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Case Report

A unique case of benign intracranial hemangioma mimicking malignant transformation

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ABSTRACT

Capillary hemangiomas are rare benign vascular lesions, commonly found on scalp, face, chest, or back of a neonate or infant. Hemangiomas of the central nervous system are very rare lesions. There are only a few cases of intracranial capillary hemangioma (ICH) arising in adults reported in the literature. We present a case of 59-year-old female with intermittent recurrent headache localized in the frontal area. Magnetic resonance imaging revealed left frontal extra-axial mass with peripheral enhancement. The patient underwent complete surgical resection of the tumor. Histopathology examination of the lesion revealed well defined vascular lesion composed of closely packed plump endothelial cells lining slit-like vascular channels containing scattered red blood cells. No evidence of infiltrative brain parenchyma was seen. Ki-67 proliferative index was low, less than 2%. The final diagnosis was confirmed to be ICH by histopathology and immunohistochemistry studies. The patient has remained healthy and free of disease 39 months since her initial surgery. ICH is a benign vascular lesion which rarely occurs in the central nervous system, particularly in the intracranial region. It can mimic malignant lesions on radiologic studies. Histopathology examination is the gold standard for diagnosis. If total resection is achieved, prognosis is generally good with no evidence of recurrence.

1. Introduction

In 1867, Virchow first described hemangiomas [1]. Hemangiomas are benign vascular tumors, or tumor like-lesions. These lesions typically grow in the skin, soft tissue, face, scalp, and trunk. Neonates and infants are the most affected age group. Ten to twelve percent shows tendency to grow within the first year of life [2]. Hemangioma can rarely grow in adults, with a slight female predominance, which undergoes remarkable size change due to pregnancy and hormonal cycles' response [3]. Clinically hemangioma goes through a proliferate phase before proceeding into an involutionary phase [4]. Most hemangiomas express spontaneous regression with age progression. Histologically, hemangiomas are classified into two main subtypes: capillary and cavernous. Capillary hemangiomas (CHs) are composed of lobules separated by variable degree of fibrous bands. These lobules are cellular due to the plump endothelial cells lining the vascular spaces and poorly defined capillary channels. Cavernous hemangiomas are large cystically dilated blood vessels with thin walls. In-

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travascular thrombosis or calcification is frequent. Immunohistochemistry highlights the endothelial cells lining in classic slit-like spaces. CHs of the central nervous system are rare diagnosed pathology [2]. Although spinal nerve roots and cauda equina are the favorable locations for CH, CH within the brain is unusual. Review of the literature revealed that most of the intracranial capillary hemangioma (ICH) was diagnosed originally by brain computed tomography and magnetic resonance imaging as meningioma prior to surgical resection. Due to studies limitation and few reported cases about ICH, the estimated rate, prevalence, and radiological findings might be underestimated. In this paper, we present a case of a 59-year-old female with left frontal extra-axial mass with peripheral enhancement, confirmed histologically as an ICH.

2. Case presentation

A 59-year-old female was admitted to the emergency department complaining of intermittent recurrent headache for the past 3 months, localized in the frontal area. The headache resolved spontaneously without medication. No associated symptoms was identified. The patient demonstrated normal vital signs and no focal neurologic deficit on physical examination. Magnetic resonance imaging of brain was performed utilizing standard technique with gadolinium administration. The study showed a peripherally located extra-axial mass lesion in the left frontal region, measured 3.4 cm Anteroposterior × 2.6 cm transverse × 1.4 cm craniocaudal. The lesion caused significant vasogenic edema in the surrounding brain parenchyma. Postgadolinium images showed strong predominantly peripheral enhancement within tumor mass with probable central area of necrotic component identified (Fig. 1A and B). Perfusion study showed significant increase within the peripherally enhancing aspect of the tumor (Fig. 1C). The top differential diagnosis was meningioma with atypical features and significant surrounding edema, or astrocytoma. The patient underwent a navigation-guided brain mass resection under general anesthesia. Solid round mass was identified, closely related to the brain parenchymal tissue, which was completely removed. Intraoperative bleeding was successfully controlled. Postoperatively, the patient was stable, doing well. Postoperative examination reveals focal hematoma without definite active tumor enhancement detected (Fig. 1D). There was a significant decrease in the surrounding vasogenic edema compared to the preoperative image. The resected specimen was sent to anatomic pathology department for prober evaluation. Macroscopic examination revealed an oval mass, well-defined, partially capsulated measures 3.5 × 2.5 × 1.5 cm, homogenous pink tan cut-surface with hemorrhage cystic spaces. No area of necrosis was seen. Microscopic examination revealed a well demarcated vascular lesion (Fig. 2A) composed of thin vascular spaces lined by delicate plump endothelial cells. These spaces are filled with red blood cells (Fig. 2B). No intervening glial parenchymal tissue was seen. No evidence of nuclear atypia, mitotic figures, necrosis, and apoptosis was seen. No evidence of intracytoplasmic hyaline body globules, extramedullary hematopoiesis was seen. Immunohistochemistry studies revealed diffuse positive antigenicity to endothelial cells markers including CD31 (Fig. 2C), CD34 (Fig. 2D), factor VIII, and vimentin, while negative for epithelial membrane antigen, S-100, human melanoma black, CD10, smooth muscle actin, desmin, D2-40, alpha-fetoprotein, neuron-specific enolase, glial fibrillary acidic protein, and signal transducer and activator of transcription 6. Ki-67 proliferative index was less than 2%. Histopathology and immunohistochemical studies were consistent with the diagnosis of ICH. The patient had a continuous follow-up after the surgery for 40 months duration, and she is healthy without any complications and/or recurrence (Fig. 1E).

3. Discussion

CHs are seen in 1–2.6% of live births [2]. These tumors demonstrate female predominance, and can undergo hormonal response changes. The pathogenesis of hemangiomas is not fully understood. No single theory can explain the predilection of hemangioma for infants, females, responses to hormonal levels, and spontaneous involution. Most theories suggested the origin of angioblasts, trophoblasts, along with defect in the cytokine regulatory pathways that can initiate the process of angiogenesis of hemangiomas [5]. Only few reports and studies are found in the literature about CH arising primarily in the brain. PubMed search until March 2018 reveals only 19 studies reporting 29 cases of ICH, confirmed by histopathology examination. Majority of ICH was diagnosed in infants and young adults with the age range from 2 weeks to 69 years old. These cases were seen in 14 male and 15 female patients [2]. ICH can arise in the cerebral lobe [6], sagittal sinus [7], cerebellum [6], sellar region [8], cavernous sinus [9], fourth ventricles [10], and anterior choroidal artery [11]. Clinical signs and symptoms are variable and depend on lesion location, ranging from asymptomatic to headache, seizure, and cranial nerve palsy [2]. Radiological differential diagnoses in most of the reported cases were meningioma, astrocytoma, and any glial tumors with high-grade features. Most of them were diagnosed as ICH postoperatively by histopathology examination. Postgadolinium studies reveal the peripheral enhancement mimicking other tumors. Therefore, it is difficult to distinguish ICH preoperatively. Generally, most of the previous cases, including this case were treated with complete surgical resection. Treatment for ICH includes surgical resection, embolization, laser treatment, β-blockers such as propranolol, corticosteroids, interferon. However, these different modalities are not clearly standard in the literature [12]. When gross total resection of the lesion cannot be achieved, the patient should be observed frequently, with consideration of adjuvant radiotherapy [3]. Histopathology examination is the gold standard for diagnosis. Hematoxylin and eosin stain can easily highlight the delicate vascular channels lined by endothelial cells. Most cases in the literature were described as capillary type intracranial hemangioma. However, one case reported a mixture of capillary and cavernous intracranial hemangioma [13].

The differential diagnoses include hemangioblastoma, hemangioendothelioma, and hemangiopericytoma. Heman-

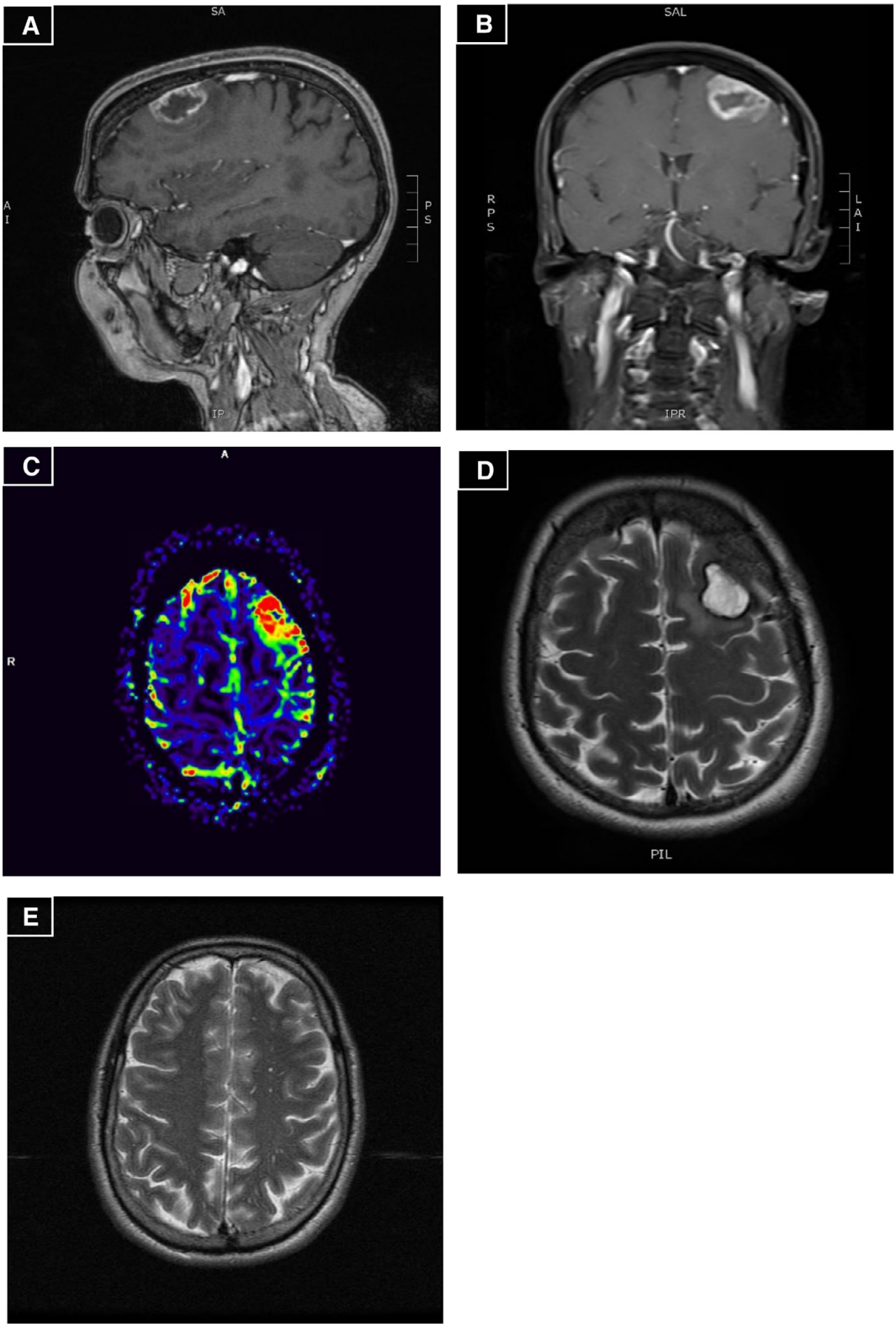

Fig. 1 – *Intracranial capillary hemangioma with avid peripheral enhancement.* (A) Sagittal and (B) coronal magnetic resonance imaging (MRI) view showing extra-axial mass lesion in the left frontal region with strong predominant peripheral enhancement. (C) Perfusion study showed significant increased perfusion within the peripherally enhancing aspect of the tumor. (D) Postoperative examination reveals focal hematoma without definite active tumor enhancement. (E) Axial MRI scan performed on the patient ~39 months following initial surgery, no recurrence of the tumor was detected.

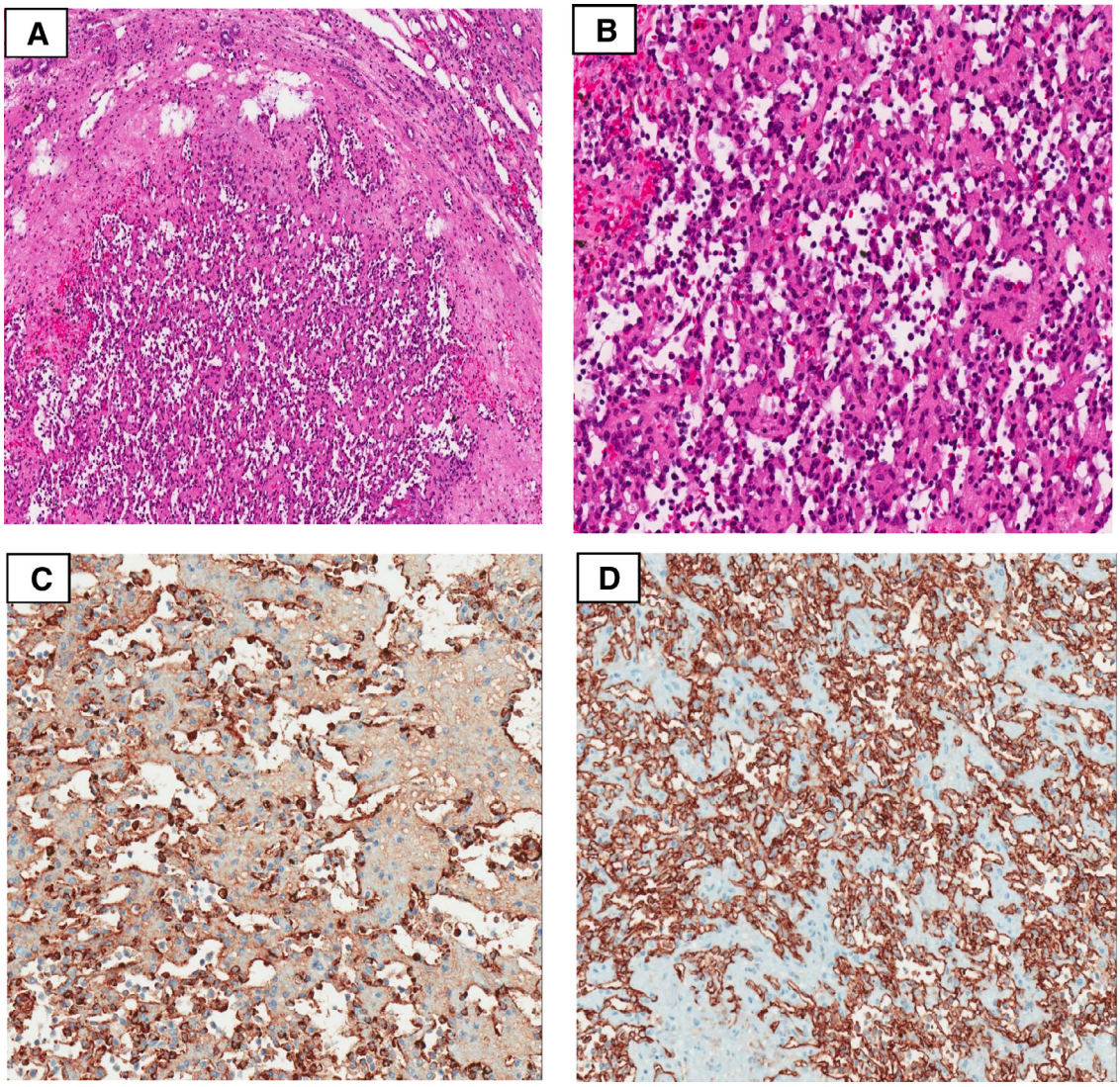

Fig. 2 – Intracranial capillary hemangioma. (A) Low-power examination reveals a well-demarcated lesion, partially capsulated vascular lesion (hematoxylin and eosin (H&E); 4×). (B) High-power examination shows vascular spaces lined by plumb endothelial cells containing red blood cells (H&E; 40×). (C) Immunohistochemistry stain reveals positivity for CD31 in the vascular spaces (40×). (D) Immunohistochemistry shows positivity for CD34 in the vascular spaces (40×).

gioblastoma is a slow growing and indolent tumor, that arise commonly in young to middle-age, typically in the posterior fossa. They can be either sporadic or associated with von Hippel–Lindau disease associated. They typically present with cyst with mural enhancing nodule and are composed of neoplastic cells showing nuclear pleomorphic changes, lipidized stromal cells, and highly vascular background. Foci of extramedullary erythropoiesis may be seen. These features were not detected in our case. Hemangioendothelioma was excluded from our differential. It shows well-differentiated nests and cords of cells with abundant eosinophilic cytoplasm and prominent intracytoplasmic lumina, commonly seen in epithelioid form. Kaposiform hemangioendothelioma are biphasic tumors with both vascular and lymphatic component, demonstrating irregular, infiltrating nodules of compressed blood vessels, with a dense hyaline stroma. Retiform hemangioendothelioma expresses a net-like pattern of infiltrative growth. Pseudomyogenic and composite subtypes are rare form of bones and soft tissue hemangioendotheliomas. All of these patterns of growth were not observed in our case [14, 15]. Hemangiopericytoma characteristically grows in a sheet like, storiform pattern, around an intratumoral staghorn blood vessel. All of the above-mentioned differential diagnoses were against our case. Overall, ICH are usually benign, slow growing tumors with good prognosis and long free survival rate.

4. Conclusion

We present a case of 59-year-old female with ICH. Perioperative radiological diagnosis is usually difficult to make. Al-

though ICH is rare to occur, it should be in the differential diagnosis of defined mass with peripheral enhancement. ICH has a good prognosis with low rate of recurrence after surgical resection.

Ethics approval\Informed consent

Verbal and written informed consent was obtained from the patient for their anonymized information to be published in this article.

Patient consent

Consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

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