Unusual manifestation of diffuse large B-cell lymphoma

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The 74-year-old man was admitted to the hospital due to visual acuity impairment. Three months earlier he underwent an uncomplicated cataract surgery. Patient presented with a significant visual acuity impairment (finger counting at 1m), irritation of the right eyeball, inflammatory cells in the anterior chamber of the eye and numerous hyperechogenic densities in the vitreous chamber. The clinical features indicated late-onset bacterial endophthalmitis. The patient was qualified for vitrectomy with silicone oil tamponade. After the procedure local stabilization was achieved. The vitreous culture was negative.

Two months later there was an acute deterioration in the patient's condition. In the anterior chamber of the eye, fibrinous blood-stained exudate accumulated (Figure 1A), and a large pink mass appeared under the conjunctiva (Figure 1B). Ultrasound examination showed an infiltrate spreading from the ciliary body under the conjunctiva (Figure 1C). A surgical specimen was obtained. Microscopical examination demonstrated population of large lymphoid cells which had nuclei with one eosinophilic nucleoli and sparse cytoplasm (Figure 1D). Neoplastic cells were CD20 positive (Figure 1E) and had high Ki67 proliferation index (Figure 1F). Histopathological diagnosis of non-germinal center B-cell-like (non-GCB) Diffuse Large B-Cell Lymphoma (DLBCL) was rendered. Computed tomography did not reveal abnormalities in other part of the body. After external beam radiation therapy (ocular irradiation with prophylactic central nervous system treatment, 30 Gy in 15 fractions), followed by chemotherapy (R-CHOP regimen), disease remission was achieved. At the last follow-up, 12 months after the end of treatment, the patient was in good general condition.

DLBCL is the most common subtype of adult non-Hodgkin lymphoma. About 60% cases present primarily with lymph node disease, as a rapidly enlarging mass, located in a part of the body with multiple lymph nodes (neck, arm pits, or groin). In the remaining 40% of the cases, the disease begins as an extranodal lymphoma, most commonly in the stomach. Other locations include the testicles, breast, uterus, ovaries, kidneys, adrenal gland, thyroid gland,
bone, or brain.[1] Primary intraocular lymphoma (PIOL) is rare malignancy and represent 1-2% of extranodal lymphomas.[2] The tendency to mimic chronic uveitis makes intraocular lymphoma difficult to diagnose. The infiltrate of the ciliary body, found in the present case, is extremely rare. Mortality rates in PIOL range between 9% and 81% in follow-up periods ranging from 12 to 35 months. Median survival of patients treated with radiotherapy alone or chemotherapy plus radiotherapy ranges from 10 to 30 months.[2]

In above case, both the history and symptoms indicated late-onset bacterial endophthalmitis – rare, but extremely dangerous complication of surgical procedures.[3] During the vitrectomy, no subretinal lesions typical for intraocular lymphoma were observed, only changes usually seen in endophthalmitis – dense vitritis and anterior chamber inflammation findings. The suspicion of the lymphoma was raised after the patient’s condition worsening and appearance of pink lesion under the conjunctiva. The example above shows how important cooperation is between the ophthalmologist, pathologist and hematologist, especially in the case of a non-standard course.

Figure 1A Fibrinous blood-stained exudate accumulated in the anterior chamber of the eye.
Figure 1B Large pink mass under the conjunctiva.
Figure 1C Ultrasound scan. Red arrow – cornea, yellow arrow – sclera, green arrow – infiltration of the ciliary body, blue arrows – margins of the infiltrate.
Figure 1D Hematoxylin and eosin staining of Diffuse Large B-Cell Lymphoma.
Figure 1E Positive CD20 staining.
Figure 1F High Ki67 proliferation index.