

A giant malignant hemangiopericytoma of the orbit

Xiao Huang¹, Ruili Wei², Jiping Cai³

ABSTRACT

An 81-year-old man with hemangiopericytoma had clinically conspicuous manifestation of a first slowly then rapidly growing, painless, and sublobe-like mass in the left orbit, proptosis, massive bleeding, and underwent orbital exenteration with keeping the eyelids. The tumor reoccurred 21 months after the operation and the patient had prolonged, serious hemorrhage in the left orbit and died of brain metastasis six months later. The etiology and inducement factors were unknown, but it is suspected that the fatal family casualty might be a more important inducement than trauma for this patient.

KEY WORDS: Malignant hemangiopericytoma, Orbit, Case report.

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INTRODUCTION

Malignant hemangiopericytoma of the orbit is rare,^{1,2} and its etiology and inducement remain unknown. To the best of our knowledge, this disease has never before been reported from China. Herein we report a new case of a giant malignant hemangiopericytoma of the orbit.

CASE REPORT

An 81-year-old Chinese man visited our clinic in September 2006, complaining of a painless proptosis of his left eye that had been gradually increasing for two years. It was first noticed after a prolonged period of crying following the loss of a family member. Eleven years before, he had lost vision in his left

eye not long after his left cornea was damaged by a chopstick. Physical examination revealed that the man's left eye was grossly proptotic with resistance to retropulsion. He could close his left eyelids well, but could not open them completely (Fig-1A top panel). Vessels in the lids were expanded. A sublobe-like immovable and irregularly shaped mass of approximately 6 × 6 × 4 cm was found in the orbit. Several bleeding dots in the engorged conjunctiva were present, which discharged blood almost every other day. His right eye was almost normal but had a mild cataract. Computed tomography demonstrated a 7 × 6 cm giant mass with iodine asymmetry enhancement and an irregular boundary in the left orbit, which pressed the eyeball into an abnormal shape (Fig-1B top and middle panels).

Then patient underwent orbital exenteration; the eyelids were retained on the patient's request. Histological examination of the tumor showed that there were rich blood vessels with different sizes and shapes throughout the tumor. Some had lumen sometimes branched like antlers, while others were pressed into crack-like or even occlusion, Vascular channels lined with flat endothelial cells, and tumor cells were arranged around blood vessels (Fig-2A). Increased cellularity and nuclear pleomorphism were found in more than 4 per 10 high-power fields (Fig-1A bottom panel) Immunohistochemistry analysis indicated that the tumor cells were round to fusiform in shape, with a dense reticulin meshwork

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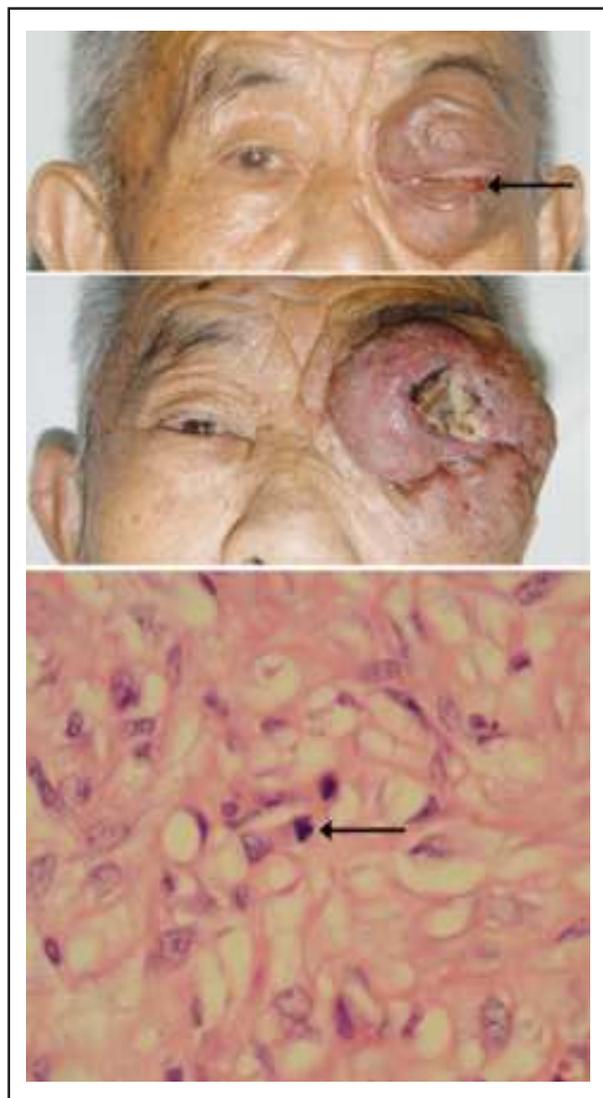


Fig-1-A: Top panel: The appearance of the patient when he first came to the clinic. The arrow points to a blood discharge dot. Middle panel: The appearance of the patient when he came to the clinic 24 months after the operation. Bottom panel: Histological examination of the tumor showed that the fusiform tumor cells were surrounded by a dense reticulin meshwork. Increased cellularity and nuclear pleomorphism (as the arrow points out) were found in more than 4 per 10 high-power fields.

surrounding the individual tumor cells, the tumor cells expressed Vim (Fig-2B), CD34 (Fig. 2C), CD99, smooth muscle antibody, and bcl-2, but not cytokeratin (Fig-2D), co-factor, epithelial membrane antigen, desmin, myogenic differentiation antigen 1, S-100, actin, or CD68.

The tumor recurred 21 months after the operation and grew rapidly. The patient came to our clinic 24 months after the operation (Fig-1A middle panel),



Fig-1-B: Top and middle panels: Computed tomography showed a 7 × 6 cm giant mass with asymmetry enhancement and an irregular boundary in the left orbit, which pressed the eyeball into an abnormal shape. The arrow points to the eyeball. Bottom panel: Magnetic resonance imaging showed a huge soft tissue mass in the left orbit which had already invaded the ethmoid sinus and the sphenoid sinus. The lateral cerebral ventricle was pressured to be flat.

complaining of intermittent unconsciousness and hemorrhage in the left orbit caused by coughing. MRI revealed a huge soft tissue mass in the left orbit which had already invaded the ethmoid sinus and the sphenoid sinus with an equal T1WI signal and an uneven equal-high-mixed T2WI signal (Fig-1B bottom panel). The patient died of brain metastasis six months later.

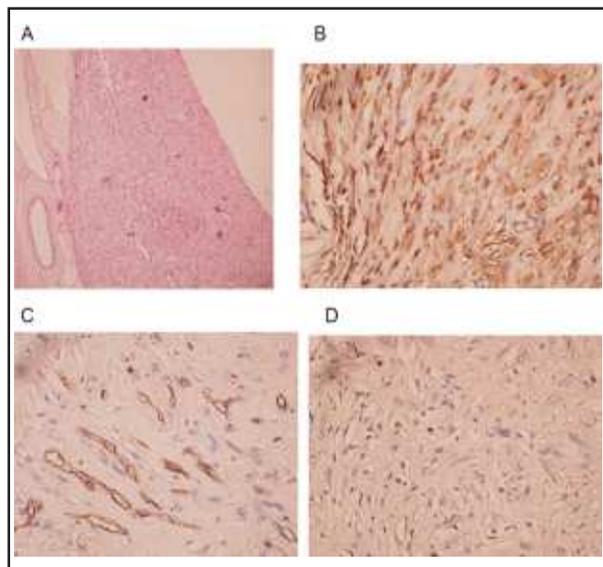


Figure-2: A: HE 100 \times : Histological examination of the tumor showed that there were rich blood vessels with different sizes and shapes throughout the tumor. Some had lumen sometimes branched like antlers, while others were pressed into crack-like or even occlusion, Vascular channels lined with flat endothelial cells, and tumor cells were arranged around blood vessels. B: VIM (+) 400 \times : Immunohistochemistry analysis indicated that the tumor cells were round to fusiform in shape, with a dense reticulin meshwork surrounding the individual tumor cells. The tumor cells expressed vimentin. C: CD34 (+) 400 \times : The tumor cells expressed CD34 too. D: CK (-) 400 \times : The tumor cells did not express cytokeratin.

DISCUSSION

The prognosis for patients with hemangiopericytomas is very poor and an effective treatment modality is lacking. Its poor prognosis is associated with large tumor size, increased mitotic rate, a high degree of cellularity, immature and pleomorphic tumor cells, and foci of hemorrhage and necrosis.³ The two previously reported cases of the disease were both female^{1,2}, with the main clinical signs being a painless swelling tumor and proptosis. Our patient was an 81-year-old man who had a first slowly and then rapidly growing, painless, and sublobe-like mass in the left orbit, which was accompanied by proptosis and massive tumor bleeding, a symptom that rarely has been reported. Trauma is one of the inducements of the disease.⁴ For our case, we suspect that the psychological trauma caused by the loss of a family member may have been a more important factor.

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