• Letter to the Editor •

Multimodal imaging in Purtscher-like retinopathy associated with sarcoidosis: a case report

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Dear Editor,

We report a case of Purtscher-like retinopathy (PLR) in the presence of acute pancreatitis secondary to sarcoidosis. To our knowledge, this is the first case report of a PLR in a patient with sarcoidosis. A 54-year-old female, hospitalized at the Internal Medicine Department for stage E pancreatitis, hypercalcemia, and mediastinal lymphadenopathies, consulted with a visual impairment in both eyes evolving for a month, especially in her left eye (LE). The best-corrected visual acuity (BCVA), was 20/32 in the right eye (RE) and 20/50 in the LE. The slit-lamp examination revealed iris pigments on the anterior lens capsule, corresponding to disrupted posterior synechiae in both eyes (Figure 1A, 1B). The vitreous was clear, whereas fundus examination (Figure 1C, 1D) showed a stage 1 papillary edema and few superficial intra-retinal hemorrhages in the RE associated with multiple peri-papillary cotton-wool spots (CWS) in both eyes. Moreover, we noted focal whitening surrounding the retinal arterioles with a clear zone on either side of the vessels (Purtscher fleken). These findings were compatible with the diagnosis of PLR. Fluorescein angiography (FA) showed papillary early hyper fluorescence (papillary edema), peri-papillary non-perfusion areas associated with late capillary staining. We also noted peri-papillary and peri-vascular hypofluorescent spots, corresponding respectively to CWS and Purtscher fleken (Figure 1E, 1F). Macular Swept-source optical coherence tomography (SS-OCT) detected a focal thickening of the retinal fiber layer corresponding to CWS in both eyes. Besides, hyperreflectivity of the inner nuclear layer consisting of a paracentral acute middle maculopathy (PAMM), was revealed in the LE (Figure 1G, 1H). However, no disruption of the ellipsoid zone or macular thickening was noted. Swept-source optical coherence tomography angiography (SS-OCTA) showed superficial and deep capillary non-perfusion (CNP) areas (Figure 2A-2D). Flow void areas were noted in the choriocapillaris, in addition to a shadowing effect due to the overlying CWS (Figure 2E, 2F). Regarding the association of ocular inflammation sequelae (disrupted posterior synechiae) with hypercalcemia and deep lymphadenopathies, the diagnosis of sarcoidosis was suspected. However, the dosage of angiotensin-converting enzyme and salivary gland biopsy was normal. Infectious causes, including tuberculosis, systemic diseases, and malignancies were ruled out. Thus, a mediastinoscope-guided biopsy of the lymphadenopathies was performed. Histopathological specimen revealed, giant cells with non-caseating granulomas, confirming the diagnosis of sarcoidosis. The patient received an immunosuppressive dose of systemic steroids, initiated by three intravenous boluses of methylprednisolone (1 g/d) followed by oral prednisolone (1 mg/kg·d). A month later, the patient's vision improved, with a BCVA of 20/25 in both eyes. The fundus examination revealed a decrease in the number of CWS and the disappearance of retinal hemorrhages (Figure 3A). The PAMM persisted in the LE (Figure 3B). However, CNP and flow void areas decreased on OCTA (Figure 3C-3E).

DISCUSSION

Acute pancreatitis is the first cause of PLR, it represents 19.1% of cases[1]. The physiopathology of this retinopathy, in the cases of acute pancreatitis, was explained by complement activation that results in leukocyte aggregates or leukoemboli, which are released in the bloodstream and occlude the retinal vessels[2]. In sarcoidosis, pancreatic involvement is uncommon, moreover acute pancreatitis due to hypercalcemia is rare[3]. The association between systemic diseases and PLR was reported in some cases, however, to our knowledge, the association

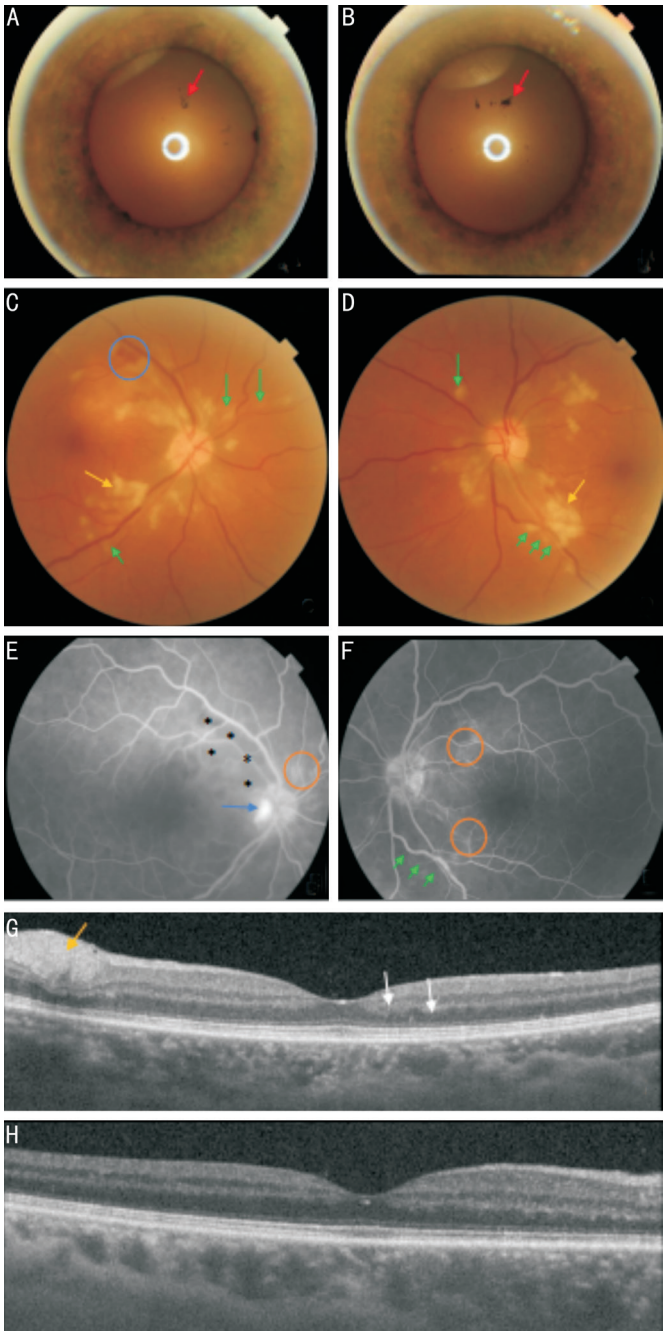

Figure 1 Anterior segment and fundus photographs, fluorescein angiography of the posterior poles and macular SS-OCT A, B: Ruptured posterior synechiae in both eyes (red arrows); C, D: Fundus photographs of both eyes: peripapillary cotton-wool spots (yellow arrows), Purtscher fleken (green arrows), scattered hemorrhage (blue circle); E, F: Fluorescein angiography of both eyes: peripapillary non-perfusion areas (black asterisks), hypofluorescent spots corresponding to Purtscher fleken (green arrows), diffuse peripapillary capillary staining (orange circles), and temporally localized papillary edema (blue arrow); G, H: Macular SS-OCT; G: Left eye. Paracentral acute middle maculopathy: thickening of the inner nuclear layer (white arrows); cotton-wool spots: hyperreflectivity in the retinal nerve fiber layer (yellow arrow).

between sarcoidosis and PLR was never described. Sarcoidosis is well known to induce occlusive retinal vasculitis[4], thus

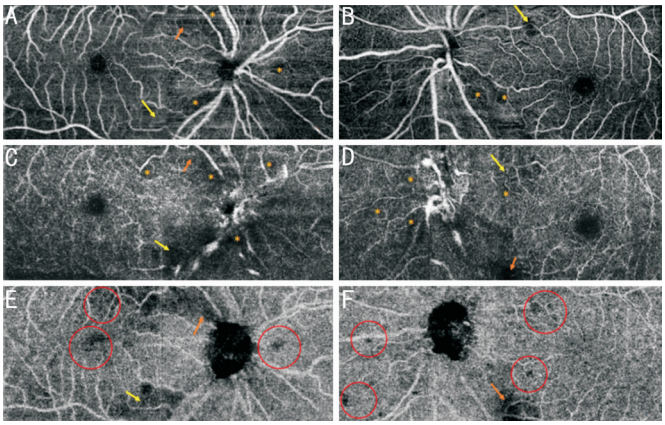

Figure 2 Composite of OCTA images (6×6 mm^2) of both eyes. A, B: Superficial capillary plexuses; C, D: Deep capillary plexuses: multiple CNP areas underlay CWS (orange arrows), shadowing effect of CWS (yellow arrows), CNP areas independent of CWS (yellow asterisks); E, F: Choriocapillaris: multiple CNP areas underlay CWS (orange arrows), shadowing effect of CWS (yellow arrows), flow void areas independent of CWS (red circles). CNP: Capillary non-perfusion; CWS: Cotton-wool spots.

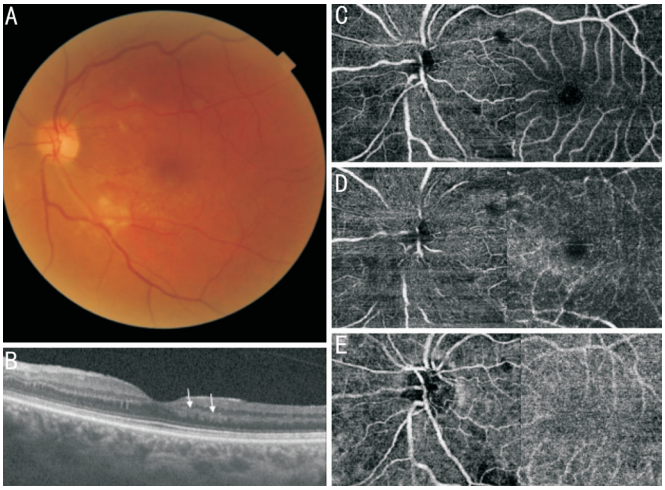

Figure 3 After 1mo of follow-up (left eye) A: The retinal lesions mostly absolved; B: Macular SS-OCT: persistence of PAMM (white arrows). C-E: Composite of OCTA images (6×6 mm^2): decreased CNP areas in the retinal capillary plexuses (C, D) and the choriocapillaris (E). CNP: Capillary non-perfusion; SS-OCT: Swept-source optical coherence tomography; PAMM: Paracentral acute middle maculopathy; OCTA: Optical coherence tomography angiography.

we hypothesized that microangiopathy that led to the PLR, in our case, could be explained by two vascular occlusive entities, sarcoidosis, and acute pancreatitis. In the context of a probable etiology, the diagnosis of PLR is based on a sudden visual impairment (bilateral in 60% of cases) and fundoscopic findings restricted to the posterior pole: CWS, retinal hemorrhages, and Purtscher flecken. Papillary edema has been observed in some cases[1]. FA shows different degrees

of CNP areas and fluorescein staining from retinal vessels, according to the severity of the retinopathy. Hypo-fluorescent spots corresponding to the CWS and Purtscher flecken are also observed[1]. In the majority of cases of PLR, optical coherence tomography shows macular edema[5]. However, in this case, the macular thickness was normal, and a PAMM was revealed. This condition in the context of PLR, might be explained by the ischemia induced by the retinal arteriolar emboli[5]. SS-OCTA is a recent multimodal imaging technique that showed, in this case, not only multiple areas of CNP in the superficial and deep capillary plexuses but also flow void in the choriocapillaris, described by Li *et al*[5] as a honeycomb-like pattern. This finding suggests the involvement of the choroid in the physiopathology of PLR. The evolution of PRL is favorable in some cases, however, no prognosis criteria have been fixed[6]. In this case, the patient regained a correct visual acuity in both eyes, despite the persistence of PAMM in her LE. This evolution might be explained by the decrease in CNP areas in the retinal capillary plexuses and the choriocapillaris as shown on SS-OCTA. The treatment of PLR is based on treating the underlying cause, corticosteroids in our patient, however, no consensus has been defined[1].

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Conflicts of Interest: Kaoual H, None; **Hachicha I,** None; **Mili Boussen I,** None; **Zhioua-Braham I,** None; **Boukari M,** None; **Zhioua R,** None.

REFERENCES

1 Miguel AIM, Henriques F, Azevedo LFR, Loureiro AJR, Maberley DAL. Systematic review of Purtscher's and purtscher-like retinopathies. *Eye* (*Lond*) 2013;27(1):1-13.

2 Behrens-Baumann W, Scheurer G, Schroer H. Pathogenesis of Purtscher's retinopathy. An experimental study. *Graefes Arch Clin Exp Ophthalmol* 1992;230(3):286-291.

3 Gebreselassie A, Mehari A, Dagne R, Berhane F, Kibreab A. Hypercalcemic pancreatitis a rare presentation of sarcoidosis: a case report. *Medicine* (*Baltimore*) 2018;97(2):e9580.

4 Pasadhika S, Rosenbaum JT. Ocular sarcoidosis. *Clin Chest Med* 2015;36(4):669-683.

5 Li B, Li DH, Chen YX. Purtscher-like retinopathy presented a honeycomb-like pattern in optical coherence topography angiography. *BMC Ophthalmol* 2019;19(1):232.

6 Vezzola D, Allegrini D, Romano MR, Pagano L, Montericcio A, Fogagnolo P, Rossetti LM, de Cillà S. Optical coherence tomography angiography in Purtscher-like retinopathy associated with dermatomyositis: a case report. *J Med Case Rep* 2019;13(1):206.