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Congenital Quadrigeminal Lipoma with Osteocartilagenous Element

- A Case Report -

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A case of congenital tectal lipoma found incidentally at an autopsy is reported. This female fetus was a product of pregnancy termination at 24 weeks of gestational age after sonographic detection of hydrocephalus. Autopsy revealed a small mass in the periaqueductal portion. The mass was composed of adipose tissue, cartilage, and mature bony tissue with hematopoiesis. The resultant diagnosis was tectal lipoma with osteocartilagenous element. It is uncertain whether the lesion represents a teratoma or hamartoma or mesenchymal metaplasia. The osteocartilagenous component suggests the latter.

Key Words : *Lipoma, Osteocartilagenous component, Tectum, Hydrocephalus*

INTRODUCTION

Choristoma or hamartoma of the brain is rare. In contrast, lipoma may be encountered anywhere along the neuraxis and can be divided into the congenital and maldevelopmental groups and a smaller neoplastic subset (Hori and Ikeda, 1982). Sometimes congenital malformative lesions contain a heterotopic component such as smooth and striated muscle, aberrant peripheral nerve fiber and a neuroglial element. Bone or cartilagenous components are relatively rare. We have added pathological details of a congenital lipoma with an osteocartilagenous component.

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CASE REPORT

The pregnancy of a 26-year-old women whose prenatal care had included an ultrasound examination revealing hydrocephalus was artificially terminated at 24 weeks of gestation. Immediately after delivery, an abnormally formed head, 24 cm in circumference, was apparent. The weight of the fetus was 620 gm.

Autopsy finding

An external examination revealed crown-to-rump length of 23 cm, rump-to-heel length of 11 cm, head circumference of 24 cm (normal 21.0 ± 1.2 cm), chest circumference of 19 cm, and abdominal circumference of 15 cm.

The brain weighed 83g (normal 59.6 ± 28.25g) and was hydrocephalic in appearance with unremarkable leptomeninges. The cerebral hemisphere revealed a unilocular cyst-like structure of hydrancephalic appearance. The cerebral convolutions were flattened. Neither macrogyria nor microgyria was found. The aqueduct of Sylvius was extremely narrowed and the brainstem showed an ill defined mass in the left lateral side of the

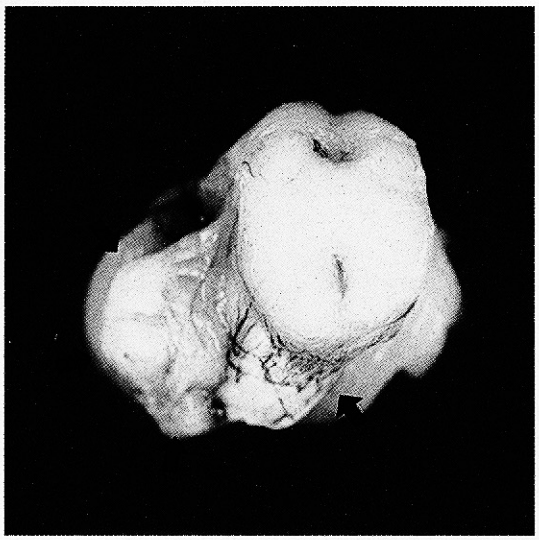

Fig. 1. Tectal area reveals a small solid mass (arrows).

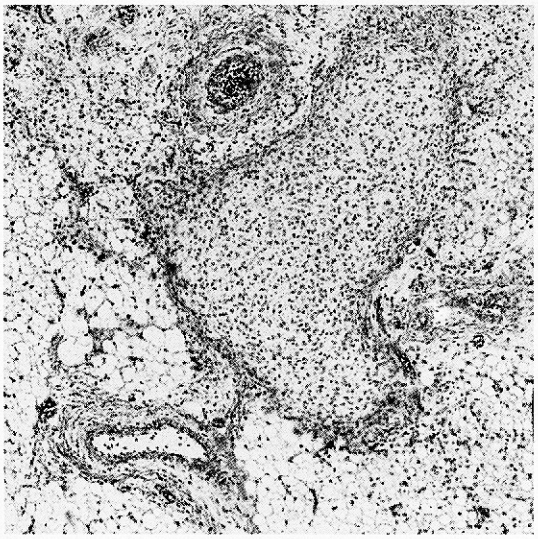

Fig. 2. The tumor is mainly composed of adipose tissue.

tectum (Fig. 1). Overall dimensions of the mass were 1.1 ×1.0×0.7 cm. It was thought to be the cause of hydrocephalus in this fetus. On section, the lesion appeared to be an ill-defined solid mass with a focal white soft area and some bony areas. However, the cerebellum and fourth ventricle occupied an extremely small portion. Other internal organs and placenta were unremarkable.

Histologic sections were taken from all organs including the brain stem. Microscopically, the tectal mass was composed of mature fat, cartilage, and bony tissue with hematopoiesis (Fig. 2 & 3). A nest of ganglion cells was also identified. Neither an endodermal nor ectodermal element was identified. The ependymal lining of the aqueduct was intact. Remaining brain tissue showed no abnormalities except for prematurity corresponding to 24 weeks of gestation. Other internal organs were unremarkable.

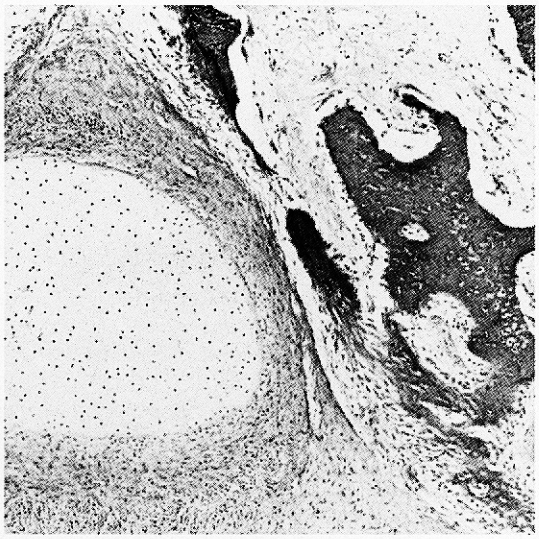

Fig. 3. Mature cartilage and bone are included in the lipoma.

DISCUSSION

In 1982, Sabet reviewed 110 cases of congenital intracranial neoplasm; according to him, 35% of these were teratoma and 31% were glioma, including four cases of glioblastoma. In descending frequency of incidence, these were followed by medulloblastoma, neuroepithelioma, craniopharyngioma, meningioma, papilloma, malignant choroid plexus tumor and mesenchymal tumor. Lipoma may be seen anywhere along the neuraxis, although the falx, hypothalamic area and dorsal brainstem are most common in location (Gouvea et al.,1989). When it involves the brainstem, the midline is a favorite site. In our case, the tumor was in a relatively uncommon location. The mass was on the left, involving quadrigeminal body and encroaching on the superiodorsal cerebellar hemisphere. Generally intracranial lipomas do not contain cartilage, muscle or bone (Hori and Ikeda, 1982). Several authors have,

however, described some lipomas as having heterotopic tissues, such as skeletal muscle (Fix et al., 1989), salivary glands (Curry et al., 1982), bone, respiratory epithelium, mucous and serous glands (Walsh and Markesbery, 1980). The mass in this case was composed of mature fat, cartilage and bone with hematopoiesis ; only the mesodermal element. Several sections revealed neither an endodermal nor ectodermal component.

The developmental origin of lipomas is uncertain. Some regard it as a form of teratoma (Walter and Kleinert, 1987), hamartoma (Budka, 1974 ; Russell and Rubinstein, 1989), metaplasia in the undifferentiated fetal tissue (Tamagawa et al.,1989) or heterotopia (Willis, 1968). Tresser et al. (1993) proposed transition between lipoma and teratoma. The osteocartilagenous component in this case favored metaplasia theory. Metaplasia is considered to be derived from the special ectomesenchyme of the neural crest (Tamagawa et al., 1989), the outgrowth of some displaced mesodermal cells (Willis, 1968) or persistent pluripotent embryonic cells (Russell and Rubinstein, 1989). Metaplastic components may include striated and smooth muscle, cartilage and bone. These mesodermal components have been found in human or animal central nervous tissue. However, it is quite rare compared with heterotopic glioneuronal tissue. Vanderahe and Niemer (1944) suggested that tumors originating during or before gastrulation could possess neuroectodermal and mesodermal components ; those developing later are composed only of more highly developed mesodermal elements and those originating last consist only of fat.

Congenital lipoma as the cause of hydrocephalus is uncommon. The incidence of congenital hydrocephalus is estimated to be between one and three of every 1000 live births ; most cases are of the noncommunicating type. Congenital malformation of the central nervous system can also be associated with hydrocephalus (Jung et al., 1990 ; Koo and Chi, 1991). They are holotelencephaly, agenesis of the corpus callosum, Arnold-Chiari malformation, Dandy-Walker malformation etc.

With the development of various antenatal examination methods, especially ultrasonography, the chances of detecting congenital anomalies are increasing. Antenatal sonography has also improved intrauterine follow-up. Increased detection of anomalies and support facilities give a few more cases which provide the pathogenesis of metaplasia in lipoma.

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