Short Reports

Oral Manifestations of Letterer-Siwe Disease

In Letterer-Siwe disease, oral and dental pathology is an unusual occurrence. Failure to consider this condition in patients with gingival inflammation, necrosis, or premature eruption of teeth may lead to delay in diagnosis.

We present two cases with these features.

Case Reports

Case 1. This boy was aged 4 months when he developed sore, swollen gums with white necrotic areas. He was treated for 5 months for stomatitis. At age 6 months he had bilateral discharging otitis and a rash over his back; these both cleared over the next 2 months. He was seen at this hospital aged 10 months, thin, miserable, and underweight. The gingivae over upper and lower jaws were swollen and necrotic with premature eruption of teeth floating in a mass of granulomatous tissue (Fig.).

Fig.—Oral cavity showed hypertrophied and necrotic gingivae with eruption of teeth.

He had crusted erythematous scalp lesions. X-ray showed an osteolytic lesion involving the alveolar margin and the body of the right mandible, extending back to the ascending ramus. Chest X-ray showed fine diffuse opacities in both lung fields.

Biopsy showed gingival, periodontal, and bony infiltration with histiocytes and eosinophils compatible with the diagnosis of Letterer-Siwe disease.

Treatment was started with vinblastine 0·1 mg/kg per week and prednisone 2 mg/kg per day.

At follow-up 3 months later the gingivae appeared normal with no hypertrophy or ulceration, and his chest X-ray showed only very minimal scattered opacification.

Case 2. This infant at birth was noted to have 'septic spots' which improved with local treatment. At the age of 2 months he was referred to hospital with the combination of difficulty in feeding related to oral ulceration and the persistence of the rash present at birth. He had swollen, necrotic, ulcerated gums, and over the next few weeks developed premature eruption of teeth floating in a mass of soft granulomatous tissue. There were also widespread pityriasisiform-like skin lesions which left dermal scarring on healing. He later developed a discharging right ear.

Skull X-ray showed translucent areas in the parietal bones. X-ray of the mandible showed alveolar absorption around 1/1 and periostral reaction around the horizontal rami.

Gingival biopsy performed when aged 3 months showed grey-white necrotic tissue and histology confirmed the diagnosis of Letterer-Siwe disease: this was also confirmed on skin biopsy.

Treatment was started with vinblastine 0·1 mg/kg per week and prednisone 2 mg/kg per day. He rapidly deteriorated despite treatment, with the development of hepatosplenomegaly and obstructive jaundice, and died aged 5 months.

Discussion

Oral and dental manifestations of Letterer-Siwe disease are uncommon, especially as presenting features. Lucaya (1971) reported 5 cases with loose teeth out of 44 cases with histiocytosis X. In 600 cases with a diagnosis of reticuloendotheliosis, histiocytosis X, Hand-Schüller-Christian disease, Letterer-Siwe disease, eosinophilic granuloma, or xanthomatosis, Blevins et al. (1959) found only 12 cases that presented with oral and dental pathology. Their most frequent findings in 27 cases with oral and dental involvement were soreness, swelling, necrosis, and ulceration of the gingivae with a grey-white purulent exudate covering the involved regions. 85% of their cases had foul, fetid, 'Vincent like' breath. The most frequent findings on X-ray were the destruction of alveolar bone and the displacement of fully or partially formed teeth.

The oral manifestations of Letterer-Siwe disease are commonly misdiagnosed as Vincent's infection, aphthous or herpetic stomatitis, or as bony cysts, or tumours involving the oral cavity.

Treatment of Letterer-Siwe disease remains
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controversial and unsatisfactory though chemotherapy and steroids are the generally accepted methods used. Beier, Thatcher, and Lahey (1963) suggest the use of vincristine sulphate, and this was the treatment in our two cases together with prednisone, as used by Hertz and Hambrick (1968) and Chantler, Milner, and Winterborn (1971).

Summary

Two cases of Letterer-Siwe disease presenting with ulcerating, necrotic gingivae are described.

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References


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Treatment of Pneumothorax in the Newborn Infant

In the newborn infant, tension pneumothorax causes acute respiratory distress and unless air is aspirated without delay it can prove fatal. It is customary to treat this condition by inserting a catheter into the affected pleural cavity and attaching it to an underwater seal (Stahlman, 1967). Various kinds of tube have been used for this purpose, e.g. intravenous cannulae, feeding tubes, etc., but each has its own disadvantage; some do not have side holes, others are narrow and block or kink easily. A larger gauge catheter can only be introduced after making a stab wound through the entire thickness of the chest wall. Nor is the underwater seal without problems. If the tube draining the pleural air is sunk too deep under-water (>2 cm) air will not drain until a high pressure builds up in the pleural space and continuous suction has to be applied to the bottle in order to prevent this. If the bottle is accidently lifted above the level of the patient, its contents will promptly syphon into the chest and could be lethal. Finally the underwater drainage bottle system is cumbersome, a serious handicap in a busy neonatal unit where space is at a premium.

The purpose of this paper is to draw attention to the availability of equipment which obviates these difficulties.

It has been our practice during the past year to drain pneumothoraces using an Argyle Thoracic Trocar Catheter size Fr. 12* which is custom-made with terminal and side holes and an X-ray opaque sentinel line. The catheter comes ready mounted on a trocar to facilitate its introduction through a small skin incision over an intercostal space. The catheter should be introduced deep enough to ensure that the side hole is within the pleural space and it is then secured to the skin by a stitch; if the side hole lies under the skin surgical emphysema develops. To the catheter a Heimlich Chest Drain Valve† (Heimlich, 1965, 1968) is attached. This is a one-way flutter valve made of rubber and enclosed in a clear plastic tube open at both ends. The catheter fits directly onto the valve and no adaptor is necessary. This valve has proved very efficient and functions equally well in the presence of fluid that may drain from the pleural cavity.

We have achieved excellent results by using the combination described above and the following case report illustrates this.

Case History

A female infant weighing 3840 g was born to a primigravida by spontaneous vertex delivery at term, 27 hours after rupture of membranes. The mother had been pyrexial and was receiving antibiotics. The Apgar score at one minute was 2 and so the baby was intubated and intermittent positive pressure ventilation started. Despite ventilation for 35 minutes, spontaneous respirations did not start and the colour remained

*Cat. number AR-2068, Sherwood Medical Industries Ltd.
The Argyle Tube is manufactured by the Brunswick Corporation (UK) Ltd., Health and Science Division, Worthing, Sussex, England.
†Cat. number 3460, Bard-Parker Company Inc., Rutherford, New Jersey, U.S.A.
The Heimlich Chest Drainage Valve is produced by Becton, Dickinson UK Ltd., York House, Empire Way, Wembley, Middlesex.