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Oral lesions in Kawasaki disease

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Abstract: *Kawasaki disease (mucocutaneous lymph node syndrome) is an acute vasculitis of childhood carrying a 1-2 per cent mortality from cardiovascular complications(1). Despite the extensive literature on Kawasaki disease in paediatric journals, there has been a paucity of documentation in the otolaryngology literature. This is despite the fact that Kawasaki disease may present as an otolaryngological emergency before the diagnosis is established. We describe thirty cases of Kawasaki disease in children, few of which presented to the ENT department of this hospital within a period of 9 months. These cases illustrate the slow evolution characteristic of the disease and highlight the difficulties of diagnosis in the initial febrile stage. We emphasize the importance of considering the diagnosis when treating a young child with a pyrexia resistant to antibiotics, as prompt introduction of therapy may decrease the risk of fatal coronary artery or cardiac complication(2). Our study emphasizes mainly on head and neck conditions along with other clinical features of Kawasaki disease,.*

Keywords: mucositis, lymphadenopathy, vasculitis

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I. Introduction

Kawasaki disease (KD), otherwise known as acute infantile febrile mucocutaneous lymph node syndrome, was first described in the Japanese literature by Dr Tomisaku Kawasaki in 1967 and in the English literature in 1974(3). The disease, a multisystem vasculitis of small and medium sized arteries ,predominantly affects children less than 5 years of age , with the peak incidence occurring in children aged 1–2 years (4). It more often occurs in the late winter and spring seasons.

Five of the following six principal symptoms are required for the diagnosis of classical KD
(1)fever of five or more days duration that is refractory to antibiotics and acetaminophen
(2)bilateral nonexudative conjunctival injection;
(3) oral cavity involvement, including fissuring of the lips, strawberry tongue and mucosal hyperemia;
(4) polymorphous rash;
(5) peripheral extremity involvement, including edema or desquamation; and
(6) acute cervical adenopathy >1.5 cm in diameter.

Atypical KD may be diagnosed on the basis of fever in addition to two or three of the additional symptoms.In addition to the aforementioned symptoms, other otolaryngologic manifestations may occur, including facial palsy, otitis media and neck abscess.

II. Materials and methods :

A retrospective review of all consecutive patients diagnosed with KD between May 2017 to Dec 2017 in PBM hospital and tertiary care centre in Western Rajasthan. Patients were reviewed to ascertain the presence of otolaryngologic symptoms of the disease mainly emphasizing on oral symptoms. Parameters reviewed included age, sex, ethnic background, a diagnosis of classical versus atypical KD, otolaryngologic manifestations, treatment, cardiac complications and outcome.

III. Results:

Between the above mentioned time period 30 cases of KD were studied. 18 were males and 12 females ranging from 9 months to 11 years of age. The mean age of the patient was 5 years. Of those patients diagnosed with KD, 21 had the classic form of the disease, with 9 having the atypical form of the disease. All 30 patients had fever for a period of at least 5 days. All 21 of the patients with the classic form of the disease and 4 of patients with the atypical form of the disease developed an oral mucosal changes. With respect to limb involvement, 20 of classic patients and 3 of atypical patients had peripheral extremity symptoms.

Table 1

Oral manifestations	male	Female	Total
Oral mucosal changes	16	10	26
Strawberry tongue	11	4	15
pharyngitis	6	12	18
Acute tonsilltis	10	5	15
epiglottitis	1	3	4
trismus	2	1	3

Table 2

Other head and neck menifestation			
Facial exanthem	15	6	21
Cervical lymphadenopa-thy	9	5	14
Deep neck abcess	2	1	3
conjuctivitis	4	2	6
Otitis media	7	2	9
Rhinitis	2	3	5
Facial palsy.	1		1

Of the total patients, 16 had abnormal laboratory values (leucocytosis, thrombocytocis and an elevated ESR) on presentation. All 30 patients had abnormal laboratory values during the course of their illness.

Table 3

Blood parameters	No. of patients
leucocytosis	18
thrombocytosis	13
anaemia	20
Raised CRP	25
Raised ESR	19
Urine analysis	Raised WBCs
Serum sodium	Decreased
Serum protein/albumin	Decreased
ECG changes	11
Echocardio pgraphy changes	9

All went on to manifest the classic form of the disease. With respect to treatment, 14 were started on antibiotic regimens and 16 were started on antipyretics other than aspirin prior to being diagnosed with KD. Upon diagnosis of KD, all patients were treated with intravenous immunoglobulin and aspirin. The list of complications appears in Table . 28 had complete resolution of their KD without any long-term sequelae. All otolaryngologic manifestations resolved without any long term sequelae. Two patients had persistent coronary artery aneurysms. There were no mortalities.

Table 4

COMPLICATIONS OF KD	NUMBER OF PATIENTS
Coronary artery aneurysm	2
Left ventricular cardiomegaly	1
Pneumonia	3
Pericardial effusion	1
Mitral regurgitation	0
Myocarditis	1
Adult respiratory distress syndrome	2
Congestive heart failure	0
Haemetemesis	1

IV. Discussion:

Kawasaki disease is an acute illness of young children and infants typically characterized by fever, conjunctivitis, oral and pharyngeal mucosal involvement, skin rash, extremity involvement and cervical lymphadenopathy. Oral manifestations seen are dry, erythematous, fissured lips that bleed easily, erythema of the oral and pharyngeal mucosa, Strawberry tongue with prominent papillae(5). As well, there are numerous head and neck manifestations of KD . The former five symptoms are present in approximately 90% of all patients(6), with cervical lymphadenopathy being the least common manifestation, occurring in 50–75% . The

presence of cervical lymphadenopathy was considerably more common in the classic versus the atypical form of KD. If the febrile child presents with cervical adenitis, KD should be considered, as this may represent an early stage of the disease.

In addition to the clinical symptoms, there are nonspecific laboratory findings in KD, including leucocytosis, thrombocytocis and an elevated erythrocyte sedimentation rate (7). Currently, there is no specific confirmatory laboratory test for KD. The current study revealed that although these elevated laboratory values may not be present initially, all patients with KD are likely to develop laboratory abnormalities during the course of their illness.

The etiology of KD remains uncertain. Various agents have been proposed as possible causes, including bacteria, viruses, carpet shampoos, pets and immunizations(8), although definitive evidence has not been established. The most commonly suspected cause is a superantigen such as toxic shock syndrome toxin-1 (9). The superantigens are proteins that bypass the normally highly antigen-specific presenting mechanisms, and instead bind to specific regions on the T-cell receptor, leading to widespread non-specific immune activation (10). This immune activation may cause excessive release of endogenous immune mediators, including interleukin-1, interleukin-6, interferon gamma and tumor necrosis factor leading to endothelial damage and subsequent development of signs and symptoms of the disease.

The vasculitis is most severe in medium-sized arteries but can also occur in veins, capillaries, small arterioles, and larger arteries.

In severely affected vessels, the media develops inflammation with necrosis of smooth muscle cells. The internal and external elastic laminae can split, leading to aneurysms. Four to 8 weeks after the onset of symptoms, inflammatory changes are less apparent and fibrous connective tissue begins to form within the vessel wall. The intima proliferates and thickens. The vessel wall eventually becomes narrowed or occluded owing to stenosis or a thrombus. Cardiovascular death usually occurs from a myocardial infarction secondary to thrombosis of a coronary aneurysm or from rupture of a large coronary aneurysm.

The vasculitis also affects other medium-sized vessels, including the renal, paraovarian, paratesticular, mesenteric, pancreatic, iliac, hepatic, splenic, and axillary arteries, resulting in systemic aneurysms. However, in certain arteries, particularly the coronary arteries, the inflammatory injury may be permanent resulting in arterial dilation(11)).

KD may be divided into three phases: acute, subacute and convalescent [12]. The acute phase lasts approximately 1 to 2 weeks with the subacute phase lasting from day 10 to 24. The convalescent phase lasts from 6 to 8 weeks in duration. Fever is typically the earliest sign, reaching as high as 40°C. The fever is typically unresponsive to antibiotics and antipyretics . Conjunctivitis, most often bulbar in nature, and oral mucosal involvement are usually commensurate with the fever. Oral changes include congestion of the oral and pharyngeal mucosa, strawberry tongue and fissuring of the lips. Extremity involvement including palmar erythema and edema begin between 5 and 10 days with desquamation beginning approximately 10–20 days after the onset of the illness. The exanthem of KD appears as a truncal eruption and may take a variety of morphologies, including morbilliform, urticarial, scarlatiniform or erythema multiforme-like lesions (13). Cervical lymphadenopathy, which is the least common of all the diagnostic criteria, may precede all other symptoms, particularly in atypical cases.

Head and neck manifestations of KD, including conjunctivitis, oral mucosal changes and less commonly cervical lymphadenopathy, are integral components of the diagnosis. In addition, there are numerous other symptoms of the disease, as reported in the literature , and as seen in the current study (Table 4). These include more common entities such as deep neck abscess, aseptic meningitis, torticollis, facial nerve palsy and rhinitis. Other symptoms such as airway obstruction, infected branchial cleft cyst, necrotizing pharyngitis and sensorineural hearing loss (SNHL) are rare.

The treatment of KD is fairly standard. IVIG dosed as single effusion at 2 g per kg and aspirin dosed at 100 mg per kg in divided doses is begun in the acute phase of the disease. Aspirin dose reduced to 3-5 mg/kg/day once fever and inflammation have subsided . Most patients experience defervescence within 24 h and by 48 h nearly 90% of patients are afebrile . Low-dose aspirin in the subacute phase issued to decrease platelet adhesiveness and further prevent cardiac sequelae. Pulse steroid therapy has also been shown to be effective (13).

The oral manifestations are typically self-limited and require only supportive management in most instances. Symptoms such as facial palsy may recover quickly or as long as several months after the acute illness. Permanent facial palsy is extremely rare . In the midst of treatment, patients should undergo electrocardiograms and echocardiograms in order to rule out cardiac involvement.

Echocardiograms should be performed routinely every year for the next 5 years in order to identify any possible cardiac complications that may have developed(14). This is especially significant as KD represents the number one cause of acquired heart disease in the pediatric patient

V. Conclusion

Kawasaki disease is an acute febrile illness of the pediatric population characterized by a systemic vasculitis. Both the classic and atypical forms of the disease have numerous common oral manifestations. Apart from them lymphadenopathy is more common in the typical form. The otolaryngologist should be aware of the possible symptoms of KD, and should include the disease in the differential diagnosis of any febrile illness refractory to antibiotic and antipyretic medications.

Early diagnosis and management with intravenous immunoglobulin and aspirin is essential in preventing the long-term sequelae of the disease, which commonly involve the coronary arteries. Other head and neck manifestations, which span the breadth of otolaryngology, are usually self limited.

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