Double-sided papilledema with normal intracranial pressure after liver transplantation

A 15-year-old girl with cystic fibrosis related liver disease (Child B, albumine 2.7 g/dl, PT 35%) with hepatopulmonary syndrome, portal hypertension and oesophageal varices graded II-III was listed for isolated liver transplantation (LTx). Pretransplant an intravitreal haemorrhage of her left eye was found which resolved after successful LTx. Immunosuppression consisted of cyclosporine and steroids. Surprisingly, ophthalmologic control showed papilledema of both eyes confirmed by optical coherence tomography (OCT).

The girl didn’t report any symptoms such as headache or blurred vision. Cerebral magnet resonance imaging and lumbar puncture with measurement of the cerebrospinal fluid pressure (5 mmHg) ruled out typical reasons for papilledema.

As papilledema increased with a progredient loss of vision the following weeks, immunosuppression was changed to tacrolimus. Papilledema resolved with improvement of her vision. Double-sided papilledema with normal intracranial pressure is a rare condition. According to Friedman et al. this constellation is called probable “normal pressure” pseudotumor cerebri syndrome. (1)

The immune-suppressant ciclosporine can be a very rare reason for pseudotumor cerebri. (2–5)
Pictures:

A) Papilledema in fundoscopy of the right eye.
B) Regression of papilledema of the right eye three month later.
C) Measuring of retinal nerve fibre layer using OCT (optical coherence tomography) confirms the diagnosis of papilledema of the right eye.

References: