Usher's Syndrome

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Picture 1. Bone Spicules: Stellate pigment hyperplasia.

Picture 3. Arteriole Narrowing: left lower part. Spared Macula: in circle.

Picture 2. Depigmented Retina.

A 34-year-old woman who was revealed to have Usher's syndrome is reported. She was diagnosed with sensorineural hearing loss necessitating the use of hearing aids at the age of 6 and started to have progressive night vision loss when she was a teenager. Few years later, she developed daytime peripheral vision loss. She had normal intrauterine life with an uncomplicated birth. She denied any dizziness or family

history of visual or hearing loss. The retinal exam findings of bone spicules, depigmented retina, and arteriole narrowing with spared normal macula (Picture A-C), confirmed the diagnosis of retinitis pigmentosa. The presence of congenital sensorineural hearing loss along with retinitis pigmentosa confirmed the diagnosis of Usher's syndrome (USH).

Usher's syndrome is an inherited condition that causes progressive sensorineural hearing loss with possible imbalance at early childhood, and progressive vision loss due to retinitis pigmentosa (1). The three main types of Usher's syndrome are: USH 1, 2 and 3. USH1 is the most severe type with fast progressing symptoms at early-life (1, 2). USH3 is the least severe type with slowly progressing symptoms that start later (2). Early diagnosis is critical to ensure early conservative management with hearing aids and special support (3). The present patient has slowly progressive hearing and vision loss and is currently doing fine with hearing aids and special support.

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