Lower lip ulceration with purulent discharge in a child with cerebral palsy

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A 2-year-old male child, presented with the complaint of ulceration of lower lip for the last few days. There was history of grouped papulovesicular eruption on the inner side of lip that was associated with high grade, intermittent fever, restlessness and excessive dribbling of saliva. This was followed by ulceration of lower lip with mucopurulent and hemorrhagic discharge and shedding of the affected part of lip within 3 days. There was history of decreased intake and weight loss of 2kg in last 1 week. Patient was known case of cerebral palsy with history of delayed milestones.

On examination, patient was pale, febrile, with 6.5kg weight and 66cm height. Submental lymph nodes were enlarged, tender and mobile. Mucocutaneous examination revealed a 3cm ulcer involving the central part of lower lip with purulent discharge at the base and hemorrhagic crusting at the margins. CNS examination revealed generalized hypertonia, hyperreflexia, and spasticity and upgoing plantars. Rest of the systemic examination was normal.

Investigation showed microcytic hypochromic anemia (Hb 8.0 g/dl), raised ESR (70mm/1hr), low serum albumin (2.1g/dl) and total protein (4.3g/dl). Renal and liver function tests, serum electrolytes, blood sugar, viral markers (HBsAg) and serology (anti-HCV antibodies), and ultrasound abdomen were normal. Histopathology of the ulcer showed dense inflammatory infiltrate comprising of neutrophils, macrophages, lymphocytes and plasma cells. No granuloma and malignancy seen.

What is the diagnosis?

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Diagnosis

Noma (cancrum oris)

Discussion

Noma or cancrum oris, infectious in nature, is a devastating disease which involves the soft and hard tissues of the oral and para-oral structures. Although it is rare even in developing countries but can affect children, predominantly between 2-16 years of age. It begins as an ulcer of the mucous membrane, extending from within out, with edema of the face. It destroys the soft tissues and bones and almost always quickly fatal. Ankylosis, partial loss of jaws, lips and cheeks are among the complications along with impairment of speech and mastication. The physical and psychological disabilities of complications in a growing child are all too obvious.

Among the risk factors malnutrition, poverty, poor oral hygiene, bad sanitation, substandard accommodation, exposure to animal and human fecal material and infectious disease like measles, scarlet fever, tuberculosis, malignancy or immunodeficiency are responsible.

The term ‘noma’ was first described by Tourdes in 1848 which originates from the Greek verb ‘numein’ meaning to ‘devour’. It presents with some unusual presentations like noma neonatorum which is a rare gangrenous form affecting the oronasal tissues in medically compromised newborns at birth or during the first month of life, and noma pudendi, involving the anogenital area and results in its necrosis.

Although in majority of cases it develops in children under 10 years of age but because of the protective role of mother milk for its protein content and antibody level, infants and young children are less susceptible. Patient discussed here was 2 years of age with poor nutrition and was not on breast milk.

Noma is not a primary disease. It is usually preceded by illnesses such as measles, tuberculosis, leukemia and AIDS. The exact bacteriology is not known but acute necrotizing gingivitis, herpes labialis, Vincent's spirochete and fusiform bacillus in symbiosis are considered to be the actual cause of the condition. In our case noma occurred in a baby with cerebral palsy after having herpes simplex infection.

Noma has high mortality rate. Enwonwu et al. reported that without appropriate treatment the mortality rate may reach to 70-90%. Nutritional deficiencies are being considered to be one of the causative factor. This is supported by a study conducted on Nigerian children, showing significantly reduced plasma concentration of zinc (< 10.8 micromol/L), retinol (< 1.05 micromol/L), ascorbate (< 11 micromol/L), and the essential amino acids, with significantly increased free cortisol level in plasma and saliva compared with their healthy counterparts. In our case patient had a dramatic recovery with acyclovir, cefixime and metronidazole for 10 days along with zinc, vitamin B complex, vitamin D, vitamin C and iron supplements. Debridement of the ulcer was done and patient was referred to plastic surgery for reconstruction of lower lip. Noma could be a fatal and life threatening disease, which must be diagnosed early, and treatment should be started at the earliest to prevent from its complications.

In our patient diagnosis of noma was made with association of primary herpes simplex virus infection, chronic malnutrition and cerebral palsy. Patient was given acyclovir, cefixime and metronidazole for 10 days along with zinc, vitamin B complex, vitamin D, vitamin C and
iron supplements. High protein diet was encouraged. Debridement of the ulcer was carried out. Patient recovered and was referred to plastic surgery for reconstruction of lower lip.

References