Anomalous Left Coronary Artery from Pulmonary Artery in an Infant with Sweet Syndrome: First Case Report

Sweet Sendromlu Bir Bebekte Pulmoner Arterden Çıkan Anormal Sol Koroner Arter: İlk Olgu Sunumu

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ABSTRACT Sweet's syndrome, or acute febrile neutrophilic dermatosis, is a rare multisystem syndrome that was first described by Sweet in 1964. The syndrome is characterized by tender erythematous skin plaques, fever, neutrophilic leukocytosis, dermal infiltrate of mature neutrophils and rapid response to corticosteroids. Moreover, this entity might be associated with malignancies, Fanconi anemia, periosteomyelitis, erythema nodosum, inflammatory bowel disease, and rarely cardiovascular involvement. The prognosis of this syndrome varies in accordance with underlying conditions and it especially becomes highly fatal in cardiac involvement. Cardiac manifestations of Sweet's syndrome are usually associated with cutis laxa that accompanies. Anomalous left coronary artery from pulmonary artery (ALCAPA syndrome) is another rare cardiac anomaly. In this report, a child with the association of two rare syndromes, the Sweet's and ALCAPA, was presented due to its unique presence in the literature.

Key Words: Sweet syndrome; child

ÖZET Sweet Sendromu veya diğer bilinen adıyla akut febril nötrofilik dermatozis nadir bir multisistem hastalık olup ilk defa 1964 yılında Sweet tarafından tanımlanmıştır. Bu hastalık eritematöz cilt plakları, ateş, nötrofilik lökositoz, dermisin matür nötrofille infiltrasyonu ve kortikosteroide hızlı yanıtı ile karakterizedir. Hastalık malignite, Fanconi anemisi, periosteomiyelit, inflamatuar barsak hastalıkları ve nadiren kardiyak tutulumla ilişkili olabilmektedir. Bu sendromun prognozu altta yatan hastalığa bağlı olmakla birlikte özellikle kardiyak tutulumlarda daha fatal olmaktadır. Sweet sendromunda kardiyak tutulum genellikle cutis laxa ile birlikte görülmektedir. Sol koroner arterin pulmoner arterden çıkış anomalisi (ALCAPA sendromu) de nadir görülen diğer bir kardiyak sendromdur. Biz burada çocuklarda nadir görülen iki sendromu, Sweet sendromu ve ALCAPA sendromu birlikteliğini, daha önce literatürde olmadığından ilk vaka olarak sunduk.

Anahtar Kelimeler: Sweet sendromu; çocuk

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Sweet's syndrome, or acute febrile neutrophilic dermatosis, is a rare multisystem syndrome that was first described by Sweet in 1964.[1]The syndrome is characterized by tender erythematous skin plaques, fever, neutrophilic leukocytosis, dermal infiltrate of mature neutrophils and rapid response to corticosteroids.[1] Moreover, this entity might be associated with malignancies, Fanconi anemia, periosteomyelitis, erythema nodosum, inflammatory bowel disease, and rarely cardiovascular involvement.

[2] The prognosis of this syndrome varies in accordance with underlying conditions and it especially becomes highly fatal in cardiac involvement.[3] Cardiac manifestations of Sweet's syndrome are usually associated with cutis laxa that accompanies.[2] Anomalous left coronary artery from pulmonary artery (ALCAPA syndrome) is another rare cardiac anomaly.[4] In this report, a child with the association of two rare syndromes, the Sweet's and ALCAPA, was presented due to its unique presence in the literature.

CASE REPORT

A previously healthy 45-day-old girl presented with fever, restlessness, and multiple erythematous rash. There was no history of drug or vaccine. Family history revealed first-degree consanguinity. A sibling with similar clinical picture had died when he was 40 days old.

Physical examination revealed a markedly distressed infant with the temperature of 38.3°C, and multiple erythematous plaques (0.4-4 cm in diameter) that were asymmetric and well- demarcated on her extremities and face (Figure 1). Cardiac examination was normal except mild systolic murmur.

Her blood count revealed leukocytosis ($14.8x10^9$/L), and thrombocytosis ($801x10^9$/L). Other blood tests, including liver function tests, electrolytes, sedimentation rate and CRP were unremarkable. Viral serology, urinalysis and chest radiograph were normal. Bone scan and bone marrow aspiration results were also in normal limits. Punch biopsy of a lesion revealed an intense dermal infiltrate of neutrophils without evidence of vasculitis that reminds Sweet's syndrome (Figure 2). The color Doppler echocardiogram showed some mosaic flows in the right ventricle that reminds the coronary anomaly. The patient was transferred to the cardiac catheterization laboratory for further evaluation. Aortic root angiography showed an anomalous left coronary artery arising from the pulmonary artery (ALCAPA) with retrograde filling through collaterals from an en-

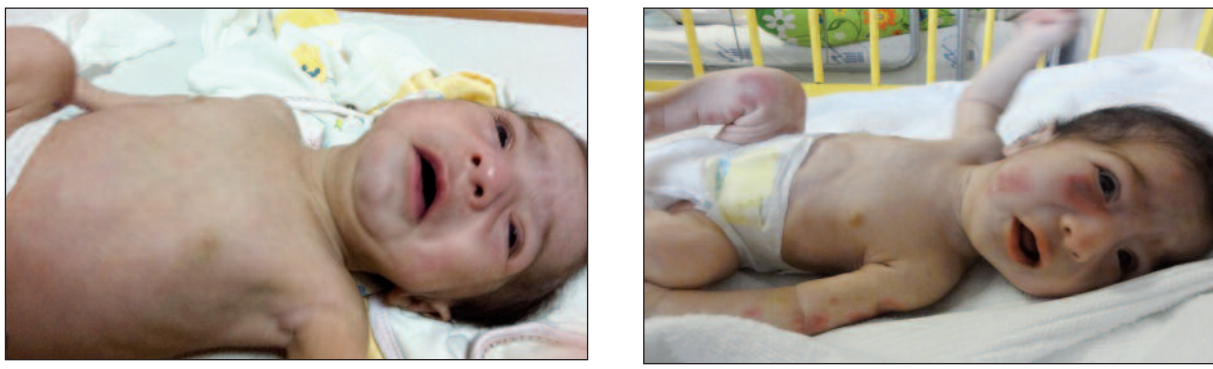

FIGURE 1a: Multiple erythematous plaques on the skin pre and after treatment.
(See color figure at http://www.turkiyeklinikleri.com/journal/pediatri-dergisi/1300-0381/)

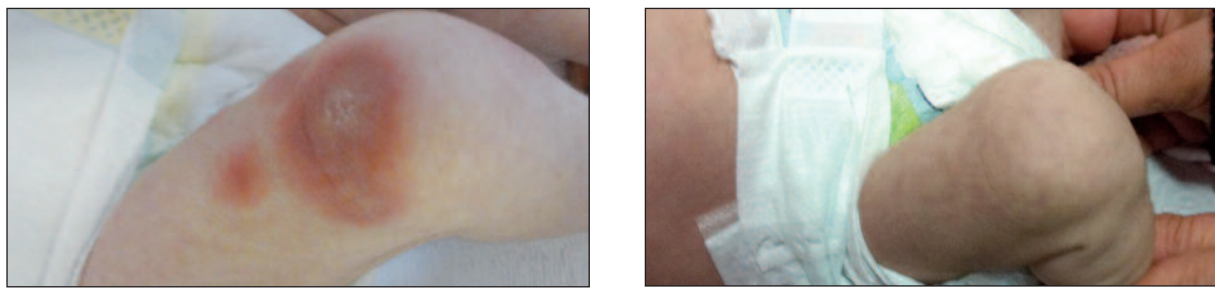

FIGURE 1b: Skin lesions recovery without scarring after methyl prednisolon treatment.
(See color figure at http://www.turkiyeklinikleri.com/journal/pediatri-dergisi/1300-0381/)

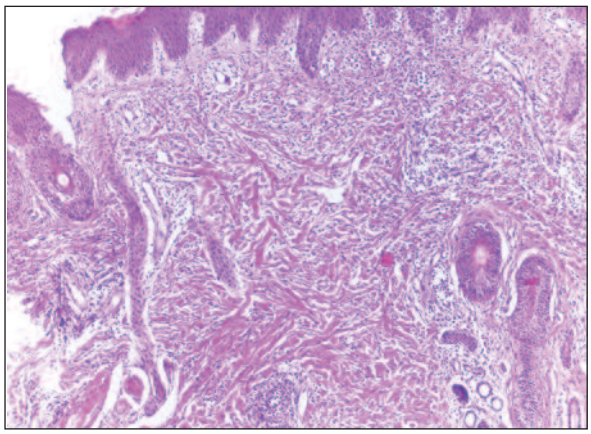

FIGURE 2: The histopathological examination of dermis shows massive neutrophilic infiltrate without vasculitis (HE, x100).
(See color figure at http://www.turkiyeklinikleri.com/journal/pediatri-dergisi/1300-0381/)

larged right coronary artery (Figure 3). Permission was given from the family for academic publication.

Prednisone therapy was started 2mg/kg/day and dramatic improvement was achieved. She was operated for ALCAPA syndrome and at present the patient is 8-month-old and she is well though she had relapse of the lesions.

DISCUSSION

Sweet's syndrome is an inflammatory disease of unknown origin that is more rare in children comparing to adults. Only 5-8% of the patients were in childhood period. Only 66 children were reported until 2009 in the literature.[1,5]

The cardiac involvement in Sweet's syndrome is rare both for adults and children; 40% of it is fatal. Myocardial infarction, aortic and coronary artery dilation, valvular disease and pericarditis are some cardiac complications, which are reported, in adult Sweet's.[1,5-7] Cardiac problems in children with Sweet's syndrome, such as mitral valve perforation and aneurysm of the sinus valsalva, are frequently associated with cutis laxa.[2,8] Muster et al. reported unexplained fatal aortic and coronary involvement in two patients with cutis laxa.[9] In the literature, familial Sweet syndrome was reported in two siblings and one of them had died from pulmonary hypertension and cardiac failure when he was 4 years old.[3] Due to the family history of another sibling with similar clinical picture (fever and rashes), it is thought that this patient might have a familial Sweet syndrome. Unfortunately he died when he was 40 days old without autopsy. Anomalous origin of the left coronary artery from the pulmonary ar-

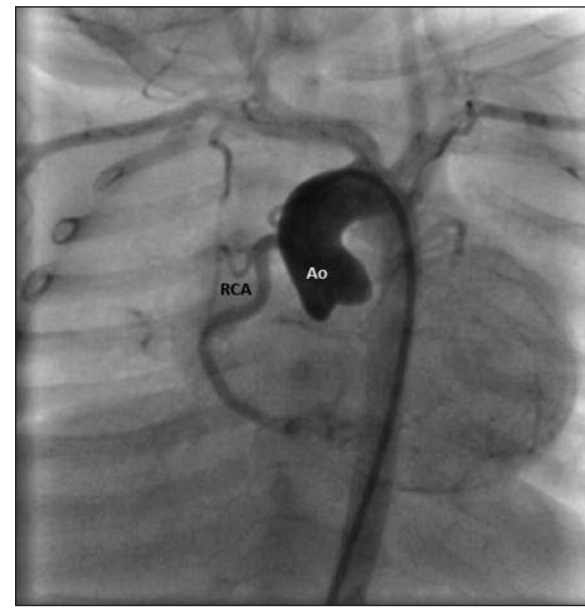

FIGURE 3A: The early phase of the injection has demonstrated enlarged right coronary artery.
Ao: Aort; RCA: Right coronary artery.

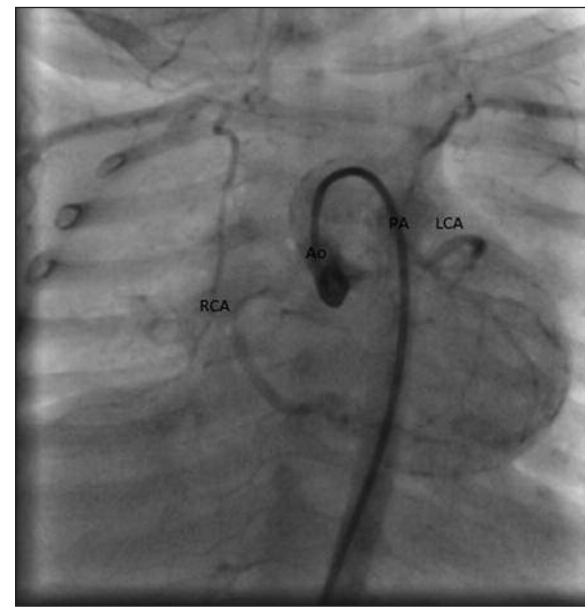

FIGURE 3B: The late phase of the coronary angiography showed an anomalous left coronary artery arising from the pulmonary artery with retrograde filling through collaterals.
LCA: Left coronary artery; RCA: Right coronary artery; Ao: Aort.

tery (ALCAPA) syndrome is another rare congenital cardiac anomaly occurring at an incidence of 1 in 300 000 live births.[10] Approximately 90% of the patients die within the first year of life because of myocardial infarction and congestive heart failure.[4] This is the first report of Sweet and ALCAPA syndrome association in the literature. Furthermore, especially cardiac involvement in children with Sweet syndrome might be highly fatal and silent.[3,6,8] For this reason, early careful physical examination, echocardiographic study and angiography if necessary might be lifesaving for these patients.

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