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Case Report

Clearing the air around spontaneous pneumorrhachis

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ABSTRACT

We present a case of spontaneous pneumorrhachis associated with minimal pneumomediastinum from a tertiary care cancer hospital in Mumbai. A 16-year-old boy who was a case of Hodgkin lymphoma undergoing chemotherapy presented to the physician with complaints of cough associated with white-colored sputum and chest pain. Computed tomography of the chest done to rule out infection revealed pneumorrhachis, that is, air lurking in the spinal canal. Radiological and laboratory investigations were done to rule out crucial and life-threatening differentials. The patient was kept on observation, and finally, we figuratively cleared the air around the finding of "air in the spinal cord." This is the first reported case in the literature of pneumorrachis in a patient with Hodgkin's lymphoma.

Keywords: Pneumomediastinum, Pneumorrhachis, Spontaneous, Spine, Computed tomography, Observation

INTRODUCTION

The Association of pneumorrachis with cancer is almost unknown, with only one case being reported previously in a patient with metastatic esophageal carcinoma. Even then, this patient had gas formation within an epidural metastatic deposit, as a likely explanation of the pneumorrachis. Our patient did not have any such deposits to the spine, and as such no obvious cause of the pneumorrachis could be determined.

CASE REPORT

A 16-year-old boy with Hodgkin lymphoma Stage IIA (early-stage unfavorable disease) was planned for chemotherapy with 6 cycles of adriamycin, etoposide, vinblastine, and dacarbazine. On day 28 of 1st cycle of chemotherapy, he presented with complaints of cough associated with white-colored sputum and chest pain for 2 days. He also gave a history of nausea and retching. He gave no history of trauma or invasive procedure. There was no history of cigarette smoking or the use of recreational inhalants.

On physical examination, his respiratory rate was 22/min, heart rate of 145/min, blood pressure of 110/62, oxygen saturation of 100% on room air, and 98°F temperature. He appeared uncomfortable and coughed excessively but no signs of laborious breathing were observed. Non-contrast computed tomography scan of chest computed tomography (CT) performed for the assessment of infective focus in the chest demonstrated a patch of consolidation in the superior lingular segment of the left upper lobe with few nodules in bilateral lower lobes. These changes were radiologically deemed to be infective in etiology. Moreover, there were substantial pneumorrachis throughout the cervical, dorsal, and lumbar spinal canal with a mild component of pneumomediastinum demonstrated in Figures 1-3. However, no evidence of pneumothorax was seen. Pneumocephalus was ruled out by taking a plain brain CT. Considering the history of nausea, retching, and chest pain, oral contrast was given to the patient to rule out occult esophageal perforation. There was no extravasation of contrast from the esophagus. No apparent cause of pneumorrhachis was found on the CT scan.

Diagnosis of spontaneous pneumorrhachis was made and since the patient was hemodynamically stable, he was kept under observation. He was treated with a course of oral broad-spectrum antibiotics and cough suppressants for respiratory tract infection. At the time of discharge, he was counseled to keep a close watch for symptoms such as sensory or motor weakness and to immediately report them to the treating physician. A repeat scan for the evaluation of pneumorrhachis was scheduled after 14 days or earlier if there were any neurological symptoms.

The patient did not develop any neurological symptoms, and on the follow-up scan after 2 weeks, complete resolution of

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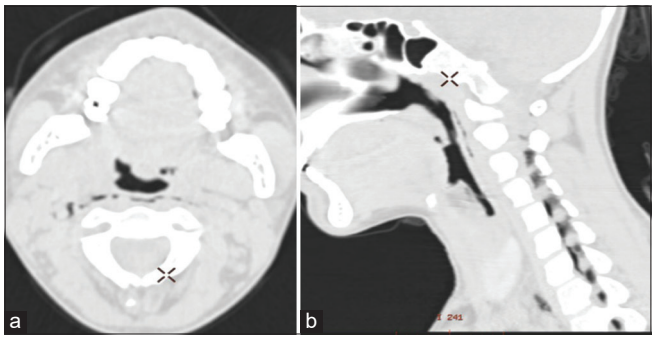

Figure 1: (a) Axial computed tomography (CT) section at the level of oropharynx showing air in the pre-vertebral space. (b) Sagittal reformatted CT section showing air in the cervical segment of spinal canal.

Figure 2: (a) Axial computed tomography (CT) section at the level of lung apices showing air tracking through spinal nerve roots to the spinal canal. (b) Axial CT section showing pneumomediastinum and air lining vascular structures.

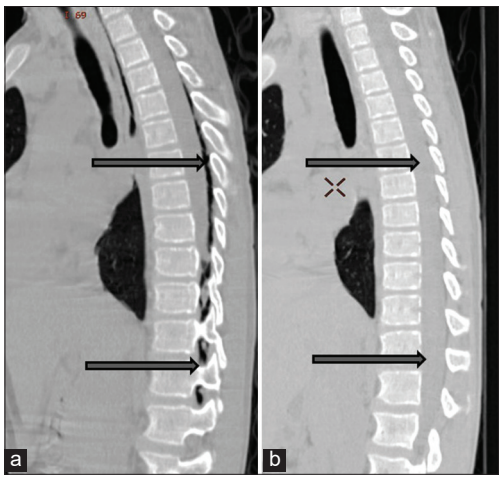

Figure 3: (a) Sagittal reformatted sections showing air in the spinal canal in the thoracolumbar level. (b) Follow-up image after 20 days shows complete reabsorption of intraspinal air.

the pneumorrhachis was seen. A comparison of previous CT and follow-up CT is shown in Figure 3b. (All the images taken from the CT scan were obtained using a 16-slice Siemens machine with 100 mAs and 120 kv).

Table 1: Summary table.

Etiology	Iatrogenic – epidural analgesia and lumbar puncture Traumatic – road traffic accidents and fall from height Non-traumatic – degenerative, inflammatory, infectious, and other
Incidence	Rare, approximately 5–10% (proportion and clinical relevance of intraspinal air in patients)
Gender ratio	No formal data are available
Age predilection	No formal data but as per previous case reports, we studied that most of the cases are<20 years
Risk factors	Predisposing lung disease (asthma and pulmonary bullae), maneuvers that raise intrathoracic pressure (Valsalva, vomiting, retching, coughing, and illicit drug inhalation)
Treatment	Management depends on the etiology and symptoms. Most cases are treated with conservative measures. Theoretical benefit of introducing high-flow oxygen. Surgical intervention if required. Control of cough, emesis, or asthma if present
Prognosis	Benign and self-limiting course with full recovery with spontaneous reabsorption of air
Findings on imaging	Air in various compartments and cavities – pleura, mediastinum, and spinal canal. In cases of underlying contributing pathology fractures (trauma) and bony destruction (infection and neoplastic) may be present

Table 2: Table of differentials.

Radiographs	Features associated with PR such as pneumomediastinum, pneumothorax, and subcutaneous emphysema are usually picked up incidentally on radiographs. PR itself cannot be diagnosed on radiographs
Ultrasound (US)	Suboptimal imaging in diagnosing PR or its underlying cause
Computed tomography (CT)	Can positively demonstrate the presence and extent of free gas in the spinal canal and extraspinal compartments. In addition, CT can help identify any underlying etiology
Magnetic resonance imaging (MRI)	Suboptimal imaging for diagnosing PR itself, but can be of value in diagnosing an underlying cause Mainly non-traumatic causes such as degenerative, inflammatory, and infectious can be identified
Scintigraphy	Suboptimal imaging for this condition
Positron emission tomography (PET).	Suboptimal imaging for this condition and rare cases of intraspinal metastatic deposit can be confirmed.

DISCUSSION

Pneumorrhachis is a rare phenomenon and is characterized by the presence of air in the spinal canal.[1] This finding has

also been described as epidural pneumatosis, intraspinal pneumocele, spinal emphysema, pneumosaccus, and aerorhacia in the literature.[2] Pneumorrhachis can be due to iatrogenic, traumatic, and non-traumatic causes.[3]

The underlying pathophysiology is postulated to be increased intra-alveolar pressure (for example, excessive prolonged coughing, forceful vomiting, asthma, etc.[4]), leading to rupture of alveoli, due to which air travels along the pulmonary perivascular interstitium and dissects along the fascial planes.[5] As the pressure in the mediastinum is lower than the lung periphery, the air dissects toward the hilum of the lung and leads to pneumomediastinum. This sequence of events is described as the Macklin effect.[6] Free entry of posterior mediastinal air into the epidural space through neural foramina can be explained by the lack of true fascial protection to the epidural space.[5] Because of the low resistance from the loose connective tissues as compared with the rich vascular network present anteriorly, the dissected air preferably collects in the posterior epidural space.[7]

Free air in the spinal canal can be localized as internal or external pneumorrhachis. Internal is intradural pneumorrhachis which means the air in the subdural or subarachnoid space and external means air in the extradural space.[6] Intradural air is generally seen after trauma and represents severe injury.[3]

Tracing the source of air in the spinal cord forms the basis of diagnostic workup once pneumorrhachis is spotted. This air can be due to surgical or invasive procedure, perforation of trachea or bronchus, rupture of the esophagus, bronchial asthma, post-intermittent positive pressure ventilation, extension of pneumoperitoneum, as a result of prolonged coughing, or can be spontaneous.[8]

Concurrent pneumorrhachis with spontaneous pneumomediastinum and without pneumothorax is an extremely rare phenomenon.[9] Rather pneumorrhachis is present in up to 10% of cases of spontaneous pneumomediastinum.[6] Pneumorrhachis is mostly asymptomatic.[6] Rarely, it can give rise to symptoms of cord compression, neurological deficits, or meningitis.[6]

Since in majority of the cases, pneumorrachis is an asymptomatic phenomenon, its diagnosis is more likely to be on imaging, instead of on clinical findings.[8] A CT scan is the best tool for diagnosis since it can depict a minute amount of air and can rule out other grim differential diagnoses leading to pneumorrhachis.[1] However, even for CT, differentiating between intra- and extra-dural air is difficult.[7]

Due to the rarity of the phenomenon, no clear guidelines exist for management. Most cases are treated with conservative measures as air is spontaneously reabsorbed by the blood.[1] There is a theoretical benefit of introducing high-flow oxygen as it can increase the oxygen content of the entrapped gas, leading to more rapid absorption.[10,11] Surgical intervention is needed in cases associated with persistent spinal fluid leak or entrapment of air in the spinal canal due to the formation

Table 3: Review of the literature (year-wise).

Author and year	Age and sex of patient	Symptoms	Predisposing factor if any	Imaging findings	Management	Outcome
Drevelengas, 1994	16 years, female	Cough with mild parasternal pain and dyspnea.	Cough	Pneumomediastinum emphysema and pneumorrhachis	Conservative	Improved completely
Oerte1, 2004	19 years, male	Cough, fever, nausea, and vomiting for 3 days	Diabetes mellitus, asthma	Pneumomediastinum external pneumocephalus and pneumorrhachis of the entire spinal canal	Conservative	Improved completely
Eesa, 2006	18 years, male	Swelling over the neck and upper chest	Mild attack of asthma previously around 24 previously	Pneumomediastinum, emphysema, and pneumorrhachis	Conservative	Improved completely
Drolet, 2007	18 years, male	Sudden onset of retrosternal pain and dyspnea	Type 1 diabetes mellitus, with a 3-day history of severe vomiting	Pneumomediastinum, emphysema, and pneumorrhachis	Conservative	Improved completely
Song, 2008	18 years, male	Dyspnea cough	Cough	Pneumomediastinum and pneumorrhachis	Conservative	Improved completely
Song, 2008	72 years, male	Progressive weakness of the lower extremities		Pneumorrhachis	C7 total laminectomy	Improved completely
Mufarrej, 2009	20 years, male	10 h history of – acute-onset neck and back pain		Pneumomediastinum subcutaneous emphysema and pneumoperitoneum	Conservative	Improved completely

Table 4: Review of literature (year wise).

Author and year	Age and sex of patient	Symptoms	Predisposing factor if any	Imaging findings	Management	Outcome
Kakel, 2010	16 years, male	Influenza-like symptoms a few days earlier, collapsed in the middle of a game of hockey but recovered within 1 min		Pneumomediastinum, emphysema, and pneumorrhachis	Conservative	Improved completely
KaraoGlan, 2010	8 years, female	Respiratory distress, non-productive violent coughing, neck swelling, and high fever	Violent coughing asthma	Pneumomediastinum, emphysema, and pneumorrhachis	Conservative	Improved completely
Kumaran SP, 2011	51 years, male	Chronic low back pain		MRI – Anterolisthesis of L5 over S1 vertebra CT – vacuum phenomenon in all the lumbar intervertebral disks, linearly placed air pockets within the spinal canal anterior to the thecal sac from L5 to S1 levels	Conservative	Improved completely
Sandhya, 2011	10 years, female	Known case of dermatomyositis dry cough without dyspnea and emphysematous changes		Pneumomediastinum, subcutaneous emphysema, basal opacities suggestive of interstitial lung disease (ILD).	Conservative	Improved completely

Table 5: Review of literature (year wise).

Author and year	Age and sex of patient	Symptoms	Predisposing factor if any	Imaging findings	Management	Outcome
Sulenan, 2012	46 years, male	Severe debilitating pain down the right leg after trivial trauma	Degenerative disease. Vacuum phenomenon	Air in the right neural foramen and spinal canal	CT-guided aspiration of air	Improved completely
Migeot, 2012	19 years, male	Chest pain 40 days followed by diffuse lower limbs paresthesias	Following episode of violent coughing and shouting	Pneumomediastinum and pneumorrhachis	Conservative	Left foot paresthesia
Carolan, 2013	16 years, male	Fever, cough, and vomiting accompanied by complaints of severe chest, neck, and back pain	Asthma	Pneumomediastinum and pneumorrhachis	Conservative	Improved completely
Ehmann, 2015	28 years, male	1 day of pleuritic chest pain and 2 days of non-productive cough and odynophagia		Pneumomediastinum and pneumorrhachis	Conservative	Improved completely
Myung-Jae Yoo, 2015	48 years, male	Fever dyspnea	Anterior/posterior fixation/ fusion on C5–C7 with infective spondylodiscitis as sequelae	Cord atrophy on lower cervical and upper thoracic spine and infective spondylitis on L4 and L5 bodies and pneumorrhachis	Conservative	Improved completely

Table 6: Review of literature (year wise).

Author and year	Age and sex of patient	Symptoms	Predisposing factor if any	Imaging findings	Management	Outcome
Ehmann, 2015	20 years, male	Neck and chest pain preceded by 3 days of cough and wheezing	Asthma, marijuana	Pneumomediastinum and pneumorrhachis	Conservative	Improved completely
Sadarangani, 2015	17 years, male	Neck and chest pain	Weight lifting session	Pneumomediastinum emphysema and pneumorrhachis	Conservative	Improved completely
Erniralioglu, 2015	12 years, male	Cough, nasal congestion and rhinorrhea		Pneumomediastinum emphysema and pneumorrhachis	Conservative	Improved completely
Jay Kirkham *et al*., 2016	21 years, male	Chest pain and shortness of breath	Asthma, wheezing, and coughing episode	Subcutaneous emphysema and pneumopericardium and pneumorrhachis	Conservative	Improved completely
Umiteroglu, 2016	46 years, female	Waist and leg pain		L5-S1 spinal extradural air	Conservative	Improved completely
Fonseca *et al*., 2016	20 years, female	2 days history of dyspnea and chest pain	Coughing and vomiting	Pneumomediastinum and pneumorrhachis	Conservative	Improved completely
Sethi, 2018	16 years, male	Slowly progressive discomfort in the neck and throat in association with concern that his voice had altered	Cannabis, shortly after inhalation, he described becoming intensely nauseated and that he had forcefully vomited several times	Pneumomediastinum and pneumorrhachis and extensive subcutaneous emphysema	Conservative	Improved completely

Table 7: Review of literature (year wise).

Author and year	Age and sex of patient	Symptoms	Predisposing factor if any	Imaging findings	Management	Outcome
Nair, 2018	18 years, female	Breathlessness and cough, painful swelling of face and neck for 3 days	Asthmatic	Pneumomediastinum pneumothorax, pneumoperitoneum, and pneumorrhachis	Conservative	Improved completely
Ramasamy *et al*., 2018	20 years, male	Neck swelling and difficulty in swallowing for a duration of 2 days	After a strenuous exercise 2 days before	Pneumomediastinum and pneumorrhachis	Conservative	Recurrence l year later
Heckman, 2018	20 years, male	Severe retrosternal chest pain, swelling in neck	Coughing and heavy emesis, occasional marijuana, and cigarette smoking	Pneumomediastinum and pneumorrhachis	Conservative	Improved completely
Liao and Wang, 2012	19 years, male	Shortness of breath and chest pain	Strenuous exercise	Pneumomediastinum emphysema and pneumorrhachis	Conservative	Improved completely
Bedolla, 2019	18 years, male	Dyspnea, chest tightness, and paroxysmal coughing	Poorly managed asthma	Pneumomediastinum and pneumorrhachis	Conservative	Improved completely
Gutierrez-Morale, 2019	20 years, male	Severe asthma exacerbation	Asthma, smoker	Pneumomediastinum and pneumorrhachis	Conservative	Improved completely

of a one-way valve.[12] Surgical intervention is also needed in thoracoarachnoid fistula and lung injuries.[1]

In our patient with Hodgkin's Lymphoma, the presence of cough most likely led to pneumorrhachis instead of underlying Hodgkin's lymphoma. In cases of spontaneous pneumorrhachis, observation and control of cough, emesis, or asthma become important if it is present.[4] A detailed review of available literature was done and is represented year-wise in the tabulated format in Tables 3-7.

CONCLUSION

Dominant pneumorrhachis in the absence of pneumothorax and the presence of only mild pneumomediastinum makes this case unique, discounting the theory proposed by Liao regarding positive alveolar pressure causing an alveolar rupture in cases of spontaneous pneumorrhachis being the predominant mechanism of air tracking into the mediastinum.

Teaching points

Pneumorrhachis:

- Asymptomatic
- Clinically non-specific
- Does not tend to migrate
- Reabsorbed spontaneously and completely
- Watch out for motor and sensory deficits.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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