Case Report

Primary Haemangiopericytoma outside Muscle Cone in Fossa Orbitalis: A Case Report and Review

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Abstract

Purpose: Orbital haemangiopericytoma (HPC) is a rare tumor with great histological variability and unpredictable clinical and biological behavior. The precise cell type origin is uncertain.

Method: A case report is provided.

Results: A 24-year-old male patient visited an ophthalmologist because of proptosis, strabismus, and movement limitation of the right eye one year ago. These symptoms worsened rapidly. Histopathology, MRI methods and microscopy coil were applied to study the features of HPC. The tumor was removed and the histological examination revealed that it was a primary HPC of the orbit.

Conclusion: MRI determined the size, location, circumscription, morphological detail, growth pattern, and relationship between masses and surrounding tissues. The genesis location of this HPC was rare, and its internal morphological characters were different from cases reported in the literature. The new type of microscope showed more details of tumor MR image characters objectively. (***Eye Science 2012; 27:205–209***)

Keywords: orbit; tumor; haemangiopericytoma; MRI; microscopy coil

Introduction

In 1942, haemangiopericytoma (HPC) was first described by Stout and Murray[1]. It is a rare soft tissue tumor arising from the pericytes of Zimmerman. On histopathological examination, the tumor was characterized by a proliferation of oval and spindle shaped pericytes, and was thought to originate from the pericyte, a specific type of cell surrounding the capillary vessels. HPCs are classified as benign, borderline, and malignant, depending on their histopathologic (mitotic activity, cellularity, and nuclear atypia) and clinical features (necrosis and tumor size)[2]. However, the variable immunohistochemical profile of this tumor and its overlapping features with solitary fibrous tumors led to the supposition that these two tumors were two ends of one process, the origin of which was still unclear, according to World Health Organization endorsement. Despite the dispute, some investigators believed that HPCs arose from pluri-potential peri-vascular cells[3]. HPCs represented approximately 5% of all sarcomatous tumors and could occur anywhere, but they occur much more often in the musculature of the extremities, retroperitoneum, pelvis (uterus, ovary, and urinary bladder), head, neck, and lungs[4,5]. Approximately one-third of all HPCs occur in the head and neck. Only 0.8%-3% of HPC primarily appear in fossa orbitalis[6]. According to the literature, only a few cases of HPCs in the orbit were described[7,8]. The aim of this report is to describe the presentation, investigation, and MR imaging diagnosis of one rare HPC outside muscle cone in fossa orbitalis.

Case presentation

A 24-year-old male presented with exophthalmos in right eye. The eyeball movement was limited and had been progressively more aggravating for one year. The outside of the right eyeball was somewhat oblique (about 25°). He presented with a firm, elastic, and hard swelling of the right side orbit, which had been present for one year and which had caused significant displacement of the eyeball. There was no pain or tenderness and a smooth surface was associated with the lesion. This had not improved with antibiotics prescribed by an ophthalmologist. The clinical examination revealed a robital mass on the right

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eye. The eyelid was obviously swollen, and the eyeball was oblique outside to 25°. Upon palpation, the mass was fixed soft, painless and non-pulsatile. Except for a retracting scar at the site of the attempted biopsy, the rest of the clinical examination was normal. Conditions of the special study were as follows: Vod 0.5 Vos 0.2 (naked eye), Vod 1.0 Vos 1.0 (corrected vision). The eyeground data were as follows: papilla optica was vertical ellipse, retina was ruddy and folded, no exudation and hemorrhage, central fovea of yellow spot existed, and the left eye was normal (Figure 1).

MRI showed an orbicular-ovate tumor with expansive growth in the gap outside the internal muscle cone in the right fossa orbitalis. Its boundary was clear (1.3×1.7×2.3 cm^3). The focus of infection signaled by plain scanning was not uniform. The signal of lesion's substantial part in T_1WI was equal, but it was a little high in T_2WI, Some long T_1 and T_2 signals with different sizes, shapes and lamellar were found inside lesions. Enhancement scanning found obviously uneven intensification. No intensification was found in the internal capsule lesion, and the intensification of lesion peplos was not obvious. The space-occupying effect of the focus of infection was relatively obvious, and the eyeball moved outside because of compression. The three-layer structure of the eyeball wall was integrated, and the MR signal was normal. The medial rectus was thinning because of compression, and gaps in the thinning fat were found between the medial rectus and focus of infection. The nasolacrimal duct was a little outside, which was caused by compression. The display of the tear sac was not good. The imaging characteristics of the tumor were non-specific. The differential diagnosis included vascularized orbital benign masses, such as mesenchymal tumors (cavernous hemangioma) and neurinogen tumors (Figure 2).

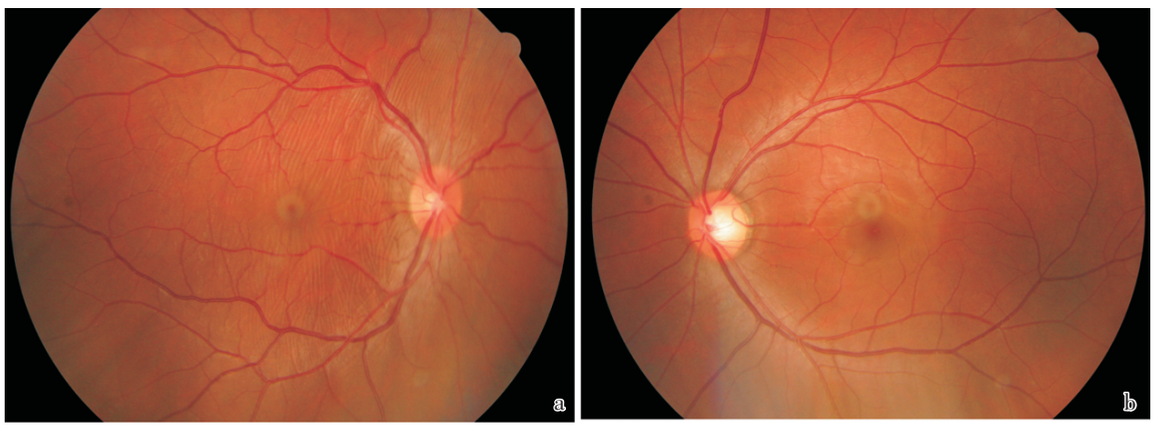

Figure 1 (a) On the right side is a picture of eyeground discus opticus presenting vertical ellipse; retina is ruddy and folded; no exudation and hemorrhage; central fovea of yellow spot exists. (b) On the left is a picture of the eyeground in a normal eyeball.

A systemic work-up, including abdominal sonography and chest CT scan revealed no other associated tumor lesions. Incisional biopsy of the lesion was performed under general anesthesia by opening the orbital tumor anteriorly with radical excision after six months. This technique had the advantages of obtaining good haemotasis and reducing the chance of recurrence. Histopathology showed that the tumor was mainly composed of fat spindle cells, and the arrangement resembled a silting vortex shape. The shapes of the tumor cells were relatively conforming, and the count of the core mitotic figure was about 6/10HPF, indicating possible malignancy. Necrosis focus was found in the tumor. There were many branching blood vessels with thin walls in the mesenchyme, which were infiltrated by many chronic inflammation cells. Folial tumor tissue was found in collagen fibers of partial borderline (Figure 3).

The various investigations led to the final diagnosis that the lesion was a hemangiopericytoma. Chemotherapy and radiotherapy on the lesion was perfromed. No perforation of the cortical plate was

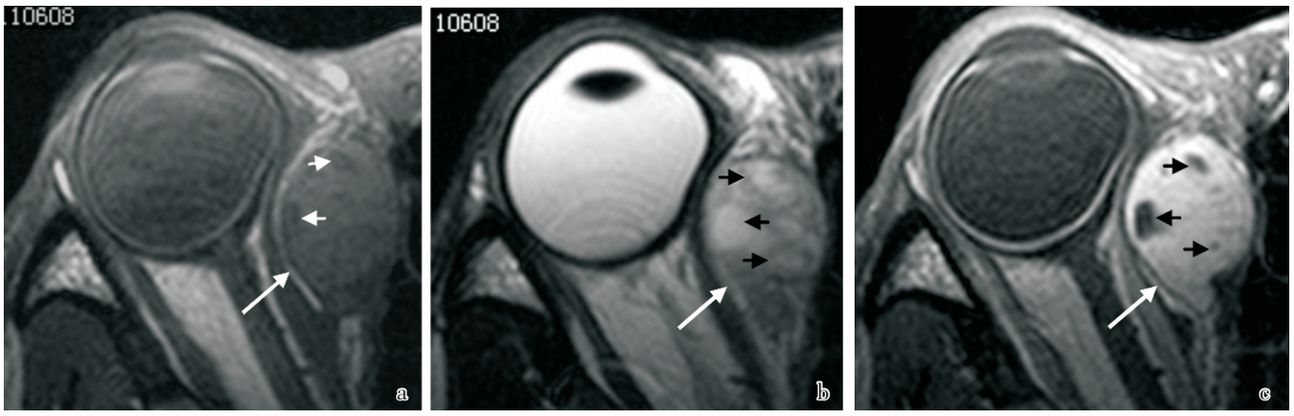

Figure 2 24-year-old male, MR plain scanning and enhancement scanning of ocular MC. (a) T_1WI lateral axis surface indicated that there was an orbicular-ovate lesion in the gap outside of internal muscle cone in right fossa orbitalis; the boundary was almost clear; some lamellar low signal zones of necrosis with different size were found in tumor (short arrow); medial rectus was thinning because of compression; the thinning fat gaps were found between medial rectus and focus of infection (long arrow). (b) T_2WI lateral axis surface showed that the signal of lesion was various; the signal in substantial part was a little high; there were many lamellas of different sizes and shape inside the lesion; their signal was from middle to obviously high (short arrow). (c) Enhancement scanning showed that a substantial part of the tumor was obviously reinforced, and there was no reinforcement in its degeneration zone of necrosis (short arrow). The long arrow shows the thinning adipose layer caused by compression, and the enhancement of lesion peplos was not obvious.

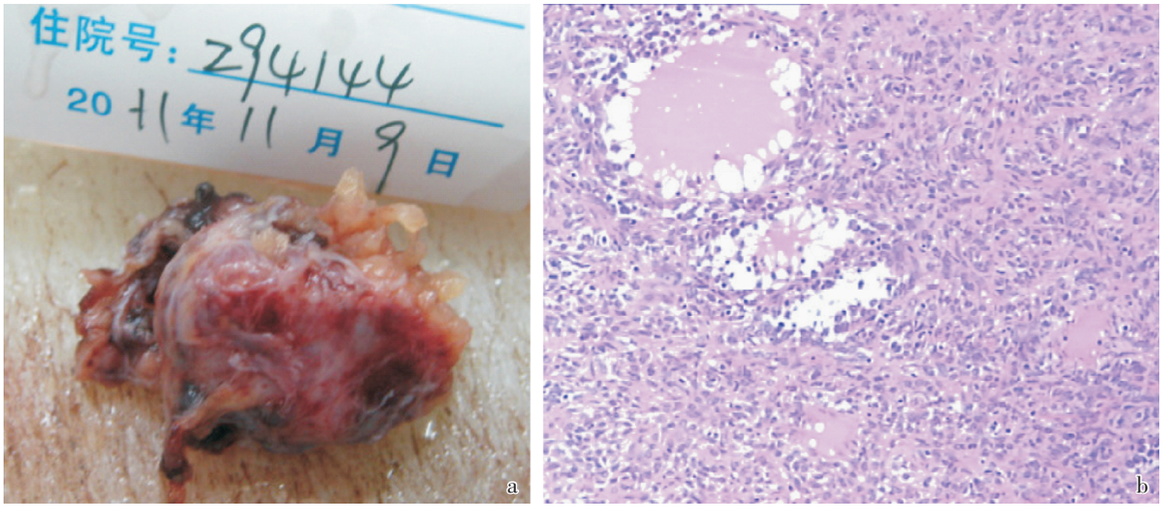

Figure 3 24-year-old male, HPC pathological results. (a) HPC general figure; (b)tumor was composed of fat spindle cells, and the arrangement resembled a silting vortex shape; the shape of the tumor cells was relatively conforming; there were many branching blood vessels with thin walls in mesenchyme; and many chronic inflammation cells were infiltrating them; folial tumor tissue was found in collagen fibers of partial borderline.

noted at the time of treatment. Currently, the patient remains well and is under regular review.

Discussion

Hemangiopericytoma is a mesenchymal neoplasm originating from pericytes—a cell type surrounding capillaries. It is classified as benign, borderline, and malignant, depending on its histopathology (mitotic activity, cellularity, and nuclear atypia). The features of malignant transformation are increased by mitotic activity, a higher cell density, an appearance of undifferentiated cells as well as necrotic and hemorrhagic zones in the tumor tissue. Cytogenic abnormalities have been found in hemangiopericytoma[2]. The HPC pathological results in this case showed that the tumor was mainly composed of fat spindle

cells, and the arrangement resembled a silting vortex shape. There were many branching blood vessels with thin walls in mesenchyme, and the lumens of blood vessels were narrow, but no hemorrhage was found under microscope. Many inflammation cells infiltrated the tumor, and the content of lattice fibers was different. There were many focuses of mucus degeneration, and a zone of necrosis was found. The count of core mitotic figure was about 6/10HPF, indicating possible malignancy. The results were almost consistent with those previously reported in the literature.

Primary orbital haemangiopericytoma is a well-defined and slowly growing mass. Clinically, it affects any age without differences in gender and race but has a greater incidence in patients between 30 and 60 years. Previous studies reported that orbital haemangiopericytoma primarily appeared in adult or child[7,9]. In addition, Ding Y et al. found that most cases of orbital haemangiopericytoma occurred in the prior orbit and downward displacement of the globe. If the tumor mass was large enough, it might produce other problems, such as[8] ambiopia, pain, hemorrhage, etc. In the case reported here, a tumor with expansive growth occurred in the gap outside the internal muscle cone in the right fossa orbitalis. Its volume was somewhat large, and the boundary was clear. The space-occupying effect of the focus of infection was relatively obvious. The eyeball moved outside because of compression. The medial rectus was thinning because of compression, and the nasolacrimal duct was slightly outside again because of compression. The display of the tear sac was not good. The main displays of the patient were anorthopia, exophthalmos, eyelid swelling, ocular movement disorder, etc. This was almost consistent with reports in literature. After a radical excision to open the orbital tumor anteriorly, eyelid swelling and exophthalmos were reduced obviously. There was no limitation of ocular movement in every quadrant, the visual activity test was similar to pre-operation. No damage to the medial rectus, superior oblique muscle, or cranial nerve was indicated.

Primary orbital haemangiopericytoma is rare, its sizes differ, and the clinical symptoms vary with different locations. In addition, the structure is multiple, and the structures of important tissues are many and tiny. In this case, many significant nerves and blood vessels penetrated the tissues, such as the optical nerve, ophthalmic artery, ophthalmic vein, and sensory nerve accompanying with extraocular muscles. Therefore, the correct evaluation of the location of HPC, its interior characteristics, and the relationship with surrounding nerves and blood vessels was the guarantee of a successful operation[10].

After tracing 17 cases of HPC ultrasonographic images, DingY et al[8]. found that the boundary of benign HPC was clear, but malignant HPC was presented as invasion change with unclear boundary. In addition, the quantity, intensity, and distribution of echos inside the tumor were closely correlated to the tissue characteristics of the lesions. If there were more blood vessel antrum cavities inside lesions, then the echo would have been greater and intensive. However, the echo was less in the tumor, which was composed of flaky tumor cells, and some areas were even without echo. Atul et al[11]. reported one HPC inside muscle cone behind the eyeball, and the main display of MR showed was placed behind the ball with aclear boundary. The signals were equally T_1 and T_2, the plain scanning signal was uniform, and there was obvious intensification with enhancement scanning. This tumor was rich in blood vessels and supplied blood by the ophthalmic artery after the angiography. Arshad and Valentini[12,13] reported HPC images separately, showing that the tumor was a cycloid soft-tissue space occupying the same isodensity or irregular shape, but the boundary was clear. There was an obvious intensification after injecting the contrast medium, which further indicated rich blood vessels inside the tumor. Much constrast medium was left inside the blood, on the other hand, because of damage to the blood-tissue barrier. Much constrast medium was exuded from the blood vessels. In this case, HPC appeared outside the muscle cone in fossa orbitalis. The relationship between the tumor and the medial rectus was close. The incidence of HPC in this location was rare, so reports on it were not found in previous studies. In addition, MC displayed more details on MR images of the tumor objectively than HC. The signal in T_1WI was equal but a little high and uneven in T_2WI. Some long T_1 and T_2 sig-

nal shadows with different sizes of lamellars were found inside it. There was uneven intensification inside a substantial part of the lesion with enhancement scanning. Because there was no intensification of abnormal signals in the area of the lamellar, we thought that signal area of lamellar was caused by some tissues left around the blood vessels. Combined with postoperative pathological results, most tumor cells died from abnormal characteristics. The new blood vessels functioned to make the tumor grow quickly. This presentation was not found in previous studies.

This report describes the long-term follow-up of a patient with a rare primary HPC arising from the orbit. As a noninvasive bio-autogyration imaging technology, MRI showed high resolution of soft tissues and the images of any section. MRI helped to determine the size, location, circumscription, morphological detail, growth pattern, and relationship between the masses and surrounding tissues. In addition, the new type of MC showed highly detailed characteristics of the tumor in MR images objectively, and provided more information for diagnosis. This study also emphasized the need to understand the rare genesis location of HPC and to observe its untypical MR imaging characteristics, combined with pathology and long-term follow-up after surgery in such tumors.

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