PAPILLOMA OF THE CHOROID PLEXUS IN CHILDHOOD

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Papilloma of the choroid plexus is an unusual but important intracranial tumor of early childhood. This tumor is interesting to the geneticist because of its occasional prenatal occurrence, to the physiologist because of its frequent association with communicating hydrocephalus and the possible role the tumor may play in overproduction or increased rate of circulation of cerebrospinal fluid, to the pediatrician because of the paucity of diagnostic symptoms and signs it produces in early childhood, to the radiologist because of the dramatic and diagnostic features of ventriculography, to the surgeon because of its common benign, noninvasive character and the frequent possibility of total excision with satisfactory end result, and to the pathologist because of its histological relation to normal ependyma and normal choroid plexus and also because of the relation of its occasionally more rapidly growing and invasive variants to other ependymal tumors.

Even when histologically benign, these tumors can prove fatal if not recognized early and subjected to definitive treatment before development of such complications as intraventricular hemorrhage and severe hydrocephalus. In general, the results of treatment of the patients reported previously have not been as good as the usual benign character of the lesion should warrant.

For these various reasons, it seemed worth while to review the literature briefly and to analyze the experience of the Children's Medical Center with 16 additional cases of papilloma of the choroid plexus seen during the period from 1941 to 1958. A few of these cases have been referred to elsewhere.29,39 There have been no other reports devoted specifically to this tumor in children since Friedman and Solomon29 collected 14 cases from the literature in 1936 and Rand and Reeves47 described 4 of their own cases in 1940.

INCIDENCE

In Cushing's31 series of 2,000 proven cases of intracranial tumor in all age groups the incidence of papilloma of the choroid plexus was 0.6 per cent. Norlén41 reported an incidence of 0.4 per cent in 3,664 cases of intracranial tumor. Zülch40 in 1956 also reported an incidence of 0.5–0.6 per cent. It is probable that papillomas of the choroid plexus are somewhat more common than these figures suggest since many cases in early childhood un-
doubtlessly have gone unrecognized and many more isolated tumors found post mortem have never been documented.

At the Children's Medical Center during the years 1941 to 1958 inclusive, there were 408 cases of intracranial tumor in children under 12 years of age, of which 16 were papillomas of the choroid plexus, an incidence in this age group of 3.9 per cent.

The possibility that this tumor is frequently of congenital origin is supported by the statistical data which indicate that it is seen most frequently in the first decade and particularly during the first 2 years of life, and gradually decreases in frequency with advancing age (Fig. 1). Posey in 1942 found that in 52 cases reported in the literature in which the age was stated, 27 cases, or 38 per cent, occurred before the age of 10 years. At the time of the present review, 48 per cent of all reported cases are in the first decade and 20 per cent are under the age of 1 year.

There is no doubt that papillomas of the choroid plexus can develop during prenatal life. Drucker reported a tumor in a newborn infant. Braunstein and Martin reported a 1-month-old infant with a history dating from birth. Matson has reported previously 1 of the present series (Case 9), an infant born prematurely with an enlarged head who had a papilloma of the choroid plexus removed before the normal expected date of birth. Another patient in this series (Case 14) had a huge tumor removed at 2 months of age.

In the present review of the literature, 67 children with well-documented papillary tumors of the choroid plexus have been identified. This includes reports from 53 different