

Ocular manifestations of SAPHO syndrome

Ayaka Takeda, Satoko Yui, Junko Hori

Nippon Medical School

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Background: Synovitis–acne–pustulosis–hyperostosis–osteitis (SAPHO) is an acronym for various osteoarticular and dermatological manifestations that can appear in the same patient. Ocular presentation associated with SAPHO syndrome is relatively uncommon. We report ocular manifestations in four cases that developed ocular inflammation associated with SAPHO syndrome.

Patients & Methods: Three female and a male (age 46-63) developed painful red eyes. Slit-lamp examinations revealed bilateral anterior diffuse scleritis in the three female patients and unilateral acute anterior uveitis (non-granulomatous iridocyclitis) in the male patient. All cases were diagnosed with SAPHO syndrome by rheumatologists after excluding other causative diseases.

Results: All cases were treated with topical steroid and/or tacrolimus eye drops, oral celecoxib, steroid, and methotrexate. Three cases (two female and a male) showed poor response to these treatments and resulted prolonged ocular inflammation. Treatment with anti-TNF mAb (Infliximab or Adalimumab) were effective for these three cases to improve ocular findings.

Case 1:

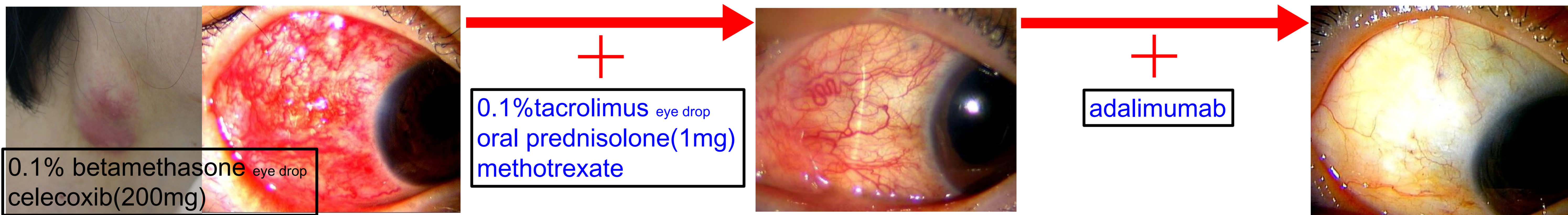
Patient: 63 year-old female

Chief complaint: bilateral red eyes, right side trachelophyma

History of illness: Bilateral red eyes appeared in 2011, treated with 0.1% betamethasone, celecoxib(200mg) or loxoprofen(60mg), but no improved. She was referred to our hospital in Sep, 2014.

Clinical findings: Vd=0.6(1.2p×S-1.75:C-0.50Ax70°), Vs=0.3(1.0p×S-2.00:C-1.50Ax80°), Tod=17mmHg, Tos=18mmHg

Clinical course: bilateral diffuse scleritis(+), lens clear, fundus n.p., painful trachelophyma(+)



During this process, she had palmoplantar pustulosis and back pain as arthritis, rheumatologists diagnosed SAPHO syndrome.

Case 2:

Patient: 48 year-old-female

Chief complain: bilateral painful red eyes, visual acuity decline

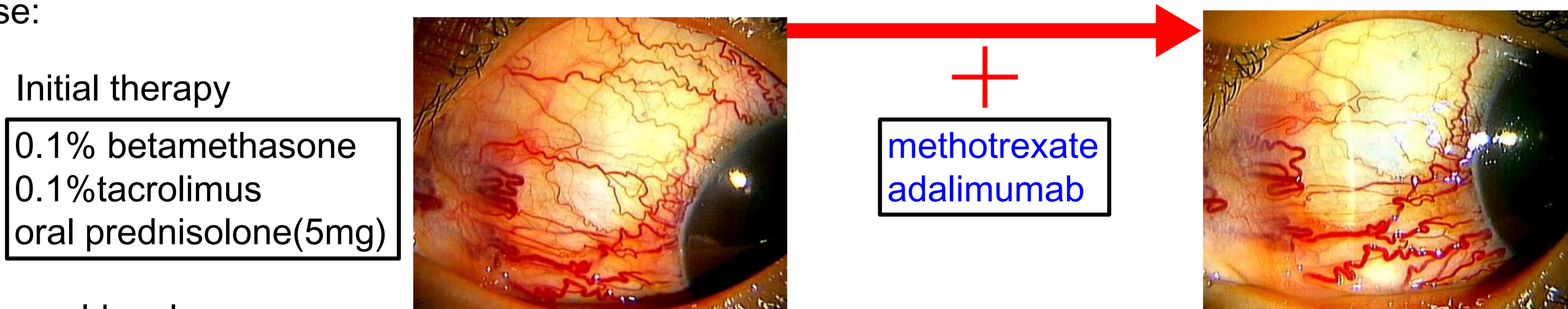
Past history: SAPHO syndrome, Takayasu's arteritis

History of illness: The patient got a check-up at our hospital with chief complain for 2 weeks in Dec 2015.

Clinical findings: Vd=0.9(1.2×S+0.00:C-0.25Ax115°), Vs=0.8p(1.2×S+0.00:C-0.50Ax50°), Tod=15mmHg, Tos=19.3mmHg

bilateral diffuse scleritis(+), lens clear, retinal hemorrhage(+)

Clinical course:



Case 3:

Patient: 53 year-old male

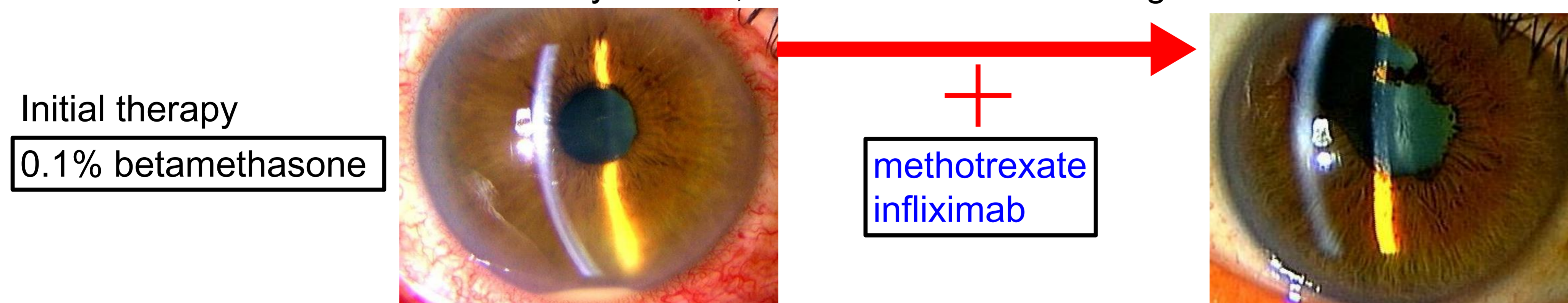
Chief complain: frequent refractory of uveitis

Past history: suspect of acute anterior uveitis (AAU)

Clinical findings: Vd=(1.2×S-1.00:C-1.00Ax90°) Vs=(1.0p×S-3.50:C-2.00Ax90°), Tod=13.7mmHg, Tos=13.0mmHg

bilateral non-granulomatous iritis(+), pus in the anterior chamber(-), fibrin(-), lens clear, fundus n.p.

Clinical course: He had several ocular inflammatory attacks, and his initial clinical diagnosis was AAU.



He developed psoriasis, swelling and pain of sternoclavicular joint, that was diagnosed as SAPHO syndrome by rheumatologists.

Case 4:

Patient: 67 year-old female

Chief complain: refractory scleritis(used 0.1%betamethasone), back pain

Past history: pustulosis palmoplantaris

Clinical findings: Vd=0.9p(1.0×S+0.25:C-1.00Ax95°)

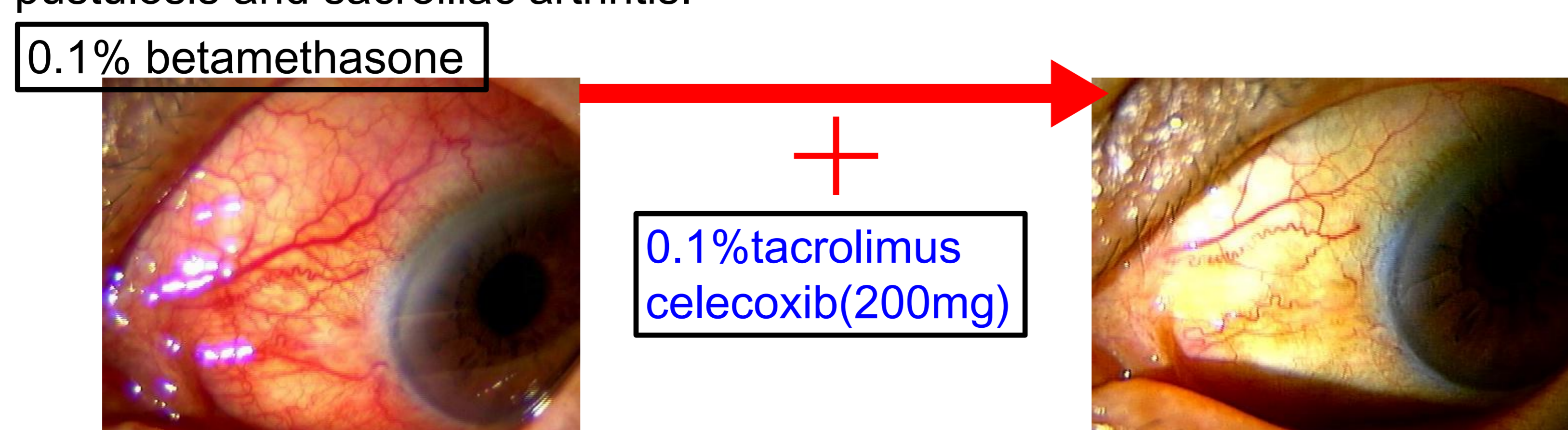
Vs=0.8p(1.2×S+0.25:C-1.25Ax95°)

Tod=21.0mmHg, Tos=20.0mmHg

anterior chamber clear, lens clear, fundus n.p., bilateral scleritis(+)

Clinical course: She was treated with 0.1%tacrolimus, celecoxib, and cured.

Rheumatologists diagnosed as SAPHO syndrome, due to palmoplantar pustulosis and sacroiliac arthritis.



Systemic Immuno-suppressant and Biologic agents that are needed

| | case1 | case2 | case3 | case4 |
|--------------|-------|-------|-------|-------|
| methotrexate | + | + | + | - |
| infliximab | - | - | + | - |
| adalimumab | + | + | - | - |

CONCLUSION: Ocular manifestations of SAPHO syndrome includes scleritis and non-granulomatous iridocyclitis, Anti-TNF therapy was effective for patients who showed poor response against immunosuppressive agents.