PULMONARY BOTRYOMYCOSIS IN A PATIENT WITH DOWN SYNDROME

Pulmonary botryomycosis is a rare chronic, pyogranulomatous infection affecting the lung parenchyma. We describe here the clinical and histopathological findings of pulmonary botryomycosis reported for the first time in a Down syndrome female who required prolonged intensive care. This case has other different unique aspects. It is the first case to present with empyema, the second case involving the right lower lobe and the first case managed by decortication.

Key words: Botryomycosis, decortication, Down syndrome, empyema

Botryomycosis was first described in a horse lung in 1870.[1] It is a rare chronic, pyogranulomatous infection caused mainly by Staphylococcus aureus and occasionally by other bacteria such as Pseudomonas aeruginosa, Proteus sp., Escherichia coli, Serratia marcescens, Neisseria sp., Moraxella sp., Propionibacterium sp. and others.[2] The term botryomycosis is derived from the Greek words botryose (bunch of grapes) and mycosis (fungus).[3] Histologically, the appearance consists of densely packed microorganisms surrounded by eosinophilic material and is similar to that of actinomycosis with characteristic granule formation.[4,5] Two forms exist; a relatively common cutaneous form (75%) and a rare visceral one (25%).[1] The lung is rarely the primary location of this disease, with only 23 reported cases in the English literature. We describe here a case of pulmonary botryomycosis in a Down syndrome female. To our knowledge, this may be the first case of botryomycosis to be reported from Saudi Arabia or in a Down syndrome patient.

Case Report

A 23-year-old Saudi Down syndrome female presented to the emergency room on 29 January 2008 with a 1-month history of fever, productive cough and worsening dyspnea. She had received medical care at different centres without any improvement. There was no history of recent travel or exposure to sick contacts. Past medical history was unremarkable apart from being diagnosed with Down syndrome and cognitive developmental delay. Physical examination revealed a sick-looking, febrile (38.5°C) female with tachypnea, tachycardia and hypotension. Oxygen saturation was 82% at room air. There was nearly absent breathing sounds over the right side of the chest. Chest radiograph revealed near total opacification of the right lung with hydro pneumothorax, lung collapse and contralateral mediastinal shift [Figure 1]. A chest computed tomography (CT) scan [Figure 1] revealed right-sided massive loculated collection associated with air-fluid levels, complete collapse of the right lung and contralateral mediastinal shift. There was also an anterior mediastinal loculated collection compressing the great vessels. Intravenous fluids and piperacillin–tazobactam were administered immediately along with other supportive measures. Initial laboratory results showed a very high erythrocyte sedimentation rate (138 mm/h), elevated blood leukocytes (17.6 × 10⁹/L) and low haemoglobin (9 gm/dL). She had low serum albumin (19 g/L) and elevated serum creatinine. The patient underwent immediate decortication (empyemectomy, multiple abscesses drainage, partial parietal pleurectomy and removal of the constricting peal) through the right posterolateral thoracotomy with lung isolation anaesthesia. The right lower lobe was found to be completely hepatized. Post-operatively, the patient required mechanical ventilation for 17 days and the antimicrobial regimens were modified to include vancomycin, meropenem and levofloxacin over variable periods due to persistent fever. The patient continued to improve clinically and a repeated chest CT scan [Figure 1] showed dramatic improvement. She was discharged after 40 days from surgery and she completed another month of oral levofloxacin. Bacterial, mycobacterial and fungal cultures of pleura, pleural fluid, blood and sputum failed to show any growth. Histopathologic examination

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of the lung tissue revealed dense nodular and mixed acute and chronic inflammatory infiltrates. Several eosinophilic granules were noticed with embedded cores of gram-positive cocci [Figure 2].

Discussion

Pulmonary botryomycosis is a rare entity that is usually associated with variable levels of immune dysfunction as in patients with diabetes, cystic fibrosis, human immunodeficiency virus infections and others. Other risk factors were also reported in literature as post-operative stress, foreign body, steroid intake, periodontal abscesses, alcoholism and malnutrition.\(^{3,6,7}\) We present here a case of pulmonary botryomycosis in a Down syndrome patient.

Figure 1: Radiological features of our patient. (a) Pre-operative chest radiograph showing right lung collapse, hydropneumothorax and contralateral mediastinal shift. (b and c) Pre-operative computed tomographic scan (lung and mediastinal windows) showing right-sided massive loculated collection associated with air-fluid levels, complete collapse of the right lung, mediastinal shift and anterior mediastinal loculated collection compressing the great vessels. (d, e and f) Post-operative radiograph and computed tomographic scans showing dramatic resolving of pre-operative findings

Figure 2: Microphotographs of the lung biopsy. (a) Mixed inflammatory cells surrounding several (arrowheads) eosinophilic granules (H and E, ×200). (b) One granule is shown (arrowheads) with centrally packed gram-positive cocci surrounded by a hyaline matrix (H and E, ×400)
Apparently, this is the first case to be reported from Saudi Arabia. The diagnosis of botryomycosis in our patient was mainly established by histopathological examination of lung parenchyma, which revealed multiple eosinophilic granules surrounding cores of dense collections of gram-positive cocci [Figure 2]. This appearance is similar to the previously described classic Splendore–Hoeppli reaction, where abscesses usually contain dense colonies of bacteria. The failure to grow bacteria from the patient’s specimens was probably due to long courses of oral antimicrobials and pre-surgical intravenous piperacillin–tazobactam. This case presents several unusual patterns about pulmonary botryomycosis. It is the first case reported in a Down syndrome patient, which might be related to the already described immune dysfunction in these patients. Unlike the other reported cases of pulmonary botryomycosis that usually presented with discrete lung masses, this patient presented with empyema and clinical sepsis. Other less common radiological variations include cavitatory lesions, consolidations or diffuse infiltrations. Review of the literature revealed that this is the second case to involve the right lower lobe. Early surgical intervention was crucial for survival of our patient. Moreover, the patient was spared any lung resection, which is the usual practice in most of the reported cases.

In summary, this case has different unique aspects. It is the first reported case in a Down syndrome patient, which might be related to the already described immune dysfunction in these patients. Unlike the other reported cases of pulmonary botryomycosis that usually presented with discrete lung masses, this patient presented with empyema and clinical sepsis. Other less common radiological variations include cavitatory lesions, consolidations or diffuse infiltrations. Review of the literature revealed that this is the second case to involve the right lower lobe. Early surgical intervention was crucial for survival of our patient. Moreover, the patient was spared any lung resection, which is the usual practice in most of the reported cases.

References


*FA Al-Rabee, WA Hayajneh, M Shorman, R Al-Hubail

Departments of Surgery (FAAR), Pediatrics (WAH), Medicine (MS), and Intensive Care (RAIH), King Fahad Specialist Hospital – Dammam, PO Box 15215, Dammam, Saudi Arabia-314 44

*Corresponding author (email: <fuadrabee@hotmail.com>)
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