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CASE REPORT

Unusual Radicular Cyst Formation Derived From Primary Teeth in Hyper Immunoglobulin E Syndrome

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Abstract

Introduction

Hyperimmunoglobulin E syndrome (HIES) is a rare genetic based multi-system disorder. It is characterized by high serum levels of IgE, pulmonary and recurrent skin infection except for these conditions abnormalities of the dentition, bones and connective tissue could be seen. The etiology of HIES is considered to be dominant-negative alterations in signal transducer and activator of transcription 3 (STAT3). STAT3 is integral to signal transduction for multiple cytokines. STAT3 is well expressed across tissue types.

Case Report

In this paper, we report a 7-year-old boy with HIES and a rare clinical manifestation regarding radicular cyst which is related to the deciduous tooth.

Conclusion

Chronic usage of antibiotics in the HIES patients can cause that sup-pressed serious lesion so that radicular cysts may be missed. Therefore, dentists should be alert on clinical and radiologic examination.

Keywords: Hyperimmunoglobulin E syndrome, HIES, Radicular cysts,

Introduction

Hyperimmunoglobulin E syndrome (HIES), also known as Job syndrome, is a rare genetic immune disorder which was first described by Davis in 1966.[1,2] Every HIES patients have increased IgE level is the 10 times greater than the upper limits of normal blood level (→2,000 IU/mL) and other common symptoms include cellulitis, staphylococcal dermatitis and folliculitis (cold abscesses), atopic eczema, recurrent pneumonia, constantly long bone fracture, osteopenia and pulmonary abscesses[1,3-6]

In patients with HIES, bacterial infections originate from Staphylococcus Group A and B, Haemophilus Influenza, Streptococci, and also other gram-negative organisms may exist. In addition, fungal infections caused by Candida albicans are most commonly seen.[3,7] Clinical findings include recurrent, severe pneumonia and furunculosis caused by Staphylococcus aureus, besides recurrent bronchitis usually occurs in infants and younger children. The abscess usually localizes in the neck, face, and scalp in that age group. Pruritic dermatitis frequently shows up around the hairline, behind the ears, and flexural areas of the body, and may turn to impetigo.[6] Sinusitis, chronic otitis media, and otitis externa are frequently seen along with "Coarse face," which consists of fleshy nasal tip and broad nasal bridge, prominent forehead, high-arched palate, irregularly proportioned cheeks and prognathism, in HIES patients (Figure 1A). Craniosynostosis case was reported infrequently. Besides recurrent fractures due to unexplained osteopenia is common in HIES patients. Oral symptoms of the HIES include retained primary teeth, cleft palate, and tongue.[1,4-6] Studies showed the failure of root resorption of primary teeth was 64%, 72%, and 75% respectively.[3,8,9]

The etiology of HIES is considered to be negative alterations in signal transducer and activator of transcription 3 (STAT3). STAT3 is integral to signal for multiple cytokines. STAT3 is well expressed across tissue types.[1] Recurrent infections and connective tissue abnormalities are linked with STAT3, which was identified in 2007.[1] T-cell differentiation-dependent processes of STAT3 lead to the failure by the alteration in

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STAT3, and this is the may be explanation of the tendency to infection in patients with HIES. Also, serum eosinophilia is laboratory finding in patients with HIES.[1,7]

Radicular cysts are known as the most common odontogenic cystic lesions of the jaws. Radicular cysts are believed to originate from proliferation of epithelial residues of Malassez as a result of inflammatory activities. Usually it is not asymptomatic until it become well developed.[10] The radicular cyst is seen mostly in the third decade of life.[11] However, studies showed that these cysts are rare in pediatric population.[12]

This report presents a patient with HIES and a rare clinical manifestation regarding radicular cyst which is related to the deciduous tooth.

Case Report

A 7-years-old boy with the diagnosis of HIES, was admitted to Kirikkale University, Faculty of Dentistry, Clinic of OMFS, regarding the prolonged retention of his mandibular incisal primary teeth. Informed consent was taken from patient's parent. The patient's medical history consisted of recurrent pulmonary infection, skin abscesses, otitis media and elevated serum IgE levels (Figure 1B). Intraoral examination revealed the presence of various dental caries and median rhomboid glossitis-like lesion distributed over less than one-third of the dorsal tongue (Figure 1C). Also, the presence of a cyst in the right mandibular corpus related to primary second molar was detected in the radiographic examination. It was assumed that the radicular cyst's size is 20mm×17mm (Figure 1D).

Ceftriaxone was admitted a day before the operation and continued five days after operation 1000 mg intravenously a day. Also, intravenous (i.v) immunoglobin (IG) was admitted two days before the operation 1000 mg for each day by pediatric staff due to consultation. The cyst and related primary teeth were removed by enucleation under general anesthesia. The removal of the cyst was performed with caution to avoid damaging neighboring vital tissues, especially the second premolar tooth germ. (Figure 1E)

The histopathologic evaluation was consistent with the radiographic appearance of a radicular cyst. The histology of the cyst showed a fibrotic cyst wall which contained proliferating strands of odontogenic epithelium and a chronic inflammatory infiltration that is rich in plasma cells. (Figure 1F)

The healing completed uneventfully, and no complications have occurred within 3 months of follow-up.

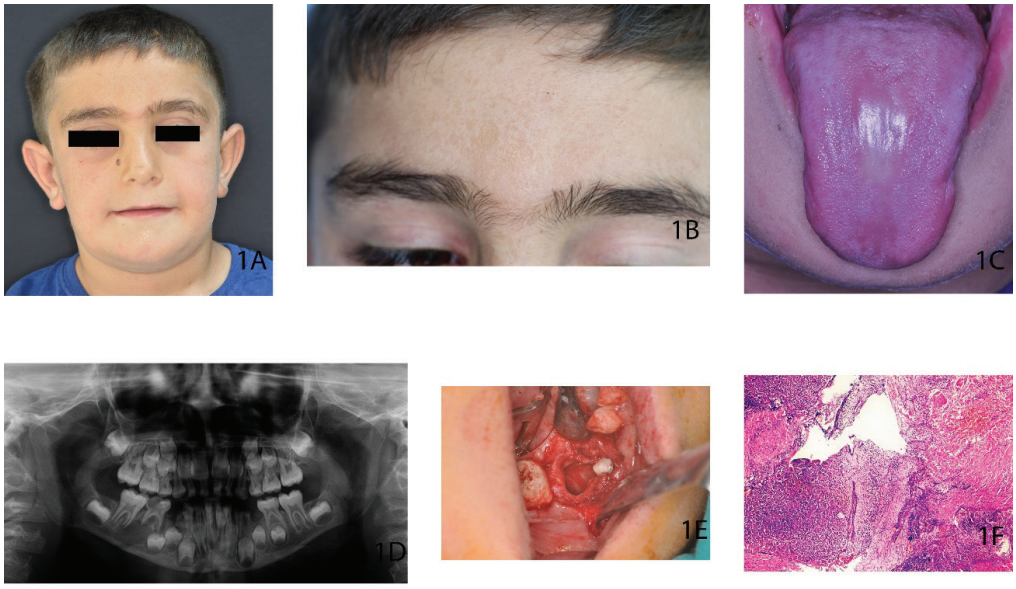

Figure 1A: HIES patient's characteristic facial appearance
Figure 1B: Patient's scars caused by the skin infections
Figure 1C: Median rhomboid glossitis-like lesion in the middle of the dorsal tongue
Figure 1D: Patient's panoramic radiograph
Figure 1E: Intra-operative vision after the removal of cyst.(there is a primary teeth germ by the cyst cavity)
Figure 1F: Chronically inflamed fibrotic wall of the radicular cyst (H-E; X100)

Discussion

HIES is characterized by high serum levels of IgE, recurrent pulmonary and skin infections, typically caused by Staphylococcus aureus and Candida albicans. Sinusitis, otitis externa and chronic otitis media are also common diseases.[4] Intermittent or long-term anti-staphylococcal antibiotics, cyclosporine, ascorbic acid and cimetidine, high-dose IVIG and interferon gamma have been used in HIES.[4] In this case, the patient has been treated with the anti-staphylococcal antibiotic, cyclosporine, and high-dose IVIG because of the recurrent skin and pulmonary infections and chronic otitis media. The patient had increased IgE level (→2,000 IU/mL).

Delayed eruption of the persistent teeth due to prolonged retention of the deciduous teeth are common oral signs of HIES patients.[8] Oral candidiasis is a common finding in the HIES patients. Also, the presence of asymptomatic midline tongue deficiency in two affected siblings has been reported.[8] HIES patients have lesions of the dorsal tongue and hard palate about 55% and 60%.[9] Our patient had prolonged retention of his mandibular incisal primary teeth and rhomboid glossitis-like lesion distributed over less than one-third of the dorsal tongue. Only 0.5-1% of all radicular cysts are associated with primary teeth.[11] This rare clinical manifestation conjunction with the primary mandibular molars (%67), respectively maxillary molars (%17), and anterior teeth.[13]

Long-term use of penicillinase-resistant antibiotics to prevent staphylococcal infection has the most successful treatment chance in HIES patients.[14] Because other antibiotics or antifungal agents are required for specific infections; they should be added, also using either trimethoprim-sulfamethoxazole or dicloxacillin with prophylaxis may prevent infections.[15] Our patient has been using IVIG regularly for 5 years and especially for his recurrent pulmonary infection that he had had recently. For the chronic otitis media treatment, he was medicated with anti-staphylococcal antibiotics and cyclosporine, so in our opinion; an acute infection was prevented unlikely the patients who are not under their control.[16] On the other hand; dentists should be aware of that an acute infection can develop in HIES patients. In the instance of infection, treatment plan should be early and aggressive, with intravenous antibiotics administration, surgical incision and drainage.[16]

Chronic usage of antibiotics in the HIES patients can cause that suppressed serious lesion so that radicular cysts may be missed. Therefore, dentists should be alert on clinical and radiologic examination.

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M. Ercüment ÖNDER : Performing the operation, writing the article
Berkan ALTAY: follow-up of the patient
Fethi ATIL: follow-up of the patient
Umut TEKİN: Performing the operation
İ. Doruk KOÇYİĞİT: writing the article
Özkan ÖZGÜL: writing the article

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Bu vaka, 12. AÇBİD Uluslararası Kongresinde "Poster" olarak sunulmuştur