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Case report

Intracranial plasmacytoma presenting as glioblastoma multiforme

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Plasma cell tumors are a heterogeneous group of neoplasms that includes multiple and isolated plasmacytomas and other immunoproliferative diseases. An extremely uncommon form of the disease are solitary extramedullary plasmacytomas (SEPs) of the central nervous system (CNS) which account for less than 1% of all malignant tumors and about 14% of hematological malignancies.

A 37-year-old man came to the casualty department due to generalized seizures and a several-week history of a numbness of the left shoulder and left upper limb. Apart from severe obesity (weight 140 kg), he was otherwise healthy. Computed tomography (CT) of the head revealed a contrast-enhanced mass of 2.5 cm in diameter with surrounding edema in the right hemisphere. The patient was administered an anti-edema treatment and a control CT revealed a contrast-enhancing mass with dimensions of 37 mm × 32 mm × 36 mm surrounded by extensive edema (Fig. 1A) which strongly suggested glioblastoma multiforme (GBM). Due to the radiological picture and the lack of response to the anti-edematous treatment the patient was referred to the urgent surgery (radical resection of the tumor) without any other preliminary diagnostics (MRI or biopsy). Surprisingly, the histopathological examination of the surgical specimen revealed the features of plasmacytoma showing immunoreactivity for CD138 (Fig. 1B), CD79A, CD3, GFAP, Ki67, vimentin, and CD10 and no reactivity for CD20, BCL6 and CD31. The bone marrow biopsy, bone scintigraphy as well as blood (calcium, β2-microglobulin, albumin, M-protein) and urine (M-protein) tests did not show any pathology.

Three months after the resection a control magnetic resonance imaging (MRI) of the head showed a 23 mm × 16 mm × 21 mm tumor with an irregular marginal contrast enhancement corresponding to either post-operative changes or a residual/recurrent mass (Fig. 1C). The positron emission tomography with 2-deoxy-2-[fluorine-18]-fluoro-D-glucose integrated with the computed tomography (18F-FDG PET/CT) did not reveal any other CNS pathology except of an enhanced glucose uptake (SUV 1.5) around the mass which indicated post-operative changes (Fig. 1D).

The patient was scheduled for radiotherapy to the volume of the post-operative pathological mass (contrast enhanced on

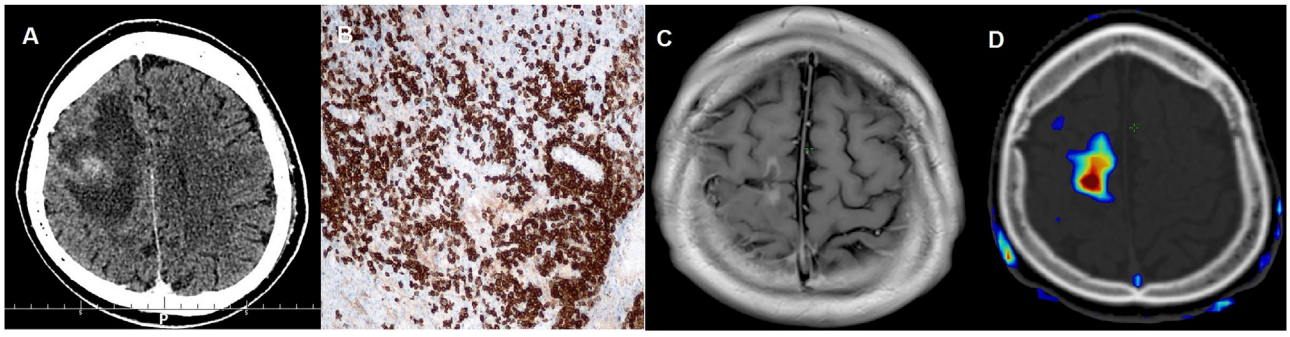

Fig. 1 – (A) CT scan showing contrast-enhanced tumor surrounded by an extensive edematous zone. (B) CD138 expression in the tumor (100×). (C) MRI after tumor resection showing surgical bed and irregular marginal contrast enhancement. (D) PET-CT scan showing enhanced glucose uptake at the site of tumor resection.

Table 1 – Clinical data of patients presenting with intracranial plasmocytoma.

Age(years)/sex	Symptoms	Localization	Treatment	Bone flap inv.	Progression to MM	Follow-up	Ref.
50/F	Motor aphasia	Left temporal lobe and thalamus	RT alone	No	No	48 mos	[1]
51/F	Diplopia	Pituitary/sphenoid	Surgery + RT	No	No	8 yrs	[5]
51/F	Mental status change	Left parietal lobe	Surgery + RT	Yes	No	2 yrs	[5]
43/F	Headache	Falx	Surgery + RT	No	No	8 yrs	[5]
30/M	Headache 6th nerve palsy	Clivus-dural	Surgery + RT	Yes	Yes	3 mos	[5]
47/M	Headache	Right parietal lobe	Surgery + RT	Yes	No	25 yrs	[5]
65/M	Hemiparesis	Right parietal lobe	Surgery alone	No	No	1 mo	[5]
75/M	Vertigo, hoarseness, swallowing difficulty	Left posterior	Surgery + RT	Yes	Yes	4 yrs	[5]
82/F	Mental status change	Left parietal lobe	Surgery alone	Yes	Yes	6 mos	[5]
64/M	Seizures, hemiparesis	(1) Frontal mass adjacent to the cerebral falx (2) In diploe	Surgery + RT	Yes	Yes	5 mos	[6]
37/M	Seizures, hemiparesis	Right temporal lobe	Surgery + RT	No	No	26 mos	Our study

Abbreviation: RT – radiation therapy.

MRI and of increased tracer uptake in 18F-FDG PET/CT) with a 2 cm margin of normal tissues and irradiated with 6 MV X-rays to a total dose of 45 Gy in 28 fractions. At the admission to the radiotherapy department, after a several week rehabilitation the patient's performance status was good (KPS 100) and did not change during irradiation. The treatment tolerance was monitored weekly based on the EORTC/RTOG post-radiation complications scale. At the completion of radiotherapy the skin reaction was estimated as a 1st degree complication; the CNS, eyes and cochlea did not present any acute complications (EORTC/RTOG-0). During a 26-month follow-up (to the last control visit) the control MRI was conducted every 3 months and did not reveal any tumor progression.

SEPs occur markedly less frequently than solitary plasmacytomas of the bone (SPBs) [1] and very rarely (15%) progress into multiple plasmacytoma [2]. Most SEPs (about 80%) involve the upper respiratory tract. The gastrointestinal tract, lymph nodes and urinary bladder are less frequent sites [3]. CNS SEPs are extremely infrequent [4] with only individual cases and small series reported. A systematic review of published cases is presented in Table 1. They may involve the meninges, parietal bones and skull base from where they may spread to or compress cerebral tissues. CNS SEPs not arising from the endocranium are casuistic cases, probably due to the fact that plasma cells are not found in brain tissues under normal conditions [1,7]. SEPs usually present radiologically as well-separated masses located around the dura mater or skull base with a homogeneous contrast enhancement and occasional calcifications. In contrast to meningiomas they are often located extra-axially [7].

In our patient radiological features of a tumor indicated glioblastoma rather than meningioma. Only one similar case of a 50-year-old female has been reported. She experienced a sudden motor aphasia and MRI of the brain showed an infiltrative contrast-enhancing tissue in the left temporal-insular lobe and thalamus with surrounding edema and a mild axial median shift. The malignancy infiltrated also the corpus callosum and diffused to the white matter of the right hemisphere. The brain biopsy and immunohistochemistry analysis revealed CD138+ elements expressing κ light-chain restriction which confirmed the presence of a mature plasma cell neoplasm. The patient received whole brain radiotherapy with 36 Gy plus a 9 Gy boost in the area of the lesion. During a 48-month follow up period there was no evidence of the disease progression [1].

Surgery is currently a treatment of choice and typically no prior biopsy is performed when an intracranial mass is considered resectable. However, Provenzale et al. suggested that a pre-operative biopsy spares in SEPs an unnecessary resection. Plasmacytomas are relatively highly radio-sensitive malignancies thus SEPs patients are better candidates for radiotherapy [7]. Nevertheless, three patients in another case series recovered after either radical or subtotal resection of CNS SEPs although two of them did not receive adjuvant radiotherapy [5]. Our experience suggests that some CNS SEPs indicate radiologically other malignancies and, consequently, an elective surgery is appropriate in patients with resectable lesions.

Radiotherapy is a well-established adjuvant treatment but should also be considered a radical treatment for non-resectable tumors [2]. A dose–response relationship has not been unequivocally established. The literature suggests 40 Gy in 20 fractions as an optimal dose in tumors measuring less than 5 cm in diameter and 50 Gy in 25 fractions for larger masses [8].

The irradiated volume usually includes the mass or its surgical bed with a 2–3 cm margin [3]. Radical whole-brain radiotherapy with a total dose of 36 Gy and up to 45 Gy boost to the tumor can be considered in patients with diffuse lesions or in those ineligible for a surgery [1].

Conflict of interest

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