CASE REPORT

Spontaneous Pneumorachis – A Case-Based Review

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Abstract: Pneumorachis is characterized by the presence of free air in the spinal canal. It is referred by different names in literature such as epidural emphysema, intraspinal air, intraspinal pneumoc(o)ele, spinal epidural and subarachnoid pneumatosis, spinal and epidural emphysema, aerorachia, pneumosaccus, air myelogram, etc. Pneumorachis can be broadly classified as traumatic, iatrogenic, or spontaneous. In this case-based review, we present a case of spontaneous pneumorachis secondary to asthma exacerbation. This is followed by a systematic review of all cases of spontaneous pneumorachis identified in PubMed. The aim of this review is to understand the pathophysiology, common causes and the management of spontaneous pneumorachis.

Keywords: pneumorachis, spontaneous pneumorachis, marijuana smoking, asthma exacerbation

Introduction

Pneumorachis is a clinical condition characterized by the presence of free air in the spinal canal.1 The presence of air in the cranial cavity, termed pneumocephalus, is a well-known clinical entity associated with skull fractures; on the other hand, pneumorachis is relatively rare. As early as 1918, air was utilized as a contrast medium for neuroimaging in a technique known as air myelogram. With the advent of less toxic contrast agents, the practice slowly died out. The first case of non-iatrogenic free air in the spinal canal (cervical) was reported in 1977 in a 20-year-old male who sustained multiple skull fractures during a motor vehicle accident. The clinical entity was initially described as “traumatic air myelogram.30” The current term pneumorachis was coined by Newbold et al in 1987.31 The condition is rare, and an accurate incidence rate is not available. As per our review, 48 case reports of spontaneous pneumorachis in adults have so far been published in the literature.

Based on the location of the air within the spinal cord, pneumorachis can be classified as intradural or extradural.90 Extradural pneumorachis is more common and is usually benign. Intradural pneumorachis, on the other hand, is indicative of major trauma. While neurological deficits are rarely associated with pneumorachis, it is more commonly associated with intradural pneumorachis even though there are a few case reports of extradural pneumorachis causing nerve root compression or cauda equina syndrome requiring decompression.90,91

Pneumorachis occurs most commonly due to either trauma or as a sequela of procedures involving the spinal canal.2 Pneumorachis is termed spontaneous when it occurs in the absence of trauma or iatrogenic insult.3 Spontaneous pneumorachis is extremely rare, and only a few cases have been reported in the literature.4 Many of them have been reported in conjunction with asthma exacerbation, but there are
no reports of marijuana-induced asthma exacerbation leading to spontaneous pneumorachis. The use of marijuana both for recreational and medicinal purposes is on the rise and the majority of current users smoke marijuana even when using it for medicinal purposes. Smoking marijuana is often associated with coughing paroxysms and is a potent trigger for asthma exacerbations making its use ideal for causing pneumorachis. With the increasing popularity of smoking marijuana and the widespread use of computerised tomography as a routine imaging modality, we believe clinicians are going to encounter spontaneous pneumorachis much more frequently in the future. In this article, we present a case of spontaneous pneumorachis and do a systematic review of spontaneous pneumorachis. Our goal is to consolidate the current understanding of this clinical entity so as to aid physicians to make the best informed clinical decisions.

Case Report
A 21-year-old male with a history of mild intermittent asthma presented with productive cough, sore throat, and neck pain for one day. The patient was smoking marijuana with his friends when he developed a sore throat, a severe bout of coughing with yellowish expectoration, post-tussive emesis, shortness of breath, and wheezing. He noticed a sharp, persistent pain starting in his neck and spreading to his anterior chest following his coughing paroxysm. He felt a sensation of swelling on his neck and could feel a crunching sensation when he touched it. He was not taking any home medications. He endorsed smoking a joint of marijuana daily and drinking 200 mL of hard liquor on weekends over the last 3–4 years.

On presentation, his blood pressure was 147/74 mm Hg, heart rate was 103 beats per minute, respiratory rate was 18 per minute, temperature was 98.7 °F, and he was saturating 94% on room air. His physical examination revealed extensive subcutaneous crepitus extending from his neck to the anterior chest wall. Auscultation of the lungs revealed diffuse expiratory wheezes with scattered rhonchi. Neurological examination did not show any motor, sensory or cranial nerve deficits.

Lab investigations are shown in Box 1. EKG revealed normal sinus rhythm. A chest X-ray showed pneumomediastinum along the left cardiac border and over the cardiomediasinal silhouette anteriorly with extensive subcutaneous emphysema in the neck’s soft tissues (Figures 1 and 2). A chest CT showed pneumopericardium and pneumomediastinum with air dissecting throughout the mediastinum, into the neck, down into the spinal canal, posterior paraspinal musculature as well as adjacent chest wall musculature (Figures 3 and 4). Air in the spinal canal extended from C1 down to the T9 level. There was anterior midline pneumothorax extending from side to side, outlining the anterior and medial margins of the lungs. A transthoracic echo (TTE) showed a small amount of pneumopericardium.

There was no clinical or radiological evidence of tension pneumothorax. Since the patient was not in any respiratory distress, he was initiated on the treatment of asthma exacerbation with intravenous methylprednisolone 80 mg every 8

| Name                  | Result            | Reference Range      |
|-----------------------|-------------------|----------------------|
| White blood count     | 13,600 cells/mm³  | 4000–11,000 cells/mm³|
| Hemoglobin            | 17.3 g/dL         | 12.5–17 g/dL         |
| Platelet count        | 163,000 cells/mm³ | 150–450 x 1000 cells/mm³ |
| Sodium                | 140 mEq/L         | 134–144 mEq/L        |
| Potassium             | 3.2 mEq/L         | 3.6–5.6 mEq/L        |
| Chloride              | 102 mEq/L         | 96–109 mEq/L         |
| Bicarbonate           | 24 mEq/L          | 20–32 mEq/L          |
| Blood urea nitrogen   | 15 mg/dL          | 5–26 mg/dL           |
| Creatinine            | 1.29 mg/dL        | 0.5–1.5 mg/dL        |
| Glucose               | 118 mg/dL         | 65–99 mg/dL          |
| Lactate               | 4.4 mmol/L        | 0.5–2 mmol/L         |
| Phosphorus            | 3.1 mg/dL         | 2.5–4.5 mg/dL        |
| C-reactive protein    | 68.28 mg/L        | 8–10 mg/L            |
hours, budesonide nebulizers twice daily, albuterol nebulizers as needed, and oxygen through a nasal cannula. Daily chest X-rays and continuous pulse oximetry were obtained to monitor the pneumorachis. Chest X-ray on day 2 of hospitalization showed progression of subcutaneous emphysema.

On day 3 of hospitalization, the patient reported significant improvement in his symptoms. On physical examination, his wheezing and subcutaneous crepitus had resolved. Chest X-ray also showed resolution of subcutaneous emphysema and mild improvement in pneumomediastinum. His oxygen supplementation was weaned off. He was prescribed an albuterol inhaler as needed and discharged home. He was counseled on smoking cessation and educated on avoiding activities that increase intrathoracic pressure.

**Systematic Review**

**Search Strategy**

We utilized PubMed’s search index for our systematic review. We used the search terms “pneumorachis” OR “spontaneous pneumorachis” under all fields to search for case reports and articles on spontaneous pneumorachis. All articles mentioned in PubMed, article bibliographies, and meeting abstracts until March 2021 were analyzed individually. Two hundred and two articles were identified, which were screened with inclusion criteria. Based on the selection criteria, 76 articles related to spontaneous pneumorachis were found, while 126 articles related to iatrogenic/traumatic pneumorachis were excluded. Of these, eight articles were written in a non-English language, two articles were inaccessible, and 18 articles on pediatric patients were excluded from the review. Forty-eight articles were included for final review and analysis, published over the past 40 years (1981-April 2021). The STrengthening, the Reporting of Observational studies in...
Epidemiology (STROBE) diagram, mentions in detail the steps of screening and scrutinizing the various articles during the selection process of the articles for this review (Figure 5).

Selection and Inclusion Criteria
The literature search was done by two independent and trained researchers. Abstracts of all the articles were screened to ensure appropriateness. Articles included were cases involving humans and published in the English language only. We excluded the articles that included pediatric cases (18 cases) and published in a language other than English (8 cases). Hence, 48 articles detailing 49 cases published from 1994 till March 2021 were included in the final interpretation and discussion (Figure 5).

Data Extraction
All selected articles were thoroughly studied: data were extracted and entered in a predefined table in a Word version 16.17 (Microsoft Corp, Redmond, WA) spreadsheet. Details on demographic characteristics, clinical features, comorbidities, management (antibiotics, surgery, oxygen supplementation), need for mechanical ventilation, and the outcome was extracted from each article.

Results
Patient Characteristics
There were 49 cases in 48 case reports (the case report by Martins et al had 2 cases). There were eight females, 40 males, and the sex of one patient was unknown. There were 12 cases between the ages of 18 to 20, 13 cases between the ages of 21 to 30, 4 cases between the ages of 31 to 40, 18 cases beyond the age of 40, and the age of 2 cases were unknown. There were no comorbidities in 25 patients. Asthma and diabetes mellitus were the most common comorbidities present in 7 patients each. There were no comorbidities in 24 patients, the comorbidities of 4 patients were unknown, and adenocarcinoma of the colon was present in 2 patients. Depression, anorexia nervosa, bullous lung disease, multiple sclerosis, Pancoast tumor, and diverticulitis were present in 1 patient each (Table 1).

The trigger was unknown in 7 patients, upper respiratory tract infection/bronchitis/common cold in 6 cases, asthma exacerbation in 5 cases, emphysematous pyelonephritis in 3 patients, cocaine use in 3 patients, vomiting caused by diabetic ketoacidosis in 3 patients, an acute bout of dry cough in 2 cases and vacuum intervertebral disc in 2 cases. Other causes include Valsalva maneuver, choking, ketamine sniffing, strenuous exercise, weightlifting, entero dural fistula, mucous plugging, pneumocystis

Figure 5 STROBE diagram depicting the selection process stepwise during the literature search for articles on nonspontaneous pneumorachis.
| S. NO | Author                          | Age/Sex | Comorbidities         | Acute Predisposition                                      | Leaks in Spaces other than the Spinal Cord | Treatment                        | Intubation | Outcome     |
|-------|--------------------------------|---------|-----------------------|-----------------------------------------------------------|-------------------------------------------|-----------------------------------|------------|-------------|
| 1     | Navriya et al<sup>(13)</sup>   | 65/m    | Diabetes mellitus     | Emphysema pyelonephritis                                  | Kidney                                    | DJ stenting                      | No         | Discharged  |
| 2     | Burns et al<sup>(62)</sup>      | 36/m    | None                  | Cocaine insufflation followed by Valsalva maneuver        | Pneumomediastinum, subcutaneous emphysema | Oxygen                           | No         | Discharged  |
| 3     | Bally et al<sup>(64)</sup>      | Mid 20s/M | None                  | Choking                                                   | Subcutaneous emphysema, pneumomediastinum | Supportive                       | No         | Discharged  |
| 4     | Llewelyn et al<sup>(7)</sup>    | 21/m    | None                  | Upper respiratory tract infection                         | Subcutaneous emphysema, pneumomediastinum | Nasal cannula                    | No         | Discharged  |
| 5     | Williams et al<sup>(8)</sup>    | 22/m    | None                  | Ketamine snifing                                          | Subcutaneous emphysema, pneumomediastinum | Nonrebreather                     | No         | Discharged  |
| 6     | Niemann et al<sup>(10)</sup>    | 19/m    | None                  | Valsalva maneuver                                         | Pneumothorax, pneumomediastinum, subcutaneous emphysema | Supportive                       | No         | Discharged  |
| 7     | Ramses Bedolla-Pulido et al<sup>(82)</sup> | 18/m | Asthma                | Acute exacerbation of asthma                              | Subcutaneous emphysema, pneumomediastinum | Steroids, oxygen                  | No         | Discharged  |
| 8     | Liu et al<sup>(69)</sup>        | 18/m    | None                  | Common cold                                               | Subcutaneous emphysema, pneumomediastinum, pneumothorax | Conservatively                   | No         | Discharged  |

(Continued)
Table 1 (Continued).

| S. NO | Author | Age/Sex | Comorbidities | Acute Predisposition | Leaks in Spaces other than the Spinal Cord | Treatment | Intubation | Outcome |
|-------|--------|---------|---------------|----------------------|------------------------------------------|-----------|------------|---------|
| 9     | Radhika Nair et al (71) | Unknown/ f | Asthma | Asthma exacerbation | Pneumomediastinum, pneumoperitoneum, pneumothorax and subcutaneous emphysema | Symptomatic treatment | No | Recovered |
| 10    | Vallyakath et al (87) | 21/f | None | Bronchitis | Pneumomediastinum, subcutaneous emphysema | Oxygen, intravenous antibiotics | No | Discharged |
| 11    | Heckman et al (89) | 20/m | None | Bronchitis | Subcutaneous emphysema, prevertebral, parapharyngeal, carotid spaces, pneumomediastinum | Azithromycin, ceftriaxone, analgesia | No | Discharged |
| 12    | Ramasamy et al (66) | 20/m | None | Strenuous exercise | Pneumomediastinum, subcutaneous emphysema | Nasal cannula oxygen, analgesics, antibiotics | No | Discharged |
| 13    | Gomez et al | 60/m | Diabetes, ventral hernia | Emphysematous pyelonephritis | Kidney | Vasopressors, Broad spectrum antibiotics | No | Discharged |
| 14    | Germino et al (14) | 58/m | None | Weightlifting | Craniocervical hyperpneumatiation, pneumomediastinum, subcutaneous emphysema | Conservative management | No | Discharged |
| 15    | Temrel et al (63) | 28/m | None | Cocaine sniffing | Emphysema, pneumomediastinum | Conservative management | No | Discharged |
| 16    | Kanu et al (70) | 19/m | None | Crack cocaine use | Subcutaneous emphysema, pneumomediastinum | Conservative | No | Discharged |
|   | First Name et al  | Age | Gender | Diagnosis | Presentation | Treatment | Discharge Status | Notes |
|---|-------------------|-----|--------|-----------|--------------|-----------|-----------------|-------|
| 17 | Schomig et al     | 67/m|        | Diverticulitis s/p clipping | Enterodural fistula caused by ingestion of dental prosthesis | Pneumocephalus, Lumbar puncture, vancomycin, ceftriaxone, Repair of entero dural fistula | No | Discharged |
| 18 | Mahajan et al     | 18/f|        | Asthma    | Mucous plugging of the right main bronchus | Pneumomediastinum, subcutaneous emphysema, right pneumothorax | Yes | Discharged |
| 19 | Patel et al       | 20/m|        | None      | Upper respiratory tract infection | Subcutaneous emphysema, pneumomediastinum, pneumothorax | No | Discharged |
| 20 | Eroglu et al      | 44/F|        | None      | Unknown | None | Anti inflammatory | No | Discharged |
| 21 | Saleem et al      | 28/m|        | None      | Pneumocystis pneumonia with new onset AIDS | Subcutaneous emphysema, pneumomediastinum | No | Discharged |
| 22 | Kirkham et al     | 21/m| Asthma | Asthma exacerbation | Subcutaneous emphysema, pneumopericardium | Observation | No | Discharged |
| 23 | Ozkan et al       | 31/m| Bullous lung disease | Spontaneous rupture of bulla | Pneumothorax, subcutaneous emphysema | Chest tube, bulla removal surgery | No | Discharged |
| 24 | Kazimirko et al   | 53/m|        | Right superior sulcus lung tumor with adjacent vertebral invasion with pathologic fracture of T1. | Bronchopleurodurosubarachnoid fistula | Pneumocephalus | C- collar and antibiotics | No | Discharged but died 3 months later from cancer progression |
| 25 | Krishnan et al    | 30/M| Unknown| Unknown | Unknown | Subcutaneous emphysema, pneumomediastinum | Subcutaneous incisions, symptomatic medications | No | Discharged |

(Continued)
Table 1 (Continued).

| S. NO | Author | Age/Sex | Comorbidities | Acute Predisposition | Leaks in Spaces other than the Spinal Cord | Treatment | Intubation | Outcome |
|-------|--------|---------|---------------|----------------------|-------------------------------------------|-----------|------------|---------|
| 26    | Jensen et al\(^{(9)}\) | Unknown | Anorexia nervosa | Bouts of self induced vomiting | Unknown | Conservative medical approach and treatment of anorexia nervosa | Unknown | Unknown |
| 27    | Moayedi et al\(^{(28)}\) | 76/f | Multiple sclerosis, diabetes mellitus | Sacral decubitus ulcer | Pneumocephalus, subcutaneous emphysema | IV antibiotics - cefepime, metronidazole; levetiracetam | Yes | Passed away 6 days later from sepsis after being made comfort measures |
| 28    | Martins et al\(^{(5)}\) | 20/m, 22/m | None | Acute bout of dry cough. | Pneumomediastinum, subcutaneous emphysema | Bedrest, oxygen, and opioids for cough control | No | Discharged |
| 29    | Liao et al\(^{(68)}\) | 19/m | None | Strenuous exercise | Subcutaneous emphysema, pneumomediastinum | Conservative treatment | No | Discharged |
| 30    | Eisa et al\(^{(84)}\) | 18/m | None | Upper respiratory tract infection | Subcutaneous emphysema, pneumothorax, pneumomediastinum | Oxygen, conservative management | No | Discharged |
| 31    | Amara et al\(^{(81)}\) | 21/m | None | Fever and vomiting in the setting of meningitis | Pneumomediastinum, subcutaneous emphysema | Ceftriaxone, lumbar puncture, bronchoscopy, laryngoscopy | Yes | Died from sepsis |
| 32    | Iacoangeli et al\(^{(21)}\) | 52/F | Adenocarcinoma of colon | Transectional cerebrospinal fluid leak | Pneumocephalus | Lumbosacral laminectomy and duraplasty; tumor removal and omental covering of the pelvis | No | Discharged home |
| 33    | Tafreshi et al\(^{(22)}\) | 66/m | Adenocarcinoma of colon T3 with history of abdominoperineal resection and recurrence with presacral mass | Enterocutaneous fistula from left natal cleft | None | Pelvic exenteration | No | Discharged |
| 34    | Lee et al\(^{(32)}\) | 76/m | Unknown | Vacuum intervertebral disc | None | Left partial hemilaminectomy | No | Discharged |
|   | Author(s)       | Age/m | History                                                                 | Diagnosis                                                                 | Treatment                                                                 | Outcome | Notes |
|---|-----------------|-------|-------------------------------------------------------------------------|---------------------------------------------------------------------------|---------------------------------------------------------------------------|---------|-------|
| 35 | Kumaran et al   | 51/m  | None                                                                    | Grade I anterolisthesis of L5 over S1                                     | None                                                                       | Spondylolisthesis correction surgery | No     | Unknown |
| 36 | Amit et al      | 60/m  | Unknown                                                                 | Perforated sigmoid colon                                                  | Pneumocephalus                                                            | Hartmann's procedure; broad-spectrum intrathecal antibiotics              | No     | Resolution of pneumocranium and pneumorachis |
| 37 | Al-Mufarrej et al | 20/m  | None                                                                    | Unknown                                                                   | Pneumoperitoneum, pneumomediastinum, subcutaneous emphysema               | Self-limited                                                                 | No     | Resolved |
| 38 | Song et al      | 18/m  | None                                                                    | Unknown                                                                   | Pneumomediastinum, subcutaneous emphysema                                | Dextromethorphan 45 mg daily                                               | No     | Discharged |
| 39 | Kim et al       | 64/m  | Unknown                                                                 | Vacuum disc L5-S1.                                                        | None                                                                       | Conservative                                                                | No     | Resolved |
| 40 | Drolet et al    | 18/m  | Type 1 diabetes mellitus                                               | Diabetic ketoacidosis                                                     | Pneumomediastinum, subcutaneous emphysema                                | Observation                                                                 | No     | Discharged |
| 41 | Eesa et al      | 18/m  | Asthma                                                                  | Asthma exacerbation                                                       | Pneumomediastinum, subcutaneous emphysema                                | Unknown                                                                     | Unknown | Unknown |
| 42 | Oertel et al    | 19/m  | Asthma, diabetes mellitus                                              | Diabetic ketoacidosis                                                     | Pneumomediastinum, pneumoperitoneum, subcutaneous emphysema, pneumocephalus | Otolaryngological exploration                                              | No     | Discharged |
| 43 | Eesa et al      | 18/m  | Asthma                                                                  | Asthma exacerbation                                                       | Pneumomediastinum, subcutaneous emphysema, retropharyngeal space         | Oxygen therapy, bronchodilators, and antibiotics                          | No     | Discharged |
| 44 | Yamamoto et al  | 63/m  | Depression, Diabetes mellitus                                          | Emphysematous pyelonephritis                                              | Right iliopsoas muscle, kidney                                            | Intravenous vancomycin 1G BID for 1 week and meropenem 1 g TID for 4 weeks | No     | Discharged |
Other Sites of Involvement

The spinal cord was the only site of air accumulation in 5 cases. Concomitant pneumomediastinum was reported in 30 cases, subcutaneous emphysema was reported in 33 cases, pneumothorax in 8 cases, pneumocephalus in 6 cases, kidney in 3 cases, deep spaces of the neck in 2 cases, and right iliopsoas muscle in 1 case (Figure 6).

Management Strategies

The management was supportive in most of the cases, except for a few cases. The supportive treatment included a combination of oxygen supplementation, analgesics, and cough suppressants. Only three patients required intubation and mechanical ventilation. It was unknown whether an intubation was required in 2 patients. Patients with asthma exacerbation were treated with steroids and nebulizers. Naviya et al report that their patient with emphysematous pyelonephritis was managed with double J stenting. The patient in Gomez et al’s case report required vasopressor and broad-spectrum antibiotics for emphysematous pyelonephritis, as did the patient reported by Yamamoto et al. Schomig et al had to do a lumbar puncture followed by repair of enterodural fistula for the treatment of pneumorachis caused by enterodural fistula. Mahajan et al’s patient required bronchoscopic removal of mucous plugging, while another patient of Saleem et al required bronchoscopy to diagnose pneumocystis pneumonia. Fonseca et al also performed a bronchoscopy, although the reason for the procedure is unclear. A total of 3 patients required chest tubes for the concomitant pneumothorax, following which pneumorachis resolved. Krishnan et al performed subcutaneous incisions to help relieve the trapped mediastinal air, subsequently leading to the resolution of pneumomediastinum. Lumbosacral laminectomy with duraplasty was employed by Iacoangeli et al for management of pneumorachis secondary to transcranial cerebrospinal fluid leak caused by adenocarcinoma of the colon. C7 total laminectomy (Song et al), otolaryngological exploration (Oertel et al.), Hartman’s procedure (Amit et al), spondylolisthesis correction surgery (Kumaran et al), left partial hemilaminectomy (Lee et al), and pelvic exenteration (Tafreshi et al) were some of the other

### Table 1 (Continued)

| S. NO | Author          | Age/Sex | Acute Predisposition | Leaks in Spaces other than the Spinal Cord | Treatment | Intubation | Outcome |
|-------|-----------------|---------|----------------------|-------------------------------------------|-----------|------------|---------|
| 45    | Pangtey et al   | 35/m    | Unknown              | Pneumothorax, pneumomediastinum, subcutaneous emphysema | Chest tube | No         | Discharged |
| 46    | Ripley et al    | 23/m    | Type 1 diabetes mellitus | Pneumomediastinum, subcutaneous emphysema | Standard treatment for DKA and supplemental oxygen | No         | Discharged |
| 47    | Fonseca et al   | 20/F    | Unknown              | Subcutaneous emphysema, pneumomediastinum | Bronchoscopy, conservative management | No         | Discharged |
| 48    | Song et al      | 72/F    | None                 | None                                      | C7 total laminectomy | No         | Discharged |
procedures employed by authors for management of the inciting factor for the pneumorachis.22-27

Outcome
The outcome was unknown in 2 cases, one patient died of sepsis, and one patient died three months after being discharged due to cancer progression.28,29 The outcome was positive in 45 out of the 49 cases, with the patients having either resolution of the pneumorachis or being discharged home.

Discussion
Pneumorachis can be classified based on etiology as traumatic, iatrogenic, or spontaneous.32 A 2010 systematic review of pneumorachis identified trauma to the respiratory tract as the causative factor in 52% of cases. Traumatic injury to the skull (especially skull base and sinus fractures) was the second most common cause.33 Iatrogenic causes include lumbar puncture, epidural analgesia, spinal surgeries, and abdominal surgeries.34,35 Epidural abscesses due to anaerobic pathogens can cause the air in the epidural space, especially in patients who have diabetes or Crohn’s disease.36 Intra-abdominal infections (especially emphysematous pyelonephritis) with anaerobic/facultative anaerobic bacteria such as Citrobacter koseri can also generate pneumoperitoneum with subsequent invasion of air into the spinal canal.15 Spontaneous pneumorachis can occur as a result of respiratory conditions and complications that incite barotrauma.37,38 Asthma is a common, chronic respiratory disease affecting 8% of adults in the U.S.A, and some of the rare but potentially life-threatening complications of an asthma exacerbation include pneumothorax and pneumomediastinum pneumopericardium, pneumorachis, tracheoesophageal fistula, and anoxic brain injury.39-41 Asthma was identified as the most common trigger for spontaneous pneumorachis in our review.42 Rarely, spontaneous extradural pneumorachis can develop as a complication of severe degenerative disc disease. Intervertebral disc vacuum phenomenon is a finding present in 1–2% of spinal radiographs and has an estimated prevalence of 20% in elderly population.93 Enlargement of this clefts in the intervertebral disc can cause a negative pressure gradient pulling gases into the space. The disk fragment containing gas could herniate into the spinal canal or the gas could get expelled into the epidural space through a weak spot in the annulus fibrosis, occasionally causing clinical presentation akin to a herniated disc.

Clinical Anatomy
Due to continuity between the cranial cavity and spinal canal, it is expected that cranial injuries communicating with the external atmosphere could cause pneumocephalus and consequent pneumorachis. However, as mentioned above, respiratory tract injuries account for most cases of pneumorachis. Defining the relations between the respiratory tract, mediastinum, and vertebral column is key to understanding the mechanism of spontaneous pneumorachis.
The mediastinum is the space within the thoracic cavity bordered superiorly by the thoracic inlet, inferiorly by the diaphragm, and excludes the lungs and pleural cavities. Within the mediastinum, the air is usually seen in the trachea, bronchi, bronchioles, alveoli, and esophagus, all of which are a potential source for pneumomediastinum and pneumorachis. Being the thinnest portion within the respiratory tree, alveolar rupture under conditions of the high trans alveolar pressure gradient is possibly the most common source of free air. If the rupture involves the visceral pleura, the patient will develop a pneumothorax. The air could also escape into the pulmonary interstitium from where it tracks along the bronchovascular bundle to reach the hilum from where it escapes into the mediastinum (Macklin effect). From here, air can dissect into the posterior mediastinum and separates the parietal pleura from the vertebral column gaining access into the epidural space via the intervertebral foramen. In the absence of any fascial barriers between the posterior mediastinum and epidural space, air can freely communicate between these compartments.

There is no direct communication between the mediastinum and the pericardial cavity. Pneumopericardium typically occurs due to macro perforation of the pericardium and communication with the respiratory or GI tract in the setting of trauma, pneumothorax, volutrauma due to mechanical ventilation, fistulas in the setting of malignancies, etc. Laparoscopic surgeries could occasionally cause pneumopericardium. This is attributed to the presence of congenital patent pathways between pleura, pericardium, and peritoneal cavities. Our patient did not have a pneumothorax or pneumoperitoneum; hence, it is likely that the large volume of air was able to push into the pericardial space near the pulmonary vein Ostia where the parietal pericardium reflects onto the visceral pericardium creating the weakest spot.

**Pathophysiology**

Literature review revealed one case of pneumorachis associated with cannabis use which was attributed to using a homemade bong with flow restricted intake, which forced the patient to inhale forcibly against high resistance, in turn creating a high negative intrathoracic pressure inducing barotrauma. This phenomenon is called Müller’s maneuver and can be considered the reverse of the Valsalva maneuver. A similar mechanism could explain pneumorachis and pneumomediastinum in our patient too. In this case, instead of an external resistance to airflow, bronchospasm induced by the cannabis smoke would have forced the patient to inhale against a tight airway generating high negative intrapleural pressures, while coughing against the resistance would have generated high positive intra-alveolar pressures, thereby increasing trans alveolar pressure gradient both during inspiration and expiration which resulted in the barotrauma. The chances of a pneumomediastinum causing pneumorachis increase with the amount of leaked air and acuity of onset.

**Clinical Presentation**

Spontaneous pneumorachis in the absence of intraspinal causes like epidural abscesses are associated with pneumomediastinum or subcutaneous emphysema. Asthma exacerbation is the most common triggering factor estimated to be present in 20–30% of all cases of spontaneous pneumomediastinum in children and adolescents. Other predisposing conditions include respiratory tract infections, activities inducing Valsalva maneuver, activities causing Müller’s maneuver, severe vomiting, esophageal rupture, foreign body aspiration, vaping and inhalational drug use. The presence of these predisposing factors along with signs and symptoms should raise the suspicion for spontaneous pneumomediastinum (SPM) and possible pneumorachis. SPM is more common among children and adolescents compared to older age groups. Males are more likely to be affected, and so does a tall, lean body habitus.

The most common presenting complaint is retrosternal, pleuritic chest pain radiating to the neck or shoulders of acute onset after an inciting event. Even when no triggering factor is present, SPM should be considered in the differential diagnosis of any young adult presenting with acute onset chest pain. Other common symptoms include subcutaneous emphysema with crepitus, especially in the neck, neck pain, dyspnea, cough, odynophagia, and dysphagia. Esophageal perforation (Boerhaave syndrome) could also present with a similar presentation and should be considered, especially in patients who have severe vomiting; but they typically present with signs and symptoms of shock.

Subcutaneous emphysema detected over the neck or precordium is an extremely sensitive and specific sign of SPM. Hamman sign, a precordial crunching sound
synchronous with systole, especially in the left lateral decubitus position, may be present in up to 18% of patients. Findings such as hypotension, severe respiratory distress, unilaterally diminished breath sounds, and distended neck veins should raise the suspicion for more serious etiologies such as tension pneumomediastinum, pneumothorax, or esophageal perforation.

**Neurological Signs and Symptoms**
Pneumorachis is asymptomatic in most cases but can cause neurologic symptoms, including spinal cord compression in about 10% of patients. As pneumorachis by itself rarely causes spinal cord compression, we should search for alternative explanations such as epidural hematomas, abscesses, herniation, etc, if neurological symptoms are present.

**Diagnosis**
Pneumorachis is almost always found in combination with air in other cavities such as pneumothorax, pneumomediastinum, or subcutaneous emphysema. Ultrasound is gaining popularity as the imaging of choice in the emergency department for the fast and accurate detection of pneumomediastinum, but it is not useful for detecting pneumorachis. Pneumorachis can be diagnosed on spinal X-ray. A CT scan is the diagnostic test of choice. An MRI is not required unless signs of cord compression are present and other etiologies such as contusion, hematoma, herniation, or compressing bone fragments need to be ruled out. The characteristic radiological features of SPM and pneumorachis, as demonstrated in our patient, are as below:

**Treatment**
There are no guidelines for the management of pneumorachis. Serial neurological examinations are recommended, while serial imaging for pneumorachis is not required unless there is a concern for cord compression. The development of neurological symptoms warrants surgical decompression; otherwise, the management is conservative.

**Clinical Outcomes**
Clinical outcomes are related to the etiology of pneumorachis. The one patient who died during the acute course of illness in our review had sepsis. Radiological resolution would occur in most cases with conservative management. Treatment of the cause will curtail ongoing air leaks allowing the intra-spinal air to get reabsorbed within hours to days. Residual neurological deficits are rare.

**Conclusion**
Clinicians should be aware of the possibility of pneumorachis in the setting of acute asthma exacerbation. Although asymptomatic in most cases, it can rarely lead to serious complications such as spinal cord compression. Addressing the underlying etiology is more important than managing the pneumorachis per se. Treatment is conservative except in symptomatic cases where intervention is required.

**Disclosure**
The authors report no conflicts of interest in this work.

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