Prosopagnosia

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This case of prosopagnosia has already been described from the clinical point of view.¹

The patient, who was 64 years old, presented with prosopagnosia, topographical disorientation, simultanagnosia, and achromatopsia. The outstanding neurological finding was left homonymous hemianopsia present initially later regressing to a left upper quadrant scotoma. These findings seemed to indicate 'a lesion of the lower lip of the right calcarine cortex'.

During a follow-up period of four years the patient's condition improved. The topographical disorientation, simultanagnosia, and achromatopsia disappeared within six months and the prosopagnosia improved, leaving only the visual field defect which persisted till his death.

It must be stressed that our patient showed no signs of dementia and there was no change in his daily life. He learned to adapt himself to his inability to recognize people by various means. Outwardly he never gave the impression of being in any way disturbed, and in the course of time learned to find means of identification other than facial expression, and thus overcame the psycho-visual defect.

In unusual circumstances, however, his disturbance became more apparent; *e.g.*, he was accustomed to frequent meetings with his lawyer for business discussions but in court faced with two lawyers, both dressed in a similar fashion, he could not distinguish between the two and discussed the proceedings of the case with his opponent's attorney, thinking him to be his own—with disastrous consequences. When his physician afterwards asked him what had happened he replied that he had always met his lawyer in the latter's office and had come to recognize the furnishings and surroundings, these serving as the means of identifying the man. But in the courtroom, presented with two persons wearing identical gowns and not in their usual setting, he was at a loss. As he himself said, 'You know that I cannot identify faces'.

He often used to ask his wife before going visiting who was due to be present and if he met people of whose presence he was not previously informed he would not recognize them.

Fatigue aggravated his disturbance. A picture which he might one day not recognize at all, he easily identified a few days later. When shown a photograph of a person

he would look at it from all angles, holding it close and then examining it at a distance, always searching for auxiliary identification marks.

His memory was in no way impaired and he was well acquainted with the current political situation. He could calculate both mentally and on paper. Neurologically, there were no objective findings. His gait was normal, his speech was unimpeded, and there were no disturbances of affect.

Latterly he began to suffer from an anginal syndrome. He was admitted to the Hadassah Municipal Hospital in Tel-Aviv with a myocardial infarction and died shortly afterwards.

NECROPSY

The body was that of a man who appeared younger than his chronological age.

The chest and back were covered with round or oval verrucose growths, 1 to 3 cm. in diameter, which were irregularly and sharply defined. Microscopic examination showed them to be *Acrokeratosis verruciformis*.

The pupils were round and equal in size, the sclerae white, and the neck veins engorged. On opening the body, 500 ml. of serous fluid was found in the peritoneal cavity. The right heart was greatly dilated with hypertrophy of the left ventricle. On the epicardial aspect of the posterolateral wall of the left ventricle a roundish, greyish white area, 3 cm. in diameter, was found. Similar areas, each 1 cm. in diameter, were found on section of the posterior wall. The heart weighed 420 g. The coronary arteries were atheromatous, particularly the posterior descending branch, to the extent that its lumen was almost completely occluded. Microscopic examination disclosed old and fresh myocardial infarctions and severe arteriosclerosis of all the vessels examined. Multiple atheromata, some of which were calcified, were found in the ascending aorta, the aortic arch, and particularly in the abdominal aorta. No infiltrates were seen in the vessel wall.

The kidneys showed arteriosclerotic, arteriolosclerotic, and secondary parenchymatous changes.

The gall bladder contained four stones, each 1 cm. in diameter, and three small stones. On microscopy the gall bladder wall showed atrophic oedematous cholecystitis. Microscopy of the liver revealed nutmeg congestion, cholangitis, and pericholangitis. The thyroid was enlarged, weighing 200 g., and on microscopic examination macro- and microfollicular adenomata were found.

¹Bornstein, B., and Kidron, D.P., 1959). *J. Neurol. Neurosurg. Psychiatr.*, 22, 124.

FIG. 1a

Fig. 195.—Sulci and gyri on the lateral aspect of the human cerebral hemisphere.

FIG. 1b

FIG. 1a and b. *The lesion found in the middle of the left inferior parietal lobule, angular gyrus, and upper part of the superior temporal sulcus.*

THE BRAIN The meninges showed severe congestion and thickening and were opalescent. The arteries at the base of the brain contained atheromata which narrowed their lumina. In the middle of the left inferior parietal lobule, in the angular gyrus, and in the upper part of the superior temporal sulcus, a yellowish-brown, slightly depressed lesion, 3 cm. in diameter, was found. The anterior border of this lesion was formed by the angular gyrus and it extended on to the parieto-occipital sulcus which also formed its posterior border (Fig. 1). These macroscopic changes in the cortex extended to a depth of 1 mm.

On microscopy signs of mild atrophy, gliosis, and rarefaction of cells were seen.

Holzer staining showed glial proliferation. On section of the brain, a narrow cystic cavity, 1 cm. in length and yellowish-brown in colour, was found in the region of the inferior border of the medial surface of the right hemisphere, 3 cm. anterior to the occipital pole, under the calcarine fissure which also formed the roof of the lesion (Fig. 2). It was 4 to 6 mm. below the cortex. The consistency of the tissue in this area was soft and gelatinous. Microscopy showed signs of encephalomalacia. The

FIG. 2. *Section of the right occipital lobe, 3 cm. anterior to the occipital pole showing the cystic cavity.*

meninges were congested, thickened, oedematous, and there was lymphocytic infiltration. The blood vessel walls in the area of the lesion were thickened.

Serial sections of the brain did not reveal any other areas of softening, and microscopic examination showed that the lesions were old.

DISCUSSION

The pathological examination revealed two lesions, one in the region of the left angular gyrus superficially situated with atrophy of the surrounding tissue, and the second in the region of the lower lip of the right calcarine fissure. These lesions were apparently due to arteriosclerotic changes in the cerebral blood vessels.

The large lesion on the right was old and was the cause of the visual disturbances. The atrophy on the left seen by the relative depression of the inferior parietal lobule as compared with that of the

opposite side indicated a long-standing process, which histologically presented as atrophy and degeneration as an expression of disturbed vascularization.

The psycho-visual changes (prosopagnosia) became clinically evident only when the lesion on the right calcarine fissure appeared in addition to the pathology in the left angular gyrus, which presented clinically only as attacks of loss of consciousness. This fact confirms to a certain extent our view that a single lesion is not enough to cause prosopagnostic disturbances. It confirms too the view expressed in the previous paper describing the clinical findings. 'Despite the fact that all the signs observed over the course of 16 months point towards a right-sided occipito-parietal lesion, it is highly likely that the two previous cerebral attacks indicated bilateral damage to the brain.'

The question is to what extent the arteriosclerotic changes, particularly of the basilar artery, were responsible for the clinical symptoms. No definite reply can be given, but, as pointed out in the clinical description, no signs of mental or psychic deterioration were noted and the patient's memory, orientation, judgment, and affect were normal until his last day. It should, however, be remembered that the

prosopagnosia, topographical memory, and the simultanagnosia improved with time, a fact which does not lend weight to the assumed arteriosclerotic basis for these changes.

It is difficult to say to what extent the clinical and pathological findings contribute to our understanding of this symptom. We know that in mental disease the anatomical findings only provide the background to our thinking. Gross and microscopic morphology cannot as yet solve the problem of functional mental disturbances. The ability to synthesize mentally the various facial features of a particular person and to recognize them as a particular face is a particularly delicate and complicated mechanism which is not related to disturbances of memory. We may therefore assume that as such under certain circumstances the capacity to recognize such features is decreased.

SUMMARY

The necropsy findings in a case of prosopagnosia are described. Two foci of cerebral softening due to arteriosclerotic changes were found, one in the right calcarine fissure and the second in the left angular gyrus. No other cerebral foci were noted.