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Case Report

Holocord spontaneous pneumorrhachis in the setting of refractory emesis

Rebecca Houston[1], Brian Fiani[1], Brian Musch[2], Emilio Tayag[1]

[1]Department of Neurosurgery, Desert Regional Medical Center, Palm Springs, California, [2]College of Osteopathic Medicine, William Carey University, Hattiesburg, Mississippi, United States.

E-mail: Rebecca Houston - r.houston.4@gmail.com; *Brian Fiani - bfiani@outlook.com; Brian Musch - bmusch476901@student.wmcarey.edu; Emilio Tayag - emiltayag@hotmail.com

***Corresponding author:**
Brian Fiani, D.O.
Department of Neurosurgery, Desert Regional Medical Center, Palm Springs, California, United States.

bfiani@outlook.com

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ABSTRACT

Background: Pneumorrhachis (PNR) is the presence of air within the spinal canal and may be either intramedullary or extramedullary in location. The etiology is most commonly iatrogenic or traumatic in nature. Treatment is dependent on underlying cause and physical exam.

Case Description: Herein, we describe the second case in the literature of spontaneous holocord PNR in a young patient without risk factors. A 22-year-old male with no past medical history presented to the hospital for 2 days of vomiting and cramping in his hands and feet secondary to severe dehydration. He recently started a new job as a manual laborer and had to leave work early 2 days prior due to overexertion working outside in heat ranging from 100 to 120 degrees Fahrenheit. CT abdomen and pelvis demonstrated spontaneous pneumomediastinum and extramedullary PNR extending upward from L3 throughout the thoracic spine to the upper limit of the scan. Subsequent CT cervical and thoracic spine showed the full length of the extradural air from C2-T12 and again at L3.

Conclusion: Spontaneous PNR is an uncommon, typically self-limited condition in which air is introduced into the spinal axis. Anatomic predisposition makes the extradural, dorsal cord in the cervicothoracic region the most common location. Patients are rarely symptomatic, and treatment is supportive in nature once secondary causes with high rates of morbidity and mortality are ruled out.

Keywords: Holocord, Intraspinal air, Pneumomediastinum, Pneumorrhachis

INTRODUCTION

Pneumorrhachis (PNR) is the presence of air within the spinal canal and may be either intramedullary or extramedullary in location. The etiology is most commonly iatrogenic or traumatic in nature, though it is not uncommon for no definitive cause to be identified. Certain acute and chronic conditions predispose patients to the development of PNR including asthma, Valsalva maneuvers such as coughing, emesis and lifting heaving objects, respiratory infections and airway obstruction or perforation.

There is no established definition of spontaneous PNR in the literature. However, classification of pneumomediastinum as spontaneous has historically been defined as the absence of history of trauma and/or iatrogenic cause.[13] Herein, we define spontaneous PNR as the presence of air in

the spinal canal (either epidural or intradural) in the absence of trauma, iatrogenic cause or predisposing condition either acute or chronic in nature. We present a case of spontaneous holocord PNR treated at our hospital with positive outcome. We also aim to examine the causes, clinical presentation, diagnostic and treatment modalities associated with spontaneous PNR.

There are no published statistics in the literature regarding epidemiology of spontaneous PNR as there is no agreed-on definition. Most non-iatrogenic and non-traumatic cases of PNR are asymptomatic. When patients do have symptoms, they are often caused by associated conditions such as pneumomediastinum and subcutaneous emphysema causing chest pain, dyspnea, dysphagia and neck pain.[1] Treatment is aimed at symptomatic relief of symptoms of associated conditions, as described.

CASE DESCRIPTION

A 22-year-old male with no past medical history presented to the hospital for 2 days of vomiting and cramping in his hands and feet secondary to severe dehydration. He recently started a new job as a manual laborer and had to leave work early 2 days prior due to overexertion working outside in heat ranging from 100 to 120 degrees Fahrenheit. CT abdomen and pelvis (later reconstructed into CT lumbar spine) demonstrated spontaneous pneumomediastinum and extramedullary PNR extending upward from L3 throughout the thoracic spine to the upper limit of the scan [Figure 1]. Subsequent CT thoracic spine [Figure 2] and CT cervical spine [Figure 3] showed the full length of the extradural air from C2-T12 and again at L3. There was no evidence of cord compression. Communication of air between the central spinal canal and neural foramen was seen at the T10 level on axial CT. Upon examination, the patient was neurologically intact without abnormal reflexes, motor, or sensory deficits. He denied neck or back pain, numbness, tingling, weakness, incontinence of bowel or bladder.

The patient was admitted for completion of gastrografin esophagram which was negative for evidence of esophageal perforation. He was discharged home the day after presentation. At 3-week follow-up, the patient remained without neurological compromise.

DISCUSSION

PNR is a rare phenomenon in which there is air the spinal canal either outside the cord or within the spinal cord itself. Extradural location is most common.[1] While many cases have no definitive cause, the most commonly identified causes are trauma and iatrogenic introduction of air, either following a procedure accessing the spinal canal such as a lumbar puncture or spinal surgery, or as a complication due

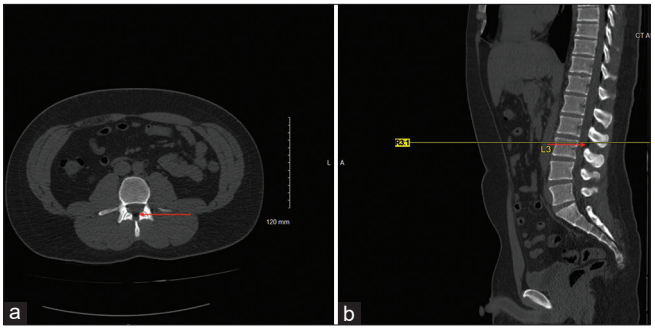

Figure 1: (a) Axial CT lumbar spine demonstrated air in the spinal canal at L3 without compression of the thecal sac. (b) Sagittal CT lumbar demonstrated air in the spinal canal extending from the lumbar region to the lower thoracic spine at the upper limit of the scan.

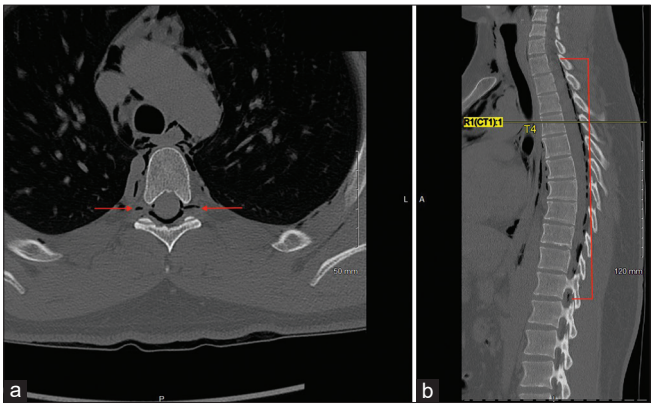

Figure 2: (a) Axial CT thoracic spine with air in the spinal canal communicating with the paravertebral extraforaminal space at the T4 level. (b) Sagittal CT thoracic demonstrates air in the spinal canal extending the entire length of the thoracic spine.

to procedures such as aberrant chest tube.[3,4] Additional risk factors for the development of PNR include asthma, inhalational drug use through increased in intrathoracic pressure (with cases documented following marijuana smoking and 3,4-methylenedioxymethamphetamine ("Ecstasy") inhalation), sacral pressure sore, sacral myelomeningocele, enterocutaneous fistula, influenza, leukemia, diabetic ketoacidosis, infection with gas-producing organisms, respiratory infection (particularly by mycoplasma pneumoniae, respiratory syncytial virus or influenza), airway obstruction from foreign body aspiration, perforated bowel and/or esophagus, and weightlifting.[3,6,11,13] Rarely, PNR can be a complication of spontaneous pneumomediastinum due to repeated, sustained Valsalva maneuver resulting in barotrauma.[6,13] In addition, there exists two cases in the literature in which the patients had none of the aforementioned underlying risk factors; the cause for each was determined to be erosion in the annulus fibrosis with vacuum disc phenomenon.[5,8]

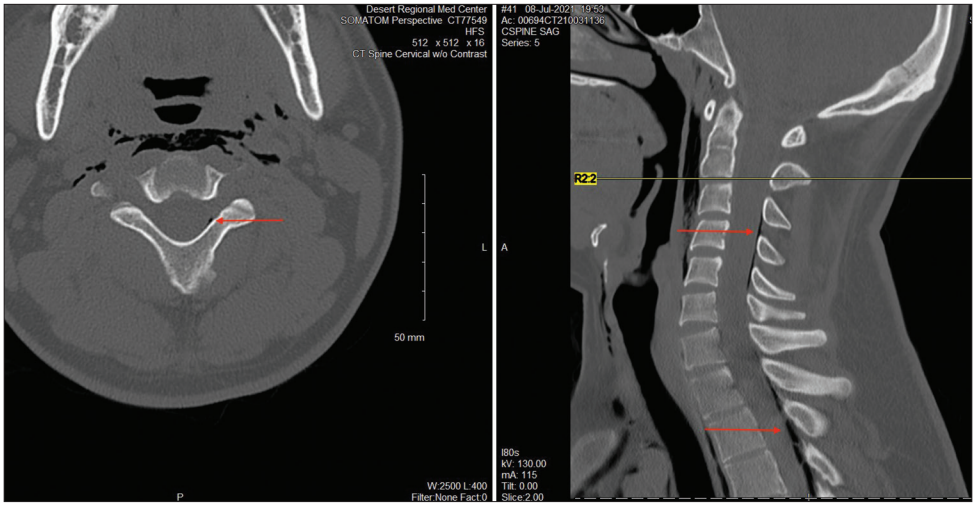

Figure 3: Axial and sagittal CT of the cervical spine demonstrating air in the dorsal spinal canal extending caudally to the C2 level.

Notably, in PNR, while air is most commonly located in the posterior spinal canal because of decreased resistance compared to the anterior epidural space which houses a denser vascular network, this patient demonstrated air in the anterior portion of the canal, proximal to the disc space. Air-filled facet cysts have also been proposed as a possible cause of PNR.[15] In instances where pneumomediastinum, pneumothorax, perforated bowel or esophageal rupture have occurred, the proposed pathophysiology for development of PNR is tracking of air through the neural foramina into the spinal canal.[1,3]

There are no pathognomonic signs or symptoms associated with PNR. There have been cases reported with neurological symptoms in the setting of iatrogenic and traumatic PNR secondary to cord compression. However, in spontaneous PNR, patients are there exists only two cases in the literature in which a patient had signs or symptoms of neurological compromise; in all other cases, patients were either asymptomatic, or complained of symptoms associated with their predisposing condition or those associated with pneumomediastinum, such as chest pain, dyspnea, dysphagia, neck pain, and cough.[9,10,13,15] In 2009 Song *et al*. reported, a case of a 72-year-old male with 1 month if bilateral lower extremity weakness and sensory deficits with a finding of epidural air at C7.[15] The patient had no history of trauma, iatrogenic cause, recent illness or predisposing condition. However, he was noted on imaging and intra-operative findings to have severe degenerative changes of his cervical spine, with hypertrophy of the C7–T1 bilateral facet joints and a thickening of the facet capsule. The proposed source of air was severe destruction of the degenerated facet joint itself. The patient made a full recovery after a C7 laminectomy. A second patient was noted to have bilateral lower extremity paresthesias in the setting of spontaneous pneumomediastinum with cervicothoracic PNR following violent coughing and shouting episodes.[10] The patient was treated conservatively and made a full recovery.

Classification of pneumomediastinum as spontaneous has historically been defined as the absence of history of trauma and/or iatrogenic cause.[13] PNR is an uncommon finding that is even more rare in healthy young adults without a history of trauma or drug use. When PNR occurs in the absence of trauma or iatrogenic causes, it is often (but not always) an epiphenomenon of pneumomediastinum. The incidence of holocord PNR, as seen in our case, is unreported. A 2021 systematic review of the literature by Alemu *et al*. that examined 339 pediatric and adult cases of spontaneous pneumomediastinum found PNR in 2.06% of cases.[1] In children, the incidence of PNR in a population of pediatric patients with spontaneous pneumomediastinum has been reported as high as 10%.[7]

In our review of the literature for cases of truly spontaneous PNR without predisposing acute or chronic conditions (such as acute respiratory illness or asthma), we found that the location of the air most commonly found in the cervicothoracic spine, with the cervical spine, alone as the next most common location The predilection for the cervical spine has an anatomical explanation; there is no fascial barrier between the posterior mediastinum and the cervical epidural space. As such, air can diffuse into the epidural space in this region if it already exists in the posterior mediastinum compartment.[12,16] The two cases of lumbosacral PNR were secondary to vacuum disc phenomenon at L5-S1.[5,8] There was only one reported case of holocord PNR, with air in the cervical, thoracic and lumbar spinal canal.[2] Our case report brings the total to two.

Treatment for spontaneous, asymptomatic PNR is most commonly geared towards treatment of the underlying condition. In the context of spontaneous

pneumomediastinum, this often means supportive care measures such as bed rest, oxygen supplementation, high-flow oxygen therapy, analgesia and in some cases, systemic corticosteroids.[12-14] The natural course of spontaneous PNR is spontaneous reabsorption through the bloodstream.[6] High-concentration oxygen therapy can speed up this process. Follow-up imaging is not indicated. Only one case has demonstrated recurrence.[12]

CONCLUSION

We present a case of spontaneous PNR diagnosed through CT in a patient with no known risk factors. There exist no guidelines for treatment of spontaneous PNR, which is often an incidental finding. As previously mentioned, all except two cases of spontaneous PNR in the literature were asymptomatic. As such, healthcare provider efforts should be directed at ruling out underlying dangerous complications of spontaneous pneumomediastinum and underlying causes of PNR, such as tension pneumomediastinum, pneumopericardium, pneumothorax, tracheal tear and Boerhaave syndrome. If no underlying cause is diagnosed and patient remains asymptomatic, the patient should be monitored under an outpatient status with observation. Serial imaging should be reserved for changes in exam or symptoms that could be related.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

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Conflicts of interest

There are no conflicts of interest.

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