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(CASE REPORT)

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A bulbo medullary hemangioblastoma revealed by hemorrhage: A case report

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Abstract

Introduction: Hemangioblastomas are highly vascularized tumors of unclear origin. Most hemangioblastomas arise in the posterior fossa. Although spontaneous hemorrhage from these tumors is extremely rare, despite their vascular nature, we describe a case of a bulbo-medullary hemangioblastoma revealed by hemorrhage.

Case report: This is a 12-year-old child, with no past-history, presented with a picture of subarachnoid hemorrhage (sudden, intense, thunderclap headaches associated with vomiting and stiff neck, for which the Neuroradiological investigation was in favor of a bulbo-medullary lesion which infracentimetric lesion was found, with anapath was in favor of a hemangioblastoma.

Conclusion: Hemangioblastomas are rare tumors that can be revealed by hemorrhage. Imagery can be misleading. In the case with very small lesion, careful neurovascular appraoch is recommended.

Keywords: Hémangioblastomas; Rare tumor; Hemorrhage; Histology; Surgery

1. Introduction

Hemangioblastomas are rare tumors, representing 2 to 3% of central nervous system (CNS) tumors, and are preferentially locatedat the infratentorial (8; 5; 16).

They can be discovered in the form of a single lesion (67%), sporadicform, or in multiple form then entering, most often, within the framework of phacomatosis(33%), Von Hippel-Lindau Disease (VHL) (8; 5; 16).

The majority of infratentorial hemangioblastomas (94, 3%) are located in the cerebellum. The cerebellar hemispheres are 4 times more frequently affected than the vermis. At the level of brainstem,the pre prepontine or pre bulbar ventral forms are exceptional (3).

Hemorrhages caused by hemangioblastomas are very rare. Duringsurgery, an infracentimetric lesion was found. (9; 10)

We report the case of a brain steam hemorrhage of the bulbo-medullary hemangioblastomas.

2. Case report

We report the case of a 12-year-old child, with no past-history, presented with a picture of subarachnoid hemorrhage (Sudden intense, thunderclap headaches associated with vomiting and stiffneck).

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The Neuro-radiological investigation was in favor of a bulbo-medullary lesion that bled., first evoking a pial fistula. (Figure-1-) (Figure-2-) (Figure-3-).

During surgery an infra centimetric lesion was found. After a neuro-radiological exploration, and whose first hypothesis in our case was a pial fistula of the bulbo-medullary junction, the decision of surgical treatment was taken. The patient was operated on based on this hypothesis (pial fistula). Our patient was operated by a classic sub-occipital approach with opening of C1.

Per operatively, we discover a lesion just below the cerebellar tonsils bulging with a thin wall reminiscent of an arachnoid layer which is continued below at the bulbo-medullary level by another hard lesion in a ball, macroscopically, it has a yellowish appearance. On exploration of the latero-bulbar vascular elements: one discovers on the left a large aberrant arterialized vein arriving in contact with the lesion in bubble, associated with a small tortuous much more slender artery. We first perform coagulation of the artery, we notice that the vein collapses and we the proceed to coagulate the vein at the foot of the blunt hook, open the lesion at the bottom of the cerebellar tonsils, with blood coming out, visualize the opposite bulb and remove all the small blood clots inside the cavity until have a good decompression.

We continue the exploration inside the cavity towards the lesion below. The latter is macroscopically yellow is in appearance, hard, and after opening the capsule, we notice a black is hemorrhagic lesion causing bleeding. The lesion is reminiscent of an AVM (arteriovenous malformation) or a cavernoma. We proceeded with the resection of this mass in block until visualization of healthy bulbar parenchyma with a white appearance (Figure 4). The final pathological diagnosis was hemangioblastoma as will (Figure 5). The post-operative evolution was marked by a very good improvement. During surgery an infra centimetric lesion was found, with anapath was in favor of a hemangioblastoma. Remote cerebro-medullary MRI is a very good radiological control (Figure 6)

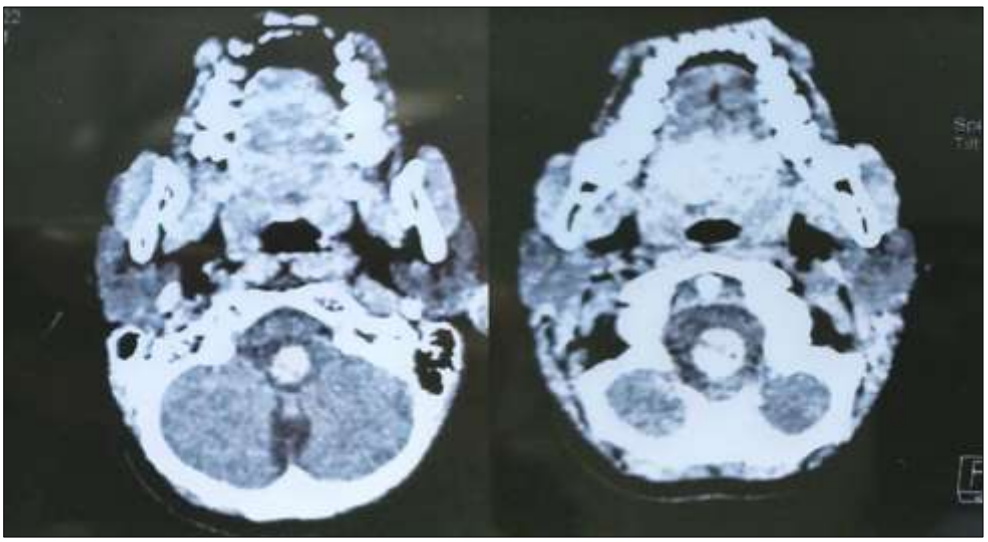

Figure 1 CT scan without injection, which shows a bulbo-medullary hematoma-

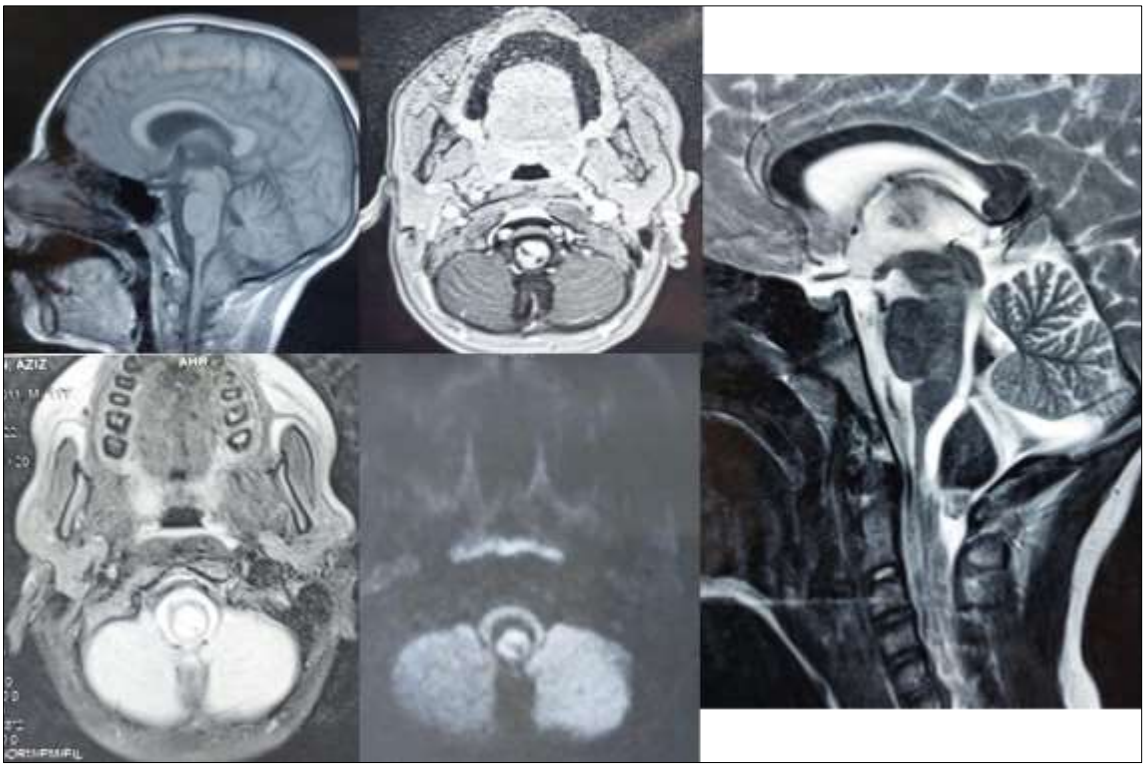

Figure 2 Brain MRI showing a bulbo-medullary lesion with hemorrhage radiologically suggestive of a pial fistula

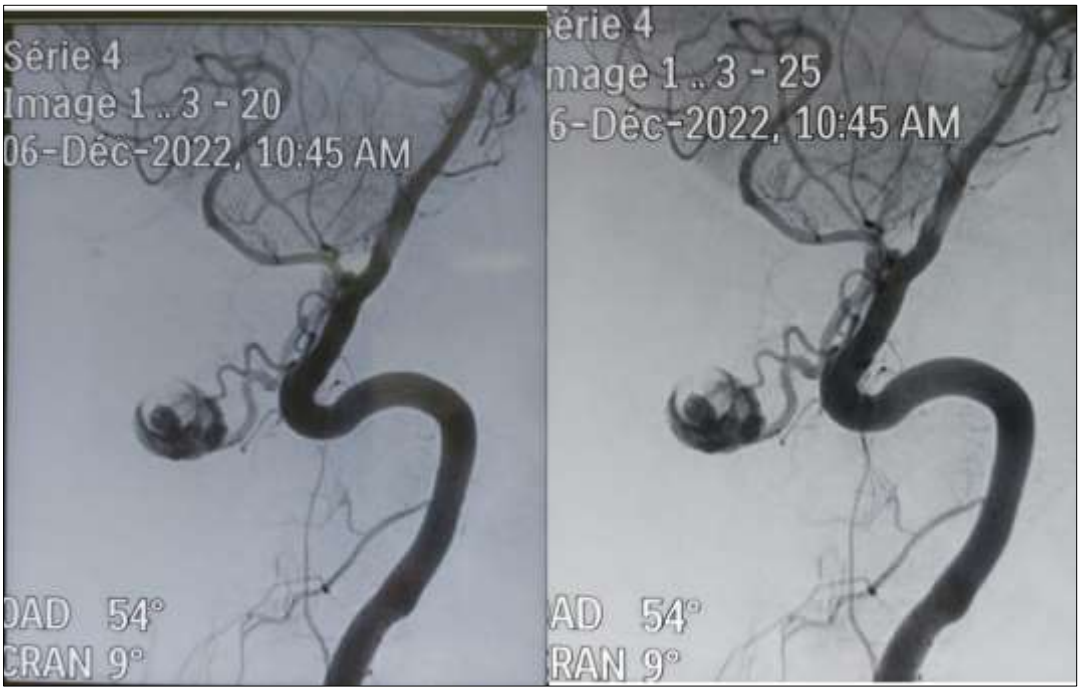

Figure 3 Cerebral arteriography, which shows: a small mass with a large arterialized vein with a small arterial branch that flows directly into the vertebral artery

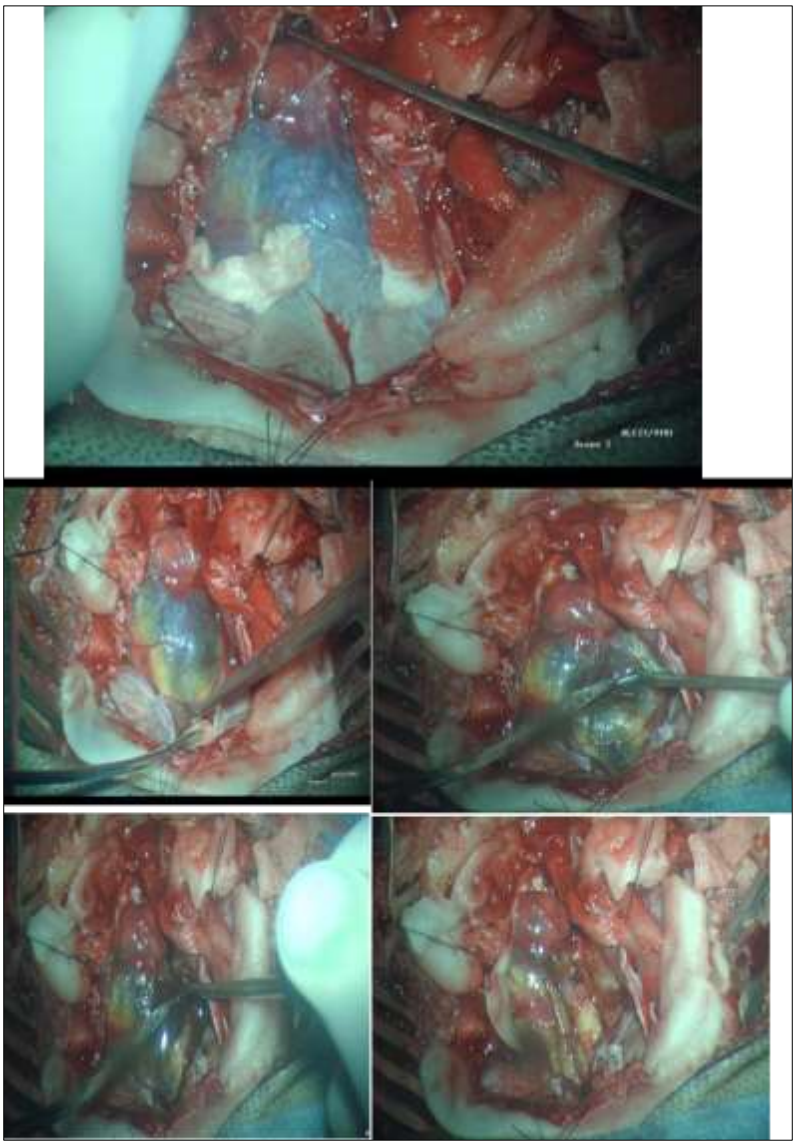

Figure 4 Intra operative images showing the appearance of the lesion ball, lesion with intratumoral hematoma

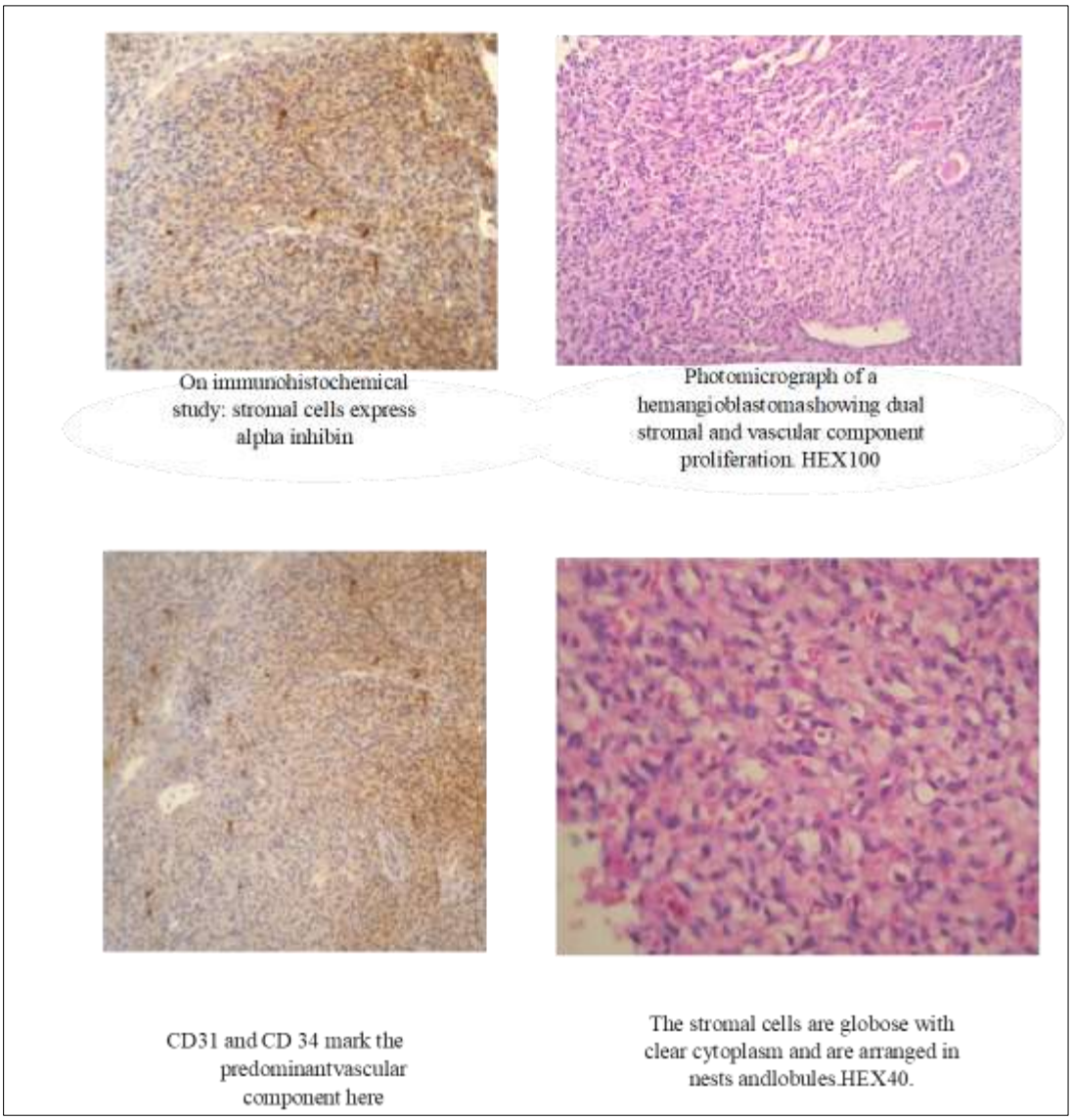

Figure 5 Pathological appearance of hemangioblastoma

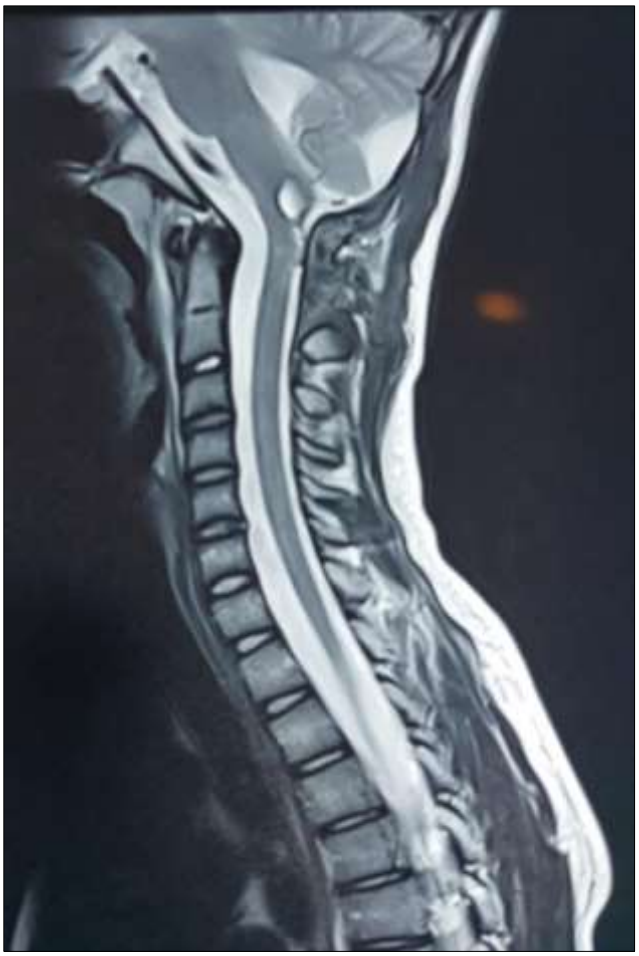

Figure 6 Post-operative MRI image of the bulbo-medullary lesion

3. Discussion

Hemangioblastomas are highly vascular lesions, representing 1.5% to 2.5% of intracranial tumors and 7% to 12% of posteriorfossa tumors. Hemangioblastoma can occur sporadically or as apart of VHL. The are usually in the cerebellum (76%) and less common in cerebral hemispheres (9%), spinal cord (7%), and brainsteam (5%) (11). Hemorrhage is rarely associated with hemangioblastomasdespite their rich vascular capillary networks (12). There are rare cases of hemorrhage from hemangioblastomasreported in the literature (15). Pathologically, hemangioblastomas are composed of a number of prominent thin-walled capillary vessels and often contain hemosiderin deposits indicating previous intra-tumoral bleeding(13). Despite being an intensely vascular tumor, as is often demonstrated angiographically and intra operatively, massive intra-tumoral hemorrhage is rare (14). In this article, we have discussed the diagnosis of the intra-tumoral hemorrhage of hemangioblastomas. You should know that the spontaneous intra-cerebral hemorrhage in patients younger than 15 years old is most frequently caused by vascular malformations, especially fistulas or arteriovenous malformations (1). Add that it is reported thathemangioblastomas with solid type, large size and spinal-radicular locations are more liable to hemorrhage (4). The presence of significant amounts of bleeding could also raise concerns about a ruptured aneurysm, AVM, or fistula; which requires a very good investigation, even a careful surgical exploration. A retrospective study and literature review by Glasker et al concluded that the risk is virtually o when the hemangioblastomas is less than 1.5 cm (6). Our case suggests the offending lesion was like by the small lesion (bulbomedullary lesion at 8mm) given the presence of intra-tumoral hemorrhage at presentation. Some more recent series have reported some cases of acute hemorrhage with tumors noted to be smaller than 1.5cm.

Note that secondary polycythemia is only very exceptionally revealing in the blood count (18). According to some authors (2, 17) polycythemia accompanies hemangioblastomas in 10 to 40% of cases. It disappears with excision of the tumor, reappears in case of recurrence and the percentage of polycythemia is then much higher. In our case, the hemoglobin was 13.7g/dl. The neuroradiological investigation was in favor of a bulbo- medullary lesion wish bled, first evoking a pial fistula. Since different causes of brain stem and bulbomedullary junction hemorrhages require different

management strategies, proper preoperative workup to exclude or confirm the diagnosis of a possible underlying lesion are of immense importance (CT, MRI) (19).

4. Conclusion

Hemorrhagic revelation of hemangioblastoma is very rare. A step-wise approach may be necessary for small subcentrimetric lesions with secondary hemorrhage, performing decompression first, followed by definitive resection.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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