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Spontaneous Pneumocephalus with Meningomyelocele: A Case Report

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Abstract | **Case Report**

Spontaneous pneumocephalus is one of the complications of open myelomeningocele with or without clinical signs, reported very rarely in the literature. Pneumocephalus It can associated with several neurosurgical procedures, lumbar puncture, or cranial trauma; But very rarely, the open neural tube defects may lead to spontaneous pneumocephalus. The radiographic appearance of intraspinal air on CT was not described previously. The following report presents a case of spontaneous appearance of internal pneumocephalus in a newborn infant with open myelomeningocele which environmental air could gain access to the cerebrum associated with bi-ventricular hydrocephalus and an Arnold-Chiari type II malformation, revealed on CT images. Spontaneous pneumocephalus should be kept in mind as a rare complication of open meningomyelocele. Any patient with open meningomyelocele should be early evaluated for pneumocephalus with a cranial imaging.

Keyword: Spontaneous pneumocephalus, open myelomeningocele, CT.

INTRODUCTION

Spontaneous pneumocephalus with or without clinical signs is one of the complications of open myelomeningocele, reported very rarely in the literature; it may have an impact on prognosis and therapy. We report a case of open myelomeningocele who developed a spontaneous pneumocephalus revealed on CT images (computed tomography). The purpose of this report is the review of the literature of the imaging of spontaneous pneumocephalus and to raise the interest of these techniques in the diagnosis and post-therapeutic follow-up.

CASE REPORT

This full-term boy was born vaginally to a 39 years old mother G3 P3, she hasn't a health problems. The pregnancy was normal without any complication, except the detection of a neural tube defect by prenatal sonography at 21 weeks of gestation. At birth, the infant weighed 3215g and had an Apgar score of 9; an open dorsolumbar meningomyelocele was confirmed. Close examination showed leakage of cerebrospinal fluid with obvious exposure of nerve roots (Figure 1).

Head circumference was increased with prominent occiput and bulging anterior fontanelle; the sutures were widely open.

Cerebrospinal fluid was dripping out of a sacral meningomyelocele that probably had been ruptured at the time of delivery or shortly after. The rest of neurological examination was normal.

A cerebral CT was ordered showing a bi-ventricular hydrocephalus with an Arnold-Chiari type II malformation, and a pneumocephalus also seen in the Clivus, the perimesencephalic and pre-punctal cisterns and in the left parietal area. (Figure 2: A-D).

An excision and plastic closure of the open meningomyelocele with placement of shunt were performed. The cranial air subsided over the next days.

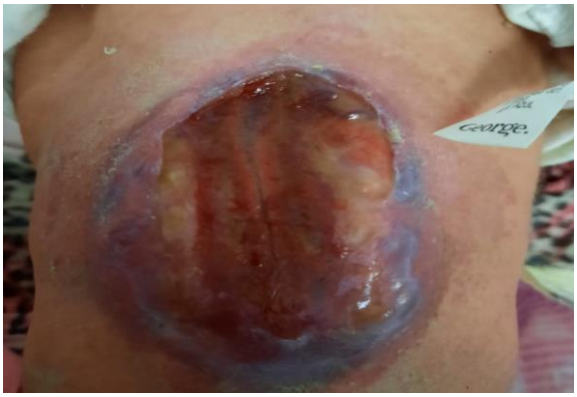

Fig-1: Image of new born baby with dorso-lumbar spina fibida, with leakage cerebrospinal fluid.

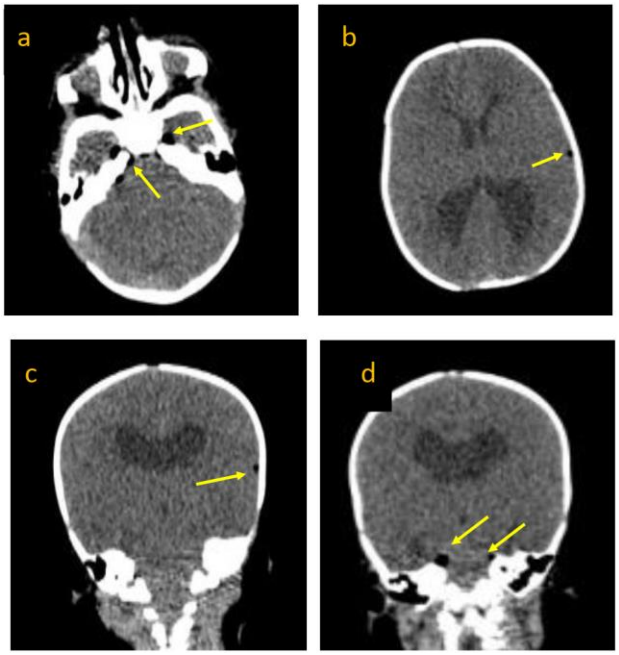

Fig-2: Axials (a,b) and coronals (c,d) Non enhanced Cranial CT Scan shows Pneumocephalus in the Clivus, the perimesencephalic and pre-punctal cisterns suprasellar air and in the left parietal area (arrows).

DISCUSSION

In the present day of antenatal screening, ultrasound, amniocentesis and folate therapy, we still are unfortunate to see babies with open neural tube defects being born occasionally. Myelomeningocele is one of this congenital malformations, it can have several complications including spontaneous pneumocephalus.

The exact incidence of pneumocephalus in myelomeningocele cannot be estimated owing to the rarity of this phenomenon. Only few case reports have been describing this finding in the literature [1]. Pneumocephalus It is associated with several neurosurgical procedures, lumbar puncture, or cranial trauma [2].

In addition, intracranial air is occurring as a complication of mechanical ventilation and tension pneumothorax. A nasopharyngeal oxygen catheter that had perforated the skull base and violated the dura mater has also been documented as a cause of intracranial air. Very rarely, the open neural tube defects may lead to spontaneous pneumocephalus [3].

Pneumocephalus, as previously described in a few case reports of newborn infants with a myelomeningocele, can increase intracranial pressure, with signs of respiratory distress and generalized muscle hypotonia [3]. But, it is unclear whether the pneumocephalus or pneumorachis played any compressive role in contributing to the clinical signs.

There are two possible pathophysiologic mechanisms for air to enter the ventricular system [3]. In addition to a ball valve mechanism, by which air is thought to be forced into the cranial cavity by coughing, sneezing, or straining [4], the most plausible mechanism of air entering the spinal canal and ascending to the cranial cavity is the so-called ‘‘inversed bottle effect’’ [5]. When a bottle is inverted and the liquid is evacuated, air enters to fill the dead space or compensate for negative pressure [3]. When a sufficient amount of cerebrospinal fluid has escaped and is not compensated by a corresponding increase in brain volume, the result is negative intracranial pressure, and air will enter via the same channel. The leakage of cerebrospinal fluid through an open myéloméningocèle creates negative pressure, allowing air to penetrate the subarachnoid space [3]. This mechanism, as demonstrated by fluoroscopy, was described by Trawoger *et al*. in a case report of a newborn with myelomeningocele [3].

Cranial findings in meningomyelocele have been reported in the literature by sonography, and magnetic resonance imaging [6, 7]. The finding of spontaneous pneumocephalus in Cerebral CT hasn’t been reported previously. This is probably the first case in the literature where this is reported.

Among these imaging techniques, transfontanellar sonography remains the most non-invasive, safe and inexpensive technique that can be performed by the bedside. Reported cranial sonographic findings in spontaneous pneumocephalus have been rarely described. The key features indicating air as the source for the bright intracranial echoes with acoustic shadows were the characteristic "ring-down" artifacts associated with gas collections anywhere within the central nervous system, distal shadowing owing to almost total sound reflection at the tissue-air interface, and the movement of loci with change in position of the patient. It can also shows associated abnormalities like hydrocephalus with dilated lateral ventricles, partial absence of septum pellucidum, third ventricle abnormalities and interhemispheric fissure...etc [6].

A cranial CT is more efficient; it can makes positive diagnosis and associated diseases. It can also show a Chiari malformation [3]. It is the same of our case.

Magnetic resonance imaging shows the spinal and cerebral malformations much better than the others image technique, in addition to pneumorachis and pneumocephalus [7].

The association of myelomeningocele with multiple cranial and spinal defects is well established. The neurological outcome of patients with this condition is determined mainly by the level of the neural tube defect [3]. Almost all case of lumbosacral

myelomeningocele is associated with Chiari II malformation [8] and a variable degree of hydrocephalus [9]. Among other cerebral and spinal malformations associated with myelomeningocele, hydromyelia is reported in about 40 per cent of cases. The respiratory complications of Chiari malformations have been well described in the literature. The poor respiratory effort is due to the hypoplastic lower cranial nerve nuclei and depressed brain stem respiratory centers and ascending reticular-activating system by the pneumocephalus [1].

Myelomeningocele is usually complicated with intracranial infections such as meningitis and ventriculitis; this complication had been reported in two cases of spontaneous pneumocephalus and had resulted in the death of the infant [1].

Treatment of simple pneumocephalus is usually conservative (oxygen, head down position, etc.). However, Massive pneumocephalus can increase the intracranial pressure and behave as life threatening condition. Drainage of air by a needle or a subdural drain may be performed. Not forgetting the cure of myelomeningocele, and drainage of hydrocephalus to reduce intracranial pressure, minimize the risk of infection and to prevent worsening of pneumocephalus [2].

CONCLUSION

Spontaneous pneumocephalus should be kept in mind as a rare complication of open meningomyelocele. Any patient with open meningomyelocele should be early evaluated for pneumocephalus with a cranial USG, computed tomography or magnetic resonance imaging, but almost importantly, early operative closure of the neural-tube defect can prevent the movement of penetrating air into the intraspinal and intracranial cavities.

REFERENCES

1. Kutty, R. K., Sreemathyamma, S. B., Sivanandapanicker, J. L., & Peethambaran, A. (2018). Spontaneous pneumocephalus secondary to leaking myelomeningocele. *Journal of pediatric neurosciences*, *13*(3), 329.
2. Ödemis, E., Aslan, Y. (2004). Indian Pediatrics, 41; 289-290
3. Trawöger, R., Strasser, K., Ellemunler, H., & Gassner, I. (1994). Spontaneous pneumocephalus in a newborn infant with myelomeningocele and hydromyelia. *Developmental Medicine & Child Neurology*, *36*(10), 924-927.
4. Markham, J. W. (1967).The clinical features of pneumocephalus based on a survey of 284 eases with report of 11 additional cases. *Acta Neuroehir, 15; 1, 78*.
5. Pop, P. M., Thompson, J. R., Zinke, D. E., Hasso, A. N., & Hinshaw, D. B. (1982). Tension pneumocephalus. *Journal of computer assisted tomography*, *6*(5), 894-901.
6. Kao, S. C. S., Brown, B. P., & Goedken, J. (1991). Sonography of intracranial air in a newborn with meningomyelocele. *Pediatric radiology*, *21*(5), 375-376.
7. De Roos, P. B., van Lindert, E. J., van der Vliet, T. A., & Sie, L. T. (2009). MRI of pneumorachis in a newborn infant with open myelomeningocele. *Pediatric neurology*, *40*(5), 377-379.
8. Caviness Jr, V. S. (1976). The Chiari malformations of the posterior fossa and their relation to hydrocephalus. *Developmental Medicine & Child Neurology*, *18*(1), 103-116.
9. Noetzel, J. (1989). 'Myelomeningocele: current concepts of management.' *Ciinirs in Perinalology, 16*, 3 1 1-329.