A CASE OF KAWASAKI DISEASE IN PORTUGAL

RUI PROENÇA, MIGUEL DE SOUSA, KARIM VAKIL, MÁRIO SEQUEIRA, ARTUR SYKES
ISABEL MARQUES, J. MALHADO

Lisbon City Hospitals. Hospital Curry Cabral. Department of Medicine and Transmissible Diseases

SUMMARY

This case report is believed to be the first case of Kawasaki disease in Portugal. An otherwise healthy 20 years old female was carefully examined and diagnosis of mucocutaneous lymphnode syndrome established, based on: typical clinical picture, exclusion of other mimicking situations and middle term evolution of this patient. The A. A. wish to emphasize their diagnosis complied on C. D. C. criteria for Kawasaki disease. A short up dated briefing on this peculiar entity and geographycal pathology are included in this article.

RESUMO

Um caso de doença de Kawasaki em Portugal

Este artigo descreve o que se julga constituir o primeiro caso do doença de Kawasaki registado em Portugal. Uma jovem do sexo feminino de 20 anos de idade, até então saudável, foi cuidadosamente examinada tendo sido estabelecido o diagnóstico de doença de Kawasaki com base nos seguintes factos: quadro clínico típico, exclusão de outras possíveis entidades nosológicas e evolução da doença a médio prazo. O diagnóstico neste caso obedeceu aos critérios internacionalmente definidos. O texto inclui uma breve revisão actualizada sobre esta entidade clínica e sua ocorrência geográfica.

INTRODUCTION

Kawasaki disease or M. L. N. S. is a clinical entity first reported in 1967 by this author[1] in Japanese children. Later on cases were reported in young adults and also children from Korea,[5] Hawaii,[8] USA.[6-9, 11] and Greece.

Its etiology remains under investigation.

Bacterial and viral agents have been speculated but unidentified. It is doubtful to consider chemical detergents or allergies implicated in this disease.

In the early 70's Japanese A. A. found cytoplamic Rickettsia-like inclusions in skin and lymphnode cells of patients aflicted with M. L. N. S.,[1, 4] but ulterior studies failed to support such a theory. Further on thorough investigation will be needed.

Nearly all cases have a seasonal prevalence between April and September involving children and rarely young adults, with great incidence during the first year of life.

It seems endemic in Japan where 1978 data report 18.182 cases. Males neatly dominate (ratio 1.5/1).

The clinical course of M. L. N. S. is variable and causing death in 2.6% (2,6) according to some series, with decreasing mortality rate through proper treatment particularly after introduction of aspirin.

High mortality risk is associated with age, being higher at lower age children and death overcomes through coronary thromboarteritis with consequent myocardial infarction. Anatomopathologic lesions pulled out strongly suggest those of I. P. N. (Infantile Poliarteritis Nodosa).

Care has been taken in the interpretation of these data since most cases have currently a benign course.

Fatalities, when occurring, are sudden, and during aparent recovery.

Myocarditis and pericarditis have been found in M. L. N. S. cases early diagnosed; endarteritis and proteinuria have been related to this disease. Of utmost importance is the evidence of coronary arteritis in fatal cases, not seldom with vessel aneurysms and/or thrombosis sometimes leading to infarction.

Latest reports with coronary arteriograms in 12 out of 20 patients with a follow-up evaluation have reinforced previous reports on myocardial dysfunction, found in 7 patients presenting coronary aneurysm.

Up dated studies on this disease have been alerting on several aspects namely electrocardiographc alterations in 74%[4] of reported cases and abnormal coronary arteriograms in 82% of patients.

Heart failure leading to death was found in 10% of the affected population and one major complication was mitral incompetence.[19, 14]

All efforts to investigate coronary artery involvement in M. L. M. S. have nor yet given a clearcut information on natural history of arterial injury. Apparent cure has also been reported.

Anatomo-pathology allowed through necropsy of fatal cases, information on arterial damage with intimal proliferation, fibrinoid necrosis and multiple aneurysms. Other than coronary arteries have been involved but less frequently.

Non-invasive methods other than coronary arteriogram have prooved helpful namely bi-dimensional ecography for aneurysm study. This device has been succesful as a diagnostic method.

This disease or syndrome remains until now produced by as unidentified agent or process; each and every case should

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be deeply studied before M. L. N. S. is diagnosed; C. D. C. criteria should be adopted and regarded when thinking of Kawasaki syndrome.

In nearly 70 % of documented survivals, mainly children, there is cardiac involvement with E. C. G. alterations (PR, QT prolongement and ST-T alterations representing poor prognosis).

Kawasaki and col. present in 1974[1] a revision of dominant manifestations with their frequency: fever (95 %); skin dequamation of extremities during recovery (94 %); Polymorphous exanthema without vesicles or crusts during onset (92 %); dry red lips (90 %); homogenous palmo-plantar erithema in early phase (88 %); conjuntival congestion (88 %);
hard edema of extremities (76 %); cervical lymphnodes (75 %).

Kawasaki disease first identified in Japan, became later a deep concern to physicians, mainly cardiologists; vasculitis and coronary arterial problems in young patients oblige investigation to exclude Kawasaki disease, nowadays.

CASE REPORT

This patient was a 20 year old healthy woman, whose illness apparently began 3 weeks before admission, when she complained of anorexia. In her past there is no history of trips abroad. On her second week of illness she disclosed right inguinal lymphnode enlargement, (diameter 2 cm), soft and painful palpation. Surprisingly her 3rd week was enriched with symptoms mainly fever with late evening uprise, sweat, voice alterations and right cervical adenomegaly. At this time she took a tablet of Optalidon (compound with 50 mg of isobutylalilbarbiturate, 125 mg of demetilamino--fenazone and 25 mg cafeine) as self prescription: she denied any other medicine taken or prescribed, before admission. Disease onset was not related to the last menses and there was no previous use of vaginal tampons.

This picture remained unaltered until the third day of this week, except for cervical lymphnode enlargement, when she discovered macular spots on her left hand palms, and progressively edema and regularly homogenous bilateral erithema of hands during next morning.

On the same day afternoon erithema and edema involved her soles, and later on nearly 5 mm maculo-papular exanthema without pruritus on whole body, except the scalp. This is the picture which determined admission on 26/5/80. At this time she had fever (below 38 °C), dry cracking red lips and tongue, cervical, axillar and inguinal lymphadenopathy, the latter ones small in size. Exanthema multiforme with bilateral regular palmo-plantar erythema, pruritus and hard oedema of lower limbs without pitting on pressure; no other alteration was found namely on throat examination and heart auscultation.

Blood assays for V. D. R. L., Weil-Felix, C.-Protein were not contributory.

Acute and recovery phase bioassays for ASO antibodies remained always below 200 U., throat and nose cultures for β hemolytic streptochoccus were negative. Urine remained normal. Immunoglobulins G, M and A were respectively — 1308, 227 and 227 mg %. Complement levels were unaltered.

Function of T and B lymphocytes revealed 65 % for the former and 26 % for the latter, without morphological abnormalities.

Acute and recovery phase serologic precipitin and agglutinating tests for ECHO, Coxsakie and cytomegalovirus were not contributory, and the same happened for toxoplasmosis. Paul-Bunnell-Davidson and anti-Epstein-Barr antibodies were also within normal values.

During acute phase (3rd day of admission), skin biopsy was performed stating hyperkeratosis, light acanthosis and lymphocitic infiltration with slight capillary enlargement. These findings have been considered non-specific.

ECG and gynaecologycal studies were unaltered. The admission follow up aspects were as follows:

4th day, apirexia without recurrence, and beginning of edema healing and palmo-plantar erithema.

5th day, dequamation, starting at finger edges.

7th onwards until third week — palmo-plantar patch-like dequamation.

This patient had a *restitutio ad integrum* in 8 weeks of illness, and remained under medical observation for 1 more year.

ECG remained unaltered. After discharge from the Hospital, tests for Optalidon compound components were performed, failing to disclose any skin sensitization to those drugs.

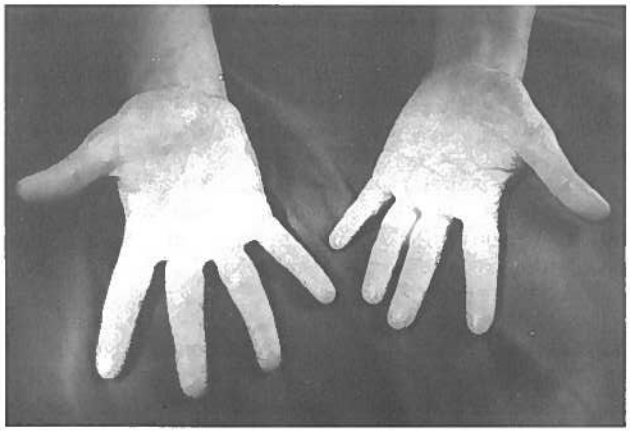

Figure 1: Maculo-papular exanthema involving the hands.

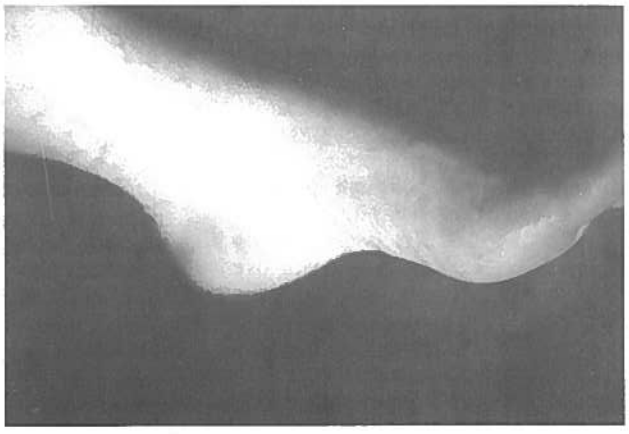

Figure 2: Maculo-papular exanthema involving the feet.

DISCUSSION

The analisis of all parameters of our patient drove us undoubtedly to Kawasaki disease; only conjuntival congestion was missing to totally fullfil disease criteria.[1]

Drug toxidermia for Optalidon or any of the components was excluded through lack of response to the tolerance tests performed.

Careful examination of the exanthema (palmo-plantar) without history of tonsilitis and pharingitis enhanced exclusion of scarlet fever. No evidence of group A streptococcus was disclosed and ASO titers remained under normal values.

Toxic shock syndrome as per CDC criteria[16] has a pattern mimicking mostly Kawasaki disease, but was carefully discarded through lack of vaginal tampons use and absent relationship with last menses. The character of the rash is different — maculo-papular in Kawasaki disease and non-papular, diffuse erythrodermic in toxic shock syndrome. The shock also was absent in the patient.[16]

Secondary stage syphilis, toxoplasmosis, infections mononucleosis, were carefully and clearly discarded through clinical, haematological and serologic determinations.

To support this diagnosis of Kawasaki disease the AA have based on four out of five CDC criteria; the only *absent sign* was conjuntival injection, which in this entity is not a dominant sign. Some reviewed papers report absence of this sign in 12% of cases.[6]

The AA maintain this patient under a careful follow-up and so far no evidence of relapse has been found or signs of late cardiac complications detected, either on clinical or ECG basis.

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Address for reprints: Rui Proença
Department of Medicine and
Transmissible Diseases
Hospital Curry Cabral
Lisboa - Portugal