Bilateral subpleural ectopic brain tissue in a 23-week-old fetus

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Bilateral lesions were seen in the subpleural region in a 23-week-old aborted male fetus. This fetus was not macerated and showed no central nervous system abnormality on physical examination and vertebral magnetic resonance imaging (MRI). Postmortem examination revealed bilateral, paravertebral, subpleural, circumscribed, yellowish-white, fluent lesions 2.5 x 1 x 1 cm in size. These lesions were localized on the upper part of both lungs and there was no other internal malformation. Histological examination of lesions showed adult neurones and well-differentiated neural tissue with white and gray matter, choroid plexus, ependymal structures and, rarely, some peripheral neural cells in addition to immature neuroectodermal cells. These cells were more mature than those in the brain tissue.

Key words: ectopic brain tissue, lung, triple test.

Central nervous system heterotopia in the lung is a rare abnormality and is commonly associated with encephalocele[1-3], anencephaly[4-9], hemicephaly[10], and rarely, no central nervous system abnormality[11,12]. The etiology of intrapulmonary neuroglial heterotopia is still obscure. Several hypotheses have been proposed, such as aspiration and implantation of detached neural fragments within the amniotic fluid[6,7], defects of neural crest migration[3,6], vascular embolization and implantation of neural tissue following central nervous system trauma[4], and primitive mesenchymal differentiation into neuroglia[11]. The purpose of this paper is to present an unusual case with bilateral, subpleural, well-differentiated neural tissue in a non-macerated fetus that differs from other cases in the medical literature.

Case Report

A 40-year-old pregnant woman was admitted to the hospital because of Down syndrome risk due to high triple test (1/80): (alpha fetoprotein 47 ng/ml (1.2 MoM), human chorionic gonadotropin 25 IU/ml (1.22 MoM), and urinary estriol 1 ng/ml (0.59 MoM) at 18 weeks' gestational age. There was a first-degree consanguinity between parents in family history. The spastic second child died at one year of age and two other siblings were healthy. The woman suffered from mitral insufficiency due to rheumatic heart disease and was under treatment (digoxin, dipyridamole, benzathine penicillin G/per month). Cloudy amniotic fluid was obtained from amniocentesis that was performed at 19 weeks' gestational age but the growth of cells was unsuccessful. For that reason chordocentesis was performed at 23 weeks' gestation, and the chromosome analysis was found normal, 46 XY. Unfortunately, spontaneous abortion occurred three weeks after chordocentesis. Weight and length of the aborted fetus were 410 g and 30 cm, respectively. The placenta was 120 g. The general appearance was completely normal except for a dimple sized 0.2 cm on the nose. Postmortem examination revealed bilateral, intrathoracic, paravertebral, subpleural, circumscribed, yellowish-white, fluent lesions sized 2.5 x 1 x 1 cm. These cystic lesions were localized on the upper part of both lungs and there was no another internal malformation (Fig. 1). Histological examination of cystic lesions showed mature neurones, well-differentiated neural tissue with white and gray matter, well-organized cerebral cortex, choroid plexus, ependymal structures (the cysts were not lined by ependyma) and, rarely, some peripheral neural cells in addition to immature neuroectodermal cells (Fig. 2-4). These cells were more mature than those in the brain tissue. Brain and cerebellum were of normal morphology for

age and there was no abnormality in connection between basal parts of the brain and occipital foramen. Likewise, postmortem vertebral X-ray and magnetic resonance imaging (MRI) were also free of any defect.

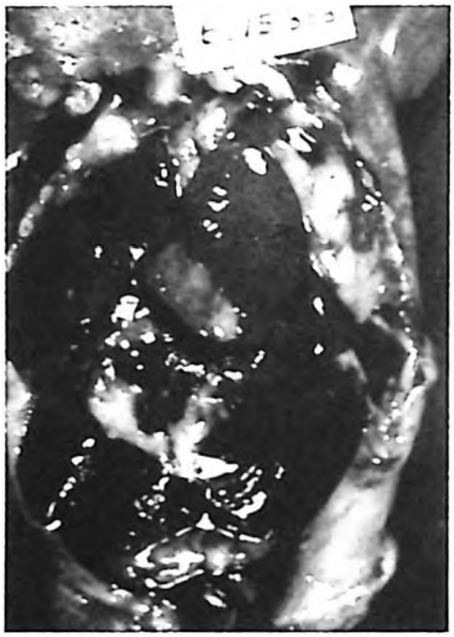

Fig. 1. Bilateral, intrathoracic, paravertebral, subpleural, circumscribed, yellowish-white, fluent lesions sized 2.5 x 1 x 1 cm. These lesions were under the parietal pleura and there was no association with the pulmonary parenchyma. No internal malformation was noted.

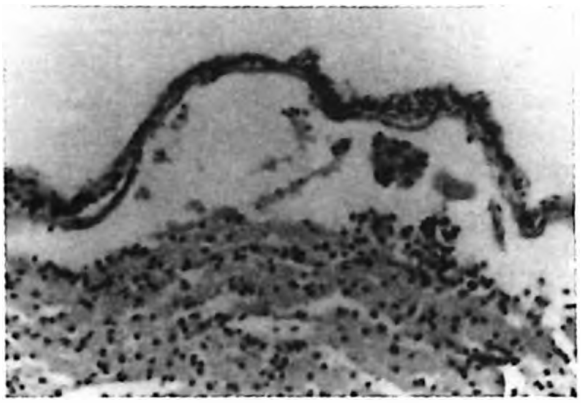

Fig. 2. Necropsy taken from the mass demonstrating arachnoidal membrane (Olympus BH-2, HEx100).

been reported; its etiology is still obscure. Several hypotheses have been proposed, such as aspiration and implantation of detached neural fragments within the amniotic fluid, defects of neuronal migration, vascular

Fig. 3. Necropsy taken from the mass demonstrating choroid plexus (Olympus BH-2, HE x 200).

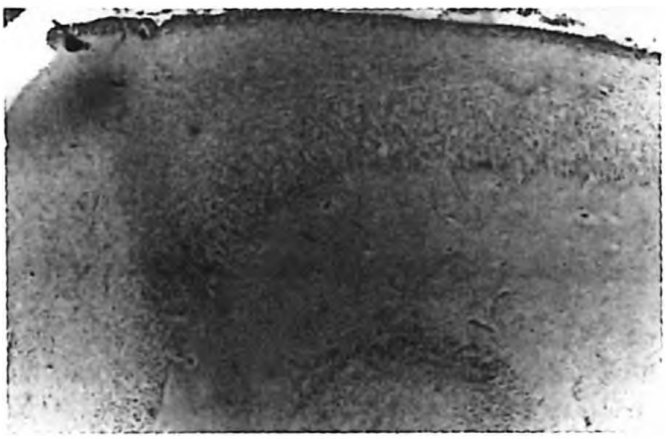

Fig. 4. Ependymal structures and a well-organized neural tissue. Ammon's horn-like appearance was noted (HEx400).

Discussion

The occurrence of brain tissue outside the cranial cavity is exceedingly rare. It is seen most frequently in central nervous system defects such as encephalocele[1-3], anencephaly[4-9], hemicephaly[10] and, rarely, with no central nervous system abnormality[11,12]. To date, 18 cases with intrapulmonary neuroglial heterotopia have

embolization and implantation of neural tissue following central nervous system trauma, and primitive mesenchymal differentiation into neuroglia. Embolization of brain tissue has been well documented after severe head trauma in adults and children. Hauck et al.[13] observed embolic cerebellar tissue in the pulmonary and coronary arteries of a male infant. Likewise, van

Noort and de la Fuente[14] reported primitive neuroectodermal tumors in macerated fetuses. The authors produced a similar lesion in a 12th week fetus by experimental compression of the skull and suggested that these tumors in macerated fetuses should be considered an artifact[14]. However, in our patient, there was neither maceration nor findings of trauma. Vertebral X-ray and MRI were also normal without any defect. The hypothesis of aspiration of neural tissue from amniotic fluid was unlikely in our case because other cases in literature all had central nervous system defects except for two cases[12,13]. Unilateral development of a teratoma is certainly possible. Moreover, intrapulmonary teratomas have been observed mostly in young adults and older individuals[15] and only rarely in infants[16]; all appeared as a distinct tumor. Valdes Dapena and Arey[17] showed a glial nodule in the lungs of a 51-year-old man with no apparent skull malformation, which was interpreted as an embryoma. Presence of mature neurones, well-differentiated neural tissue with white and gray matter, well-organized cerebral cortex, choroid plexus, ependymal structures and, rarely, some peripheral neural cells in addition to immature neuroectodermal cells suggested to us primitive mesenchymal differentiation was likely possible in the etiology. Until the mechanisms of cellular differentiation are better understood, these cases will remain in question. Further observations will enlighten the etiology of these rare abnormalities.

REFERENCES

1. Askanazy M. Beiträge zu den Beziehungen zwischen MiB- und Geschwulstbildung anläBlich einer Beobachtung einer eigenartigen Schädelhernie mit Lungengliom. Arb Ge Pathol Anat Inst Tübingen 1908; 6:433-499.
2. Hückel HR. Über Gliaektopien in der Lunge bei angeborener vorderer Hirnhernie. Verh Dtsch Ges Pahol 1929;24:272-279.
3. Gruenwald P. Emboli of brain tissue in fetal lungs. Am J Clin Pathol 1941; 72: 281-285.
4. Potter EL, Young RL. Heterotopic brain tissue in the lungs of two anencephalic monsters. Arch Pathol 1942; 34: 1009-1015.
5. Okeda R. Heterotopic brain tissue in the submandibular region and lung: report of two cases and comments about pathogenesis. Acta Neuropathol 1978; 43: 217-220.
6. Kanbour AI, Barmada MA, Klionsky B, Moossy J. Anencephaly and heterotopic central nervous tissue. Arch Pathol Lab Med 1979; 103: 116-118.
7. Chen WJ, Kelly MM, Shaw CM, Mottet NK. Pathogenic mechanisms of heterotopic neural tissue associated with anencephaly. Hum Pathol 1982; 18: 179-182.
8. Rakestraw MR, Masood S, Ballinger WE. Brain heterotopia and anencephaly. Arch Pathol Lab Med 1987; 111: 858-860.
9. Kershisnk MM, Kaplan C, Craven CM, Carey JC, Townsend JJ, Knisely AS. Intrapulmonary neuroglial heterotopia. Arch Pathol Lab Med 1992; 116: 1043-1046.
10. Campo E, Bombi JA. Central nervous system heterotopia in the lung of a fetus with cranial malformations. Wirchows Arch (Pathol Anat) 1981; 391: 117-122.
11. King WI. Tumor (embryoma) of lung containing brain tissue. Med Bull VA 1938; 15: 181-183.
12. Gonzales-Crussi F, Boggs JD ,Raffansperger JG. Brain heterotopia in the lungs. A rare cause of respiratory distress in the newborn. Am J Clin Path 1980; 72: 281-285.
13. Hauck AJ, Bambara JF, Edwards WD. Embolism of brain tissue to the lung in a neonate. Report of a case and review of the literature. Arch Pathol Lab Med 1990; 114: 217-218.
14. van Noort G, de la Fuente AA. So-called primitive neuroectodermal tumor in macerated fetuses: a confusing artifact. Pediatr Pathol 1988; 8: 359-365.
15. Collier FC, Dowling EA, Plott D, et al. Teratoma of the lung. Arch Pathol Lab Med 1968; 68: 138-142.
16. Pound AW, Willis RA. A malignant teratoma of the lung in an infant. J Pathol 1969; 98: 111-114.
17. Valdes Dapena M, Arey JB. Pulmonary emboli of cerebral origin in the newborn. Arch Pathol Lab Med 1967; 84: 643-646.