Papillary Carcinoma of Choroid Plexus

Light and Electron Microscopic Study

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Summary Two cases of papillary carcinoma, one in a 23-month-old girl and the other in a 25-month-old boy who both died within a relatively short time after operation, were studied histologically and electron microscopically. Both tumors originated in the right trigone of the lateral ventricle and spread widely via the cerebrospinal fluid. Histologically, the tumors consisted mostly of a differentiated papillary architecture closely resembling choroid plexus papilloma. Some carcinoma cells, showing cellular atypism, displayed a multilayer arrangement. The amount and distribution of PAS-, Alcian blue- or orcein-positive substances on the cell surface and/or in the interstitial elements of the carcinomas differed from that of choroid plexiomas examined in our laboratory. Electron microscopically, the carcinoma cells in some areas showed a loss of apical-basal polarity, and the formation of both microvilli and desmosome-like structures was indistinct. Papillary carcinoma is reviewed on the basis of the literature from 1906 till 1980.

Key words: Choroid plexus carcinoma – Choroid plexus papilloma

Tumors originating from the choroid plexus are rare. The incidence of choroid plexus tumors is reported to be about 0.4 to 0.8% of all verified intracranial tumors (Cushing 1932; Norlen 1949; Grant 1956; Zülch 1965; Arai 1976); they account for 1.5 to 3.9% of tumors in children (Bodian and Sawson 1953; Matson and Crofton 1960; Sato and Sano 1975), and 1.1 to 2.0% of intracranial gliomas (Cushing 1932; Ringertz and Reymond 1949; Russell and Rubinstein 1977). The tumors occur at any age, but the majority are found in young subjects, particularly in the first decade of life (Friedman and Solomon 1936; Turner and Simon 1937; Posey 1942; Matson and Crofton 1960; Bohm and Strang 1961; Zülch 1965; Rubinstein 1970). It is generally accepted that lateral ventricle papillomas develop in children while fourth ventricle papillomas occur mostly in...
adults (Wagenen 1930; Ringertz and Reymond 1949; Bohm and Strang 1961), and in males in particular (Tooth 1912; Kahn 1952). Most tumors of the choroid plexus are benign in character, and malignant tumors are extremely rare. Almost all malignant tumors have been reported to occur in the lateral ventricle in childhood (Russell and Rubinstein 1977). Although several choroid plexus carcinomas have been reported in adults, the diagnosis in most cases was considered to be uncertain (Lewis 1967). Zülch (1965) postulated that the few cases described as having been malignant from their very inception might well have been metastases from some primary malignancy elsewhere in the body. The basic pattern of the histological features of plexus carcinoma in children was described in detail by Lewis (1967), but more cases of the same genus would be necessary to generalize on the basis of his findings. Plexus carcinoma is a distinct clinicopathologic entity and should be distinguished from other benign or malignant neoplasms.

In our laboratory, 8 tumors originating from the choroid plexus were encountered from 1960 to 1980. They accounted for 0.8% of all verified intracranial tumors (995 cases), and 2.8% of all intracranial gliomas (287 cases). Two of them, from a 23-month-old girl and a 25-month-old boy, showed malignant features, and both cases were autopsied. The present study reports these 2 primary carcinomas of the choroid plexus which were studied histochemically and electron microscopically and compared with benign papillomas and normal choroid plexus examined in our laboratory. Also, papillary carcinoma is reviewed on the basis of the literature from 1906 till 1980.

Case Reports

Case 1. A 23-month-old Japanese girl was admitted in August 1978. She was unable to walk or stand up; her external ocular movements appeared full and without nystagmus. Babinski's and Chaddock's signs were positive on the right side. After admission her general condition became poor, and she was drowsy and inactive just before the operation. X-rays of the skull showed separation of the sutures and a CT brain scan revealed a large, well demarcated lobulated, high density mass in the right temporo-parieto-occipital area and a round high density mass in the mid-brain, with dilatation of the left lateral ventricle and third ventricle. There was a shift of the septum to the left. The right carotid arteriogram showed a large mass in the right temporo-occipital region.

On 1st September, 1978, a right temporal craniotomy was performed. A massive tumor, encountered 1 cm below the cortex, was reddish, granular, fragile and well demarcated from the brain substance. The tumor was not resectable, and small biopsy specimens were removed for light and electron microscopic examination.

On 10th September, 1978, the 2nd operation was performed for marked bulging at the site of the craniotomy. Gradual deterioration occurred along with decerebrate posture, anisocoria and dilatation of the pupils before death on 4th November, 1978. Autopsy revealed that the right hemisphere contained a lobulated, partly encapsulated tumor, 9 cm anteroposteriorly, 13 cm transversely and 7 cm vertically. It was partly gray-pink and finely granular, suggesting a papillary structure with widespread necrosis and hemorrhage. The cerebral cortex was thin, atrophic and necrotic in part. The right thalamus and the corpus striatum were invaded by the tumor. The trigone of the right lateral ventricle was occupied completely by the tumor tissue. The anterior half and the inferior horn of the right lateral ventricle and the left lateral ventricle were generally dilated, and the third ventricle and the cerebral aqueduct were closed under pressure from the tumor. On sections through the rostral part, the pons was almost replaced by the tumor mass (Fig. 1). Sections through the pons near its caudal border showed massive tumor deposits in the subarachnoid space, and the pons was compressed from outside and atrophied. The medulla oblon-