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Intracranial Reticular Hemangioendothelioma: Report of a Rare Case

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Abstract

Reticular hemangioendothelioma is a rare intermediate vascular tumor. Only about 70 cases have been reported in the literature. The tumor develops in the skin and subcutaneous tissues, and there are no reports of RH with intracranial origin. In this article, we report the clinical data of a patient with intracranial reticular hemangioendothelioma and summarize the diagnostic and therapeutic features of intracranial reticular hemangioendothelioma in the light of the literature.

Key words: Reticular; Vascular Endothelioma; Intracranial; Rare

Abbreviations: RH: Retiform Hemangioendothelioma; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; WHO: World Health Organization

Introduction

Reticular hemangioendothelioma (retiform hemangioendothelioma RH) is a rare intermediate-type tumor of vascular origin. The tumor consists mainly of an elongated, branching testis-like vascular network with peg-like endothelial cells. It was first reported by Calonje et al. in 1994 [1], and only about 70 cases have been reported in the national and international literature [2]. The tumor is most commonly found in the skin and subcutaneous tissues, with a few reports of occurrence in rare sites such as the pleura [3]. There are no reports of RH with intracranial origin. The aim of this study is to analyze the pathological features, clinical manifestations, imaging presentation and treatment of this disease based on previous reports in the literature.

Case Reports

The patient, a 19-year-old female, was admitted to the Department of Neurosurgery, Banan Hospital of Chongqing Medical University on September 8, 2023, the patient started to have headache 2 weeks ago, which was mainly characterized by left-sided head swelling and pain. The patient did not pay attention to it and did not go to the hospital. Four hours before admission, the patient had a sudden convulsion with loss of consciousness in front of his home, which stopped on its own after a few minutes. The ambulance sent to the local hospital after the examination of cranial CT suggests left temporal lobe occupational lesion possibility, bleeding to be drained. Re-transferred to our hospital. Admission examination showed no obvious neurological abnormalities. After admission levetiracetam antiepileptic and mannitol decongestive therapy were given. Examination of MR showed that a mass of about 44x34mm in size was seen in the left temporal region (Figure 1), which was considered to be more likely to be an angioepithelioma, with meningiomas to be drained. From the imaging, the intracranial tumor blood supply was very rich, and the surgery might bleed a lot. The treatment plan was chosen to perform preoperative angiography to clarify the blood-supplying arteries of the tumor, and then interventional embolization of the blood-supplying arteries to reduce intraoperative bleeding before total resection of the intracranial tumor. Cerebral angiography + embolization of tumor blood-supplying arteries was performed under local anesthesia at 09:00 on August 25, 2023 (Figure 2). After successful embolization, craniectomy for left temporal lobe occupying lesion was performed on August 26, 2023, at 1005 hours under general anesthesia. The operation went well and the tumor was seen to originate from the meninges at. Surgical total resection of the

tumor was performed, and the tumor envelope is intact (Figure 3A). After surgery, levetiracetam antiepileptic treatment was continued. Postoperative follow-up cranial MRI showed that the tumor was completely resected (Figure 4A and B). Postoperative pathology showed extensive vascular proliferation within the lesion with lobulated structures, significant anastomosis of the proliferating vessels, and papillary proliferation of endothelial cells in some areas. A fibrous peripheral envelope was seen around the lesion, which was clearly demarcated from the brain tissue. Immunohistochemistry: EMA(-), Vim(+), PR(-), GFAP(-), S100(-), SOX10(-), CD34(+), hCD31(+), ERG(+), Desmin(-), SMA(weakly +), actin(weakly +), Calponin(-);EMA(-), CK(-) , Vim(+), PR(-), ER(-), S100(-), Ki67 (~2%+) (Figure 3B-F). No postoperative radiotherapy was performed because the surgery had been total excision. The tumor did not recur during the six-month follow-up after surgery (Figure 4C and D).

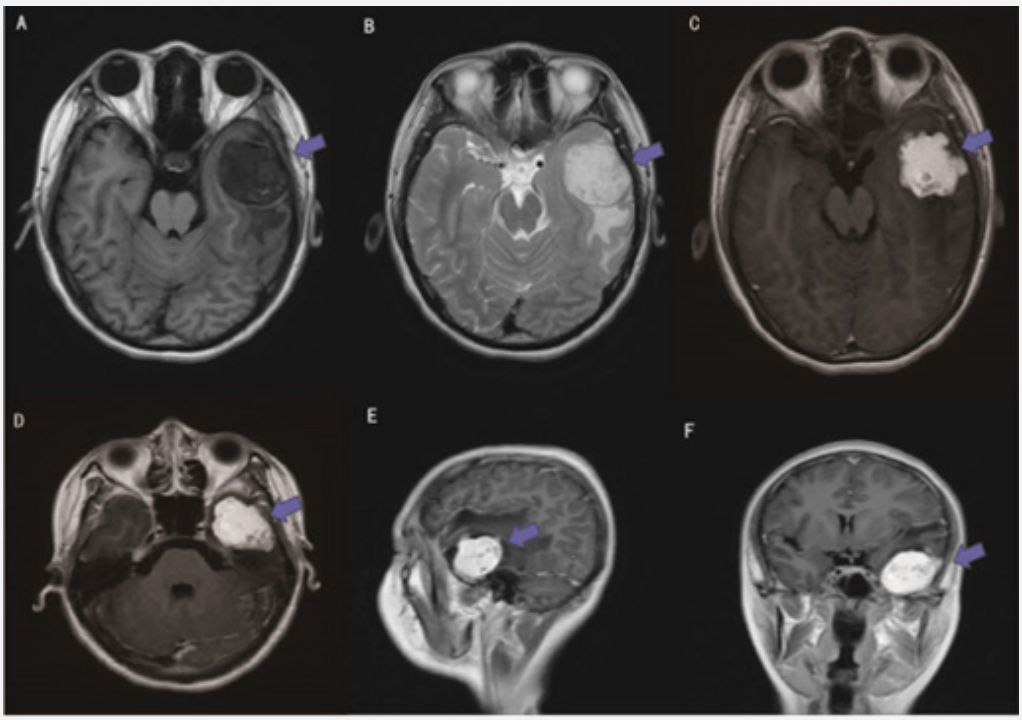

Figure 1: A The tumor is low signal in T1-weighted image with clear boundary. The tumor is surrounded by a large edematous band of high signal in the T2-weighted image. C-F enhancement scan, the tumor is significantly strengthened.

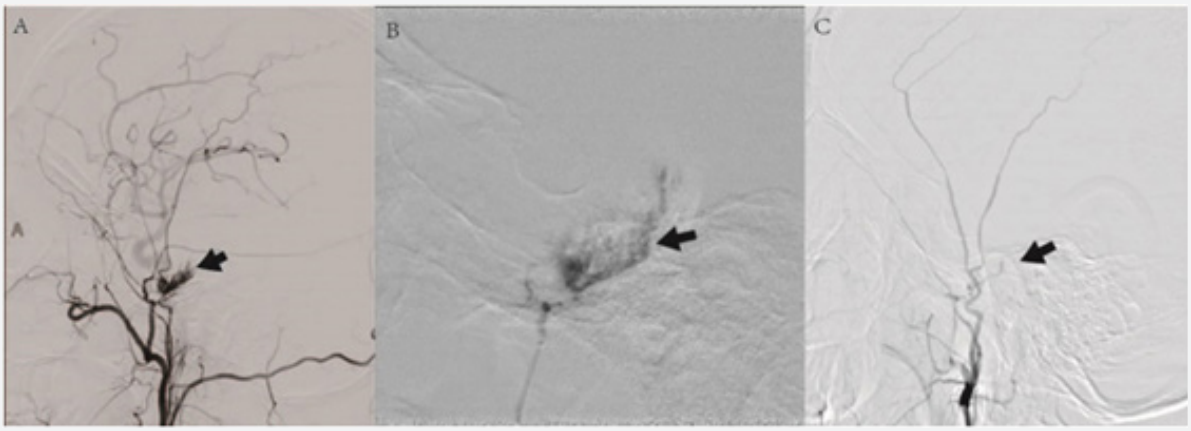

Figure 2: A The involvement of the superficial temporal artery branch of the external carotid artery in the blood supply of the tumor was seen on imaging. B The tumor-supplying artery was embolized with PVA particles. C The tumor was not visualized on imaging of the external carotid artery after embolization.

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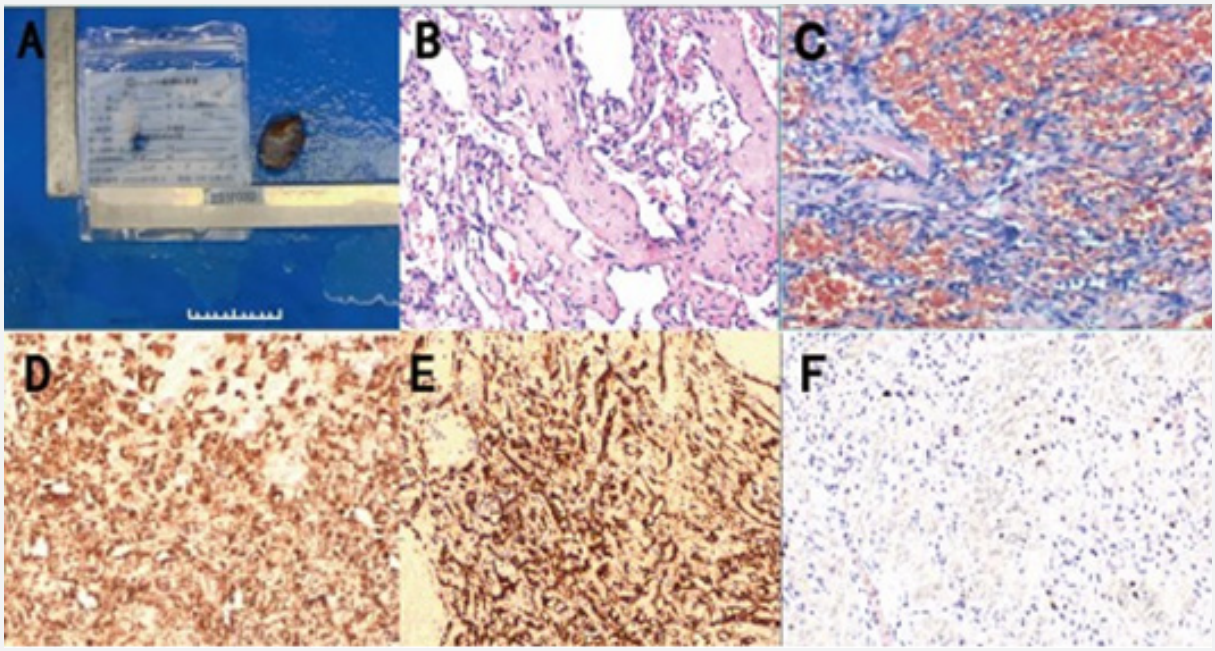

Figure 3: A Grayish grayish brown irregular mass 4x2.6x2.2cm, with multiple nodule-like protrusions on the surface, grayish brown in section, medium texture, and intact peritoneum; B Microscopic H&E staining showed irregular vascular meshwork with anastomosing vessels ; C Vascular endothelial cell hyperplasia in the form of a typical “shoe peg” shape; D,E CD34 and CD31 staining showed abundant blood vessels; F Ki67 staining showed scattered positive cells (2%).

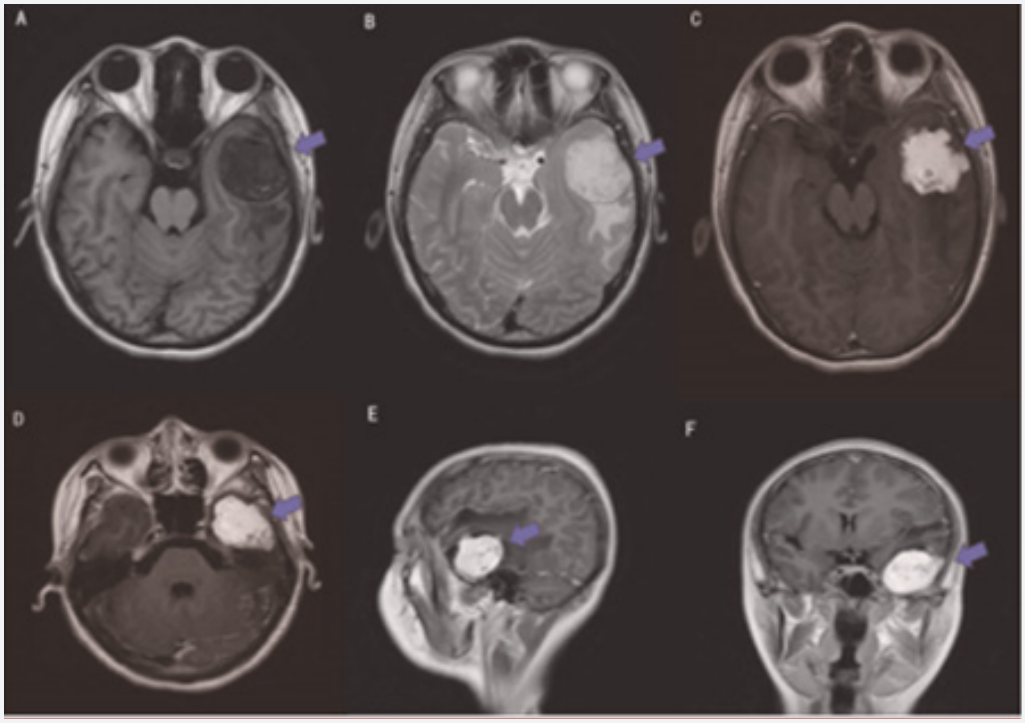

Figure 4: A and B. 3-day postoperative review shows that the tumor has been completely resected; C and D. 6-month postoperative review shows that the tumor has not recurred.

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Discussion

The 5th edition of the WHO Soft Tissue Tumor Classification released in 2020 defines RH as a rare intermediate vascular tumor that rarely metastasizes [4]. Intermediate (rarely metastasizing). RH was first reported to occur in all age ranges with no apparent gender predilection [1]. However, recent reports have shown that RH occurs predominantly in young and middle-aged people, with more females than males [3,5,6], which is consistent with the case reported herein.

RH occurs in the skin and subcutaneous tissues throughout the body, mostly as exophytic or patchy skin lesions, usually as solitary nodules or plaques of blackish red to brownish, purple color with pain, or the skin may show only thickening, elevation, and swelling, and there may be fusion of the nodules, thickening of the skin, or formation of vesicles or ulcers [7,8]. There have been 2 cases reported to occur in the pleura and both patients had dry cough and dyspnea [9,10]. There are no reports of RH with intracranial origin, and only two cases of RH originating from the skull have been reported worldwide [11,12]. These two cases originated from the parietal bone and the pterygoid bone, respectively, and both of them had bone destruction. The tumor in the case reported in this paper originated in the meninges, with destruction of the meninges, without affecting the skull, and the tumor compressed the nerves causing epileptic symptoms.

Imaging manifestations of RH vary depending on the origin. CT manifestations of pleural RH are mainly irregular masses [9,10]. The imaging manifestations of RH of bone origin all have osteolytic bone destruction [11-13]. The present case is the first reported case of intracranial RH, which has some of the features of benign and malignant intracranial tumors. The CT presentation of RH was a well-defined slightly low-signal mass. Like other intracranial tumors, the main preoperative diagnosis relied on MRI. The mass was low signal in T1-weighted images and high signal in T1-weighted images with clear borders. Due to the slightly rapid growth of RH, mixed signal shadows may appear within the tumor. Due to the very rich blood supply of tumors of vascular origin, obvious enhancement of the tumor can be seen on enhancement scan. MRS can be used as an auxiliary diagnostic test, and RH tumors have low NAA wave peaks and no neuronal markers, suggesting non-neuronal brain tumors. Decreased Cho and increased ML peaks are in line with the characteristics of intermediate tumors.

The diagnosis of RH is mainly based on pathological examination, and its histological features mainly include Extensive blood vessels in the tumor are irregularly proliferating and anastomosing with each other to form a reticulum. Vascular endothelial cells are uniform in size, with little cytoplasm, and some endothelial cells have protruding nuclei, which appear as “boots nails” into the lumen of the vessels. There is no nuclear heterogeneity or mitosis. Immunohistochemistry showed positive expression of vascular endothelial cell markers CD34, CD31 and ER G [13,14]. This case showed typical reticular vascular hyperplasia and the “boot nail” sign, which, combined with immunohistochemistry, was consistent with the pathologic diagnosis of RH.

Surgical total resection is the treatment of choice for RH. Surgery is challenging because of the very rich blood supply of RH and the relative difficulty of intracranial hemostasis. Preoperative cerebral angiography is recommended, and if branches of the external carotid artery can be found to be involved in the blood supply of the tumor, these blood-supplying arteries should be embolized immediately. Postoperative adjuvant therapy needs to be decided according to the type of pathology and surgery. Local radiotherapy combined with cisplatin chemotherapy is recommended for patients with incompletely resected tumors and with lymph node metastases [7]. Postoperative pathology should focus on Ki67. For patients with completely resected tumors, local radiotherapy is recommended if Ki67 is >15%. For patients with Ki67>30%, local radiotherapy combined with chemotherapy is recommended. Metastasis of RH is rare, but recurrence is possible, and strict follow-up is needed after surgery. For recurrent RH, surgery combined with postoperative radiotherapy is recommended.

Conclusion

In summary, RH matters a rare intermediate-type vascular tumor, and intracranial RH is very rare. It is difficult to diagnose clinically, and the diagnosis should be confirmed by pathology. Surgical resection is the first choice of treatment, and patients can benefit from preoperative embolization of the blood-supplying arteries because of the rich blood supply of the tumor. Radiotherapy is effective in postoperative adjuvant treatment of RH, depending on the type of pathology and the surgical situation, and RH has the potential to recur, requiring strict long-term follow-up after surgery. Nowadays, the number of reports on intracranial RH in the world literature is low, and the human exploration of the diagnosis and treatment of this disease is still continuing, which requires more case experiences and persistent follow-up by scholars around the world.

Consent for Publication

The patient has given written consent to the publication of this case report and any accompanying images.

Author’s Contribution

BXH and KYL performed literature review and drafted the manuscript. JHH helped to collect and review radiological data. CFG performed the histological and immunohistochemical evaluation.LY operate on the patient and revised the manuscript. All authors contributed to this paper and approved the submitted version.

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Conflict of Interest

The authors declare that this article does not contain any commercial or financial relationships with potential conflicts of interest.

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