

## Pneumomediastinum, pneumorachis, subcutaneous emphysema: An unusual complication of leukemia in a child

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**Core tip:** Pneumomediastinum, pneumorachis, subcutaneous emphysema is an unusual complication in leukemia. Although not mentioned in literature, this case may be an eye opener to look for this complication in this scenario. Acute lymphoblastic leukemia (ALL) is not mentioned as a cause of this condition but medical science has always made us learn from case to case. This case may be an index for this complication as all mentioned secondary causes were ruled out by all available investigations. Whether it is related to chemotherapy or not, it is definitely a case of ALL with this complication.

### Abstract

Pneumorrhachis (PR), or epidural emphysema, denotes the presence of air in the spinal epidural space. It can be associated with a variety of etiologies, including trauma; recent iatrogenic manipulations during surgical, anesthesiological and diagnostic interventions; malignancy and its associated therapy. It usually represents an asymptomatic epiphenomenon but also can be symptomatic by itself, as well as by its underlying pathology, and rarely can be fatal. The pathogenesis and etiology of PR are varied and can sometimes be a diagnostic challenge. As such, there are no standard guidelines for the management of symptomatic PR and its treatment is often individualized. Here, we present a case of a 14-year-old boy treated for leukemia who developed this complication and whether chemotherapy related or not, it proved to be fatal for him. To our knowledge, this is the first case in the literature of this complication with acute lymphoblastic leukemia.

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### INTRODUCTION

Pneumomediastinum or air in the mediastinum may originate from the esophagus, lungs or bronchial tree. As suggested by a handful of small case series in the literature<sup>[1-4]</sup>, spontaneous pneumomediastinum is an uncommon, self-limiting condition. It results from alveolar rupture, otherwise known as the Macklin phenomenon. Alveolar rupture results from high intra-alveolar pressures, low peri-vascular pressures, or both. Air escaping from the alveoli tracks into the mediastinum during the breathing cycle as the pressure in the mediastinum de-

Table 1 Investigation chart

Hb (g%)	TLC	PLT	Neutrophil/lympho	KFT urea/creat	PO <sub>2</sub>	SPO <sub>2</sub>	Ca <sup>2+</sup> (mg/dL)	Blood culture	Urine culture
11.2	4.2	68	73/21	33/0.98	77	95	9.8	Sterile	Sterile
10.5	3.7	63	69/26		68	91			
11.3	3.4	61	77/20	39/1.1	64	92	8.9		
10.9	3.2	58	78/19		67	94			
11.2	3.3	57	66/22	31/0.78	69	90	9.1	Sterile	Sterile

TLC: Total leucocyte count; PLT: Platelet count; KFT: Kidney function test.



Figure 1 Computed tomography showing pneumomediastinum and pneumorachis and subcutaneous emphysema.

creases relative to the pulmonary parenchymal pressure. From there, air may track into the cervical subcutaneous tissues, epidural space<sup>[5]</sup>, pericardium<sup>[6]</sup> and/or peritoneal cavity<sup>[7,8]</sup>. A handful of small case series in the literature have suggested the benign course of this condition<sup>[1-4]</sup> but there are still no clear guidelines regarding the diagnostic and therapeutic interventions needed.

## CASE REPORT

A 14-year-old male with a case of precursor B cell acute lymphoblastic leukemia (ALL) (Bcr-Abl-negative), who was on chemotherapy (as per UKALL-XII protocol), developed a cough for 5 d which was non-productive, brassy and with no nocturnal worsening. In addition to antibiotic policy as per IDS guidelines, he also received nebulisation and antitussives. Over a period of time, he developed pain and swelling of the chest and face, had crepitus on the chest wall and parotid area and complained of neuropathic pain of the lower limbs. Examination showed a pulse of 102 beats/min, blood pressure 110/72 mmHg, respiratory rate 18/min and temperature 99F. Crepitus was noticed on the anterior chest wall, parotid area bilaterally and neck. The chest showed an occasional wheeze in the right infrascapular area. Neurological examination showed that the neuropathic pain and reflexes were exaggerated in the lower limbs only. Computed tomography showed pneumomediastinum, pneumorachis and subcutaneous emphysema (Figure 1). The patient was managed in consultation with the cardiovascular and neurosurgery team who preferred a conservative line of management in the form of high flow oxygen, respira-

tory care and monitoring blood gases and electrolytes. Investigations showed hemoglobin 11.2 g%, total white cell count  $3.3 \times 1000/\text{mm}^3$  and platelets  $57000/\text{mm}^3$ . Oxygen saturation was 90% on room air. Kidney and liver functions were normal, as shown in Table 1. Blood and urine cultures were sterile and no sputum was possible even after giving mucolytic nebulisation and chest physiotherapy by an expert. The patient was managed conservatively but after 7 days of conservative management he developed respiratory arrest, was intubated and moved to the intensive care unit, but expired 2 d later. No apparent or definite cause could be ascertained.

## DISCUSSION

The presence of pneumomediastinum implies that there is or has been a breach of an air-containing mediastinal structure. Air in the mediastinal tissues may originate from the respiratory tract, such as after blunt or penetrating trauma to the facial bones, pharynx, hypopharynx, trachea and main stem bronchi. Dental procedures using compressed air may result in facial and neck subcutaneous emphysema and pneumomediastinum. Severe straining, the Valsalva maneuver and free perforation of the gastrointestinal tract may be responsible for the appearance of pneumomediastinum and subcutaneous emphysema<sup>[9]</sup>. Surgical emphysema was reviewed in 1957 by Shovelton<sup>[10]</sup> who reported 13 cases after endodontic treatment. Epidural space pneumorrhachis usually occurs by the two following mechanisms: atmospheric air passes through a spinal needle into the epidural space or air moves through the posterior mediastinum into the epidural space. Air may unintentionally enter the epidural space during a lumbar puncture or may enter intentionally during lumbar epidural anesthesia to locate the epidural level. These conditions are self-limited<sup>[11]</sup>. If air is present in the posterior mediastinum, it may dissect along fascial planes from the posterior mediastinum (or retropharyngeal space), through the neural foramina and into the epidural space. No true fascial envelope protects the epidural space. Mediastinal air moves into the epidural space behind the driving pressure of a tension pneumothorax or pneumomediastinum. In case reports, there are associated causative factors, such as trauma<sup>[12,13]</sup>, strenuous exercise<sup>[14]</sup>, asthma and violent coughing<sup>[15,16]</sup>.

Spontaneous pneumomediastinum is a self-limiting and benign condition that is frequently over-investigated and overtreated<sup>[17,18]</sup>. However, clear guidelines regarding

the diagnostic and therapeutic interventions are currently unavailable. The management of these patients has to be decided on an individual basis and often requires an inter- and multi-disciplinary approach. There is little literature showing any direct relationship between pneumomediastinum or pneumorachis and leukemia but there are a few case reports showing mediastinal necrosis of a mass causing mediastinal emphysema and pneumorachis can happen later, as already discussed, through fascial planes through the neuro foramina to the epidural space<sup>[19]</sup>.

Unusual complications like this must be kept in mind in leukemic patients and treated with a multidisciplinary approach as we did, but unfortunately even although the literature says this is a treatable condition, we lost the patient without any apparent cause. More work needs to be done on this condition and a helpful working plan needs to be charted out. Autopsy could have solved many issues but because of religious reasons and policies in this part of world, it was not possible for this case. ALL is not mentioned as a cause of this condition but medical science has always made us learn from case to case. This case may be an index for this complication as all mentioned secondary causes were ruled out by all available investigations. Whether it is related to chemotherapy or not, it is definitely a case of ALL with this complication.

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