



Pneumomediastinum – Uncommon Presentation of Rabies

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Abstract

Rabies is caused by neurotropic RNA virus of rhabdoviridae family, transmitted mainly through the saliva of infected domestic or wild animals, resulting in incurable, fatal encephalomyelitis. Of the two varieties of its presentation, classical furious form is most often diagnosed readily with features of hydrophobia, aerophobia, aggressive behaviour, whereas the atypical paralytic presentation may pose dilemma in diagnosis.

Here we have a case of an adolescent male admitted for evaluation for agitation, dysphagia, and breathlessness with incidentally detected finding in Computed Tomography of head prompted us to review the chest radiograph, which showed pneumomediastinum.

Keywords: Rabies; Rhabdoviridae; Pneumomediastinum; Encephalomyelitis; Negri bodies.

Case report

A 17-year-old male from tribal area of northern Kerala, India was brought to the emergency department with complaints of breathlessness, agitated behaviour of one day duration. He was in his usual good health, prior to the onset of symptoms. At the time of admission patient was conscious, oriented and anxious. His vitals were stable and maintained 98% saturation in room air. On neurological examination, Glasgow Coma Scale was (15/15).

Plain Computed Tomography (CT) of head showed air in the retropharyngeal space extending between C1-C4 vertebral body levels (figure 1), a finding picked up incidentally prompted us to review the chest radiograph and history.

The lateral scout image of skull with neck showed air in the retropharyngeal space extending between

C1-C4 vertebral body levels. The plain axial CT sections of brain showed fairly normal brain parenchyma (figure 2). Axial CT sections of skull base in bone window settings showed air involving the deep neck spaces spanning retropharyngeal, parapharyngeal, carotid and posterior cervical space (figure 3). On reviewing the frontal chest radiograph, there was streaky air lucencies outlining soft tissues of lower neck bilaterally, extending down along the mediastinum, with band of air lucency lifting the mediastinal pleura laterally on the left side and outlining and lifting the thymic remnant (figure 4).



Figure 1: Lateral scout image of head and neck showing air in the prevertebral space (black arrow).

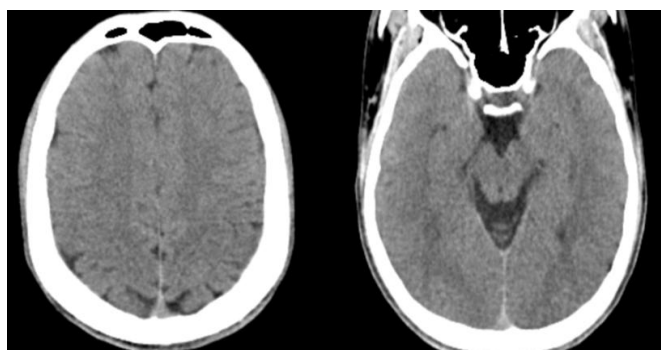


Figure 2: Axial CT sections showing normal brain parenchyma

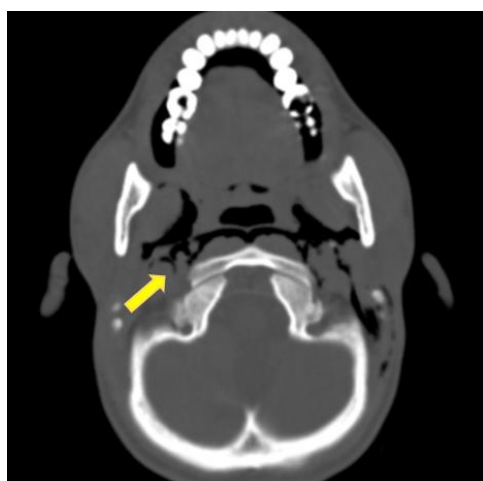


Figure 3: Axial CT sections of skull base showing extensive air in the deep neck spaces (yellow arrow)

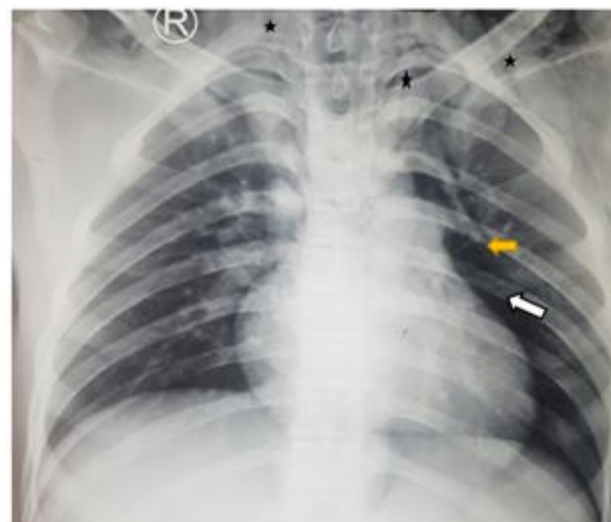


Figure 4: Chest x-ray streaky air lucencies outlining soft tissues of lower neck bilaterally (asterisks*), extending down along the mediastinum, with band of air lucency lifting the mediastinal pleura (white block arrow) laterally on the left side and outlining and lifting the thymic remnant (yellow arrow).

History was reviewed. He had history of stray dog bite to his left index finger about two and a half months back; following the incident the dog died next day, for reasons not known. He had not sought medical advice and not taken post exposure prophylaxis.

Discussion

The pneumomediastinum is diagnosed in 1/44,500 of accident and emergency attendances or 1/100,000 of natural births^[1].

This rare condition is confirmed via chest X-ray or computed tomography (CT) scanning of the thorax. Severe central chest pain is the most common symptom. Other symptoms include breathlessness and subcutaneous emphysema. A "crunching" sound timed with the cardiac cycle (Hamman's crunch) is the auscultatory feature. Out of the overwhelming aetiologies of pneumomediastinum, one of the forgotten entities is due to rabies.

The basic pathophysiology of pneumomediastinum and interstitial air leak in rabies still remains elusive, however un-self-controlled, violent spasm of pharynx and the larynx resulting in acute and transit obstruction of airway and the oesophageal lumen,

can cause alveolar or oesophagus rupture under a transient high pressure. Boerhaave's syndrome has been reported in rabies^[2]. Rabies is a highly fatal zoonotic viral disease^[3] caused by neurotropic RNA virus of genus *Lyssavirus*, family rhabdoviridae^[4]. It has variable and uncertain incubation period, transmitted to man by the bite of a rabid animal or the contamination of scratch wounds by virus infected saliva. From the site of inoculation, the virus replicates, migrating along the peripheral nerves from neuromuscular junction and finally reaches central nervous system (CNS), with resultant widespread dissemination^[3].

The disease is invariably fatal for humans, in the absence of post exposure prophylaxis prior to the onset of symptoms. Broadly based on clinical manifestation, rabies can be classified into classic furious and atypical paralytic type. The furious type has dramatic clinical presentation in the form of aerophobia, hydrophobia, hypersalivation and aggressive behaviour, and often easily diagnosed even in the absence of clinical history of exposure. Nevertheless, the paralytic form most often has atypical clinical presentation or complication leading to delayed detection and misdiagnosis, mostly due to its similarities with the Guillian-Barré syndrome and neuropsychiatric illnesses^[2].

Often diagnosis is made based on clinical history and presentations. Hence imaging in rabies is seldom done, with imaging findings in rabies encephalitis showing non-specific areas of hypoattenuation in the basal ganglia, hippocampus, brainstem, and periventricular white matter, and late cases showing diffuse cerebral swelling and haemorrhage^[5].

Definitive diagnosis can only be ascertained by laboratory tests conducted post mortem on central nervous system tissue by the presence of Negri bodies, and positive staining by direct fluorescence assay (DFA) and reverse transcriptase -polymerase chain reaction (RT-PCR) or using the samples of saliva, serum, and skin biopsies of hair follicles at the nape of the neck^[3].

In our patient with agitation, the clinician investigated for encephalitis. Incidentally picked up retropharyngeal and deep neck space air in CT head, prompted to further evaluate for pneumomediastinum and search for underlying pathology.

Teaching Point

Rabies, though best known with catastrophic clinical manifestation, uncommon presentation like pneumomediastinum or extensive subcutaneous emphysema, should prompt the clinician to have index of suspicion to prevent misdiagnosis or delay in diagnosis of the atypical presentation of rabies.

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