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Case Report ALLERGY & IMMUNOLOGY

Successful treatment using agalsidase alfa of a patient with Fabry disease who had anaphylaxis after agalsidase beta: A case report

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ABSTRACT

Fabry disease is a rare genetic disease caused by a deficiency of α-galactosidase A gene (α-Gal A). Two intravenous enzymes administered every two weeks, agalsidase alfa and beta can slow disease progression and increase survival if administered early, before organ damage occurs. In this case report, we present a patient with a history of anaphylaxis to agalsidase beta who was successfully treated with agalsidase alfa.

Keywords: Agalsidase alfa; agalsidase beta; anaphylaxis; drug allergy; Fabry disease.

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Fabry disease is a rare genetic disease caused by a deficiency of α-galactosidase A gene (α-Gal A). A decrease in α-Gal A activity can cause increase in globotriaosylceramide in various organs and tissues such as vascular endothelium, smooth muscle cells and cardiomyocytes, leading to multiple organ failure and death [1, 2].

Two intravenous enzymes administered every two weeks, agalsidase alfa and beta, can slow disease progression and increase survival if they are administered early before organ damage [3]. Sensitization to agalsidase beta has been demonstrated by skin prick test (SPT) or allergen-specific immunoglobulin E (sIgE) [4]. In this report, we present a patient with a history of anaphylaxis to agalsidase beta who was successfully treated with agalsidase alfa.

CASE REPORT

A 24-year-old male patient who had been receiving agalsidase beta at 70 mg/2 weeks for five years due to Fabry disease was referred to our allergy clinic due to the development of an allergic reaction during the last three infusions. Shortness of breath, itching, swelling all over his body, and hypotension (70/40 mmHg) developed 20 minutes into the drug infusion. During the last two infusions, a similar reaction occurred despite the administration of premedication (pheniramine 22.7 and methylprednisolone 40 mg intravenously). Pheniramine 22.7 mg (intravenous), methylprednisolone 40 mg (intravenous), and adrenaline (0.5 mg, 1:1000 intramuscular) were administered to the patient each time to treat the allergic reaction. At the time of admission to our allergy clinic, baseline tryptase level of the patient was 4.4 (normal: 0–11.4 ng/mL). The other laboratory parameters were normal. Six weeks after the last allergic reaction, intradermal test (IDT) and SPT were performed with agalsidase beta. The SPT (undiluted solution) was negative and IDT (1/1000 dilution, 5×5 mm wheal and erythema) was positive with agalsidase beta. SPT (undi-

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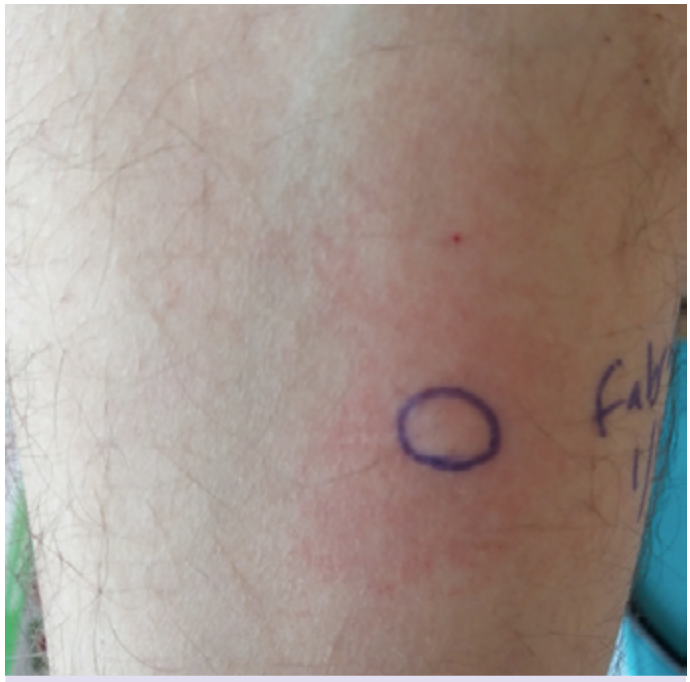

FIGURE 1. Positive IDT with agalsidase beta (1/1000 dilution, 5×5 mm wheal and erythema).

luted solution) and IDT (1/1000, 1/100 and 1/10 dilution) were also performed with agalsidase alfa. The SPT and IDT were negative with agalsidase alfa. The positive IDT with asalsidase beta is shown in Figure 1. A drug provocation test was performed with agalsidase alfa. A total of 14 mg of agalsidase alfa was administered via slow intravenous infusion over two hours. No allergic reaction was observed during the infusion or during the six-hour surveillance after it. The patient has continued to receive agalsidase alfa therapy without any issues. Informed patient consent was obtained for this case report.

DISCUSSION

In this case report, we present a patient with Fabry disease who had a history of anaphylaxis to agalsidase beta and showed tolerance to agalsidase alfa. To the best of our knowledge, this patient is the first in the literature to have a history of anaphylaxis to agalsidase beta and tolerance to agalsidase alfa.

Allergic reactions to agalsidase beta are rare and have been reported in the literature as case reports. DuBuske et al. [5] successfully desensitized a patient who developed anaphylaxis to agalsidase beta using omalizumab premedication. Talreja et al. [6] reported that they successfully performed desensitization to agalsidase beta in a patient who developed anaphylaxis after receiving it. Aydin et al. [7] reported that they successfully desensitized two brothers with Fabry disease who had a history of allergic reactions to agalsidase alfa. Tanaka et al. [8] successfully treated a Fabry patient with agalsidase alfa who developed purulent eczema and fever after administration of agalsidase beta. In the present case report, contrary to the case reports in the literature, we did not perform desensitization in a patient who developed anaphylaxis to agalsidase beta. We performed SPT and IDT with an alternative drug, agalsidase alfa. Because the SPT and IDT were negative, we performed a drug challenge test with agalsidase alfa. We did not observe any allergic reaction at the end of the drug challenge test with agalsidase alfa. Further application of agalsidase alfa treatment has not caused any problems for the patient.

In conclusion, in patients with a history of allergic reactions to agalsidase beta, SPT, IDT, and drug provocation tests should be performed with agalsidase alfa as an alternative drug before considering a desensitization protocol.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Conflict of Interest: No conflict of interest was declared by the author.

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