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Oligophrenia with the Hallermann-Streiff Syndrome

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Summary. A case of oculomandibular dyscephaly (Hallermann-Streiff syndrome) associated with severe oligophrenia and consequent behavioral disorders is reported. The not infrequent association of oculomandibular dyscephaly and oligophrenia suggests that the same unknown embryopathy could damage the structures derived from the first branchial arch and the CNS.

Key words: Oculomandibular dyscephaly – Hallermann-Streiff syndrome – Oligophrenia – Ullrich-Fremerey-Dohna syndrome.

Zusammenfassung. Es wird über einen Fall von oculo-mandibulärer Dyscephalie (Dyscephalie-Katarakt-Hypotrichosesyndrom; Hallermann-Streiff-Syndrom) berichtet, bei welchem auch eine ausgeprägte Oligophrenie mit entsprechenden Verhaltensstörungen vorlag. Das relativ häufige gleichzeitige Vorkommen einer Oligophrenie mit dem oculo-mandibulären Dyscephaliesyndrom spricht dafür, daß die gleiche bisher unbekannte Embryopathie sowohl die vom ersten Kiemenbogen abgeleiteten Strukturen als auch das zentrale Nervensystem lädieren könnte.

The Hallermann-Streiff syndrome is an oculomandibular dyscephaly characterized by: (1) dyscephaly with bird-like face and hypoplastic mandible, (2) proportionate nanism, (3) congenital cataract, (4) microphthalmia, (5) hypotrichosis, (6) dental anomalies, (7) cutaneous atrophy limited to the face and/or scalp, (8) frontal or occipital bossing, (9) open sutures and fontanelles, (10) high arched palate, and (11) nystagmus [5].

François [2] considered the first seven features as essential.

We have recently observed a case of typical Hallermann-Streiff syndrome associated with severe oligophrenia.

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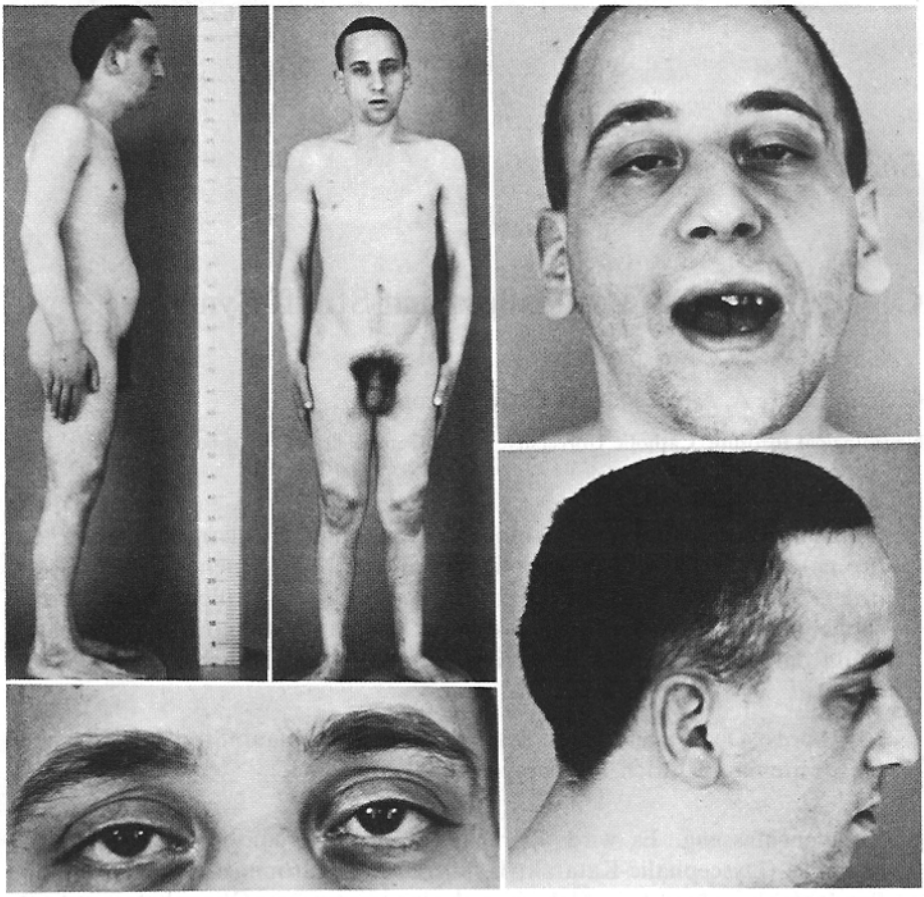

Fig. 1

Case Report

The patient, a 23-year-old man (Fig. 1), was admitted to the clinic drunk and completely unable to give his history, which was later obtained from his relatives.

The father is an alcoholic; the mother, two sisters and two brothers are healthy. There is no history of malformation in the family. Prenatal course and birth had been normal. His motor development was reported as slightly delayed and his psychic development as markedly delayed. He stopped schooling at the age of 12, still in the first elementary class. Since childhood he showed behavioral disorders such as alcohol abuse, motor hyperactivity, restlessness, aggressiveness, wanderlust, and was repeatedly referred to psychiatric hospitals for many years.

On general examination he was found to have: (1) moderate proportionate dwarfism (157 cm tall), (2) dolichocephaly with bird face and high arched palate, (3) frontal bossing, (4) mandibular hypoplasia, (5) multiple dental malformations such as malposition, absence of enamel and many caries, (6) bilateral anterior lenticonus with bilateral central cataract and many myelinized peripapillar fibers, (7) hypotrichosis of the chest and temples with normal pubic hair, (8) severe oligophrenia.

Wechsler Adult Intelligence Scale (WAIS) test results showed a very low intelligence quotient (IQ = 54), with a widespread involvement of all items and no signs of mental deterio-

Table 1. Wechsler Adult Intelligence Scale

	Raw score		Scaled score
Information	3		3
Comprehension	4		2
Arithmetic	4		4
Similarities	3		4
Digit span	7		4
Vocabulary	11		4
Verbal score			21 (17.5)
Digit symbol	19		4
Picture completion	3		3
Block design	10		4
Picture arrangement	7		3
Object assembly	7		2
Performance score			16
Total score			33.5
Verbal score	17.5	I.Q.	58
Performance score	16	I.Q.	55
Total score	33.5	I.Q.	54

ration (Table 1). From a behavioral point of view the patient's relations with other people were featured by psychomotor hyperactivity, exaggerated expansiveness verging on hypomania, and hyperbolic confabulations sometimes leading to an adventurous pseudodelirium. Vain mythomania based on self-accusation sometimes added to this pattern.

The clinical examination of all other organs was uneventful. All laboratory findings, tests for syphilis, EEG, ECG, karyotype were normal. The skull X-ray showed dolichocephaly with decreased basal angle and flattened squamooccipital, short palate and increased mandibular angle. Periodontal spaces were widened, alveolar laminae had some osteolytic areas and many interalveolar septa appeared to be reabsorbed.

Comments

Our patient had the most important features of the Hallermann-Streiff syndrome and severe oligophrenia. In their review Judge et al. [4] quoted 34 cases of the Hallermann-Streiff syndrome in whom intelligence was also studied, clinically and/or by testing. Four of them had severe oligophrenia, 14 a mildly or moderately retarded development. Central nervous system involvement was therefore present in more than 50% of patients. Hopf et al. [3] and Volpi et al. [6] subsequently reported two other associations of the Hallermann-Streiff syndrome with oligophrenia, while Crevits et al. [1] presented one case of a H.S. patient who was epileptic and had a normal intelligence.

Our observation of one additional case of Hallermann-Streiff syndrome associated with severe WAIS tested oligophrenia and some typical consequent behavioral disorders further suggests the possibility that the same unknown *embryopathy* could damage the structures derived from the first branchial arch and the central nervous system.

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