A case of bilateral temporal lobe agenesis

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SUMMARY A 76-year-old man with bilateral temporal lobe agenesis producing clinical features resembling the Robinson syndrome is described. The malformation was discovered during a routine CT examination after the appearance of a homonymous visual field defect. The patient was examined by (neuro) psychological testing. The findings are compared with other reported cases and discussed with regard to cerebral localisation.

Isolated cerebral lobe agenesis unassociated with defects of other structures and without apparent functional disorders were usually not detected in life before the introduction of cranial computed tomography (CT).

On the basis of 10 cases Robinson¹ established the so-called "temporal lobe agenesis syndrome" as a clinical entity. These cases were distinguished by a defect on one side and discovered on the occasion of a thorough medical examination. The syndrome was "four times commoner in the male than the female. Most patients present clinically between five and 21 years of age but incidental cases have been found at autopsy up to 74 years of age. Headache is the leading symptom and in some the temporal bulging had been apparent to the parents soon after birth. Usually neurological signs are minimal or absent and there may be a generalised prominence of the temporal area of the skull. In a few patients the headaches are severe, and papilloedema and a mild hemiparesis are found. The lesions are commoner on the left with a left/right ratio of 3:2."² Radiological signs are an elevation of the lesser wing of the sphenoid bone and the middle cerebral artery in anterior-posterior projection. Other authors add a facial asymmetry and slight exophthalmos of the side concerned. The ventricles of the side involved are usually small. Seur and Kooman³ mention a hyperplasia of the sphenoid sinus (pneumosinus dilatans) as a prominent sign on plain skull radiographs. Other terms for this syndrome are

"cerebral arachnoid cyst," "subarachnoid cyst" and "subarachnoid pouch." Reports before 1955, the time when Robinson first described the syndrome, classify it under "porencephaly," "pseudoporencephaly," or "cystic hygroma."⁴

Further case reports after Robinson who by 1968 had added 10 more descriptions to the 18 already known in publications were given by Aguilera, Blázquez and Obrador⁵ who reported on three cases, and by Seur and Kooman³ who contributed another two cases. The latter authors presented two male patients aged 15 and 19 years without neurological deficit who supposedly originally had a temporal lobe agenesis leading to secondary manifestations in the form of a bulging of the temporal squamae, excessive pneumatisation of the sphenoid sinus, and an abnormal course of the intracerebral blood vessels. Kolberg⁶ who published five additional personal observations of patients between two-and-a-half and 57 years of age assumed a developmental disorder with compression of the vein of Rosenthal through strangulation of the hippocampus in the tentorial hiatus as the underlying disorder. Kolberg regarded the temporal lobe agenesis as a porencephaly and therefore as secondary, so he proposed the name "incisural porencephaly of the temporal lobe." Without anticipating any specific pathogenesis we will refer to "temporal lobe agenesis."

There are several examples of one-sided temporal lobe agenesis revealed by CT: without further description Shields, Isherwood and Pullan⁷ presented a transverse axial tomogram of a left-sided arachnoidal cyst covering the area of the left temporal lobe. The 22-year-old male patient of New and Scott⁸ suffering from headaches of

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several months duration had "a large, well-defined, cavitory-appearing lesion . . . in the right fronto-temporal region with an absorption range of 6-18 units. . . . Considering the size of this lesion, there [was] a rather modest mass effect upon the ventricular system (suggesting an underlying loss of cerebral substance)." Lange, Grumme and Meese⁹ presented the tomogram of a one-year-old child whose left temporal region was filled by CSF.

As far as we know, only three cases of bilateral temporal lobe agenesis have been reported, none of them shown by CT. The first is that described by Faris¹⁰ who found in a 41-year-old woman suffering from a psychiatric illness an unusual bitemporal bulging and thinning of the squamae, an elevation of the lesser sphenoid wing on both sides, marked dilatation of the third and lateral ventricles, and an abnormal course of the middle cerebral arteries, which may be described as a bilateral form of the Robinson's syndrome. The second case is that of Tuynman, Hekster and Pauwels¹¹ who reported on a 14-month-old girl with bilateral temporal arachnoid cysts who was found to be blind. Again the plain skull radiographs indicated a bilateral expanding lesion and arteriography showed the typical upward course of both middle cerebral arteries. Routine Tc-scintigram demonstrated a pathological accumulation of radionuclide on the right side only. Shaw and Alvord¹² among 5000 necropsy specimens mention a case of bilateral temporal arachnoid cysts without further comment.

We present a patient with bilateral temporal lobe agenesis who not only was examined by CT but also from psychological and neuropsychological aspects.

Case report

The patient is a 76-year-old business man who is now retired but is still active on a voluntary basis. After training in electrical engineering and mechanics he underwent commercial apprenticeship and became a state official before he decided to become self-employed. Besides philately, hiking in the mountains, sailing and motor-cycling his hobbies comprised active music making on at least five musical instruments. He had been fully employed up to the age of 75 years.

Neuro-ophthalmological findings

Nine weeks before we examined the patient, he complained of visual disturbance and a homonymous field defect was discovered by perimetry (fig 1). Arteriosclerotic retinopathy and arterial hypertension were present, which led to the assumption that there was a vascular disorder. Slow filling of the retinal vessels was detected by fluorescence angiography, but as the

patient objected to a carotid arteriography, the suspected stenosis of the right carotid artery could not be demonstrated.

Fig 1 Visual field (a) left eye, (b) right eye. The homonymous visual field defect on the left includes the blind spot. The roman numerals mark the area of the test spot ($I=1/4 \text{ mm}^2$; $II=1 \text{ mm}^2$), the arabic numerals the relative intensity ($3=0.315$; $4=1.00$).

Roentgenological findings

Radiographs of the orbits and the sella turcica showed an excessive pneumatization of the sphenoid sinus which extended to the base of the dorsum sella, from which it was only separated by a thin bony lamella.

Isotope encephalographic findings

After radionuclide examination had shown a discrete area of increased concentration of radioactivity in the

occipital region which corresponded to the presumed area of infarction, CT was performed, and bilateral temporal lobe agenesis was found. No neurological abnormalities other than hemianopia were discovered at any time.

Fig 2 Plain radiographs show marked pneumatisation of the sphenoid sinus extending to the base of the dorsum sellae, from which it is separated only by a thin bony lamella.

Fig 3 Static radionuclide scan with ^{99m}Tc -pertechnetate shows an occipital paramedian elevated concentration of radioactivity of the right side as a result of infarction.

Fig 4 (a-f) Bilateral temporal defects of a slightly higher density than CSF are shown. The temporal poles on both sides are missing, on the right side a small rim of temporal tissue can be seen. The cisterna pontis and cisterna laminae quadrigeminae are slightly enlarged as is the fourth ventricle.

Psychological findings

During and even at the end of the interviews the patient was calm, friendly, fresh and enterprising. He took part in the first four-hour-examination willingly without asking for pauses although he had had a one hour journey to the clinic driving his own car. He answered questions frankly, was spontaneous and talkative. The IQ as measured by the HAWIE (Wechsler-Adult-Intelligence-scale, German version)¹³ was 129 points, verbal and performance IQ were about the same. The relatively low score on the picture-arrangement-subtest could be explained by the visual field defect. The IQ level was corroborated then by the MWT (Multiple-Choice-Vocabulary-Test by

Lehrl),¹⁴ scoring 130 points. The non-verbal Raven progressive matrices,¹⁵ too, were clearly above average with a percentage of over 90. The "Biographisches Inventar zur Diagnostik von Verhaltensstörungen" ("Biographical Inventory for the Diagnosis of Behavioural Disturbances")¹⁶ was answered in a way which showed marked ego-strength, mental as well as physical robustness and level-headedness, great social activity, extraversion, social openness, sociability and impulsiveness. High scores for mental stability on the neuroticism scale and medium scores on the extraversion scale in Eysenck's Personality Inventory (German version by Eggert)¹⁷ corresponded with these findings. The SKT (Syndrom-Kurz-Test) by Erzigkeit¹⁸ disclosed no impairment of attention, concentration and short-term-memory. The Defect Test by Kinzel,¹⁹ validated with regard to defect syndromes after cerebral contusion, revealed no difference from normal.

Neuropsychological findings

The Benton visual retention test²⁰ gave hints of an organic cerebral defect; this however was most likely solely a consequence of the visual field defect. The difference between the number of correctly as well as incorrectly reproduced figures and the results expected was abnormal. Mistakes by rotation were the most frequent; furthermore distortions, one omission and one misplacing were noted. The patient stated that he was forced to move his head to and fro during the examination in order to discover all the relevant details. The testing for amusia was carried out by means of the Seashore-Test for Musical Abilities (German version by Butsch and Fischer).²¹ The scores were—as compared with a sample of normal 19-year-olds—average or above average. This excluded the presence of such a disturbance, as could be expected from the excellent musical skills already mentioned. The clinical impression of totally undisturbed linguistic abilities was confirmed by faultless performance in aphasia screening tests, the Token-Test by De Renzi and Vignolo (German version by Orgass)^{22 23} and the "Drei-Figuren-Test" (Three-Figures-Test) by Peuser.²⁴ At no time was there an indication of prosopagnosia: the patient recognised without hesitation all examiners even after a period of six months.

Discussion

Classical clinico-neuropathological correlation implicitly suggests in a reductionistic way that mental functions consist of a number of relatively independent, elementary functions that are bound to circumscribed cerebral areas, so-called centres. Typical consequences of this concept are models such as the well-known Lichtheim-Wernicke aphasia schedule, and Henschen's²⁵ or Kleist's²⁶ cerebral maps where the abilities of acoustic attentiveness, melody, word, sentence and name recognition, noise, sound and tone perception are

allocated to the temporal lobe. More recent concepts, however, emphasise the importance of a lesion's dynamics; mental abilities are not innate in certain cerebral areas, but are acquired during lifetime. Cases have been reported showing lesions of the dominant hemisphere without signs of aphasia or amusia. No symptoms of a Klüver-Bucy syndrome nor of prosopagnosia or memory deficit could be detected in our case as would have been expected if the lesions had developed rapidly during adult life. Karvounis, Chiu, Parsa and Gilbert⁴ emphasised the psychological normality of their 48-year-old patient, including clear consciousness, intact speech and undisturbed memory, although he was lacking the left temporal lobe. Tuynman, Hekster and Pauwels¹¹ stressed the normal physical and intellectual development of their young patient with bilateral temporal lobe agenesis, while the psychiatric symptoms of Faris's¹⁰ patient apparently were not related to her cerebral lesion.

The malformation in our patient may be an example of Robinson's syndrome, in view of the temporal bulging, pneumatization of the sphenoid sinus and the CT findings, although a genetic defect as the underlying disorder also must be considered. Temporal lobe agenesis is very rare and bilateral temporal lobe agenesis even rarer. A review of 650 patients by Paxton and Ambrose²⁷ contains no example, nor does a survey of CT findings in cranio-cerebral anomalies by Harwood-Nash.²⁸ In a review of 5000 human brains, Shaw and Alvord¹² discovered one example of bilateral temporal arachnoid cysts. Since almost all of these malformations were found incidentally only an increase in CT examinations is likely to disclose their true rate of occurrence.

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