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Case report

Regression of Benedikt's syndrome after single-stage removal of mesencephalic cavernoma and temporal meningioma: A case report

Rodolfo Maduri[a,b], Giuseppe Barbagallo[c], Giuseppe Iofrida[a], Mariagrazia Signorelli[a], Francesco Signorelli[a,b,*]

[a] Cattedra ed U.O. di Neurochirurgia, Università degli Studi di Catanzaro "Magna Græcia," Catanzaro, Italy
[b] Hopital Neurologique et Neurochirurgical, Lyon, France
[c] Cattedra ed U.O. di Neurochirurgia, Università degli Studi di Catania, Italy

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1. Introduction

We herein report a unique association of midbrain cavernoma and Benedikt's syndrome (BS) that was reversed after microsurgical removal of the lesion. BS was originally described by Moritz Benedikt in 1889, characterized by the presence of an oculomotor nerve palsy and cerebellar ataxia including tremor and ascribed clinical constellation of signs to a lesion of the cerebral peduncle [1]. Subsequently, Charcot [2] and de la Tourette and Charcot [2] reported other cases, and in the latter case, involvement of the red nucleus, substantia nigra, third nerve root and sparing the cerebral peduncle were reported [3]. In this report, the author questioned the involvement of the corticospinal tract as a key feature of BS [3]. By that time, midbrain ischemic lesions and tuberculomas appeared to be the most frequent causes of BS, which was never reversible when midbrain structures were disrupted.

Our case helps to elucidate the clinicopathologic characteristics of BS, the potential reversibility of which may indicate surgery even in severely disabled patients with noninfiltrating midbrain mass lesions.

2. Case report

A 49-year-old woman with a 3-year history of generalized seizures presented with recent onset of diplopia, difficulty swallowing and language problems associated with progressive right-sided weakness and a coarse right upper limb tremor. Neurological examination confirmed a complete left third cranial nerve deficit, oropharyngeal dysphagia, severe mixed aphasia, right-sided hemiparesis with no movement against resistance, right-sided hyperreflexia and dysmetria, and a right arm tremor present at rest, at posture and with intention. This tremor sometimes took the appearance of choreoathetosis. The patient also displayed left exteroceptive and proprioceptive hemihypoesthesia. The clinical picture fit the definition of Benedikt's syndrome [1–3], with the addition of a left hemicorporeal sensory deficit not included in the original description of the syndrome [1], and language and swallowing difficulties.

Magnetic resonance imaging revealed a cavernous malformation located in the left midbrain tegmentum. T1 and T2 hyperintensity indicated subacute intralesional hemorrhage surrounded by hemosiderin deposition that extended to the ipsilateral cerebral peduncle. A left temporal convexity meningioma associated with enlargement of the sylvian fissure cistern due to local blockage of CSF (cerebrospinal fluid) circulation and a prerolandic left parasagittal meningioma were also present (Fig. 1).

The patient underwent a single-stage resection of the left temporal meningioma and mesencephalic cavernoma through a left

* Corresponding author at: Cattedra ed U.O. di Neurochirurgia, Università degli Studi di Catanzaro "Magna Græcia," viale Europa, Campus "S. Venuta," 88100 Germaneto, Italy. Tel.: +39 3381876962.
E-mail addresses: fsignorelli@unicz.it, signorelli2007@gmail.com (F. Signorelli).

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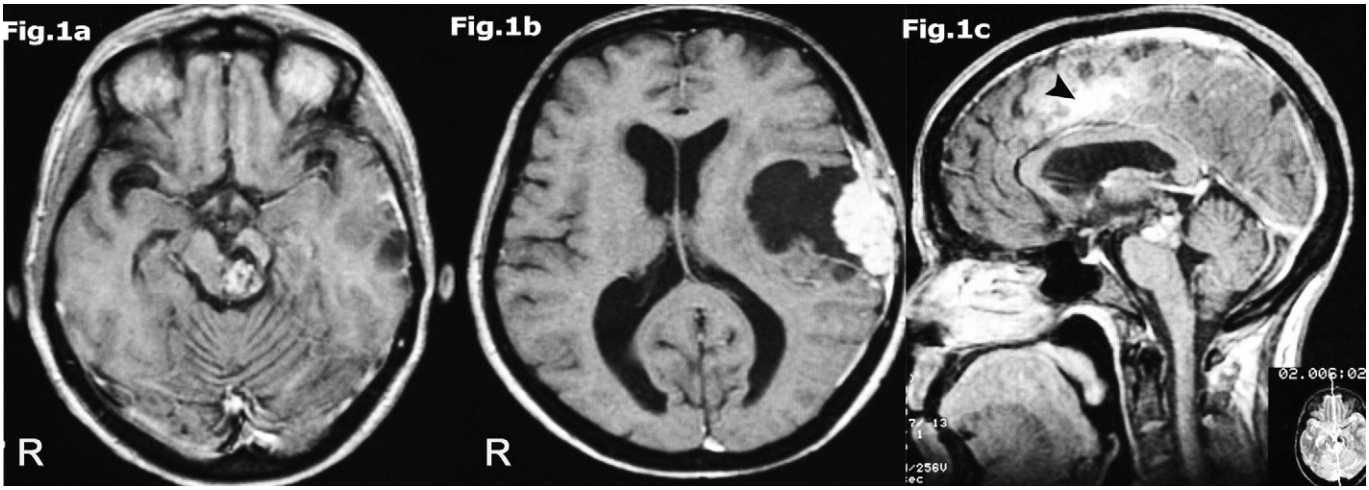

Fig. 1. Preoperative axial (a, b) and sagittal (c) T1-weighted enhanced MRI revealed a cavernoma in the left midbrain tegmentum with subacute intralesional hemorrhage and a hemosiderin ring that extended to the left cerebral peduncle. The cavernoma was associated with a prerolandic left parasagittal meningioma, whose dural implant is shown in the picture (arrowhead), and a left temporal convexity meningioma that excluded the sylvian fissure cistern.

temporal craniotomy and subtemporal approach. Exposure of the lateral mesencephalic surface was facilitated by removal of the temporal meningioma and opening of the superficial sylvian fissure, which also minimized the retraction of the Labbé vein (Fig. 2). Once the cisterna ambiens was reached, the trochlear nerve was recognized and followed anteriorly along the tentorial edge. The superior petrosal vein was seen emptying into the superior petrosal sinus in front of the point of entry of the trigeminal nerve into the Meckel's cave and was spared.

An incision of the lateral free tentorial edge that extended posteriorly to the entrance of the trochlear nerve widened the exposure of the entire lateral surface of the midbrain. The cavernoma surfaced into the ambient cistern behind the cerebral peduncle. After evacuation of the associated hematoma, the cavernous malformation could be exposed and dissected from adjacent tissue with no additional rectraction. Complete resection of the cavernoma was performed, preserving the surrounding hemosiderin ring, as shown by the control MRI (Fig. 2). Dura mater was then reconstructed with a microporus polyester urethane dura substitute (Neuro-Patch®, Aesculap, Tuttlingen, Germany), fixed with sutures and sealed by human fibrin glue (Tissucol®, Baxter Healthcare, Milan, Italy).

Histopathology of the supratentorial tumor revealed a World Health Organization (WHO) grade I transitional meningioma,. Histological examination of the midbrain lesion showed multiple dilated vascular spaces lined by endothelium and containing red blood cells, with recent and old hemorrhages in the stroma and no intervening brain parenchyma within the lesion, thus confirming the diagnosis of cavernous angioma. Postoperatively, the patient's upper limb tremor resolved immediately and the ptosis improved before discharge. She was then referred for physiotherapy as an inpatient. The parasagittal meningioma was removed three months after the first surgery. At the 7-year follow-up, the patient showed recovery from aphasia and dysphagia and the oculomotor palsy regressed. To date, the patient has a moderate right hemiparesis and hemihypoesthesia with a modified Rankin Scale score of 2.

3. Discussion

To our knowledge, an association between BS and midbrain cavernoma has not been reported to date. The mesencephalic tegmentum is a rarely documented site of expanding mass lesions. Loseke at al. reported a case of red nucleus syndrome featuring only a unilateral involuntary tremor caused by a solitary metastatic lesion from a glandular epithelioma of the prostate [4]. Ono et al. reported a case of a 26-year-old man who presented with an unusual multilobulated cyst in the cerebellopontine angle

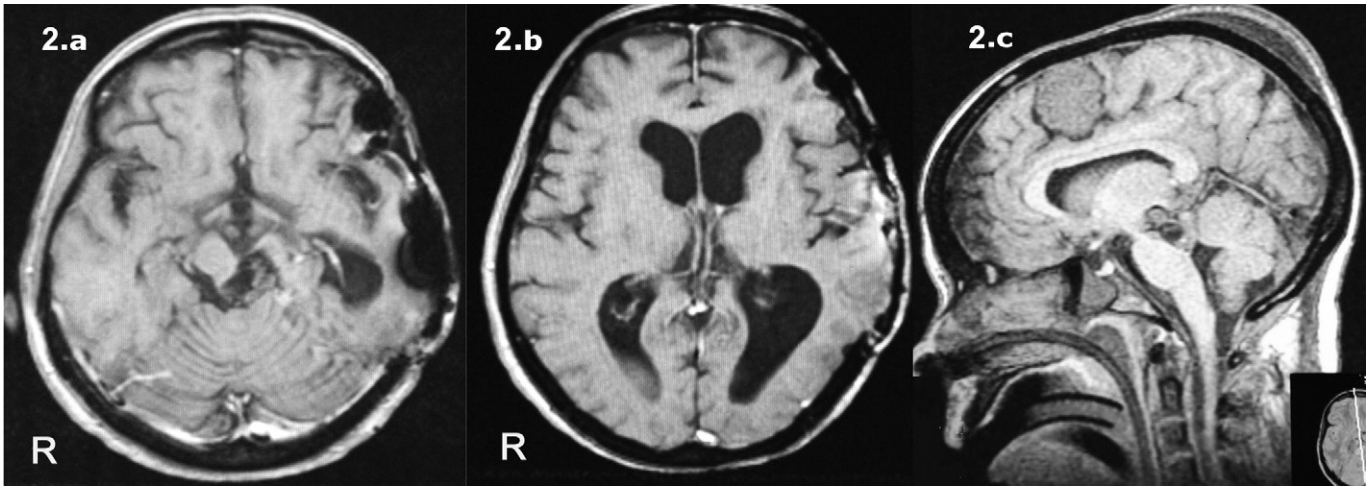

Fig. 2. Postoperative axial (a, b) and sagittal (c) T1-weighted MRI showing complete resection of the cavernoma with preservation of the surrounding hemosiderin ring and complete removal of the left temporal meningioma, with resolution of sylvian cistern exclusion.

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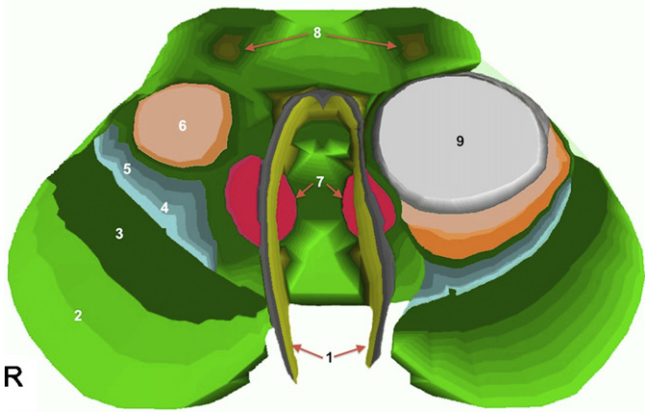

Fig. 3. Artist's drawing showing the anatomic relationship between the cavernoma and surrounding functional structures. The graphic model represents the tissue distortion according to a tridimensional projection of pixels based on an axial MR scan. Oculomotor nerves (1); cerebral peduncle (2); substantia nigra (3); medial lemniscus (4); lateral spinothalamic tract (5); reticular formation (6); red nuclei (7); superior collicular nuclei (8); cavernoma (9). Note that the perspective is from the top looking down; the patient's left is the observer's right.

extending into the brain stem. The clinical picture was dominated by normal pressure hydrocephalus and delayed onset of BS due to enlargement of the cyst [5]. In our case, BS was caused by a mesencephalic cavernoma that was associated with a left temporal and parasagittal meningioma.

Surgical resection of the mesencephalic mass was indicated in our case because of the accessibility of the lesion, its symptomatic nature (including hemorrhage), and the patient's young age. Surgical removal of the cavernoma led to drastic improvement of symptoms and signs, similar to the case described by Ono et al. in which the patient's involuntary movement, oculomotor nerve palsy and right hemiparesis were improved after removal of a midbrain multilobulated cystic formation [5]. We opted for a subtemporal approach because removal of the meningioma and emptying of the associated blocked cistern would facilitate exposure of the left middle incisural space. Otherwise, a supracerebellar infratentorial approach, in its extreme-lateral variant [6], that avoids left temporal lobe retraction and Labbé vein injury, would have been more suitable. The incision of the free edge of the tentorium further widened the surgical exposure cranially, revealing the cavernoma that edged out from the pial surface of the midbrain posteriorly to the cerebral peduncle, thus providing itself a safe entry zone in the mesencepahlic tegmentum. No attempt was made to remove the hemosiderin ring surrounding the cavernous malformation, which would have jeopardized the nearby functional tissue.

In our case, the anatomical substrate of the tremor (Fig. 3) was the compression of the ipsilateral red nucleus and its connections with the contralateral anterior horn of the spinal cord, which decussate in the ventral tegmentum and travel down the cord, partly mingling with the lateral corticospinal tract. Hemiparesis was obviously caused by the involvement of the corticospinal tract at the cerebral peduncle, which also caused weakness of the facial and lip muscles that are involved in coordinated mastication. However, swallowing problems may have been multifactorial in our case given the involvement of the insula and brain stem. Moreover, the oculomotor palsy was derived from the lesion of the nerve fascicles as they emerge from somatomotor nuclei in the midbrain tegmentum, which are not involved in BS because they are more medially located. The particular clinical feature of our case was the contralateral sensory deficit, which has not described in association with BS to date. It was ostensibly caused by the involvement of the medial lemniscus, situated just anterolateral to the red nucleus.

4. Conclusion

BS is a highly localized syndrome rarely caused by expanding mass lesions. In our case, the deficits were regressive after removal of the etiological agent, a midbrain cavernoma. Even highly disabled patients with BS may be improved by surgery if the cause is a noninfiltrating lesion, as in our case.

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