

Retinal pigment epithelial detachment post radiation therapy in metastatic ocular infiltration of non-Hodgkin's lymphoma: a case report

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Abstract

• A 32-year-old lady, diagnosed with anaplastic large cell non-Hodgkin's lymphoma of the mediastinum, presented with bilateral floaters and reduced right eye vision 3 years post chemotherapy. Ophthalmic examination revealed bilateral panuveitis with multiple deep seated choroidal lesions in the left eye. Computed tomography scanning of the orbit showed enhancing and bulky optic nerve sheath at the retrobulbar part of both optic nerves. The patient was treated with involved field radiation therapy of the orbit, with cumulative dose of 30Gy. Eight months post radiation therapy, she developed retinal pigment epithelial detachment at the macula and deep chorioretinal degeneration at superotemporal and inferonasal regions of the right fundus. There was evidence of vitreoretinal traction at the margin of chorioretinal degeneration areas, thus barricade lasers were performed in the affected eye. Her visual acuity remains 6/6 in both eyes. Retinal pigment epithelial detachment is a possible complication of radiation therapy in non-Hodgkin's lymphoma with intraocular metastasis. It is essential to alert the managing ophthalmologists about this rare complication.

• **KEYWORDS:** non-Hodgkin's lymphoma; metastatic ocular infiltration; involved field radiation therapy; retinal pigment epithelial detachment

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INTRODUCTION

The orbit is rare secondary site of lymphoma dissemination, and only a few reports exist on the course and characteristics of involvement in these sites^[1]. Ophthalmic manifestation is rarely seen unless the disease is clinically and hematologically active. It involves the vitreous and retina and frequently represents a diagnostic challenge to the clinicians.

Retinal pigment epithelial detachment had been reported previously as an intraocular presentation of systemic form of large cell non-Hodgkin's lymphoma and primary central nervous system lymphoma^[2]. To the best of our knowledge, it has never been reported as a complication of radiation therapy of such cases. Here, we present a patient with metastatic ocular infiltration with non-Hodgkin's lymphoma in remission stage, treated successfully with involved field radiation therapy (IFRT) and subsequently developed retinal pigment epithelial detachment post radiation therapy.

CASE REPORT

A 32-year-old lady, previously healthy presented with pyrexia of unknown origin, cough and loss of appetite for six months. She had reduced breath sound in the right lung and palpable right axillary lymph nodes. Chest radiograph showed lobulated opacity in the right hemithorax overlying the hilar region, which was associated with obliteration of upper part of the cardiac border suggesting of right mediastinal widening. Computed tomography (CT)-scan of the thorax confirmed a right mediastinal mass. CT-guided tissue biopsy from right mediastinal mass and fine needle aspiration from right axillary lymph nodes were highly suggestive of anaplastic large B cell lymphoma. Fortunately, there was no evidence of lymphoma infiltration in bone marrow aspiration studies. She received 6 cycles of chemotherapy which consisted of cyclophosphamide, adriamycin, vincristine and prednisone (CHOP regime) and 18 courses of external beam radiation therapy. Unfortunately, she developed multiple lymphadenopathies as she continued with the treatment. A repeat CT-scan of the neck and thorax showed progression of the lymphoma stage IV. The mediastinal mass has extended into the right hemithorax and the anterior chest wall. There were multiple lymphadenopathies noted in the neck region. She was started with the salvage chemotherapy for 2 cycles.

Three years after completing the regime, she presented with blurring of vision and floaters in both eyes. Ocular examination revealed visual acuity of 6/9 in the right eye and 6/6 in the left eye. There were 3+ cells in the anterior chamber, numerous fine keratic precipitates and iris pigments on the anterior lens capsule in both eyes. The pupillary response and intraocular pressure were normal in both eyes. Severe vitritis was noted in the right eye that hindered further fundus details. However the vitritis was mild in the left eye thus enabled a detailed fundus examination. Multiple small deep seated choroidal lesions were observed at superotemporal

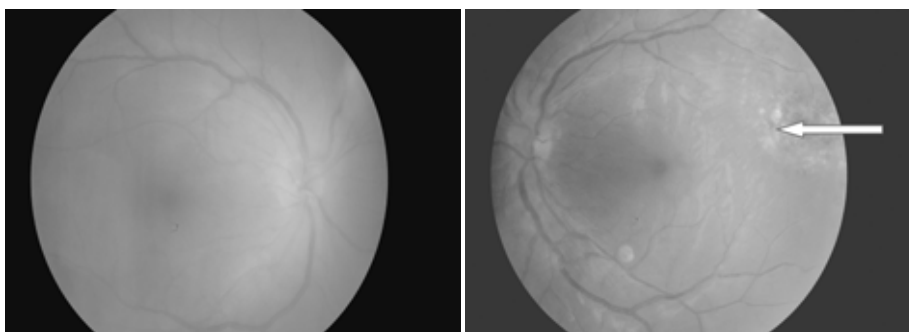
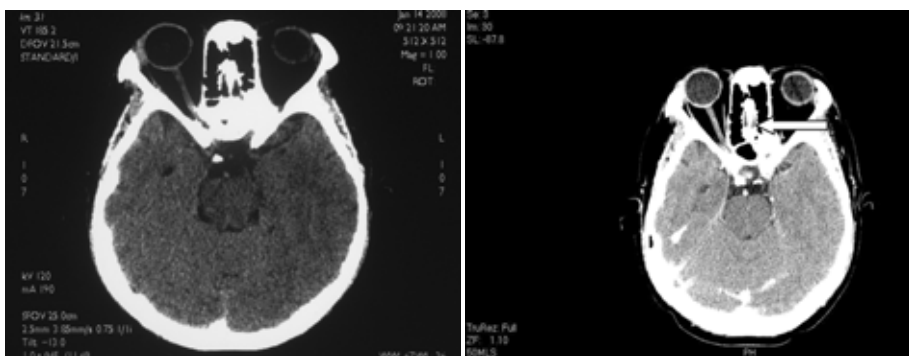


Figure 1 A hazy retinal view due to severe vitritis in the right eye and choroidal lesions (arrow) in the left eye.



Non-enhanced

Enhanced

Figure 2 The right optic nerve sheath is enhancing and slightly thickened at the medial part. Retrobulbar part of both optic nerves are bulky, worse in the right eye(CT-scan).

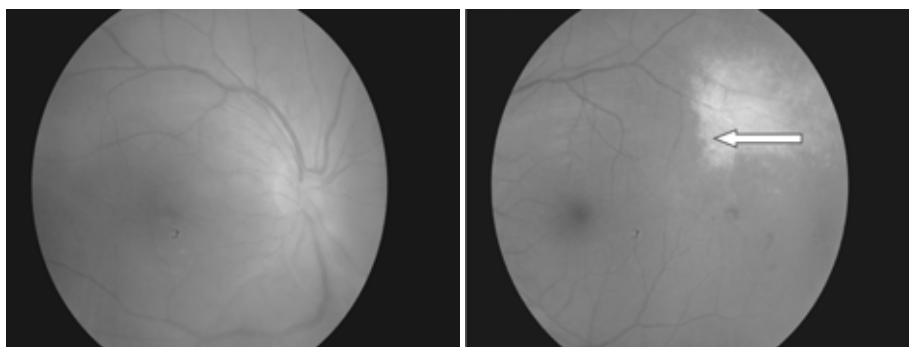


Figure 3 Resolved vitritis in the right eye and subretinal fibrosis (arrow) with residual intraretinal hemorrhages in the left eye post IFRT.

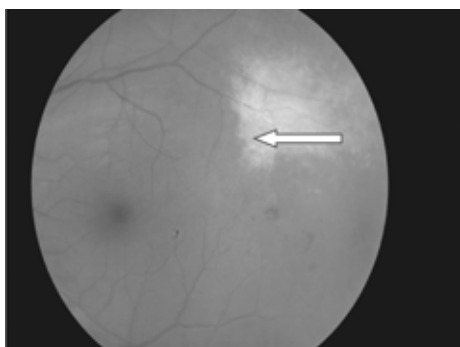


Figure 4 Maculopathy in the right eye 8 months post radiation therapy.

area of the left fundus, which was associated with intraretinal hemorrhages and ghost vessels (Figure 1). CT-scan of the orbit revealed enhancement of the right optic nerve sheath, which was slightly thickened at the medial part with a smooth

outline. Retrobulbar part of both optic nerves were bulky, which was worse in the right eye. There was no definite enhancing lesion within the optic nerves (Figure 2). The above findings were consistent with bilateral lymphomatous infiltration of the optic nerves.

She refused diagnostic vitrectomy. Thus, she was treated with IFRT of the orbit for two weeks and received a cumulative dose of 30Gy in 15 fractions with 6mv photons. The technique used for radiation delivery was lateral field technique with no lens blocking. Floaters were reduced tremendously and her visual acuity improved to 6/6 in both eyes 2 months later. The anterior uveitis resolved and vitreous was clear. There was no residual choroidal lesion except for minimal intraretinal hemorrhages in the left eye. The right fundus was normal (Figure 3). A repeat CT-scan of the orbit showed cessation of the previous findings. She again complained mild blurring of vision and floaters in the right eye 8 months post radiation. Visual acuity was 6/7.5 in the right eye and 6/6 in the left

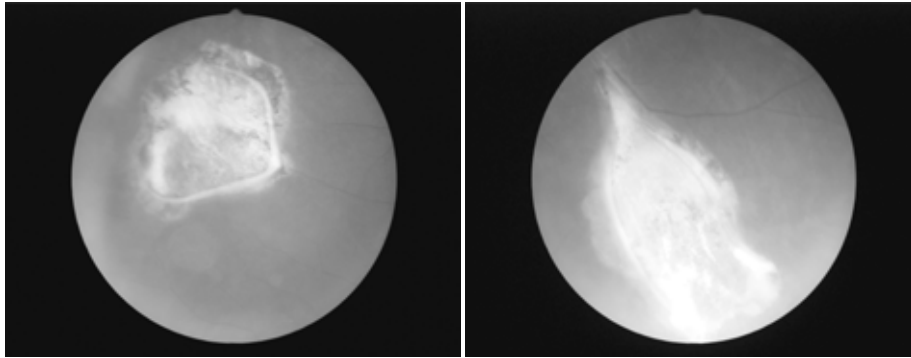


Figure 5 Chorioretinal scars with pigmentary degeneration and fibrosis were noted at superotemporal and inferonasal region of the right fundus.

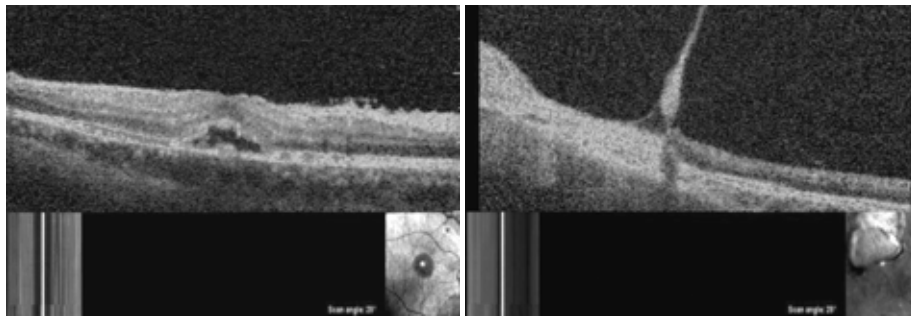


Figure 6 Retinal pigment epithelial detachment at the macula region (left) and vitreoretinal traction at the margin of the chorioretinal scar (right) of the right eye (optical coherence tomography).

eye. There was evidence of maculopathy (Figure 4) and chorioretinal scar with pigmentary degeneration at inferonasal and superotemporal region. Retinal fibrosis was noted at the margin of lesions in the right eye (Figure 5). The intraretinal hemorrhages totally resolved in the left fundus. Optical coherence tomography revealed retinal pigment epithelial detachment at the macular region with evidence of vitreoretinal traction at the margin of chorioretinal scars in the right eye (Figure 6). In view of risk of retinal detachment, two to three rows of laser photocoagulation were applied surrounding the lesions. At one year's follow-up, the visual acuity was 6/6 in the both eyes. There was no sign of re-infiltration. Both the macula and the chorioretinal scars in the right eye remained similar appearance with no further progression.

DISCUSSION

It has been reported that non-Hodgkin's lymphoma patients responded very well to local radiotherapy, for example in orbital and CNS lymphomas, 1500-4500 rads in 10-30 fractional doses on the CNS and orbit. Our patient received IFRT with cumulative dose of 30Gy in 15 fractions with 6mv photons. The technique used for radiation delivery was lateral field technique with no lens blocking. It is an option for delivering radiation to only those areas of the body involved by lymphoma^[3]. In contrast, extended field radiation therapy delivers radiation to larger areas of the body, including regions which are not immediately involved by lymphoma^[3]. The duration of treatment depends on the dose delivered. As IFRT is commonly given after chemotherapy, the dose is often based on the remaining disease following chemotherapy. In intraocular metastasis, combination of chemothe-

rapy and radiotherapy or radiotherapy alone may result a better outcome^[1, 4-6].

Numerous radiation complications, which included dry eyes, radiation induced cataract, punctate keratopathy, radiation retinopathy, neovascular glaucoma and optic atrophy had been reported^[4,7]. Hayreh has listed numerous radiation retinopathy changes by fluorescence fundus angiographic study such as obliteration of the retinal vessels, microaneurysm, retinal telangiectases and neovascularization, cotton wool spot, white retinal deposits, retinal hemorrhages, retinal oedema, sheathing of large retinal vessels, perivasculitis, optic disc changes, vitreous hemorrhages and retinal detachment. Amaro *et al*^[2] reported two cases of different forms of retinal pigment epithelial detachment as presenting features in intraocular large cell non-Hodgkin's lymphoma both in systemic and primary central nervous system lymphoma. In contrary, our patient developed retinal pigment epithelial detachment at the macula 8 months post radiation therapy. We did not encounter sign of residual or recurrent primary disease occurring in the affected eye. It is postulated that retinal pigment epithelial detachment occurs due to reduction of hydraulic conductivity of the thickened Bruch membrane. This will impede the movement of fluid from the retinal pigment epithelium towards the choroids.

Though retinal pigment epithelial detachment causes non-visual threatening condition, its subsequent course may lead to various complications, such as detachment of the sensory retina, choroidal neovascularization, tear of the retinal pigment epithelium or geographic atrophy of retinal pigment epithelium following spontaneous resolution. Our patient

showed evidence of vitreoretinal traction at the margin of the chorioretinal lesions. This can contribute to the development of retinal detachment. Thus, we performed barricade laser to stop the progression and to prevent the development of retinal detachment.

In conclusion, retinal pigment epithelial detachment is an unusual presentation after radiation therapy in non-Hodgkin's lymphoma with ocular metastasis. Thus, it is important to alert all who involve in patient care.

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非霍奇金淋巴瘤转移性眼浸润放射治疗后视网膜色素上皮脱离 1 例

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摘要

患者,女,32岁,被诊断为纵隔未分化大细胞非霍奇金淋巴瘤,化疗后3a出现双眼悬浮物和右眼视力下降。眼科检查发现带有多发深在的左眼脉络膜病变的双侧全葡萄膜炎。眼窝电脑断层扫描显示两侧球后视神经部分加强和大块的视神经鞘。该患者接受眼窝受累区域放射治疗,累积剂量为30戈瑞。放射治疗8mo后,发展为黄斑部视网膜色素上皮脱离和右侧颞上和鼻下区眼底深脉络膜视网膜变性。在脉络膜视网膜变性区边缘有玻璃体视网膜牵引迹象,从而路障激光在受累眼睛进行。双眼视力仍然6/6。视网膜色素上皮脱离可能是放射治疗非霍奇金淋巴瘤眼内转移时的并发症。至关重要提醒了眼科医生这个罕见的并发症。

关键词:非霍奇金淋巴瘤;转移性眼浸润;受累区域放射治疗;视网膜色素上皮脱离