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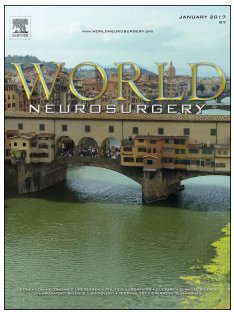

Pneumoventricle of Unknown Origin: A Personal Experience and Literature Review of a Clinical Enigma

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Pneumoventricle of Unknown Origin: A Personal Experience and Literature Review of a Clinical Enigma.

Short Title: A case of Pneumoventricle of Unknown Origin

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Abstract

Pneumocephalus (PC) is an uncommon and life-threatening neurological condition. Air within the ventricular system of the brain is also known as Pneumoventricle (PV). It requires emergency treatments to prevent catastrophic neurological outcomes. Head injury is the most common cause of PV, but there are other well-recognized etiologies in case there is no clear radiological evidence of skull discontinuity. Although this clinical entity has been well described in Literature, our report presents the unique feature of describing a purely ventricular PC without evidence of skull base or cranial vault fracture. Therefore, this case presentation explores mysterious causes of fistulous connections with the atmosphere that may lead to air trapped in and around the cranial vault.

The aim of the present paper is to report a case of post-traumatic PV without radiological signs of skull base or convexity fracture in a 72-years-old man, underlining the diagnostic and clinical features, and review the relevant Literature.

Keywords: Pneumoventricle, Pneumocephalus, Head Trauma, MRI, HRCT.

Pneumoventricle of Unknown Origin: A Personal Experience and Literature Review of a Clinical Enigma.

Introduction

Pneumocephalus(PC) defines the presence of air within the cranial vault, and this may involve the extradural, subdural, subarachnoid, intracerebral, and intraventricular compartments[1]. Air within the ventricular system of the brain is also termed Pneumoventricle(PV). The accumulation of intracranial air can be acute(<72 h) or delayed(≥72 h)[1-2]. More commonly, an intracranial air collection is a benign and asymptomatic condition. Tension pneumocephalus(TPC) occurs when intracranial air causes intracranial hypertension generating a mass-effect with neurological deterioration. However, the conversion of a PC into TPC is a rare event. PC could per se increase the risk of developing meningitis also[3].

Most cases of PC with involvement of ventricles occur as a result of discontinuity in the cranium, including skull fractures which let the atmospheric air access the intracranial compartment through a direct communication; such fractures involve cranial vault and/or skull base fractures including air-containing sinuses such as the frontal, ethmoid, sphenoid, or maxillary sinuses. It can occur as a result of an head trauma in 3.9–9.7% of cases and after supratentorial craniotomy either, but can also be a result of non traumatic causes[3].

Cases of spontaneous non-traumatic PC remain highly uncommon; previously reported causes include malignancies, meningeal infections with gas forming bacterial strains, and nasopharyngeal carcinomas[6]. Although a relationship between misdiagnosed skull base diseases and PC has been hypothesized, there is no Literature reporting clear evidences of an association between infection involving the skull base and its sinuses and the development of such conditions.

The aim of this paper is therefore to report a unique case of pure PV of unknown origin without evidence of air collection at the convexity or in the fissures and without a radiological evidence of infection or skull fracture, discussing the most salient radiological clinical and prognostic features of such a yet poorly understood and widely unexplored clinical condition.

Case report

A 72-years old man, accessed the Emergency Department of our Institution reporting a clinical history of 48 hours headache, vertigo and confusion. Such symptoms preceded an incidental fall with a minor head-trauma without loss of consciousness. Neurological examination showed no focal sign or evidence of cranial nerve impairment, GCS score at first evaluation was 14. No obvious cerebrospinal fluid(CSF) rhinorrhea or othorrhea were observed. An head and neck High Resolution Computed Tomography(HRCT) ruled out the presence of skull fractures. However, a significant collection of intraventricular air was found bilaterally in the frontal horns, in anatomical contiguity with the anterior aspect of the third ventricle, moderately compressing the brain (Figure 1). No air was disclosed in the basal cisterns, in the posterior cranial fossa, in the overlying cortical giry or in the Sylvian and Interhemispheric fissures. The supratentorial ventricular complex was straight in the midline and partially dilated. Furthermore, CT disclosed no intracerebral bleeding. The temporal bones, including the mastoid processes, were completely devoid of fluid collections and nevertheless showed a well-developed pneumatisation. A detailed petrous bone HRCT scans, demonstrated no continuity between petrous bone, orbit, ethmoid sinus, sphenoid sinus and the intracranial compartment (Figure 2).

The patient was admitted in the Neurological Department of our University Hospital and treated by means of a conservative approach with absolute bed rest with head lying in a straight position, intravenous antibiotics(Ceftriaxone at 2 g per day dosage) and intravenous corticosteroids(Dexamethasone at 4 mg twice per day dosage) were also administered. An in-depth ENT examination, including nasal endoscopy and pure tone audiometry, were normal.

A Contrast-enhanced brain Magnetic resonance imaging(MRI) scan, performed the following day, better defined the intraventricular location of the PC, and confirmed the absence of any other lesions involving the brain. Nine days after, the patient underwent a new cranial CT scan that yielded unmodified results, and the patient was discharged with tapered corticosteroid treatment and the recommendation to avoid physical efforts. During the entire period, the patient presented no fever, besides his laboratory findings were within the normal range, no rhinoliquorrhea or otholiquorrhea were observed also.

The patient remained asymptomatic and gradually resumed regular physical activity. He underwent periodic radiological follow-up examinations to verify the status of the PV. Cranial CT scans at one, two and six months, and a temporal bone CT scan at 3 months all showed a strong reduction of the air collection until a normal ventricular complex aspect was restored.

Discussion

The presence of intracranial air normally points out to the presence of an open communication between intracranial compartment and the atmospheric air; it should be considered as a form of cerebrospinal fluid (CSF) fistula[9]. Air enters the intracranial space through dural tears even without a direct brain laceration. It is a serious condition and a neurosurgical emergency treatment may be required especially when associated with clinical deterioration. TPV, which indicates the presence of air trapped under pressure within the ventricles, is a rare incident. It is occasionally observed following craniofacial trauma and its incidence has been estimated to range between 0.5% and 1.0% of all head injuries[5]. Nontraumatic factors causing PC include intracranial tumors, intracranial infection and complications of craniotomies and spheno-ethmoidal surgeries. It may follow spinal/epidural anesthesia, positive pressure ventilation, and administration of hyperbaric oxygen[4].

1. *Pathogenesis*

Some authors advocated that two requirements are needed for the development of s PC:

1. The presence of a CSF diversion system that causes a decrease in intracranial pressure, and
2. The presence of a craniodural defect with or without an obvious CSF leak.

Two mechanisms have been postulated to account for the entrance of air into the cranial vault:

- The “ball-valve mechanism” was previously described as a causative agent when a fistula allows atmospheric air (at an higher pressure value in respect to the ICP) to enter the intracranial space[8]. This process continues until the ICP value overcomes the atmospheric pressure and the brain and dura mater are forced over the craniodural defect. Under such

circumstances a "valve mechanism" is established, such a mechanism does not allow air to outflow,

- A second mechanism establishes when a continuous CSF leak from an enclosed space is present, the subsequent loss of CSF creates a "void space" and relative negative pressure, allowing air to bubble in and fill the void[9,10]. This scenario is called the *"Coke-bottle mechanism."*

The term *spontaneous or non-traumatic PC* refers to condition of *unknown origin*, whereby air accumulates intracranially[7] without an obvious causative agent. Iatrogenic factors predominate among atraumatic PC. Lee et al.[15] reported a patient who developed extensive TPC that required surgery after undergoing a diagnostic spinal tap. Nitrous oxide as an anesthetic agent can diffuse into air-filled spaces causing expansion of any trapped air loculi. This condition has been linked to intraoperative onset of TPC[9].

Small volume collections of intracranial air(<2 ml) present usually an asymptomatic course and resolve without treatment over about 2–3 weeks[14]. Larger volumes can be encountered with major skull base fractures involving the air-containing sinuses and/or the cribriform plate, TPC will ensue in such scenario unless other extracranial communication occurs. It is a serious complication evolving in a neurosurgical emergency when associated with clinical and neurological deterioration.

2. Symptoms

When a PC is limited in extent, clinical features are usually few, with headache being the most common symptom. However, forceful spread of air into intracranial spaces may have serious consequences. Clinical signs of massive intracranial air include a headache, CSF rhinorrhea, pupillary changes, agitation, delirium and altered level of consciousness, focal neurological deficit, convulsion, frontal lobe syndrome, and cardiac arrest[1-2]. Episodes of bradycardia may also be seen with or without hypertension. The rapid increase of intracranial pressure may be difficult to compensate, giving rise to intracranial hypertension, with symptoms such as agitation, delirium, loss of consciousness, stiff neck, lethargy, disorientation and papilloedema[11].

3. Diagnoses

An high level of clinical suspicion and confirmation through Neuroimaging are important in reducing mortality and morbidity. Head and Neck HRCT scans can play a critical role in determining the precise location of the gas collection along with its relationship with the exact location of the skull base fractures, the number and extent of the air bubbles, and of course the severity of the intracranial mass effect[6,7,12].

Magnetic resonance imaging (MRI) can also demonstrate the presence of intracranial air, of course with longer image acquisition time and greater costs. On MRI, intracranial air collections will appear completely dark (signal void) on all sequences.

Despite progress in MRI, CT definitely remains the "gold standard" diagnostic tool for tension PC. HRCT scans are also able to detect as little as 0.5 cm^3 of air. CT is used to easily identify the amount and location of air and fluid, presence of a craniodural defect, and the effect of trapped air on the brain[12]. The diagnosis of otogenic PC is usually straightforward, although some misinterpretation may occur, and the PC may be interpreted as a hyperpneumatisation extending to bones contiguous with the temporal bone[11-12].

4. Treatment

Treatment is conservative in most cases. A ventriculostomy may be required to relieve ICP in cases of tension PC or PV. Endoscopic or Conventional Surgical approaches to repair the dural breach may be considered whenever an associated CSF leak persists for more than a week. Although the prognosis is fair in most cases, the presence of multiple pockets of air has a worse prognosis compared with solitary intracranial air bubble[1] and an overall mortality rate as high as 10% is reported[5]. Since the development of TPC as a complication of a standard PC, can take a considerably variable amount of time, it is strongly suggested to consider an early repair of the fistula, especially if conservative treatments, such as bed rest, are not successful.

To the best of our knowledge, this is the first case reported in Literature in which no apparent cause of PV was retrieved both from the clinical history and from the results of the radiological and clinical investigation. The course of our patient was relatively benign and, after the onset with confusion

symptoms, which quickly recovered after bed rest and intravenous treatments, was uneventful. HRCT is reported to have an extremely high accuracy in determining the exact location of the skull base fractures in regards to, also small, skull fractures involving the base or the cranial vault[3]. In other terms the best of our currently available technology is, in fact, in a minority of cases not enough accurate in confirming the diagnosis of a small skull fracture, with a fracture related dural tear which is responsible for a CSF leak and subsequent PC/PV. In such cases, when there's no neurological deterioration it is always advisable to admit the patient for in depth investigations including HRCT and Gadolinium enhanced head and neck MRI scan, and nasal endoscopy. Even in case of no evidence of causative agent there is a clear necessity of a close range radiological and clinical follow-up to early detect the potential recurrences. In the author's opinion it is also important to inform the patient about the associated symptoms, such as oto and rhinoliquorrhea that could fleetingly appear and resolve in a time span of seconds. Such procedures do not yield a sure diagnosis but increase the probability to convert a case of PC/PV of unknown origin in a treatable patient.

Conclusions

Symptoms of PV are related to the amount of air that is within the cranial cavity. Although small amounts can be asymptomatic, larger amounts produce catastrophic neurological outcomes, consequently a early and accurate diagnosis is mandatory. It is essential that any precipitating factors is eliminated immediately and adequate investigations are promptly carried out to rule out bony infections[7]. There is a general lack of formal guidelines for the management of a non-traumatic PC and thus further evaluation of best management and follow-up are the gold standard for any patient.

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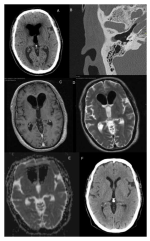
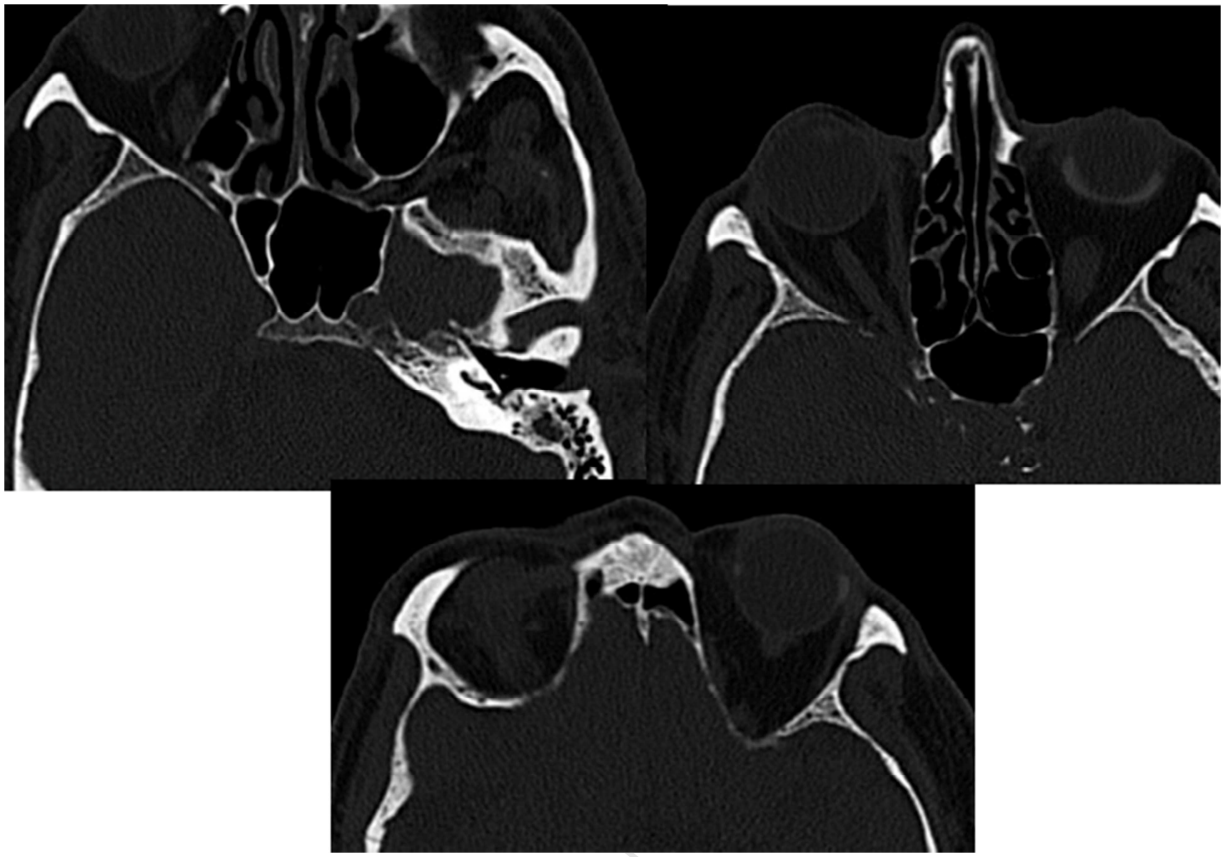
Figure Captions:

Figure 1: Gaseous distention of the frontal horn of the lateral ventricles was seen with mass effect on adjacent brain parenchyma in CT SCAN (A). The temporal bones, including the mastoid processes, were completely devoid of fluid collections and nevertheless showed a well-developed pneumatisation (B). Both anterior ventricular horns were also gas distended with demonstrable air-cerebrospinal fluid (CSF) levels specifically visualized in MRI T1-weighted and T2-weighted exam (C-D). The DWI confirmed the absence of any other lesions involving the brain (E). The CT-scan was repeated 1 month later and showed a significant reduction of the air collection, the normal ventricular complex aspect was restored.

Figure 2: No evidence of fracture or any comunication between orbit, sphenoid sinus, ethmoid sinus and intracranial space in CT SCAN.





Abbreviations: Magnetic Resonance Imaging (MRI), High Resolution Computed Tomography (HRCT), Pneumoventricle (PV), Pneumocephalus (PC), Tension Pneumoventricle (TPV), Tension Pneumocephalus (TPC), Ear Nose Throat surgeons (ENT), Cerebrospinal Fluid (CSF), Intracranial Pressure (ICP).