A SELECTIVE IgA DEFICIENCY IN A BOY WHO PRESENTED RECURRENT PAROTITIS

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Recurrent parotitis is a non-obstructive, non-suppurative inflammatory disease which is characterized by unilateral or bilateral parotid gland swelling attacks. It is also known as juvenile recurrent parotitis. Although the etiology is unknown, congenital malformations of the ductus, genetic predisposition, infections, allergies, autoimmune diseases, and some immune deficiencies are blamed. Here, we present a case report of recurrent parotitis with selective immunoglobulin A deficiency in a six-year-old boy. The patient was presented to us with a new episode of swelling of left parotid region. In the last 2 years, the patient suffered from recurrent parotitis which lasted for approximately 5 days in ten individual episodes.

Keywords: selective IgA deficiency, recurrent parotitis, childhood

Introduction

Recurrent parotitis is described as recurrent parotid inflammation, usually associated with non-obstructive sialiectasis of peripheral ducts of the parotid with periductal lymphocytic infiltration of the parotid gland [1]. This disease is characterized by recurring episodes of unilateral or bilateral swelling and/or pain in the parotid gland, usually accompanied by fever and malaise which last from 1 day to 2 weeks. Recurrent parotitis usually starts early in life with a peak at 3–6 years of age and occurs more frequently in boys [2, 3]. In most cases, recurrent parotitis resolves spontaneously after puberty without leaving behind any permanent sequel [4]. Recurrent parotitis in children has been observed in correlation to human immunodeficiency virus (HIV) disease, various forms of phagocytic dysfunction, and rarely in cases with deficiencies of humoral immunity. Some cases with isolated IgG3 subclass deficiency, common variable immunodeficiency (CVID) or selective IgA deficiency have also been reported [5]. Conservative approaches such as analgesic, warmth, and massage are usually preferred in exacerbation treatment of disease [6]. In this case report, we present a six-year-old boy with selective IgA deficiency who had ten episodes of recurrent parotid swelling. The aim of this report is to describe a case of selective IgA deficiency with recurrent parotitis, in the absence of an autoimmune disorder.

Case report

A six-year-old boy presented with swelling of his left parotid gland and pain, especially during mastication. He had experienced ten such episodes lasting 10–15 days each in the previous 2 years. Physical examination revealed a swollen left parotid gland with local tenderness and erythema of the overlying skin. The rest of the examination was unremarkable. There was no history of dry mouth, dry eyes, joint pain, swelling, or skin rash, which could have been suggestive of autoimmune disease. There was also no family history of recurrent parotitis. The boy was afebrile, and there was no erythema around the stenson duct, whereas mucous discharge was seen when pressure was applied to the gland. The patient's blood count and erythrocyte sedimentation rate were within normal limits. The child's HIV serology was negative, and his serum amylase and lipase were normal. Levels of complement (C3, C4), serum antinuclear antibody (ANA), and rheumatoid factor (RF) were within normal limits. Ultrasonography of bilateral parotid glands showed heterogeneous texture, hypoechogenic areas and increased intraparotid lymph nodes in both parotid glands as well as increased volume in the left gland. There were no calculi nor duct dilatation. Antibodies including anti-Sjögren's syndrome A (SSA) and anti-Sjögren's syndrome B (SSB) for Sjögren's syndrome were negative. Immunological evaluation revealed IgA level

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Table 1. The laboratory findings of patient

	Patient results	Normal values
Sedimentation (mm/h)	15	10–20
C3:	130	90–210 mg/dl
C4:	20	14–40 mg/dl
Amylase	50	30–130 u/l
Lipase	20	0–60 u/l
ANA	Negative	
RF	Negative	
Anti-SSA	Negative	
Anti-SSB	Negative	
HIV serology	Negative	
Phagotest	Normal	
White blood cell (/mm ³)	5400	3700–11100 [14]
Lymphocytes (/mm ³)	2400	1500–7600 [14]
IgA (mg/dl)	5.1	36–268 [15]
IgG (mg/dl)	1260	527–1590 [15]
IgM (mg/dl)	56	30–220 [15]
IgE (iu/ml)	18	0–200

of less than 7 mg/dl (normal range: 36–268). Remaining immunoglobulin subgroups, including IgE, IgM, IgG, and IgG, were within normal limits (*Table 1*). Phagotest for disorders of the phagocytic system was normal. A culture from mucous discharge obtained from orifice of the left parotid duct revealed normal oral flora.

After examination, analgesic (paracetamol) and massage were recommended to the patient. Local inflammatory signs disappeared within 2 days, and swelling was completely reduced within 1 week.

Discussion

IgA deficiency is defined as decreased or absent level of serum IgA in the presence of normal serum levels of IgG and IgM in a patient older than 4 years of age. Selective IgA deficiency is an isolated absence or near absence (<7 mg/dl) of serum and secretory IgA, with a prevalence of 1/143 to 1/18,500 in different populations [7]. IgA deficiency is sometimes associated with IgG2 and IgG4 deficiency [7]. Recurrent parotitis in children has occasionally been associated with immunodeficiency. Marsman and Sukhai [8] described a five-year-old girl with recurrent parotitis and isolated IgG3 deficiency. In the literature, recurrent parotitis with selective IgA deficiency has been published from different countries. Shkalim et al. [9] reported a nine-year-old boy with recurrent parotitis and IgA deficiency. In another study, Fazekas et al. [5] identified five of 17 patients with recurrent parotitis as having selective IgA deficiency. In another report, Conley et al. [10] described a two-year-old boy and a five-year-old girl with common variable immunodeficiency (CVID) who

had episodes of recurrent parotitis. Primary Sjögren's syndrome (PSS) is an autoimmune disorder characterized by dryness of the eyes (keratoconjunctivitis sicca or xerophthalmia) and mouth (xerostomia) and associated with autoantibodies to the nuclear antigens SS-A/Ro and SS-B/La. A combination of positive ANA, RF, SS-A, and SS-B; hypergammaglobulinemia; elevated amylase (parotid or pancreatic); and elevated erythrocyte sedimentation rate (ESR) is suspicious for PSS [11]. Some children with recurrent parotitis have autoantibodies and symptoms that suggest PSS [5]. However, our patient's autoantibodies and IgG (including IgG1, G2, G3, G4), IgM, and IgE levels were normal for immune deficiencies, PSS, and CVID.

In normal individuals, 80–90% of the B lymphocytes in the parotid gland produce IgA [9]. B lymphocytes are lacking in IgA-deficient persons. Because IgA is the most common immune deficiency, a coincidental association between IgA deficiency and recurrent parotitis is possible. However, since IgA is also the major immunoglobulin secreted into the mucosal surfaces (more than 95% of secretory IgA is produced locally), including the mucosa of the respiratory and gastrointestinal tracts, and into the saliva [5, 7], and selective IgA deficiency is associated with various inflammatory disorders, one can speculate that lacking IgA in the parotid gland may be associated with an inability to completely clear inflammatory stimuli. This in turn may subsequently lower the threshold of developing inflammatory pathology. Thus, in some cases, it can be speculated which IgA deficiency may play a role in the pathophysiology of recurrent inflammation of the salivary gland. Plenty of IgA patients are symptomless, and recurrent parotitis prognosis is good as mentioned above, but we know that some patients with IgA deficiency have autoimmune dis-

ease, malignancy, and asthma in their course [7]. Thus, the follow-up of these patients is important for these problems.

The diagnosis of recurrent parotitis is made on a clinical basis and can often be confirmed by ultrasound, showing the classical features of sialiectasis. These consist of multiple hypoechoic areas that correspond to the punctate pools seen on sialogram. Sialograms typically show widespread, multiple, sharply demarcated, round areas (1–3 mm) of contrast, which persist in delayed films. Histologically, periductal lymphocytic infiltration is seen [12].

Therapeutic management is controversial in recurrent parotitis. Ductal dilation with sialendoscopy and hydrocortisone injection may reduce swelling but will not prevent recurrences. Treatment of the acute episode aims with analgesic to deliver relief of symptoms and to prevent damage to the gland parenchyma. The use of antibiotics is controversial, and preventing dehydration is very important. Since most of the cases resolve by puberty, reassurance of the family about the benign course of disease is enough [6, 11–13].

In conclusion, recurrent parotitis is an uncommon condition in pediatric population. This is the first case report of recurrent parotitis with selective IgA deficiency from Turkey. Some etiologic factors such as genetic inheritance, allergy, viral infection, and immunodeficiency can be related with recurrent parotitis. Thus, if the etiology of recurrent parotitis is not found, we recommended that serum IgA and other immunoglobulins levels should be measured in pediatric patients.

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