Chest manifestations of Kaposi’s Sarcoma in HIV-infected children

S. Theron, S. Andronikou, J. Du Plessis et al

Introduction

• Kaposi’s Sarcoma (KS) = major complication of adult AIDS
• AIDS-defining disease (1980’s)
• Most common neoplasm in adults with AIDS
• KS classically affects skin and mucous membranes and presents with red or purple raised plaques or papules
• KS can however involve other organ systems, eg GIT, lungs, liver

Kaposi’s Sarcoma in adults

• Mucocutaneous KS asymptomatic
• Pulmonary KS presents with dyspnea, cough, chest pain and hemoptysis
• Pulmonary KS is:
  – rare if CD4 count > 150 cells/ul
  – usually seen if CD4 count < 100 cells/ul
• Pulmonary KS in adults is rare without mucocutaneous involvement
• 25% of adults with cutaneous KS have lung involvement and it is considered the most life-threatening form of the disease

Kaposi’s Sarcoma in children

• KS is rarely seen in children from developed countries
• BUT is not uncommon in HIV-children from endemic regions without access to HAART
• In Sub-Saharan Africa KS is one of the leading childhood tumors
• Paediatric KS has an aggressive and fulminant behaviour
• Pulmonary involvement rare
  – 5 documented paediatric cases in literature
• Difficult to differentiate from other neoplastic / infective conditions
• Median survival without treatment = months

Pathology

Kaposi’s Sarcoma
• Angioproliferative tumor
• Characterized by the presence of
  – Proliferating endothelial cells
  – Fibroblasts
  – Infiltrating leucocytes
  – Spindle shaped tumor cells

Three Recognized types

• Classical
  – Elderly Mediterranean males
  – Slowly progressive
  – Typically only cutaneous lesions
• Endemic
  – Young males from equatorial Africa
  – Frequent visceral and lymphatic involvement
• HIV Related
  – HIV infected individuals
  – Affects males and females as well as adults and children
  – Aggressive with early spread to organs and lymphatics
Radiology

- Adult patterns on CXR well described
- 2 main patterns
  - Diffuse pulmonary linear interstitial infiltrate
  - Ill defined nodular pattern with coalescence
- Central or peri-hilar predominance is typical and associated pleural effusions common (50%)
- Lymphadenopathy uncommon
No description of the radiological appearance of paediatric pulmonary Kaposi’s Sarcoma could be found in the literature.

**Our Study**
- 6 HIV positive children with Kaposi’s Sarcoma and pulmonary involvement
- **Diagnosis**
  - 4 Histological (specimens: skin lesions, lymph nodes, tongue mass)
  - 1 FNA
  - 1 Autopsy
- In all 6 the diagnosis of pulmonary involvement was made clinically and radiologically and in 1 case it was confirmed with autopsy
- Sequential CXR’s in all 6 and Chest CT’s in 4 were evaluated

**Clinical**
- Only 2 had typical skin lesions at presentation and 1 developed them after diagnosis
- CD4 count 162 – 646 cells/ul
- 3 had CD4 count above 400 cells/ul
- All 6 had presenting complaints
  - Chronic cough
  - Dyspnoea
  - Generalized Lymphadenopathy

**Radiological Findings: CXR**
- Combination of air-space (100%) and reticular opacification (83%)
  - Predominantly mid and lower zone
- Usually bilateral (83%)
- Pleural effusions common (83%)
  - Often bilateral (50%) and large (50%)
- Lymphadenopathy common (50%)
- Typically progressive air-space involvement on follow up (100%)

**Radiological Findings: CT**
- Typically bilateral peri-hilar air-space opacification (100%)
  - Lower zone involvement common (75%)
- Reticulonodular opacification usual (75%)
- Pleural effusions common (75%)
  - Often bilateral (50%)
- Lymphadenopathy major feature
  - Mediastinal (100%), Axillary (100%) and bilateral hilar (75%)
- Bronchial compression (100%)
Differential Diagnosis

- **PCP**
  - CXR: Perihilar reticular opacification, can progress to airspace
  - CT: ground glass opacity, pleural effusions and nodes rare

- **PTB**
  - CXR: Ghon complex, progress to multilobar consolidation
  - CT: Large nodes, central necrosis and "ghost-like" enhancement

- **Cryptococcosis**
  - CXR: Single or multiple pulmonary nodules

- **Aspergillosis**
  - CXR: Pulmonary nodule(s)
  - CT: "Halo" surrounding the nodule, can have central necrosis
Take Home Message

• Immuno-compromised child with non-resolving respiratory symptoms
• Suspicion of Kaposi
  – Combination reticulonodular + airspace opacification (bilaterally, predominantly mid and lower zone)
  – Pleural effusions
• Higher index of suspicion
  – Progression on follow-up X-rays
• Absence of cutaneous lesions and a high CD4 count do not exclude the diagnosis