Pomeranian J Life Sci 2022;68(1):35-36 doi: 10.21164/pomjlifesci.776

Giant plasmocytoma of frontal cranium as a rare first manifestation of multiple myeloma – case report

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ABSTRACT

Introduction: Plasmocytomas are lesions that are rarely encountered in neurosurgical practice. Whereas multiple myeloma (MM) occures primarly in bone marrow, solitary plasmocytomas present exclusively extramedullary or as a solitary bone plasmocytoma. Although typical first manifestation of MM are osseus lesions, there are only few case reports in medical world literature of first MM presentation as a single cranial tumor of the skull vault.

Clinical presentation: A 63-year-old male with a 4-month history of painless scalp swelling was sent to our department after an unsuccessful attempt to remove the tumor. The patient was neurologically intact and his general condition was good. Bifrontal craniectomy was performed in order to obtain a total resection of the lesion. The tumor was histologically diagnosed as plasmocytoma. Postoperative bone marrow biopsy demonstrated myeloma cells. The subsequent combined therapy had been successful and the patient is in a good condition almost 3 years after the procedure.

Conclusion: As an uncommon type of solid tumors, plasmocytomas may mimic other neoplasms, which can lead to delay of diagnosis and proper treatment. Satisfying effects of treatment might occur only with proper diagnostic schema, which should be employed despite the rarity of plasmocytomas in daily clinical practice.

Keywords: skull neoplasms; diagnosis; plasmocytoma; surgery; primary bone tumor; skull vault.

INTRODUCTION

Plasma cell dyscrasia is characterized into 2 main groups: solitary plasmocytoma – occurring primarily as a single mass of clonal plasma cells with no or minimal bone morrow plasmocytosis and with no other symptoms other than those derived from the tumor mass [1], or multiple myeloma (MM) – primarily localizing in bone morrow and predominantly associated with hematological disorders (typically normocytic normochromic anemia), fatigue and weight loss or osteolitic bone changes [2]. An uncommon primal manifestation of MM is a single solid form of plasma cell neoplasm – plasmocytoma. Most medical literature about cranial plasmocytomas report cases of tumors in the base of the skull, the mandible, maxilla, orbit or temporal bone. As far as the authors are aware, there are only a few cases in medical journals of an initial presentation of MM as a single neurocranial tumor.

CLINICAL PRESENTATION

A 63-year-old male presented in February 2017 with swelling over the midline of the frontal bones. The mass was not tender on palpation and the overlying skin tissue was unchanged. He reported a 4-month history of an incremental spreading of the swelling. The general condition of the patient was good, he was conscious, fully cognizant, and did not present any neurological deficits. The patient did not report any chronic diseases, nor complaints in other systems, and his family history for neoplasms was negative. The first attempt at removal of the lesion was made in the General Surgery Department. Since the patient did not experience pain, the mass was initially suspected to be a benign neoplastic lesion, thus the surgeons decided to remove the tumor under local anesthesia. The resection became impossible due to severe haemorrhaging. While magnetic resonance imaging (Fig. 1) revealed a 9.7 x 7.9 x 8.1 cm enhanced mass lesion over the midline of the frontal region with skull erosion, the general surgeons decided to transfer the patient to the Neurosurgery Department. While preparing for the surgery, laboratory blood tests were performed, which revealed normocytic normochromic anemia, high inflammatory parameters and impairment of coagulation, with normal renal parameters and calcium levels.

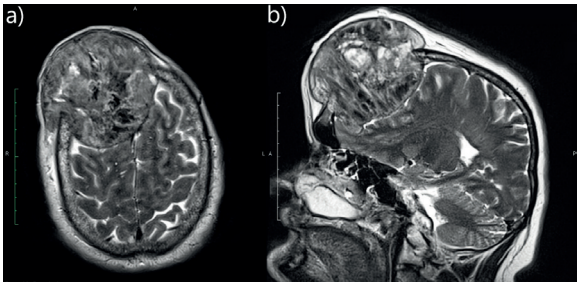

FIGURE 1. Preoperative magnetic resonance imaging: a) axial view on T2; b) sagittal view on T2

Two days after the unsuccessful attempt at the resection of the mass, the patient was operated on again the neurosurgery operating room. Bifrontal craniectomy was presented. The tumor adhered periphally and centrally exceeding the dura mater. The sagittal sinus was closed with clips frontally of the coronal suture.

The neoplastic mass was completely removed along with the invaded skull and dura. For the duraplasty, Neuropatch was used to replace the fragment infiltrated by the tumor (Fig. 2). No adverse events were reported during the course of hospitalization and the patient was discharged from the hospital in a good general condition, conscious, fully aware without any neurological deficits (Fig. 3). The wound was healed completely without complications.

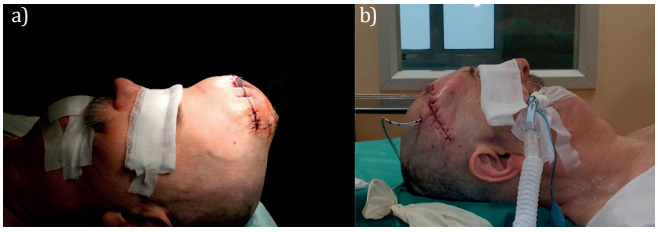

FIGURE 2. Photo from the operating room: a) preoperative photo. Patient in supine position, well exposed tumor mass; b) postoperative photo

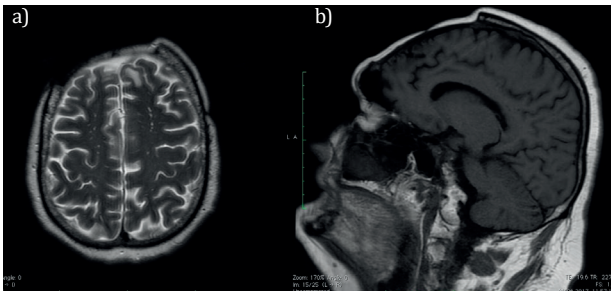

FIGURE 3. Postoperative magnetic resonance imaging: a) axial view on T2; b) sagittal view on T1

Histopathology of the lesion revealed a tumor consisting of plasma cells positive for CD138 and vimentin, which corresponded to MM. As the mass had formed a solitary tumor it was diagnosed as plasmocytoma.

The patient presented to the Hematology Department for a follow-up examination 15 days after he was discharged from the neurosurgery department. Further hematological examination uncovered abnormalities typical of MM. The neurosurgical procedure combined with accurate hemato-oncological treatment resulted in a positive outcome: in a 3-year follow-up, the patient is in good general condition with a controllable underlying disease and satisfying health level.

DISCUSSION

Neuroranial plasmocytomas are extremely rare. They constitute only 0.7% of all solid plasma cell tumors and although their typical primal location is red marrow-containing bones, there are a few case reports of neurocranial plasmocytomas [3, 4]. This rarity of neurocranial plasmocytomas indicates difficulties with proper differentiating and diagnosis. Upon initial diagnosis, and without any radiological imaging, plasmocytomas may mimic old subcutaneous hematomas, abscesses or giant atheromas [5, 6]. Both puss and old hemolised blood reservoirs are quite characteristic during examination. Abscesses typically cause pain and irritation of the overlying skin tissue, accompanied by high levels of inflammation and frequently a fever. Similar symptoms might also occur in hematomas, but most significant is the medical history, which often reveals past head trauma. Giant atheromas may seem to be an insidious differential diagnosis – a slow growing mass that is not tender on palpation without any prior injury and no signs of inflammation. Radiological imaging can help in a proper diagnosis.

Both the magnetic resonance imaging and computed tomography scans revealed a neoplastic mass invading the frontal bone as well as the underlying tissue (dura mater and brain). Tumors, which can cause similar changes are categorized as those derived from the bone tissue (e.g. osteosarcomas) or from dura mater (meningiomas), as well as metastatic carcinomas [3, 7]. Plasmocytomas are characterized by the dissolution of bone tissue with no sign of sclerosis and sharp borders of the lesion. They enhance heterogeneously after contrast injection and do not show any satellite lesions. Osteosarcomas are another very rare type of skull tumor, but rather occur almost exclusively extracranially. While plasmocytomas grow as a solitary mass, slowly superseding healthy tissue, metastatic carcinomas of the bone usually appear as multiple lesions.

CONCLUSION

Plasmocytomas of the neurocranium as a primary manifestation of MM are very rare. As an uncommon type of solid tumor, they may mimic other neoplasms, which can lead to delays in diagnosis and proper treatment. Proper identification of the underlying disease requires cooperation of both neurosurgeons and hemato-oncologists. Knowledge of this insidious presentation of plasmatic cell dyscrasia may shorten the time needed for implementation of recommended therapies.

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