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Unusual location of a central neurocytoma: Particularities of a rare brain tumor

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ABSTRACT

Introduction: Central neurocytoma is a typically rare intraventricular glioneuronal tumor in young adults. Clinically, it is revealed by symptomatology of intracranial hypertension. Intra-parenchymal, extraventricular, and intramedullary localizations are exceptional. Central neurocytoma is considered a benign tumor that can recur locally, but craniospinal dissemination remains as an exception.

Case Report: We report a case in a 35-year-old male subject who consulted for quadriparesis and genito-sphincter dysfunctions. The clinical examination noted a syndrome of intracranial hypertension, a kinetic and static cerebellar syndrome, spastic tetraplegia with crural predominance, and anesthesia going up to the sixth thoracic vertebra (T6). A magnetic resonance imaging (MRI) showed a heterogeneous left cerebellar lesion. This lesion was excised, and the sample was freshly sent to the pathological anatomy laboratory. Extemporaneous cytology using the Smear technique was performed and was consistent with a hemangioblastoma. The standard histological examination also revealed the diagnosis of hemangioblastoma, and an additional immunohistochemical (IHC) study was performed for better diagnostic accuracy. The study was conducted in a North African laboratory and was conclusive of a neurocytoma. The evolution was marked by cerebellar tumor regrowth at five months, then the death of the patient at eight (08) months after surgery.

Conclusion: Central neurocytoma is a rare tumor that can be confused with several other tumors of the central nervous system. Immunohistochemistry is essential for diagnostic confirmation.

Keywords: Brain tumor, Central neurocytoma, Cerebellar region

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INTRODUCTION

Central neurocytoma is a rare intraventricular glioneuronal tumor. It can occur at any age but is more common in young adults [1–3]. A histologic investigation commonly demonstrates monomorphic round cells tumors in a fibrillar background, which can be confused with several other tumors depending on the topography

and clinical data. Thus, additional techniques are often required for diagnostic accuracy. We report a case of central neurocytoma to highlight the anatomoclinical particularities and the diagnostic difficulties in a laboratory with limited resources.

CASE REPORT

The study was carried out on a 35-year-old patient who was admitted for headaches of increasing intensity, walking disorders, weakness in all four limbs, genital sphincter disorders, and seizures. The clinical examination found a preserved general condition with a Glasgow score (GCS) of 15, an intracranial hypertension syndrome, a kinetic cerebellar syndrome, spastic tetraplegia, and anesthesia going back to the sixth thoracic vertebra. An MRI revealed a heterogeneous left cerebellar lesion with overlying hydrocephalus and a hypervascularized thoracic spinal cord lesion ranging from the seventh to the eighth thoracic vertebra. However, a direct correlation or link between the two lesions was established. Only the cerebellar lesion was subjected to a surgical operation and not the spinal cord lesion due to technical platform insufficiency. The fleshy component was intensely enhanced after the injection of the contrast agent. The standard histological examination of the cerebellar lesion found a tumoral proliferation consisting of more or less monomorphic cells, small to medium in size, pale cytoplasm, and a rounded or ovoid nucleus, with fine chromatin, sometimes nucleolated, without atypia, or obvious mitotic activity. Mitoses are evaluated at less than one mitosis per 10 fields at 400 magnification. These cells were arranged in small clusters or cords surrounded by a large network of fine capillaries. There were a few congestive vessels, and in places, there was a pseudocystic appearance; there was no tumor necrosis (Figure 1). In the periphery, there was healthy cerebellar parenchyma. These morphological aspects also made it possible to suggest the diagnosis of hemangioblastoma of the cerebellar region, which led to an IHC study for better diagnostic accuracy and possible molecular classification. As neuronal and glioneuronal antibodies were unavailable for this study, the paraffin blocks were sent to a pathological anatomy laboratory in North Africa for additional immunohistochemistry. In the IHC study, the tumor cells did not express the anti-HHV8, CAIX, CD34, and GFAP antibodies, thus invalidating the diagnostic hypothesis of hemangioblastoma. On the other hand, these tumor cells intensely and diffusely expressed anti-NSE and anti-Synaptophysin antibodies, allowing the diagnosis of WHO grade 2 neurocytoma of the cerebellar region (Figure 2).

The progression was further confirmed by headaches and convulsions two (02) months after the surgery. The CT scan at four (04) months after the procedure showed tumor continuation. The patient died eight (08) months after the surgery.

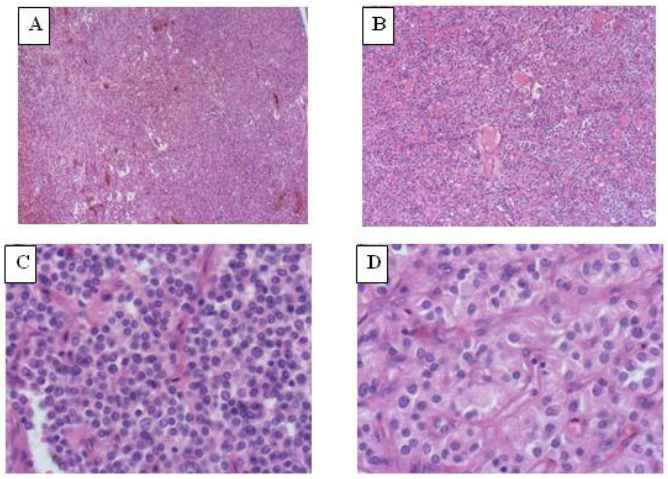

Figure 1: Histological samples stained with hematein-eosin (A) and (B): magnification 40 showing many capillaries of varying sizes, separated by more or less extensive bays or ranges of a cell population. (C) and (D): 100 and 400 magnification showing vacuolar cells with a rounded or elongated nucleus, which motivated the diagnosis of hemangioblastoma.

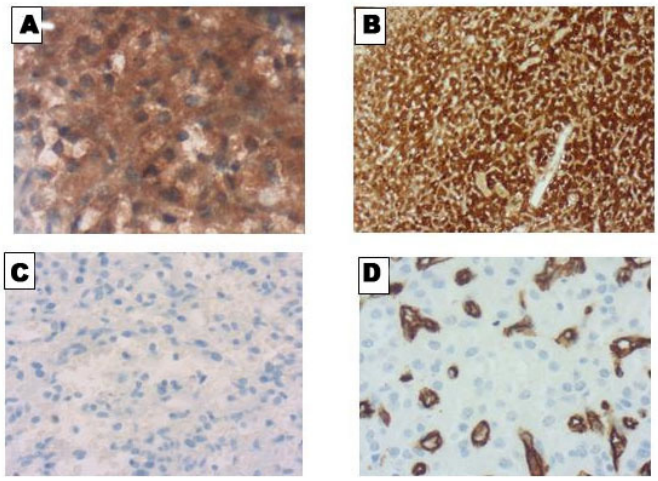

Figure 2: Immunohistochemical images (Synaptophysin, NSE, HHV8, CD34). A: immunostaining with anti-Synaptophysin antibody: diffuse high intensity labeling of cytoplasmic tumor cells. B: Immunostaining with anti-NSE antibody: diffuse high intensity labeling of cytoplasmic tumor cells. C: Immunostaining with anti-HHV8 antibody: absence tumor cells labeling. D: Immunostaining with anti-CD34 antibody: absence of labeling of tumor cells. Positive internal control.

DISCUSSION

Central neurocytoma is a rare glioneuronal tumor that was first described in 1982 by Hassoun et al. [1]. It is a pathology of young adults aged between 20 and 40 with an average age of 30 years, affecting both sexes equally [1–4]. Although the age of our patient (35 years old) is within the range described in most studies, the location remains unusual [4–9]. Indeed, central neurocytoma is typically described as an intraventricular tumor (third ventricle and lateral ventricle). It often develops from the septum pellucidum, buds in the lateral ventricles, and frequently obstructs the foramen of Monro [1, 3, 8, 10].

Extraventricular and intramedullary localizations have been reported by some authors [11–13]. Intra-parenchymal localizations outside the ventricles are exceptional. And for Hassoun et al. most of the reported cases of central neurocytomas of intra-parenchymal development would in fact correspond to dysembryoplastic neuroepithelial tumors (DNTs) [1].

Neurocytoma is classically revealed by symptoms of intracranial hypertension (headaches, nausea and vomiting, drowsiness, both impaired vision and consciousness, and hydrocephalus). These clinical symptoms (HIC) are related to the effect of the tumor mass and cerebrospinal fluid (CSF) outflow obstruction [4, 5, 7, 8, 12–15]. In addition to the intracranial hypertension syndrome found in our patient, the clinical examination found a kinetic and static cerebellar syndrome, spastic tetraplegia with predominantly crural anesthesia going back to the 6th thoracic vertebra (T6), and genital sphincter disorders. The cerebellar syndrome in our patient could be explained by the cerebellar location of the tumor. Other symptoms, such as static paraplegia and sphincter disorders, could arise from the hypervascularized spinal cord lesion of T7–T8, as shown by the MRI.

Based on histological analysis, the differential diagnosis of central neurocytoma comprises all intraventricular tumors, ependymoma, oligodendroglioma, choroid plexus papilloma, and hemangioblastoma. Fayçal et al., out of twelve (12) confirmed cases of central neurocytoma, concluded that there were 2 cases of ependymoma and one (01) case of papillary glioneuronal tumor based on standard histology. These data show the importance of IHC analysis to confirm the diagnosis of central neurocytoma. In our case, it was the first morphological diagnostic discrepancy. Indeed, on a purely morphological level, the main differential diagnoses of neurocytoma are oligodendroglioma, ependymomas and subependymomas, astrocytomas, and to a lesser extent, hemangioblastoma [7, 8, 16]. Different approaches, including epidemiology (young adult male), imaging (heterogeneous cystic and fleshy tumor, hypervascularized with contrast uptake), and topography (cerebellar region), the main probable diagnoses are metastasis, hemangioblastoma, astrocytoma, and ependymoma. In our case, faced with a tumor located in the cerebellum, with small to medium-sized cells of diffuse architecture without epithelial component, we first thought of hemangioblastoma. In the microscopic observation, there were many capillaries of varying sizes that were separated by more or less extensive bays or ranges of vacuolar cells with rounded or elongated nuclei. These tumor cells often had a spongy appearance due to the abundance of intracytoplasmic vacuoles. A fine network of reticulin fibers surrounded the capillaries and tumor cells (Figure 1). These cells were long considered as derived from endothelial cells of capillaries; hence the classic term “vascular tumors” and their origin remains unclear.

The diagnosis was rectified by IHC examination. This examination makes it possible to make a definitive diagnosis due to the anti-NSE (neuro-specific-enolase) and antisynaptophysin antibodies (the latter not marking either the oligodendroglioma or the ependymoma) and the negativity of the vascular and glial markers. Indeed, central neurocytoma is a regular round-cell tumor presenting neuronal differentiation with synaptophysin expression but not chromogranin or neurofilament [5, 8].

The prognosis following the surgery of the tumor is excellent in the event of complete macroscopic surgical excision. Similarly, the prognosis after the surgery in the event of incomplete resection is generally good because tumor regrowth is very slow (“inactivity” is sometimes possible for 15–20 years) [4, 7, 8]. Headaches can be persistent for many years due to scarring trauma after the surgery [4]. The probability of recurrence is approximately 20% in the event of macroscopically complete resection [8, 15, 17–19]. In the study by Fayçal et al., the immediate postoperative course was simple in 75% of cases. Five-year survival was 83% of cases in their series. No recurrence has been reported [4]. In the study by Peltier et al., the postoperative course was simple in 52% of cases [19].

Our patient’s evolution was marked by headaches and convulsions two (02) months following the surgery. The computed tomography (CT) scan four (04) months after surgery showed a tumor lesion. This lesion could correspond either to a tumor remnant, an evolutionary recovery, or a tumor recurrence. It is difficult to provide a clear answer because no imaging was performed early after the surgery (D1–D2). Given the early presence of a lesion on the CT scan control, the absence of histopathological signs of malignancy, and the absence of early postoperative control imaging, we believe that the resection must have been probably incomplete. This situation, associated with the spinal cord injury, could explain the worsening of the patient’s condition until death.

CONCLUSION

The central neurocytoma shares morphological aspects with several intracerebral tumors making complementary immunohistochemistry essential for diagnostic confirmation.

REFERENCES

1. Hassoun J, Gambarelli D, Grisoli F, et al. Central neurocytoma. An electron-microscopic study of two cases. Acta Neuropathol 1982;56(2):151–6.
2. Chuang MT, Lin WC, Tsai HY, Liu GC, Hu SW, Chiang IC. 3-T proton magnetic resonance spectroscopy of central neurocytoma: 3 case reports and review of the literature. J Comput Assist Tomogr 2005;29(5):683–8.

3. Baallal H, Akhaddar A. Neurocytome central: À propos d´un cas. PAMJ - Clinical Medicine 2020;3:10.
4. Abbad F, Sellami S, Hazmiri F, et al. Central neurocytomas: Clinical and radiopathological correlations: About 12 cases. [Article in French]. Pan Afr Med J 2017;27:222.
5. Favereaux A, Vital A, Loiseau H, Dousset V, Caille J, Petry K. Histopathological variants of central neurocytoma: Report of 10 cases. Ann Pathol 2000;20(6):558–63.
6. Choudhri O, Razavi SM, Vogel H, Li G. Atypical and rare variants of central neurocytomas. Neurosurg Clin N Am 2015;26(1):91–8.
7. Figarella-Branger D, Pellissier JF, Daumas-Duport C, et al. Central neurocytomas. Critical evaluation of a small-cell neuronal tumor. Am J Surg Pathol 1992;16(2):97–109.
8. Kerkeni A, Ben Lakhdher Z, Rkhami M, et al. Central neurocytoma: Study of 32 cases and review of the literature. [Article in French]. Neurochirurgie 2010;56(5):408–14.
9. François P, Wager M, Vasilijevic A, Roche RH. Neurocytome central révélé par un saignement intratumoral. Neurologie.com 2009;1(1):30.
10. Coquil O, Rolling AC. Les tumeurs du cerveau, Cancer info. Institut National du Cancer, 2010. p. 47.
11. Louis DN, Ohgaki H, Wiestier OD, Cavenee WK. WHO Classification of Tumours of the Central Nervous System. 4ed. Lyon: International Agency for Research on Cancer; 2016.
12. Morsli A, Houari L, Meziani S, Saadi M, Benbouzid T. Neurocytome central: á propos de 06 cas. Journal de Neurochirurgie 2015;22(2):22–7.
13. Mnif I, Chaker M, Daoud E, Ben Mahfoudh K, Mnif Z, Mnif J. Central neurocytoma: Three case reports. J Neuroradiol 2008;35(1):56–9.
14. Alexandre V, Anne J. Classification et «grading» des tumeurs du SNC. Neuro-Oncol 2014;86:7–9.
15. Martin-Duverneuil N, Mokhtari K. Neurocytome, in Les tumeurs intracrâniennes de l'adulte. Sauramps Médical 2009. p. 593.
16. Peltier J, Baroncini M, Le Gars D, Lejeune JP. Central neurocytomas of the lateral ventricle. A series of 35 cases with review of the literature. [Article in French]. Neurochirurgie 2011;57(4–6):215–9.

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Author Contributions

Aimé Sosthène Ouédraogo – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

FAHA Ido – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Souleymane Ouattara – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Assita Sanou Lamien – Design of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.