



Case Report

Sphenoidal Plasmacytoma Associated with Multiple Myeloma Mimicking Meningeal Tumor in a Geriatric Patient

Wei-Yuan Pan^a, Tao-Yeuan Wang^{b,d}, Cheng-Chia Tsai^{c,d*}

^a Section of Critical Care Medicine, MacKay Memorial Hospital, Taipei, Taiwan, ^b Department of Pathology, MacKay Memorial Hospital, Taipei, Taiwan, ^c Department of Surgery, MacKay Memorial Hospital, Taipei, Taiwan; ^d Mackay Medical College, New Taipei City, Taiwan

ARTICLE INFO

Accepted 7 March 2019

Keywords:

meningioma,
multiple myeloma,
plasmacytoma,
sphenoidal

SUMMARY

A 76-year-old woman without any neurological symptoms or discomfort was revealed to have a sphenoidal lesion with erosion, destruction of the adjacent bone structure, and extension to the middle left cranial fossa via neck computed tomography (CT). The lesion characteristics resembled a meningioma and she was advised to undergo surgery, which she refused. At the 3-month follow-up, the lesion was seen to have enlarged, and a new osteolytic lesion was found in the frontal skull vault. She was diagnosed as having skull plasmacytoma after frontal bone biopsy. Later, immunofixation test, bone marrow smear, and bone survey revealed multiple myeloma (MM). The patient received systemic chemotherapy and responded well after seven months of treatment. The case is unusual as sphenoidal plasmacytoma is rarely present in MM, and the patient presented neither neurological symptoms nor discomfort. The skull lesion was found incidentally. Moreover, the initial neuroradiological finding mimicked that of meningioma, but the diagnosis of a plasmacytoma associated with MM was made later. Although intracranial plasmacytoma is rarely associated with MM, the neuroradiological findings lack specificity and do not differ from those of meningioma, metastasis, etc. Proof based on tissue analysis was extremely important for ensuring accurate diagnosis before proceeding with therapy.

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1. Introduction

Plasma cell neoplasm is the proliferation of neoplastic monoclonal plasma cells, which can appear as solitary or multiple plasmacytoma, and may arise from the skeleton (medullary) or soft tissue (extramedullary), but rarely occur in the skull base region, where the sphenoid and clivus are more affected as reported previously.^{1–7} When the skull base is involved, plasmacytoma can induce a variety of discomforts, such as headache, eye pain, dizziness, epistaxis, rhinorrhea, nasal obstruction, cranial nerve palsy, according to the affected site.^{4,5,8} Plasmacytoma may be associated with or progress to MM with multifocal osteolytic lesions, pathological fractures, hypercalcemia, and anemia, and occurring mostly in the elderly. However, there are only a few studies describing intracranial plasmacytoma associated with MM. Without an existing diagnosis of plasmacytoma or MM, imaging finding easily misled the initial diagnosis. Here we present another case with sphenoidal plasmacytoma, presenting no neurological symptoms or discomfort, which based on imaging results, was believed to have meningioma. The patient was suspected to have a meningioma initially, was found to have plasmacytoma later and was diagnosed with multiple myeloma finally.

2. Case Report

A 74-year-old woman was referred to the neurosurgical clinic because CT scan incidentally revealed an intracranial lesion of 1.6*1.9*3.0 cm³ involving the left sphenoid ridge with erosion, destruction of the adjacent bone confined and also an extension to the anterior aspect of the left middle cranial fossa (Fig. 1). In addition to the focal brain lesion, she also had hypertension and a history of lumbar spondylolisthesis. She presented full muscle power of all four extremities, normal deep tendon reflexes of four limbs and had no pain, malaise, fatigue, visual disturbances, etc. Otherwise, she lost

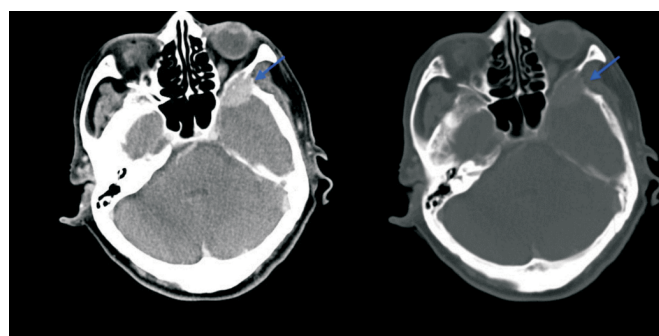


Fig. 1. Brain CT with contrast enhancement and bone window showed a 1.6*1.9*3.0 cm³ intracranial lesion involving the left sphenoid ridge with erosion, destruction of the adjacent bone, and the extension to the anterior aspect of left middle cranial fossa.

* Corresponding author. MacKay Memorial Hospital, No. 92, Sec. 2, Zhongshan N. Rd., Taipei 10449, Taiwan.

E-mail address: angle@mmh.org.tw (C.-C. Tsai)

more than 15 kg in two years. The lesion was suspected to be a meningeal tumor or dural metastasis, so an operation was suggested. However, she decided to attend the follow-up. On the following visit after three months, brain CT revealed a new osteolytic lesion in the frontal bone and the sphenoidal lesion enlarged with a size of 2.0*2.3*3.2 cm³ (Fig. 2a, Fig. 3a). A frontal bone biopsy was performed for tissue proof, and the result suggested it was plasmacytoma composing of monotonous sheets of neoplastic plasma cells with a small volume of normal host tissue. The tumor cells were immunoreactive with VS38c, CD79a, κ light chain and negative for λ light chain & leukocyte common antigen (LCA) (Fig. 2b, Fig. 2c, Fig. 2d). Further study of the bone survey showed multiple osteolytic lesions in the skull vault, thoracic, lumbar, right costco-vertebral junction, ribs, humeral, femur, ilium, etc.

Laboratory test showed normocytic anemia with a Hb level of 7.8 g/dl and leukopenia with WBC level 2200/μl. Calcium and creatinine tests revealed normal results. Immunofixation test showed a monoclonal protein pattern of IgA (5727 mg/dl) and free light Kappa chain (496 mg/dl). The Bence Jones protein urine test was negative. Beta 2 microglobulin level was 7.11 mg/dl. A bone marrow smear showed hypocellularity with a ratio 9:1 of myeloid to erythroid precursors and a total plasma cell of around 30–40%. Multiple myeloma with bone metastasis diagnosis was established. A targeted combined biological therapy based on bortezomib, thalidomide, and prednisolone was immediately initiated, and after seven months of treatment, the sphenoidal lesion appeared to shrink and to respond to therapy (Fig. 3b). The patient attended follow-up in the clinics.

3. Discussion

MM is a bone marrow malignant disease, which is found in the spectrum of plasma cell dyscrasias, most frequently diagnosed among people aged 65–74 with 29.8% of all new cases, and is more common in males with an incidence rate of 8.4/100,000 comparing to that of 5.3/100,000 in females.⁹ Asian/Pacific Islanders have the lowest incidence rate comparing to other ethnic groups.⁹ MM is

characterized by the proliferation of a single clone of plasma cells, monoclonal immunoglobulins in the serum or urine, accompanied by end-organ damage-hypercalcemia, renal failure, anemia, and bone lesions. After being diagnosed with MM, 50.7% of patients survive five years, and the median age at death is 75.⁹ According to the report of the National Cancer Institute (NCI) Surveillance Epidemiology and End Results program (SEER) of the U.S., the death proportion was the highest among individuals aged 65–74, 29.8%, and of 23.7% among people aged 75–84.⁹ MM revealed by a plasmacytoma has been described previously, mostly presented as intracranial plasmacytoma.^{1,3,6,10,11} However, there have not been many cases of MM revealed by sphenoidal plasmacytoma.^{3,10}

When the skull base is involved, and the tumor is enlarging, it may cause tumefaction, headache, pain, nasal congestion, dizziness, epistaxis, rhinorrhea, cranial nerve palsy, etc.^{4,5,8} In our case, the tumor size was around 1.6*1.9*3.0 cm³ in the first scan and progressed to 2.0*2.3*3.2 cm³ in the following CT. However, the patient encountered none neurological symptoms or other general discomfort except abruptly body weight loss in two years and anemia, which is different from previous cases and not compatible with the clinical manifestations of plasmacytoma or multiple myeloma.

The patient was diagnosed with intracranial meningeal tumor or dural-meningeal metastatic lesions initially, since it lacked specificity and was not differentiated from neuroradiologic findings of intracranial plasmacytoma and meningioma. Moreover, meningeal intracranial tumors are prevalent among elderly women and the symptoms often appear after a prolonged subclinical phase. It is challengeable to accurately diagnose plasmacytoma based only on neuroradiologic findings, especially if there are no other clinical manifestations. Cases of dural-based plasmacytoma resembling meningioma have been reported previously,^{6,11–14} most of which were solitary cranial plasmacytomas and presented with clinical symptoms. Even so there was still the case of plasmacytoma/MM not being diagnosed until the operation had been performed.⁶ In this case, the diagnosis of plasmacytoma was based on tissue biopsy and histopathological findings after following CT imaging, to be later diagnosed with MM through further analyzes, which is a similar situation to other cases. Solitary plasmacytoma occurs rarely¹⁵ and is sometimes presented as the manifestation or concurrent of MM.^{11,14}

The management of plasmacytoma and multiple myeloma are somewhat different. Radiotherapy is the treatment of choice for solitary bone plasmacytoma and transits to chemotherapy if patients do not respond. The role of surgery is merely to accurately diagnose. When MM is the diagnosis, consideration of transplanta-

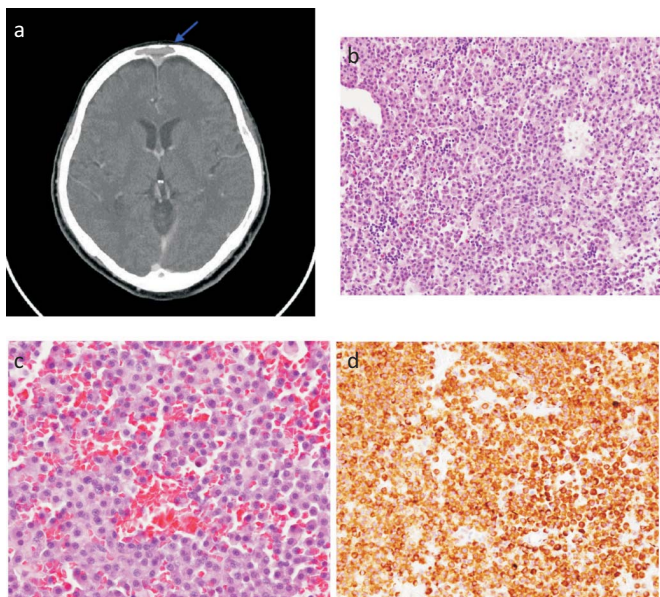


Fig. 2. (a) Biopsy site of new osteolytic lesion in the frontal bone. (b) H&E staining, 200X. (c) H&E staining, 400X. (d) VS38c staining, 200X. The plasmacytoma is composed of monotonous sheets of neoplastic plasma cells with a small volume of normal host tissue.

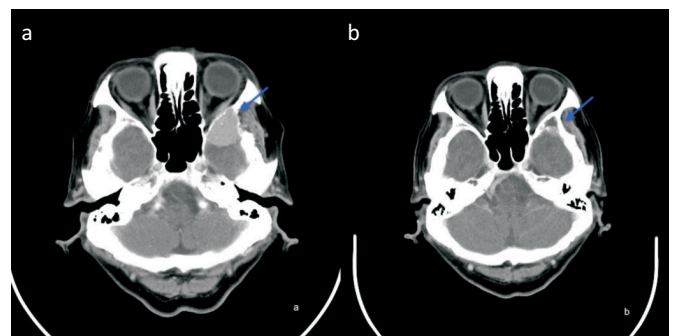


Fig. 3. (a) Brain CT with contrast enhancement after three months revealed a new osteolytic lesion in the frontal bone while the sphenoidal lesion enlarged with a size of 2.0*2.3*3.2 cm³. (b) After seven months of chemotherapy, brain CT with contrast enhancement showed a reduced size of the intracranial sphenoidal lesion.

tion or not, patients' general conditions, and primary induction or maintenance therapy, much systemic chemotherapy, such as bortezomib, thalidomide, lenalidomide, doxorubicin, corticosteroids etc. can be chosen.¹⁶ Hence, accurate diagnosis is crucial to the treatment. Although MM is a malignant disorder among the elderly, the survival rates have improved significantly. It has been reported that the percent of a five-year survival increased from 35.5% in 2000 to 51.8% in 2010.⁹ Despite the age of the patient, under accurate, early diagnosis and suitable therapy, the patient with intracranial skull base MM recovered well and remained in good physical shape.

Skull base plasmacytoma diagnosis is based on the exclusion of other diagnostics by tissue biopsy of the suspected site and clinical manifestations because it is so uncommon. Here the patient had no neurological symptoms or other manifestations, which increased the difficulty of accurate diagnosis based only on radiologic findings without further follow-up. Although neuroradiological findings can help to diagnose, it is difficult to differentiate other skull base tumors from plasmacytoma based only on their results. Therefore, it is critical to consider plasmacytoma into differential diagnosis when encountering a cranial image mimicking meningioma.⁸ Generally, quantitative immunoglobulins (serum IgG, IgA, IgM) can be considered for differentiating the skull plasmacytoma or multiple myeloma from intracranial meningioma with skull invasion. The results may show low levels of one or more immunoglobulins. Also, a full workup before proceeding with therapy is suggested if plasmacytoma is one of the differential diagnosis.

Conflicts of Interest

The material in this manuscript has not been previously published and is not concurrently submitted elsewhere.

All authors declare no potential financial and non-financial conflicts of interest.

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