

Definitive Intensity Modulated Radiotherapy For Rare Extramedullary Sphenoclival Plasmocytoma- A Clinical Case From Our Practice

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Abstract

Sphenoclival extramedullary plasmocytomas are a rare disease. We present a 58-year-old patient with headache, ophthalmoparesis and ptosis of the right eyelid, symptoms dating back for 2-3 weeks. Brain MRI with contrast revealed clivus involvement by an irregularly shaped soft tissue tumor with a heterogeneous structure. Biopsy confirmed the diagnosis of solitary plasmocytoma after ruling out systemic spread by the initial assessment. The patient conducted definitive intensity modulated radiation therapy (IMRT) in the tumor area up to total dose 50 Gy with daily dose 2 Gy.

Through this rare clinical case, we want to show the difficult pathohistological and differential diagnosis, as well as the need for IMRT, since it is a radiosensitive tumor.

Keywords: Extramedullary Plasmocytoma, Solitary Sphenoclival Plasmocytoma, Pathohistological Diagnosis, Immunohistochemistry, Intensity Modulated Radiotherapy

Introduction

Solitary plasmocytoma is a rare tumor that constitutes <10% of plasma cell neoplasms [1,2]. The two forms of the disease, solitary bone plasmocytoma (SBP) and extramedullary plasmocytoma (EP), are distinguishable by sites of origin and prognosis [3]. Extramedullary plasmocytoma commonly arises from the head and neck region, nasal cavity, and nasopharynx [4]. Plasmocytoma originating from the skull-base is an extremely rare tumor, and very few cases have been described in the literature [3,5]. We present a rare clinical case with extramedullary sphenoclival plasmocytoma to expand the literature on the difficult pathohistological diagnosis requiring immunohistochemistry and the need for definitive radiation therapy (RT).

Clinical Case

We present a 58-year-old patient with headache, ophthalmoparesis and ptosis of the right eyelid, symptoms dating back for 2-3 weeks. Brain MRI with contrast revealed clivus involvement by an irregularly shaped soft tissue tumor formation with a heterogeneous structure. The finding has data on the restriction of the diffusion of water molecules and the post-contrast scanograms increase its signal intensity inhomogeneously. The formation has sagittal dimensions of 56/32 mm. It ventrally fills the sphenoidal

sinus, cranially exerts compression on the pituitary gland and narrows the suprasellar cistern, dorsally narrows the prepontine cistern and is stenodopic to the basilar artery, and bilaterally laterally engages the two cavernous sinuses. There is no MRI evidence of occlusive internal and external hydrocephalus. Conclusion: A tumor of considerable size, filling the sphenoidal sinus with destruction of the bone structures in the area (Figure 1). Intraoperative: An endonasal transsphenoidal biportal craniotomy was performed using endoscopy. A tumor filling the sphenoidal sinus was encountered, profusely bleeding with macroscopic features of a malignant tumor of the lining of the sphenoidal sinus. The tumor completely destroyed the clivus, lateral and medial optic-carotid recesses, the anterior wall, the ostiums of the sphenoidal sinus and part of the anterior cranial fossa. Without a clear border, the formation entered the cavernous sinuses bilaterally, and instead was tough and non-aspirable. Due to the lack of anatomical landmarks, the tumor was removed as much as possible and a biopsy was taken for histological examination. Histological result: Fragments of tumor with plasmocytoma morphology after immunohistochemical / IHC examination - CD138 - positive reaction in tumor cells; MUM1- positive nuclear immunolabeling; Ki-67- over 80% nuclear immunolabeling. In this case, we excluded MM by serum electrophoresis and urine

examination for Bence–Jones protein. Laboratory tests revealed absence of hypercalcemia and renal failure, as well as low serum concentration of M-proteins. Bone marrow biopsy showed only 1% polyclonal plasma cells. Whole-body scintigraphy showed no other bone changes. Tumor markers such as carcinoembryonic antigen, a-fetoprotein and squamous cell carcinoma-related antigen were all within the normal range.

After the necessary above investigations, confirming a solitary extramedullary plasmacytoma, we considered carrying out definitive intensity modulated radiation therapy (IMRT) in the

tumor area up to total dose (TD) 50 Gy with dayli dose (DD) 2 Gy. In Figure 2, we present the delineation of the target volumes (GTV and CTV) after fusion of the preoperative MRI and planning CT. In Figure 3, we present the IMRT by the VMAT method in the tumor area up to TD 50 Gy with DD 2 Gy. Against the background of anti-inflammatory and anti-edema therapy with Dexamethasone/ 4 mg twice a day/ the patient tolerated radiation therapy very well without significant side effects on the part of the central nervous system. A follow-up MRI is recommended two months after completion of definitive radiotherapy.

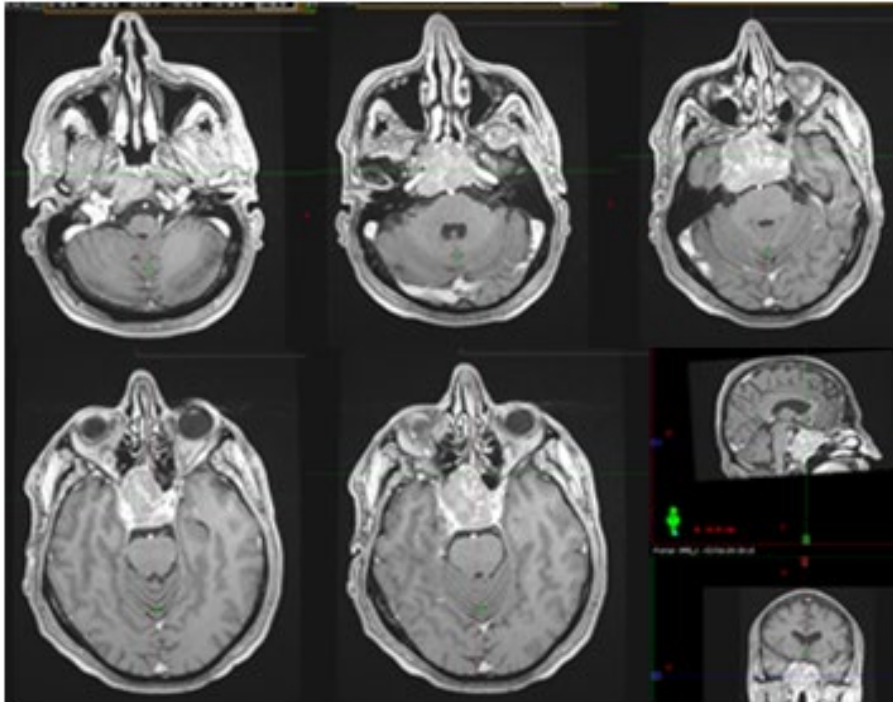


Figure 1: Brain MRI with contrast revealed clivus involvement by an irregularly shaped soft tissue tumor formation with a heterogeneous structure. The formation has sagittal dimensions of 56/32 mm. It ventrally fills the sphenoidal sinus, cranially exerts compression on the pituitary gland and narrows the suprasellar cistern, dorsally narrows the prepontine cistern and is stenodopic to the basilar artery, and bilaterally laterally engages the two cavernous sinuses.

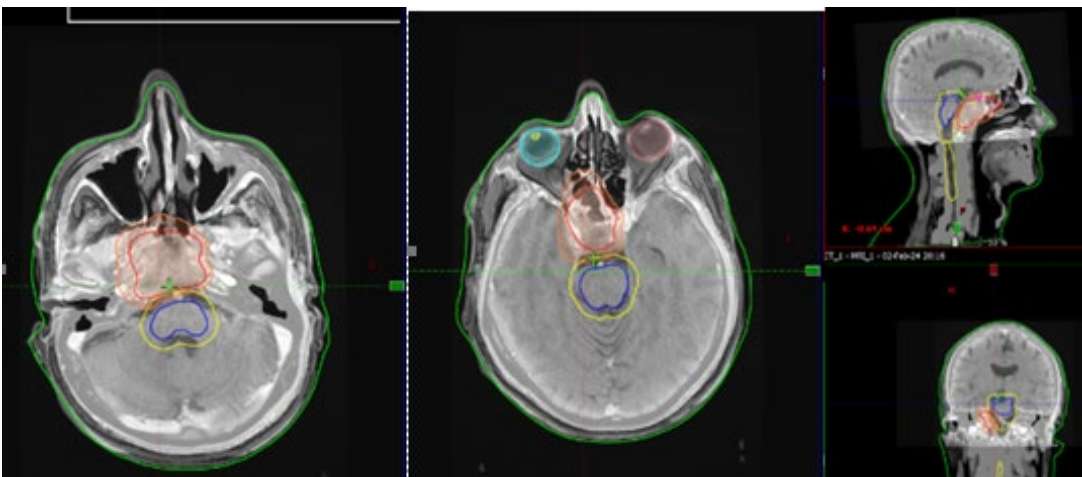


Figure 2: The delineation of the target volumes (GTV and CTV) after fusion of the preoperative MRI and planning CT.

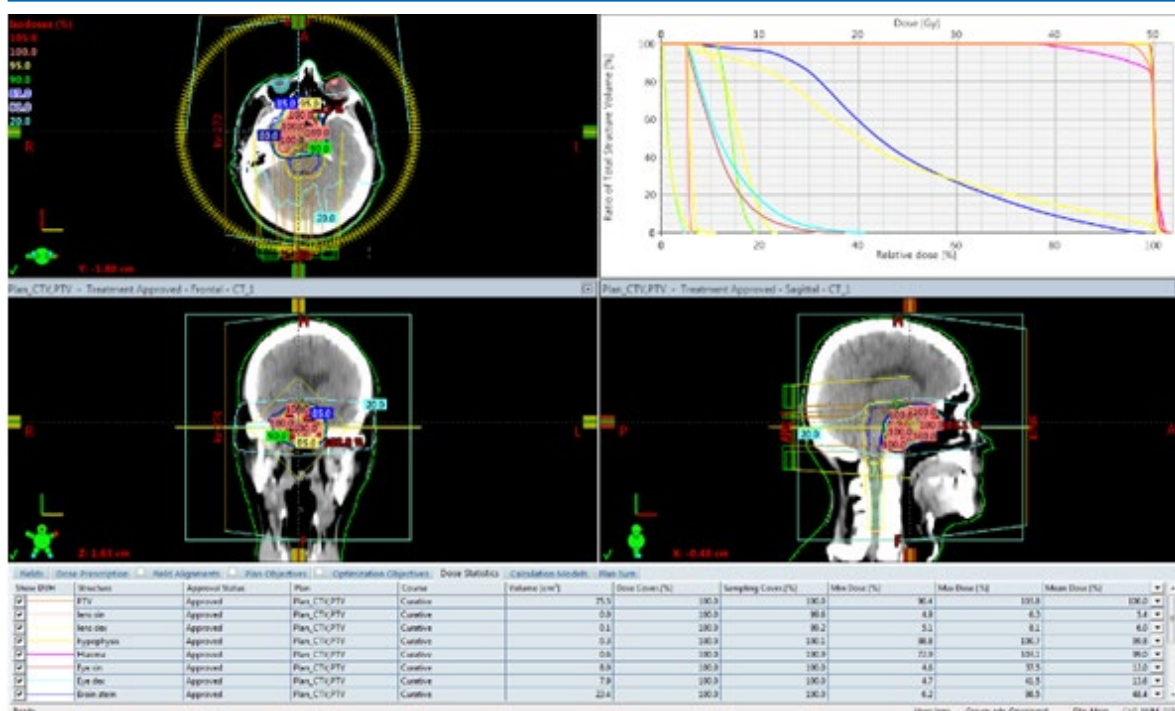


Figure 3: The IMRT by the VMAT method in the tumor up to TD 50 Gy with DD 2 Gy.

Discussion

The most common sites of primary extramedullary plasmacytomas (EP) arising from the skull are the parietal bone and the skull base bones [6]. The skull base site that is most frequently affected-representing 18% of all head and neck cases is the nasopharynx [7]. Less frequently seen are sphenoidal, clivus, and petrosal apical presentations. The diagnosis of solitary bone plasmacytoma (SBP) is based on a radiologically solitary bone lesion, plasma cells in the biopsy specimen, fewer than 5% plasma cells in bone marrow, less than 2.0 g/dl monoclonal protein (M-protein) in the serum when present, negative urine test for Bence Jones protein, no evidence of hyperglobulinemia and hypercalcemia and absence of anemia [8-10]. Neurologic symptoms due to plasmacytomas located either in the base of the skull or at intracranial locations are extremely rare [11]. We observe the following symptomatology in our patient -headache, ophthalmoparesis and ptosis of the right eyelid, symptoms dating back for 2-3 weeks. The differential diagnosis of a clival lesion includes chordoma, chondrosarcoma and meningioma [12]. The radiological differential diagnosis of the skull base plasmacytoma includes carcinoma nasopharynx, chordoma, meningioma, osteosarcoma, lymphoma, pituitary adenoma, metastatic carcinoma, eosinophilic granuloma, and multiple myeloma [13]. Although findings from a computed tomography (CT) scan or magnetic resonance imaging (MRI) may lead to the diagnosis of plasmacytoma, differentiating from other skull base tumors based only on radiologic findings is difficult [14]. The MR appearance is either isointense, hyperintense or heterogeneous intensity compared with the brain parenchyma on T1-weighted images and homogeneous enhancement by intravenous administration of Gd-DTPA [9,15-17]. Only total body

imaging work-up, tissue diagnosis, and immunohistochemical (IHC) staining determined the final diagnosis of plasmacytoma [3]. In the presented clinical case, MRI of the brain with contrast revealed involvement of the clivus by an irregularly shaped soft tissue tumor with a heterogeneous structure. The finding has data on the restriction of the diffusion of water molecules and the post-contrast scanograms increase its signal intensity inhomogeneously (Figure 1). The IHC analysis in extramedullary plasmacytoma reports CD-138 positive, CD-38 positive, CD-56 positive, and CD-45 negative cells [5]. Through the IHC analysis of the tumor cells, a strict differential diagnosis is made with other primary tumors of the skull base such as carcinoma nasopharynx, chordoma, meningioma, osteosarcoma, lymphoma, pituitary adenoma, metastatic carcinoma and eosinophilic granuloma. The presented inoperable solitary extramedullary sphenoclivus plasmacytoma is subject to definitive radiotherapy (RT). Local RT is the primary mode of treatment for EP, occasionally followed by surgical resection of the residual tumour [12]. Radiotherapy up to total dose 50 Gy in five weeks is the treatment of choice for the solitary skull base plasmacytoma, as it is a radiosensitive tumor [13]. The radiation dose ranged between 3,000 cGy and 5,400 cGy, which compares with the radiation dose recommended of 5,000 to 6,000 cGy [18,19]. After the necessary above investigations, confirming a solitary extramedullary plasmacytoma, we considered carrying out definitive intensity modulated radiation therapy (IMRT) in the tumor up to total dose (TD) 50 Gy with daily dose (DD) 2 Gy (Figure 2 and 3). Against the background of anti-inflammatory and anti-edema therapy with Dexamethasone/4 mg twice a day/ the patient tolerated radiation therapy very well without significant side effects on the part of the central nervous system.

Conclusion

Solitary extramedullary sphenoclivical plasmacytoma is a rare inoperable tumor. A biopsy is imperative, and the pathohistological diagnosis is proven after a mandatory immunohistochemical analysis. After the necessary laboratory tests and imaging diagnostics, excluding multiple myeloma, definitive radiotherapy requiring a total dose of 50 Gy, as EP is a radiosensitive tumor.

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