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CASE REPORT

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PNEUMOMEDIASTINUM AND PNEUMORRHACHIS. RARE COMPLICATIONS IN PEDIATRIC AGE: CASE REPORT AND ITS MANAGEMENT

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ABSTRACT

Pneumomediastinum (PM) is an unusual and rare event in children. It is usually secondary to alveolar rupture in the pulmonary interstitium, followed by dissection of gas towards the hilum and mediastinum. Many events can lead to alveolar rupture, but the most common trigger factors in children are asthma and upper airway infections. Extremely rare is pediatric PM related to cardiac diseases, lung diseases such as pneumothorax, pulmonary embolism, thoracic traumatism, central airway perforation or digestive tract perforation and foreign body aspiration.

The clinical diagnosis is based on the concomitant presence of chest pain, dyspnea, and subcutaneous emphysema that may affect face, neck and chest. In severe cases, pneumomediastinum may lead to a cardiac tamponade, induced by an increase in pressure in the mediastinal compartment to develop a severe obstacle venous flow back to

the heart or in case of bacterial over-infection PM can lead to a mediastinitis. The diagnosis is confirmed by chest radiography and/or chest computerized tomography (CT).

In most patients the air in the mediastinal compartment is slowly reabsorbed by neighbors' tissues, favoring the spontaneous resolution of this condition. This process is also favored by the inhalation of high concentrations of low flow oxygen. In most cases conservative treatment such as bed rest and analgesics led to a rapid resolution of PM. The invasive surgical approach is necessary only in selected cases. It's important to identify and treat all the possible underlying causes (if identified) and predisposing factors should be identified and controlled to prevent recurrence of PM.

The combination of pneumomediastinum with pneumorrhachis (PR) rarely occurs in children. The present case report describes the presence of pneumomediastinum, subcutaneous emphysema, and pneumorrhachis in a child who had a history of persistent dry cough. A 9-year-old male child presented to our emergency service with respiratory distress, persistent dry cough, neck and chest pain. A chest X-ray and CT were performed and showed extensive pneumomediastinum with subcutaneous emphysema in neck area with no pneumothorax and concomitant air was in vertebral canal in the epidural space. Both clinical presentation and instrumental exams were consistent with those reported in the literature. The patient received noninvasive monitoring, analgesia, low flow oxygen, nebulized bronchodilators, intravenous steroids, and intravenous empiric antibiotics.

This case highlights how PM and PR can be successfully managed conservatively and how an early diagnosis and management of the underlying cause is essential and important.

KEY WORDS

Pneumomediastinum; asthma; children; subcutaneous emphysema; pneumorrhachis.

INTRODUCTION

Pneumomediastinum consists in the presence of free air within the mediastinum. Pneumomediastinum can be divided into spontaneous pneumomediastinum (SPM) without any obvious primary source and into secondary or traumatic pneumomediastinum with mediastinal organ injury or other known events such as trauma, surgery or medical procedures (1). Alveolar rupture leads to air infiltration along the bronchovascular sheath with free air finally reaching the mediastinum (2). Furthermore, if the air travels along tissue planes and spreads through the neck, face, abdomen or even the limbs, it can lead to subcutaneous emphysema. When the presence of air is in the spinal canal, we can observe pneumorrhachis (PR). The air may spread through fascial planes from the posterior mediastinum, through the neural foramina, and into epidural space. PR is usually asymptomatic and improves spontaneously.

Pneumorrhachis (PR) is characterized by the presence of air within the spinal canal. The air may spread along fascial planes from the posterior mediastinum, through the neural foramina, and into epidural space. PR is usually asymptomatic, doesn't tend to migrate and it is generally regarded as a self-limited and relatively benign process. Early diagnosis and management of the underlying cause it is essential. The causes of PR can be divided into iatrogenic, non-traumatic and traumatic. It is an exceptional but eminent radiographic finding, accompanied by different etiologies and possible pathways of air entry into the spinal canal. Since PR is usually asymptomatic, it is often a radiographic diagnosis and not a clinical one (3).

In the pediatric age the most frequent cause of PM is asthma and airway infection. PR associated with asthma is extremely rare in pediatric age and only very few cases are reported in the literature (4).

We describe the case of a pediatric patient with pneumomediastinum, subcutaneous emphysema and pneumorrhachis associated with asthma.

CASE PRESENTATION

A 9-year-old boy was admitted to the emergency department with a persistent dry cough, dyspnea and severe chest/neck pain, no fever reported. Family history: mother with allergic rhinitis associated with sensitization to *Olea Europeae*; Personal history: term birth, hyperreactive airways with multitrigger wheezing since the first year of life. The adenotonsillectomy was performed at 4 years of life. Not referred to allergic sensitizations. No trauma referred. No additional comorbidities were present.

At first clinical evaluation the patient was not well appearing, with signs of respiratory distress. Body temperature was normal, 35 breaths/min, 115 beats/min, sat O2 97% with low flow oxygen 1-2L/min, normal blood pressure for age, absence of cyanosis, normal peripheral perfusion. Auscultation of his chest revealed pathological sounds: reduced vesicular breath sound and bilateral wheezing and inspiratory substernal retraction. Normal heart sounds. Subcutaneous emphysema on the right and left side of his neck and upper chest was detected. Blood exams were performed: arterial hemogasanalysis: pH 7.40, pCO2 42 mmHg, pO2 59 mmHg, HCO3 24 mmol/l, normal C reactive protein; normal renal, hepatic function tests and electrolyte levels. Hemoglobin 12.7 g/dl; white blood cell count 16.4 × 10^3/μL; neutrophil count 47.7%; lymphocyte count 40.4%, monocyte count 9.8% platelet count 410 × 10^3/μl.

A chest X-ray showed extensive pneumomediastinum with subcutaneous emphysema in supraclavicular and neck area with no pneumothorax. Chest computerized tomography (CT) was performed and revealed the presence of air in all compartments of the neck: both in the visceral compartment (retropharyngeal area, carotid area and submandibular area)

and in the non-visceral ones, in the supra, infra and subhyoid bilaterally moreover at the level of the masticatory space and infratemporal fossa. Air was even in the supra and infraclavicular areas, axillary muscles, subcutaneous adipose tissue and on the anterior chest wall. Severe mediastinal emphysema was appreciable in all recesses of the mediastinum and between the chest wall and the pericardium with a slight compression of mediastinum structures.

Air was also appreciable in the retrocrural area and in the left posterior extrapleural space, between the erector spinae muscles and the subcutaneous adipose tissue. Air was also detected in vertebral canal in the epidural space, predominantly left approximately at C6, C7, D1, D2 and D3 (**Figures 1**, **2, 3**).

The patient was admitted and received noninvasive monitoring, analgesia, low flow oxygen (1-2 l/min) for 4 days, nebulized bronchodilators (salbutamol and ipratropium), intravenous steroids, and intravenous empiric antibiotics for preventing mediastinitis.

Cardiological consult was performed without pathological results. Neurological consult was performed to exclude neurologic involvement related to compressive events. Sputum analysis and culture was performed to exclude lower respiratory tract infections. Also, SARS-CoV-2 infection was excluded performing oropharyngeal swab and IgM and IgG against SARS-CoV-2 were absent. Normal dosage of alpha1 antitrypsin. Serological tests for the detection of *Mycoplasma Pneumoniae*, *Chlamydia Pneumoniae* and *Bordetella Pertussis* infection were normal. Immunological evaluation excluded congenital and acquired immunodeficiency.

Paper Radio Immunosorbent Test (PRIST) and ImmunoCap ISAC assay documented serum total IgE levels (575 kU/l) and positivity of specific IgE for *Dermatophagoides*

pteronyssinus, *Dermatophagoides farinae* (Der p1 1.00 kU/l and Der p 2 0.48 kU/l) and *Cynodon dactylon* (nCyn d 1 0.67 kU/l).

After 72 hours, a chest X-ray showed a reduced amount of gas in the neck and mediastinum but persistence of gas into medullary space without clinical neurological signs and symptoms. He continued noninvasive monitoring, analgesia, low flow oxygen (1-2 l/min), ~~inhaled~~ nebulized bronchodilators (salbutamol and ipratropium), intravenous steroids, and intravenous empiric antibiotics (Cefotaxime and Clarithromycin). He showed a progressive improvement during hospitalization and was discharged on day 12. It was prescribed therapy at discharge included fluticasone dipropionate 125 µg twice daily with spacer, home environmental interventions and he advised not to perform extreme physical activity to avoid barotrauma.

DISCUSSION

PM is an uncommon disease in pediatric age that usually shows a self-limited and not complicated course. It should be treated conservatively unless a complication requires using invasive procedures (5). Asthma exacerbation and lower airway infection appear to be the most frequent risk factors for PM (6). PM and PR have already been described as potential complications of virus infections, also in healthy children. Common flu virus infection also can cause pneumothorax, pneumomediastinum, subcutaneous emphysema and pneumorrhachis in a healthy child (7). Most cases appear to occur in teenagers and no obvious differences in incidence have been reported between the sexes. The probable reason of the major incidence in teenagers is described in many manuscripts and some authors reported that their mediastinal tissue is looser than adults, who have a fibrosed sheath that make air migration more difficult (8).

The clinical diagnosis is based on the association of chest pain, dyspnea, and subcutaneous emphysema that may affect face, neck and chest. Chest X-ray is increasingly being replaced by CT to confirm diagnosis. Lateral neck X-ray appear to be useful only in doubtful cases before CT exam (9).

Therapy was mainly based on supportive care, bed rest, low flow oxygen therapy, analgesics, steroids, and bronchodilators. Oxygen therapy has been recommended in most previous reports because it is considered that the consumption of oxygen increases the diffusion pressure of nitrogen in the interstitium, promoting absorption of free air in the mediastinum. Many studies report the use of empirical antibiotics to prevent possible infection such as mediastinitis even if it remains debatable whether antibiotic treatment is essential. Since PM may occur in asthmatic children, it's very important to obtain control of asthma. Clinical course in reported cases has been generally favorable with spontaneous resolution being achieved after hospital admission and supportive care.

Since asthma and upper and lower air infections are being described as the most common causes of PM it is very important to detect allergies, to detect viral and/or bacterial infections. Alpha1-antitrypsin deficiency screening has been recently recommended in patients with PM for differential diagnosis purposes (10).

A rare complication associated with pneumomediastinum is pneumorrhachis (PR) that consists in the presence of air within the spinal canal. It can be classified into internal or intradural and external or epidural. The causes of PR can be divided into iatrogenic, nontraumatic and traumatic (11). Although PR is usually asymptomatic and improves spontaneously, early diagnosis and management of the underlying cause is essential and important.

Most cases of epidural space pneumorrhachis are usually benign and improve spontaneously when the underlying cause is treated. In our case, pneumorrhachis almost disappeared without any intervention after ten days. Rarely, symptomatic PR with neurological deficits has been reported. Our case report represents an extremely rare case of pneumorrhachis secondary to pneumomediastinum with bronchial asthma in pediatric age. Spontaneous resolution occurs in most cases of epidural space pneumorrhachis, which allows conservative management in this benign occurrence (12).

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COMPLIANCE WITH ETHICAL STANDARDS

Conflict of interests

The Authors declare that they have no competing financial interests.

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Authorship

Each Author listed on the manuscript has seen and approved the submission of this version of the manuscript and takes full responsibility for the manuscript. All Authors approved the final manuscript.

Author contributions

Wrote the manuscript: AF and GC; contributed to the discussion: FF, MO and LT; collected the references: NZ, VA and MC. Reviewed the manuscript: FF, MO and LT. Each author listed on the manuscript has seen and approved the submission of this version of the manuscript and takes full responsibility for the manuscript. All authors read and approved the final manuscript.

Ethical approval

Human studies and subjects

The manuscript was written according to Good Clinical Practice and compliance with the Declaration of Helsinki with successive amendments.

Animal studies

N/A.

Data sharing and data accessibility

The data presented in this manuscript are available on request from the Corresponding Author.

Publication ethics

Plagiarism

All original studies are cited as appropriate.

Data falsification and fabrication

All the data corresponds to the real.

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Figure 1. *CT shows the presence of air in all compartments of the neck.*

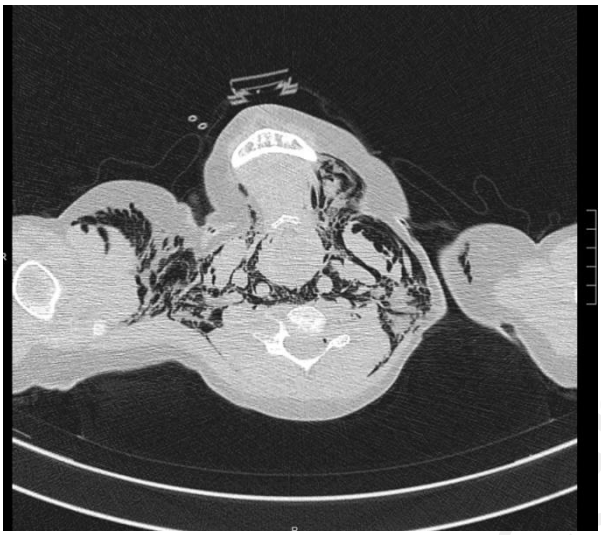

Figure 2. *CT shows the presence of air in all recesses of the mediastinum and in vertebral canal in the epidural space.*

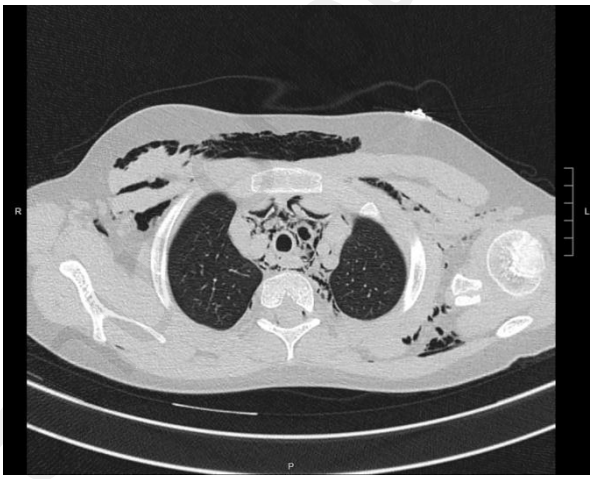

Figure 3. *CT shows the presence of air in vertebral canal in epidural space.*