KAWASAKI DISEASE

Martin Wong & Lee Wai Seah

Introduction

Kawasaki disease (KD) is an acute systemic febrile syndrome of the early childhood characterised by generalised vasculitis involving small and medium-sized arteries, formerly known as mucocutaneous lymph node syndrome. It was first described by Dr Tomisaku Kawasaki in year 1967. The most significant complication of KD is coronary artery aneurysm with risk of coronary artery occlusion, myocardial infarction and sudden death. It has replaced rheumatic heart disease as the leading cause of acquired heart disease of children in developed countries. There is no single diagnostic test which is specific to KD and diagnosis is established based on a combination of various clinical criteria; supported by laboratory tests. Prompt recognition and treatment is essential to decrease the risk of coronary complications and mortality.

Epidemiology

KD affects children of all ethnic origin but the incidence is highest among Asians (ranges from 3.4 to 218.6 per 100,000 children below 5 years of age). It is mostly observed in children below 5 years although rare cases among teenagers and adults have been reported. The peak age of occurrence is between 18 to 24 months.

Pathology

KD is a generalised systemic vasculitis involving blood vessels throughout the body with predilection of coronary arteries involvement. Early disease stage is characterised by necrotising inflammation of the vessel wall with neutrophil infiltration and oedema of media and intima which is followed by proliferation of lymphocytes and plasma with release of inflammatory cytokines and enzymes which cause disruption of elastic lamina. This leads to weakening of vessel wall, ectasia and aneurysm formation. During the convalescent phase, the acute inflammatory cells are gradually replaced by myofibroblasts and monocytes with deposition of fibrotic tissues which eventually result in vessel wall thickening and obliteration of vessel lumen.

Aetiology

The exact aetiology of KD remains unknown. However, the clinical and epidemiological features strongly support an infectious origin in genetically susceptible individuals.

Clinical Features

The clinical course of KD can be divided into 3 clinical phases: acute, subacute and convalescence phase (Table 36.1).

Typical Kawasaki Disease

Typical or classical KD is characterised by presence of ≥ 5 days of fever and ≥ 4 of the following main clinical criteria (Table 36.2).