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CASE REPORT

Malignant hemophtalmus as a first sign of orbital rhabdomyosarcoma in adult

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Abstract: We report on a rare case of an adult patient with rhabdomyosarcoma treated at the Dpt Ophthalmology, Comenius University, Medical School in Bratislava as a single case since 1968 *(Fig. 2, Ref. 5)*. Full Text in free PDF *www.bmj.sk.*
Key words: malignant hemophtalmus, orbital rhabdomyosarcoma in adult, tumor.

Orbital rhabdomyosarcoma is an extremely rare tumor in adults. From the standpoint of an ophthalmologist, it is the most common malignant orbital tumor in children. Nevertheless, the situation in adults is different. During his/her life, an ophthalmologist does not have to encounter an adult patient with rhabdomyosarcoma. The reason is that most of the cases are misdiagnosed and treated under other entities.

Case report

We report on a rare case of an adult patient with rhabdomyosarcoma treated at the Dpt Ophthalmology, Comenius University, Medical School in Bratislava as a single case since 1968.

A 78-year-old man was sent by an ophthalmologist to our Dpt for counseling on his right eye problems. The patient complained only about his glasses. The first examination in January 06 revealed that best corrected visual acuity (BCVA) in his right eye was 6/9, and 6/12 in his left eye. The ultrasound A,B mode showed a solid tumor-like mass on the posterior pole and secondary retinal detachment. The patient was suspected to have an intraocular malignant melanoma, and was sent to computer tomography and to complete the check-up for distant metastasis.

The local findings revealed normal intraocular pressure, incipient cataract, and opacitates corporis vitrei. One month later, the A and B ultrasound scan examination showed a large tumor mass in the posterior pole and retrobulbar space. Computed tomography and magnetic resonance revealed a large intrabulbar lesion. The clinical status of the patient was good, he refused therapy. Two months later he was admitted to our Dpt with an increased intraocular pressure (over 50 Torr) and red painful eye. The ultrasonographic examination showed hemophtalmus and secondary retinal detachment.

A radical surgical treatment was indicated, i.e.enucleation of the eye. The operation was without any complications.

One week after enucleation there was no presence of intraorbital complications or tumor masses. The patient was recommended to get an individual prosthesis.

One month after enucleation a solid painless tumorous mass was arising from the cone. It was classified as stage T4 of orbital tumor, Nx, M2(?). Upon ultrasound checking of abdomen, a liver metastasis was present.

Prolapse of intraorbital tumorous masses was significant six weeks after enucleation.

The tumor was confirmed by histopathology of a specimen of the mass. It showed a non-differentiated tumorous sarcoma arising from the orbit, (ICD 8800/3), while histogenesis was not set, the tumor developed from undifferentiated mesenchymal cells and was infiltrated with granulocytes. Positive immunophenotype showed D45RO-, CD20-, CD79a-, CD30-, ALK of non-lymphoma origin, and CHE-, CD68-, myeloperoxidase of non-myelocytic origin, while positive S-100 was suspected to be of melanoma origin; NSE, HMB45 and melanA were negative while melan A was not suspected to be of melanoma origin.

The patient was recommended to undergo external radiotherapy (ERT) -Co^{60}, with a total dose of 35Gy in 7 fractions. Three weeks after radiotherapy, the reaction was dramatically positive while the prolapsed tumorous masses became necrotized and there was no presence of prolapsing masses over the closed eyelids.

At the end of ERT, the patient was in good clinical stage, survival was four months after radiotherapy; he died due to liver metastasis in his regional hospital (stage T4 N2 M4) (Figs 1, 2).

Discussion

Clinical features of rhabdomyosarcoma in adults can present insidiously. The most characteristic presentation is a rapid onset

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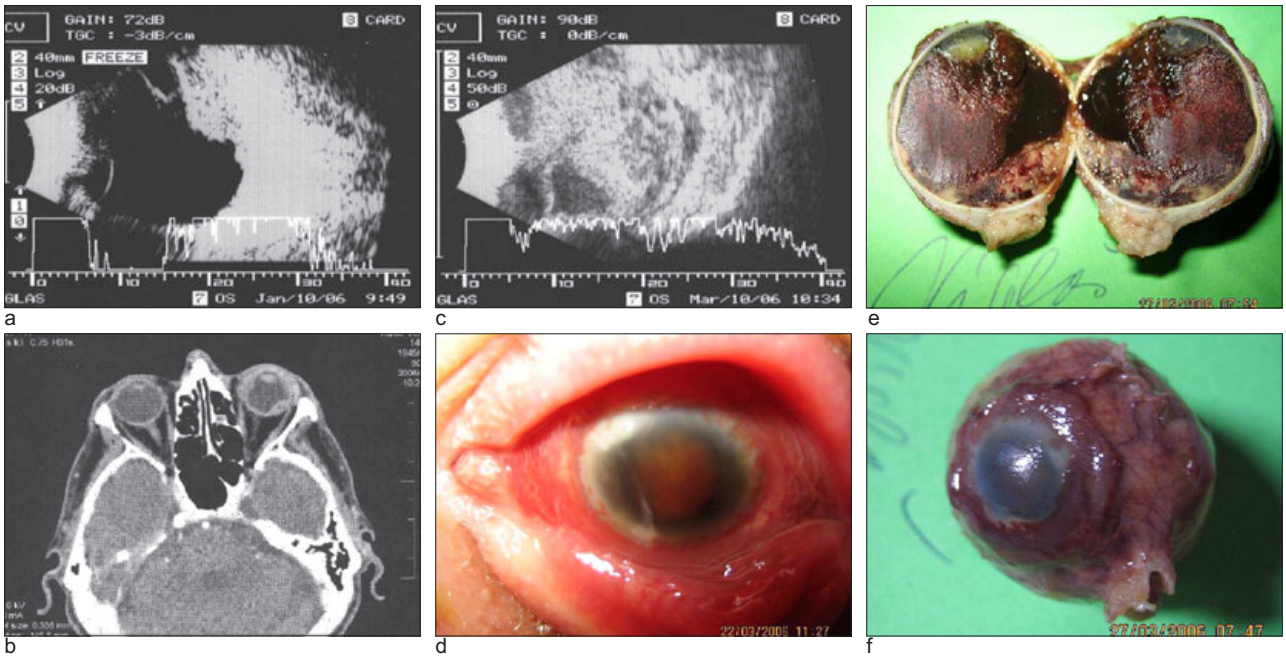

Fig. 1. a) Ultrasound examination (A, B mode) – hemophtalmus, b) Computed tomography showed a solid mass on the posterior pole, maximum elevation of 5 mm, c) Ultrasound examination after three months – hemophtalmus, d) Anterior segment of the eye – infiltration of the conjunctiva, cataract, low anterior chamber, e) Specimen of enucleated eye globe – macroscopically only hemophtalmus without the tumorous mass, f) Enucleated eye globe – no presence of extrascleral dissemination.

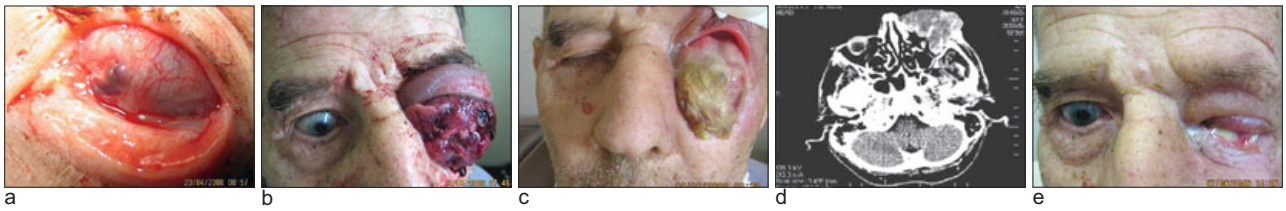

Fig. 2. a) Conjunctival sac after enucleation – no presence of intraorbital expansion, b) one month after enucleation – a solid painless tumorous mass was arising from the cone, c) computed tomography of the site of the tumor mass, d) prolapsed tumor mass over the eyelids, e) local findings at the beginning of radiotherapy, f) local findings after radiotherapy.

and progression of proptosis and displacement of the globe. It can clinically develop into unilateral exophthalmus. Symptoms depend on the origin and site of lesion while posterior tumors can cause edema of the optic disc. Other localizations of the tumor mass such as in the inferior part of the orbit cause chemosis and eyelid swelling. The clinical differential diagnosis includes most causes of proptosis in childhood; in adults the onset may be most radical (1, 2). Computed tomography and magnetic resonance imaging are important to localize the tumor mass and progression while the definitive diagnosis is always based on histopathologic confirmation (3).

Mesenchymal orbital tumors are rare in adults. Advanced techniques in radiotherapy and chemotherapy improved the survival rates in young patients, nevertheless radiation therapy may lead to undesirable side effects and the mortality rate of patients is very high (4). The data based upon prospective, randomized multicenter trials are necessary in order to improve the clinical outcome of adult rhabdomyosarcoma patients (5).

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