 7400

Fig. 7. Electron lucent and dense segments are present at various intervals in the filaments. Electron dense globules were precipitated within the filaments (◄). × 180000

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References

Brauman, J., Verbanck, M.: Processus intervenant dans la genese des calcifications cerebrales. J. belge Radiol. **56**, 271–282 (1973)

Cameron, D. A.: The ultrastructure of bone. In: The biochemistry and physiology of bone, vol. I, p. 191–236. New York: Acad. Press 1972

Cervós-Navarro, J., Matakas, F.: Struktur und Zusammensetzung von Kalk und Pseudokalkablagerungen im Gehirn. Vortrag, 58. Verh. Ges. Dtsch. Path. (1974)

Earle, K. M.: X-Ray diffraction and other studies of the calcareous deposits in human pineal gland. J. Neuropath. exp. Neurol. **24**, 108–118 (1965)

Frigyér, L., Máttyus, A., Molnár, L.: Zur Kenntnis der Sturge-Weberschen Krankheit. Mschr. Psychiat. Neurol. **126**, 118–134 (1953)

Guseo, A., Boldizsár, F., Gellért, M.: Elektronenoptische Untersuchungen in "striatodentaler" Calcification (Fahr). Acta neuropath. (Berl.) (In Druck)

Hassler, O.: Calcification in the spinal arachnoid in man. A microradiological and X-ray diffraction study. Acta neuropath. (Berl.) 8, 163–170 (1967)

Spatz, H.: Über den Eisennachweis im Gehirn, besonders in Zentren des extrapyramidalmotorischen Systems. Z. ges. Neurol. Psychiat. **77**, 261–390 (1922)

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