FRONTO-ETHMOIDAL ENCEPHALOMENINGOCELE

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Meningocele in the anterior part of the head is a rare condition in Europe and America, but appears to be common in Thailand. We have found 25 cases in the past 3 years. During this period 10 spinal and 3 occipital meningoceles were encountered. Meningoceles situated in the anterior part of the skull are sometimes referred to as sincipital encephalomeningoceles and are usually classified according to the site of the protruding mass. The findings at operation or autopsy in 20 of our cases, however, showed that the persistent site of the cranial opening in all cases was at the junction between the frontal and ethmoidal bones. Consequently we believe that these encephalomeningoceles can be identified as a fronto-ethmoidal type.

Our 25 patients with fronto-ethmoidal encephalomeningocele, present many interesting problems which have to be taken into consideration in their management.

Clinical Features

All of our cases were congenital in origin; the lesions were found at birth but the patients came to us at ages ranging from newborn to 25 years. The lesions were of variable size, shape and location. Some only showed swelling and widening of the root of the nose. Some presented as a mass or masses at many sites ranging from the middle part of the forehead, the root of nose or the side of the base of nose. The eye was often displaced laterally. The size of the mass varied from 1–9 cm. in greatest diameter. The lesion frequently increased in size as the child grew. Some of the meningoceles had no skin covering and the protruding brain was exposed. The covering skin when present varied from thin and shiny or thick and wrinkled. In patients with mere widening of the root of the nose, the covering skin was normal. A congenital scar on top of the mass was seen in 6 patients.

Only occasionally was the mass cystic and compressible. The majority were solid and firm. Transmitted pulsation from the brain was not frequently seen. In some cases the mass increased in size or became tense with crying. Some were apparently sensitive since some children cried when the lesion was palpated.

There was no problem in the diagnosis of the classical cases. This could, however, be difficult in patients where the opening was small and the communication between the sac and the intracranial cavity was not readily detectable. The lesion had to be differentiated from tumors and cysts indigenous to the region. Aspiration of the mass and determination of the sugar content of the fluid was helpful in some patients. Plain roentgenography of the skull was very helpful, the diagnosis of the encephalomeningocele being suggested by the abnormally wide distance between the 2 orbits. As a rule, the opening could not be seen in a newborn infant due to poor calcification of the cranium, but in older children it was usually visible. Our radiologist suggested posteroanterior projections at various angles which proved to be helpful because the opening was best seen only at certain angles. Occasionally tomography was helpful in defining the opening more clearly.

Radiography after injection of air into the encephalomeningocele was done in 3 patients but in none was air found intracranially and was, therefore, of no help. The failure to demonstrate the communication could be explained by the frequent operative finding of arachnoid adhesions around the brain at the site of the opening. Similarly, no air was seen entering the encephalomeningocele in pneumoencephalography.

Many sequelae and complications of encephalomeningocele were encountered:

1. Associated brain damage. In our 25 patients, 6 (Cases 4, 5, 18, 22, 23, 25) showed microcephaly. In 2 of these a relatively large amount of brain tissue had herniated into the sac and the head was consequently small. In the other 4 patients there was probably congenital under-development of brain.

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Two-year-old girl with a mass about the size of a finger tip at birth and progressively enlarging.

The mass was covered with thick, hairy skin and contained the herniated tip of the left frontal lobe. The wall of the sac was about 1 cm. thick, and was composed of fatty, fibrous tissue.

The covering skin was thin and dark colored. The patient fell and hit the mass on the ground 1 month before admission. She then had leakage of cerebrospinal fluid from the mass and recurrent fever.

Note the wide intercanthal distance and the scar-like marking. The head was small and the anterior fontanel was closed.