Morbus Fahr and genetic transmission

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Published in:
Proceedings of the 26th Dutch Federation meeting, Amsterdam, 11-12 April 1985

Publication date:
1985

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Morbus FAHR and genetic transmission

Morbus Fahr(idiopathic nonarteriosclerotic cerebral calcifications) can now be detected easily and accurately with computerized tomography (CT). A family exhibiting an unusual pattern of autosomal dominancy (10 of the 24 examined patients were affected) were examined. During a follow-up period of more than 5 years no changes in symptomatology could be shown. None of the family members, aged from 6 to 70, displayed any neurological sign or symptom, except one with intermittent gait disturbance of unusual type. Serum calcium, phosphorus and parathormone values were normal, although such laboratory findings do not exclude pseudohypoparathyreoidism.
The family tree suggested autosomal dominant heredity with a penetrance rate of 100%. The most important CT findings were symmetric calcification in nucleus caudatus, globus pallidus, putamen, nuclei thalami, nucleus dentatus and subcortical regions of the occipital lobe.
There was a tendency toward increased area of calcification with increased age. As far as we know this case is the second familial cerebral calcification type without any circumscribed neurological symptomatology, nor psychiatric disturbances.