Research Paper Medical Science

Choroidal Malignant Melanoma

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ABSTRACT

The authors present a patient with intraocular tumor, incidentally found, associated with senile cataract in the other eye. At the on set the patient complained of decreased visual acuity. Laboratory tests, complete ocular examination (visual acuity, intraocular pressure, fluorescein angiography, Ultrasound Bscan), Nuclear Magnetic Resonance (MRI) sustained the positive diagnosis of intraocular tumor. Histopatological examination revealed choroidal malignant melanoma type Spindle A and B. We performed enucleation on this eye with good evolution after the surgery. Also, we performed on the opposite eye extraction of the cataract with intraocular lens implantation.

Keywords : Choroidal malignant melanoma, cataract, enucleation

Introduction
Malignant choroidal melanoma is a tumor of cells that produce pigments and it is the most frequent primary tumor found in adult (Calugaru, M. et al., 2001). Latest technology in imagistic has improved the means in putting a positive diagnosis. Of course the confirmed diagnosis is done by anatomopathologic exam. An early diagnosis of malignant choroidal melanoma is very important in the vital prognosis. Current treatment modalities are strictly based on the size and extension of the tumor and also on the anatomoclinical form of the tumor (Calugaru, M. et al., 2001).

Patient and Methods
We present the case of S.I., 77 years old, complained of decreased visual acuity(VA) at the right eye (RE). From his personal history we focus on his Parkinson's disease and prostate hypertrophy.

Ocular examination revealed the following data: RE VA = 1/50 without correction, LE VA = 0.4 with correction -1.25 D (left eye LE). Perception and light projection: both eyes (BE) present. Pupillary reaction: BE present.

Ocular refraction: RE +4 sf -0.5 cyl axis 90 ° LE - 1.25 sf -0.5 cyl axis 90 º.

Tonometry (intraocular pressure): IOP RE = 10 mmHg, LE = 12 mmHg.

Endothelial cells count RE: 2448/mm ² and LE: 1775/mm ².

Slit lamp examination: BE: nuclear lens opacities more advanced at LE.

On fundus examination RE we found an inferotemporal tumor, well defined, brown, with fixed retinal detachment, and the LE vital optic disc, increased reflex of arteries, Sallus II, macula with reflex.

The first step clinical diagnosis was RE: Suspicion of intraocular tumor. Secondary retinal detachment. Medium hyperopia. BE: Senile nuclear cataract in evolution.

To confirm the diagnosis we performed the following examinations:

Laboratory tests: ESR = 5 mm/h, lymphocytes = 1.33 UM, UM monocytes = 13.1, glucose = 90 mg/dL and urine examination - negative melanuria.

Ocular ultrasound, showed in axial and transverse section a hyperreflective tissue growth at RE, looking as a dome and overlying retina off in the bottom, and LE was within normal parameters. Conclusion of ultrasound was RE: intraocular tumor with secondary retinal detachment. (fig. 1, fig. 2)

Figure. 1 Ocular ultrasound RE - axial section

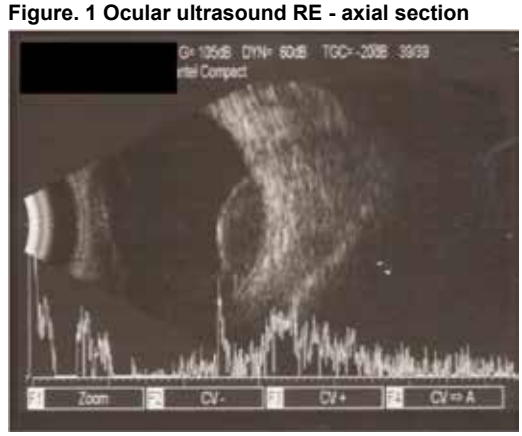
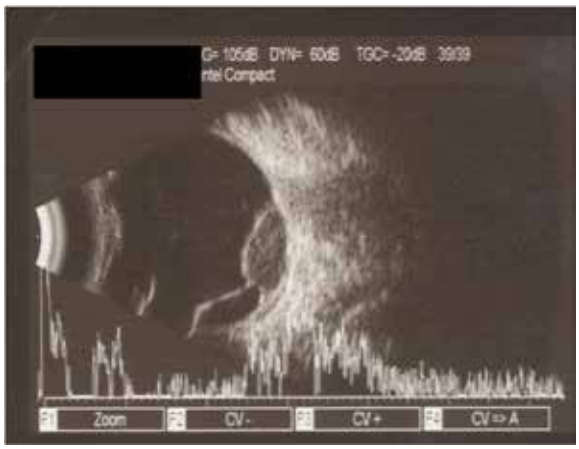


Figure. 2 Ocular ultrasound RE - transversal section

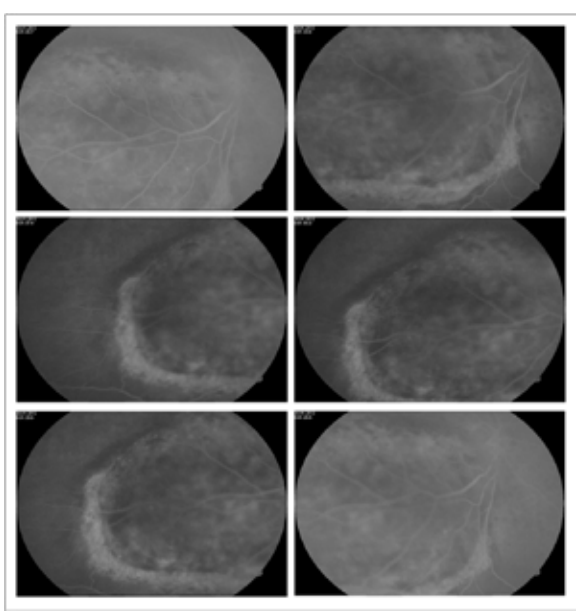


Fluorescein angiography at RE reveals:
Arterial time: Placard about 6/7 DD spread, dark, over the inferotemporal arch. Double retinal and choroidal vascular network (tumor circulation). Arterio-venous time: hyperfluorescent looking placard lesion, pinpoint spots in the periphery. Central and upper absolute screen.

Early venous time: moderate increase in fluorescence of the spots in the lower half of the placard.

Very late venous time: Placard lesion with moderate hyperfluorescence with screen areas especially in superior sector, bounded below by a rupture of the pigment epithelium. (fig. 3)

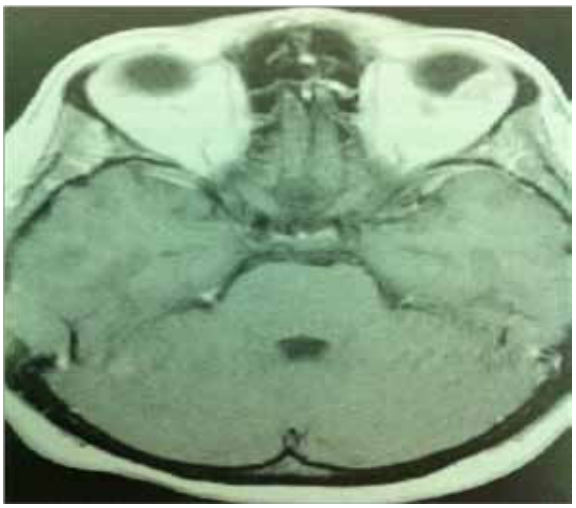
Figure. 3 Angiofluorography RE



Liver ultrasound examination and liver enzyme levels were normal.

Cerebral contrast MRI, revealed an intraorbital tumor, intrabulbar RE, discoid, size of 06/12/11 mm hypointense T2 and izointense T1, located at the posterior pole, intense and homogeneous with secondary retinal detachment. MRI showed probably choroidal melanoma.(fig. 4)

Figure. 4 Cerebral MRI



Cardiological examination shows a newly diagnosed essential hypertension, supraventricular extrasystoles.

Positive diagnosis after history, ocular and imagistic investigation was RE: intraocular tumor (suspected choroidal melanoma). Secondary retinal detachment. BE: Senile nuclear cataract in evolution.

Differential diagnosis was done with primary retinal detachment, choroidal metastasis, choroidal hemangioma, choroidal osteoma, melanocytoma, melanocytic choroidal nevus, choroidal granulomas, benign reactive lymphoid hyperplasia, choroidal tuberculoma, sarcoidosis granuloma, retinal hamartoma, subretinal hemorrhage, posterior scleritis by ophthalmoscopic appearance, ultrasound and fluorescein angiography.

Evolution without treatment of the case would lead to secondary glaucoma, orbital and general metastases (liver, brain).

The aim of the treatment was removal of intraocular tumor, specifying the anatomopathological type of tumor. Other ophthalmologic therapeutic procedures were local irradiation (brachytherapy of episclera, teletherapy), surgical excision of the tumor (sclerochoroidectomy). We choose enucleation with metacrylate ball implant at RE, under local anesthesia.

The eyeball was sent to the pathology laboratory. Postoperative, the patient received local treatment with antibiotics, steroids and general non-steroidical drugs.

Results
The macroscopic anatomopathologic results: eyeball 2.6 / 2.6 / 2.6 inches with a 13 mm optical nerve. At the level of optic nerve in the posterior pole there is a black tumor of 13/05/14 mm, lenticular. It does not exceed the eye wall; the tumor is developing towards the eye. Microscopic examination revealed between the retina and fibrous connective layer one well defined tumor with sarcomatoid appearance, composed of elongated nuclei, many with brown pigment in high quantity. There were no vascular invasion by malignant cells, but they try to slip between retinal and the fibrous layer, perivascular, towards the anterior pole. Section through the optic nerve revealed no malignant elements, vessels around the optic nerve shows no malignant infiltration.

Histological examination: posterior pole uveal melanoma of cell type fusiform spindle A and B, pT2a, Nx, Mx, St II. (fig. 5)

Figure. 5 Microscopical aspect of the tumor

Six months after enucleation presents LEVA = 0.3 w.c.-1.5 sf. Anterior pole examination reveals: nuclear lens opacitie. Cerebral contrast MRI at 6 months postoperatively, describes a status of RE enucleated for malignant melanoma and prosthetics, the posterior fossa has an arahnoidian cyst of 20/26 mm without other particularities.

Cataract surgery is recommended at LE by phacoemulsification technique with artificial lens implantation. Ocular biometry showed an axial length of 24.25 mm and the dioptric power of the artificial lens is 20.0 D for emmetropia. Patient choice was monofocal hydrofobic Acrysof IQ lens.

A month after cataract surgery in LE, the patient presents LE VA = 1 without correction and ocular refraction LE: +0.5 -1.75 axis 75 °. Optical correction for near vision: +3.5 sf.

Discussion

Choroidal melanoma is the most common ocular malignant tumor. Most common age is between 60-65 years. Early diagnosis of the disease is particularly important, because these tumors untreated develop metastasis: orbital and general- liver, lungs, brain and bones. Histopathological appearance is important for prognosis, considering the fact that these tumors metastasize exclusively by blood.

Sommer, quoted by Calugaru (2001) correlates the histologic aspect with cell origin, considering that there are three different cytological types: melanosarcoma - uveal cromatofore cell origin, leucosarcoma - originating in nerve bulb, angiosarcoma - originating in uveal vascular elements.

Folberg et al. (1993) have described several types of melanomas with different microvascularization aspects, with prognostic implications. The study concluded that the presence of vascular loops and network anastomosis as unfavorable prognostic factors are at high risk of metastasis. Carmeliet et al (2000) have shown some links between the network and the type of vascularization VLA-2 antigens, involved in extracellular matrix organization in the process of tumor metastasis, without yet applied clinically. Folkman et al (1995) analyzed the vascularization of the melanoma and have shown a significant vascularization, consisting of a network and a network of lakes capillary blood, the wall is formed directly from tumor cell membranes.

The presented case, because of the histopathological appearance, has a good prognosis, requiring further evaluation of the patient.

Choroidal melanoma is a disease with high mortality rate, usually irrespective of the choosen treatment modality. About 30-50% of patients with choroidal melanoma will die ten years from diagnosis and treatment. Death is usually secondary to distant metastasis and the risk is greatest in larger tumors (COMS report no. 22, May 2004; COMS report no. 4, Jul 1997). The Collaborative Ocular Melanoma Study found that the ten years rates of death secondary to metastasis were 45% in the preenucleation irradiation group and 40% in the enucleation alone group (COMS report no. 6, Jun 1998; COMS report no. 11, Sept 1998; COMS report no. 10, Sept 1998).

From the prognostic point of view considering the original classification Callender (Calugaru, M. et al., 2001), groups with cells of fusiform AB type have better prognosis with a mortality of 22%, whereas groups of mixed type of cells (epitheliod and necrotic) have a poor prognosis with a higher mortality of 62% due to metastasis (Calugaru, M. et al., 2001). Severe life prognosis is correlated with larger size of the tumor, anterior location, transcleral extension, growth through the Bruch membrane, optic nerve extension, lack of pigmentation and histologic characteristic (COMS report no. 4, Jul 1997; COMS report no. 5, Dec 1997). Our case has a good prognosis due to its histopathologic aspect and needs further evaluation.

Extraocular involvement usually occurs in large tumors (Shields J.A., 1991) and appears through emissary scleral channels. Some authors consider the incidence of extension outside the ocular globe to be 8-17%. Extraocular extension darkens the vital prognosis of the patient and has to be stabilized before any surgical treatment.Therefore ultrasound and topographic support in evaluating the extraocular extension is very important. De Potter et al (1992) recommended orbital exenteration in cases where the extraocular extension are more than 3mm. In our case, imagistics revealed only an intraocular tumor therefore we choose enucleation as our surgical option.

Several ways of treatment are available for choroidal melanomas, but the choice of the treatment take into account several factors: visual acuity of the affected eye, visual acuity of the opposite eye, size of the tumor, age and general health of the patient, ocular structures involved and presence of metastasis (Garcia-Valenzuela E., 2013).

Enucleation of the ocular globe in case of malignant choroidal melanoma has been an elective treatment for over a century and has been the preferred treatment for large (basal diameter >15 mm and height >10 mm) and complicated tumors which compromise visual function and for which other therapies tend to fall (Garcia-Valenzuela E., 2013); Chang M.Y. et al., 2013). For sure applying this method remains difficult in cases where vision is still present. Thus some authors proposed alternative treatment in cases of small malignant choroidal melanoma such as: local external irradiation, with charged particles or episcleral brachytherapy with Cobalt, Rutheniu, Iridium plate, resection of the tumor, laser photocoagulation and hyperthermia (Shields J.A. et al., 1991). Plaque brachytherapy is indicated in the treatment for medium sized posterior uveal melanomas (<10 mm in height and <15 mm in diameter).

The most common material used in modern plaques is Iodine-125, because of its lower energy emission(lack of alpha and beta rays), its good tissue penetration and its commercial availability (Boggs W. et al., 2013; Semenova E. & Finger P.T., 2013). Detorakis et al (2005) found that after this therapy, iris and anterior chamber angle neovascularisation developed in 23% of eyes.

Chang and colab (2013) showed that patients survival after treatment of medium sized melanoma is similar when plaque radiotherapy is compared with enucleation.

External beam irradiation using charged particles (protons or helium ions) is frequently used alternative method for the treatment of medium sized choroidal melanomas (<10 mm in height and <15 mm in diameter) (Phillips C., 2013). Patients treated with external beam irradiation seem to have a survival rate comparable to those treated with enucleation (Garcia-Valenzuela E., 2013).

Laser photocoagulation and transpupillary thermotherapy are used to treat selected small choroidal melanomas when the lesions are located away from the fovea and are less than 3 mm in thickness (Garcia-Valenzuela E., 2013).

Orbital exenteration is a radical treatment reserved for cases with wide spread orbital extension (Garcia-Valenzuela E., 2013).

Conclusions

Particularities of this case are:

- accidental discovery of intraocular tumor, the patient knowing he has cataract in BE;
- advanced age of the patient.

Vital prognosis depends on the histological type. Histopathologically fasciculated cells type A has the best prognosis (Calugaru, M. et al., 2001) whereas advanced age, intense pigmentation and extrascleral extension form a poor prognosis.

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