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Posterior fossa hibernoma: A rare case in unexpected location radiologically mimicking arteriovenous malformation

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A B S T R A C T

Background: Hibernomas are rare tumors histologically originate from the fetal brown adipose tissue. Here, we reported the first hibernoma case located in posterior fossa radiologically mimicking arteriovenous malformation (AVM), presenting with abrupt onset of dizziness, and hiccups. This is the first case of hibernoma location in the posterior fossa.

Case description: A previously well 32-year-old man, presented with a headache for three days and abrupt onset of dizziness, nausea, vomiting, and hiccups. He had left-sided muscle weakness and hypoesthesia. Non-contrast enhanced computed tomography revealed hemorrhagic lesion at the inferoventral portion of the posterior lobe of cerebellum with compression to medulla oblongata with partial obstruction of fourth ventricle. Magnetic Resonance Imaging (MRI) revealed a mass lesion neighbouring at medulla oblongata which had intratumoral large vessels with branching segments. On T1 weighted images the lesion was isointense, on T2 weighted images it was hyperintense. On contrast-enhanced MRI images, heterogeneous contrast pattern was observed which is highly suspicious for AVM. Emergent surgery was performed and total excision was achieved. Histopathological analysis with Hematoxylin and eosin staining revealed yellow coloured, minimal hemorrhagic areas and well-defined encapsulated solid structure. The specimen showed brown adipose tissue content, with well-defined cytoplasm, large granular or multivacuolated cytoplasmic cells. Besides, small groups of univacuolar adipocytes were also observed. The histopathologic diagnosis was typical hibernoma. The postoperative period was uneventful and the patient was discharged in the fourth postoperative day with full neurological recovery. His neurological examination at the sixth postoperative month was completely normal. Radiological evaluation revealed total excision without any recurrence.

Conclusions: Hibernoma located in the posterior fossa is a unique condition, which may cause acute deterioration of the patient. These lesions may radiologically mimic vascular malformations. The neurological surgeons must kept in mind that hibernoma could also be seen in posterior fossa.

1. Introduction

Hibernomas are rare tumors histologically originate from the fetal brown adipose tissue which resembles the special fat deposits of hibernating animals [1]. Hibernomas occur over a wide age range, usually seen in adults with mean age of 38.0 years and account for about 1.6% of lipomatous tumors [1]. In the literature, the largest clinical series of all hibernoma cases were included 170 cases [1]. The most common anatomic locations included the thigh, shoulder, back, neck, chest, arm, and abdominal cavity/retroperitoneum. Hibernomas are usually presented as slow-growing, painless, soft, mobile, palpable masses, or as an incidental imaging finding. They also often show hypervascularity, sometimes accompanied by locally increased skin temperature [1]. Central nervous system (CNS) localization for hibernoma is a unique condition, where to best of our knowledge only one intracranially located hibernoma case has been previously reported [2].

Abbreviations: AVM, arteriovenous malformation; CNS, Central nervous system; CT, computed tomography; GFAP, Glial fibriller acidic protein; H&E, Hematoxylin and eosin; HU, Hansfield Unit; MRI, Magnetic Resonance Imaging; NSE, Neuron specific enolase; PAS, Periodic acid-schiff; WI, weighted images

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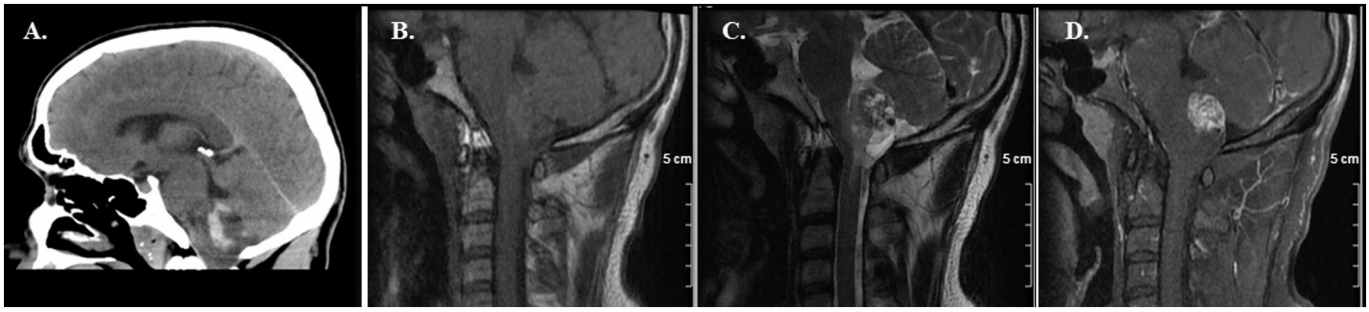

Fig. 1. A. Non-contrast enhanced CT revealed hemorrhagic lesion at inferoventral portion of the posterior lobe of cerebellum. B–D. Magnetic Resonance Imaging (MRI) revealed a mass lesion neighbouring at medulla oblongata which had intratumoral large vessels with branching segments. B. On T1 weighted images (WI) the lesion was isointense. C. On T2-WI it was hyperintense. D. On contrast enhanced MRI images heterogeneous contrast pattern was observed.

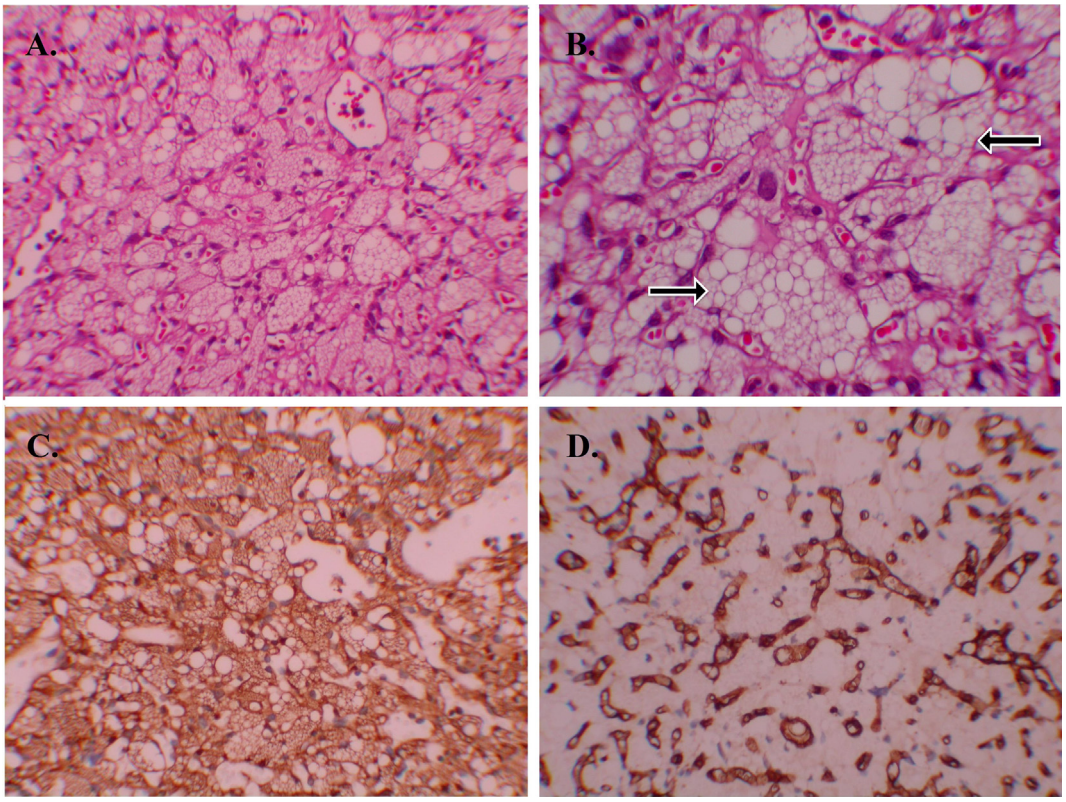

Fig. 2. Representative Hematoxylin and eosin (H&E) and immunohistochemistry results for the typical hibernoma. A, B Low-power (A) and high-power (B) H&E views of the typical hibernoma. C, D High-power views of S100 (C) and CD-34 (D) immunohistochemistry.

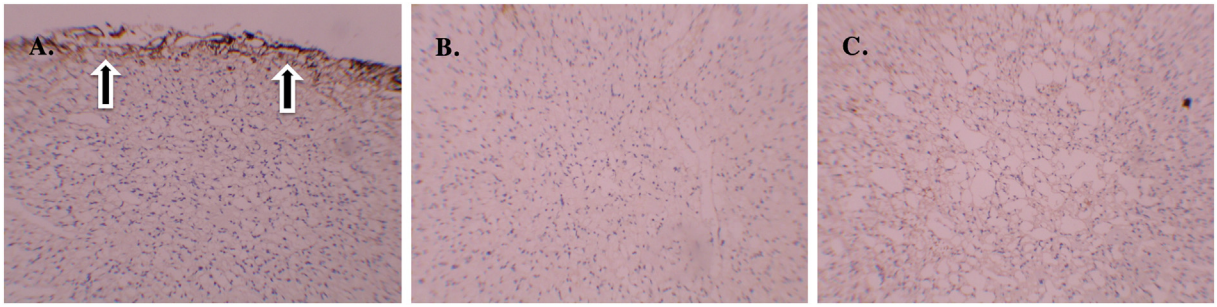

Fig. 3. Representative **a-c** high-power views of glial fibriller acidic protein (GFAP) (**a**) inhibin alpha (**b**) and neuron specific enolase (NSE) (**c**) immunohistochemistry. Only peripheral glial tissue was stained minimally with GFAP (**a**).

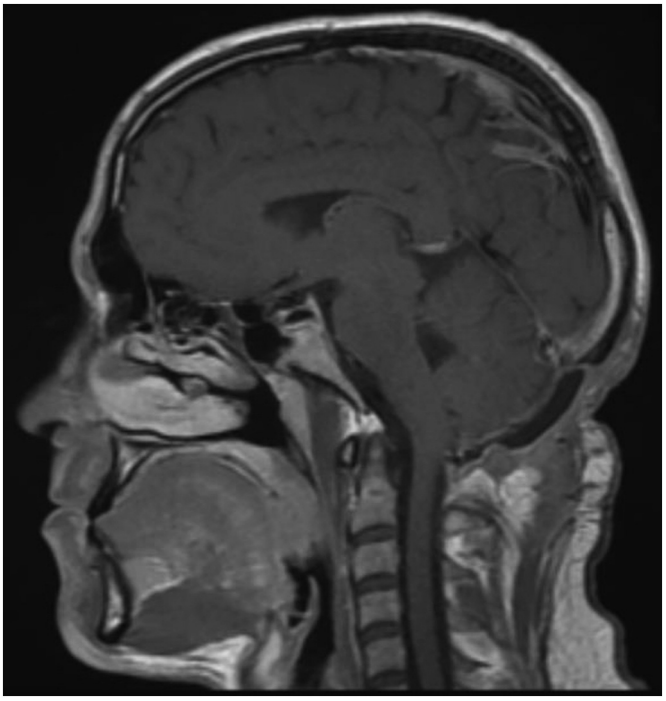

Fig. 4. On postoperative contrast enhanced MRI images there was no residual lesion.

In the majority of cases, noncontrast computed tomography (CT) demonstrates a fat-containing lesion, although it is important to note that some pathology-proven hibernomas do not show visible fat at CT. The lesion often contains internal septations. On T1 weighted Magnetic Resonance Imaging (MRI) they show up as hypointense to fat, and on T2 images they are usually found to be isointense to subcutaneous fat [3]. Typically, hibernomas can be easily separate from adjunct tissues, surgical treatment is the treatment of choice.

Here, we reported the first hibernoma case located in posterior fossa radiologically mimicking arteriovenous malformation (AVM), presenting with abrupt onset of dizziness, and hiccups. This is the first case of hibernoma location in posterior fossa.

2. Case report

A previously well 32-year-old man, presented with a headache for three days and abrupt onset of dizziness, nausea, vomiting and hiccups. He had left sided muscle weakness and hypoesthesia. Non-contrast enhanced CT revealed hemorrhagic lesion (Hansfield Unit (HU): 73.8) at the inferoventral portion of the posterior lobe of cerebellum with compression to medulla oblongata with partial obstruction of fourth ventricule (Fig. 1). MRI revealed a mass lesion neighbouring at medulla oblongata which had intratumoral large vessels with branching segments. On T1 weighted images (WI) the lesion was isointense, on T2-WI it was hyperintense. On contrast enhanced MRI images heterogeneous contrast pattern was observed which is highly suspicious for AVM. Angiography was planned because of high suspicion of vascular lesion but due to neurological deterioration of the patient emergent surgery was performed.

In concort position, midline skin incision was performed. Following suboccipital craniectomy, the mass was firstly devascularized from its vascular suppliers from anterior inferior cerebellar artery and posterior inferior cerebellar artery, and totally resected. Histopathological analysis with Hematoxylin and eosin (H&E) staining revealed yellow coloured, minimal hemorrhagic areas and well-defined encapsulated solid structure (Fig. 2). The specimen showed brown adipose tissue content, with well-defined cytoplasm, large granular or multivacuolated cytoplasmic cells. Besides, small groups of univacuolar adipocytes were also observed. Immunohistochemical staining for S100 and vimentin were both immunoreactive (Fig. 2). CD34 was not stained outside of the vessels and CD68 was stained at macrophage level. It was also negative for glial fibriller acidic protein (GFAP), inhibin alpha, neuron specific enolase (NSE), pankeratin, periodic acid-schiff (PAS) stainings (Fig. 3). The histopathologic diagnosis was typical hibernoma. The postoperative period was uneventful and the patient was discharged in the fourth postoperative day with full neurological recovery. His neurological examination at the sixth postoperative month was completely normal. Radiological evaluation revealed total excision without any recurrence (Fig. 4).

3. Discussion

To best of our knowledge, we reported the first case of hibernoma in posterior fossa presenting abrupt onset of dizziness and hiccups. Due to its vascular content, radiologically vascular pathologies were highly suspected in the differential diagnosis of this case. Hibernomas are a very rare lipoma variant within the CNS, composed of uniform granular or multivacuolated cells with small, centrally located nuclei, resembling brown fat. It is found in humans during the 21–24th weeks of the fetal life in various locations, such as the neck, axilla, paraspinal region, mediastinum, and retroperitoneum. The amount of brown fat normally diminishes in volume shortly after birth, but it may persist in these locations; hibernomas can occur anywhere that brown fat remains (Fig. 4).

The radiographic appearance of hibernomas is similar to that of lipomas or angiosarcomas, depending on the vascularity of the lesion. Hibernomas usually enhance in homogeneously; a dominant feeding vessel and/or large intratumoral vessel is often visible [4]. Murphey et al. [4] discussed the presence of prominent high-flow and low flow vascular structures within masses that show MR signals similar or identical to fat as being an important feature for distinguishing hibernoma from liposarcoma. On T1 weight MRI they show up as hypointense to fat, and on T2 images they are usually found to be isointense to subcutaneous fat [3]. Nevertheless, even if the CT and MRI findings may not be pathognomonic, hibernoma should be strongly considered if a mass shows negative HU measurements or MR signal intensity close to that of fat and contains large, branching blood vessels. In the present case, CT revealed hemorrhagic lesion (HU mean: 70) at inferoventral portion of the posterior lobe of cerebellum with compression to medulla oblongata with partial obstruction of fourth ventricule. MRI studies revealed a mass lesion neighbouring at medulla oblongata which had intratumoral large vessels with branching segments with heterogeneous contrast pattern which is highly suspicious for AVM. Angiography will also show the hypervascularity of the hibernoma along with possible arteriovenous shunts [5]. Cranial angiography was planned in our case because of high suspicion of vascular lesion, however due to acute neurological deterioration of the patient during follow-up emergent surgery was performed.

Hibernomas are typically removed surgically because histology and pathology cannot be confirmed by imaging and can even mimic more ominous tumors on imaging studies. Differential diagnosis includes granular cell myoblastoma, round cell liposarcoma, lipoblastomatosis, sebaceous adenoma, pleomorphic lipoma, atypical lipoma, chondroid lipoma, adult rhabdomyoma, and normal brown fat accumulation [1]. The imaging findings of hibernoma are not specific; other differential diagnostic considerations for a mass with signal similar to fat or containing large intratumoral vessels could include angiolipoma, hemangioma, lipoma, liposarcoma, alveolar soft part sarcoma, clear cell sarcoma, hemangiopericytoma, and hemangioblastoma. A definitive diagnosis is needed to exclude the possibility of malignant neoplasm; many advise excision rather than biopsy alone because of the risk of hemorrhage at biopsy of these vascular tumors.

Four morphological subtypes of hibernomas were described: typical,

myxoid, lipoma-like, and spindle cell, and they were added to the WHO Tumor Classification [1,3]. These four subtypes are determined based on ratio of multivacuolate adipocytes that are commonly seen in brown fat and univacuolate adipocytes that are commonly seen in white fat. Tumors with ≥70% multivaculoate adipocytes are considered to be non-lipoma hibernomas (typical), while < 70% multivacuolate adipocytes are considered lipoma-like hibernomas [1,3,5]. The non-lipoma or typical subtype is considered the most common (82%) and has three histological appearances: the eosinophilic variant, the pale variant, and the mixed variant. In the present case, it has been diagnosed as typical hibernoma. Approximately 85% of all hibernomas were found to contain S100 protein on immunohistochemical staining which is also positive in our case [3]. This case were negative for negative for GFAP, inhibin alpha, NSE, PAS stainings which differantiates our case from hemangioblastoma.

4. Conclusion

Hibernoma located in the posterior fossa is a unique condition, which may cause acute deterioration of the patient. These lesions may radiologically mimic vascular malformations. The neurological surgeons must kept in mind that hibernoma could also be seen in posterior fossa.

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