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## Case Report

# Solitary Skull Base Plasmacytoma with Sella Turcica Extension

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## SUMMARY

A case of 65-year-old female with intrasellar mass was operated on under the diagnosis of pituitary adenoma. The final diagnosis of plasmacytoma was made by immunocytochemical stain of IgG and electron microscopical findings. Postoperatively, the radiological studies of systemic skeletal bone and cytology of bone marrow were normal. However, the serum IgG was high, and the urinary Bence-Jones protein (kappa type) was positive. Although these laboratory data normalized after operation and irradiation, the patient converted to severe multiple myeloma and expired one and half year after operation.

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## 1. Introduction

Plasmacytoma is a pathological diagnosis of multiple myeloma which involves mainly bone marrow and skeletal system. Approximately 40% of cases present neurological complications for their metabolic disorders of biochemical, immunological alterations<sup>1,2</sup> and nerve compressions<sup>3</sup> or meningeal myelomatosis.<sup>4</sup> However, only 3% of cases represent as an intracranial or cranial mass<sup>5</sup> which originate from calvarium, dura or cerebral tissue.<sup>6</sup> We are reporting a case of intrasellar plasmacytoma mimicking those of pituitary tumor.

## 2. Case report

A 65-year-old menopausal female complained of bilateral visual disturbance. The initial symptoms were bilateral hyposphagma and tearing 6 months before admission. The visual disturbance deteriorated progressively 3 months before admission. The movement of left eyeball became restricted.

### 2.1. Physical and laboratory examinations

Physical examinations revealed palsy of left sixth cranial nerve and atrophy of left optic disc. Her left visual acuity was totally lost and the right O.D. was 0.2 with a central defect of visual field. There was neither galactorrhea nor other signs of hyperpituitarism. The nasopharyngeal examination was within normal limits. The laboratory data showed slightly high level of total protein (8.9 gm/dl) with 51.3% albumin. The immunoglobulin fractions were  $\alpha_1$  2.8%,  $\alpha_2$  6.7%,  $\beta$  5.5% and  $\gamma$  33.7%, indicating an increase of gamma globulin (normal 11.3–24.3%). Hemoglobin was 13.5 gm/dl. The

electrolytes were normal. There was proteinuria. The pituitary hormones including prolactin (70 ng/ml), LH (25 mIU/ml) and FSH (44 mIU/ml) were slightly higher than normal values. Absence of end-organ damage, such as hypercalcemia, renal insufficiency, anemia and bone lesions (CRAB) attributed to a plasma cell proliferative disorder was observed in this studied subject.

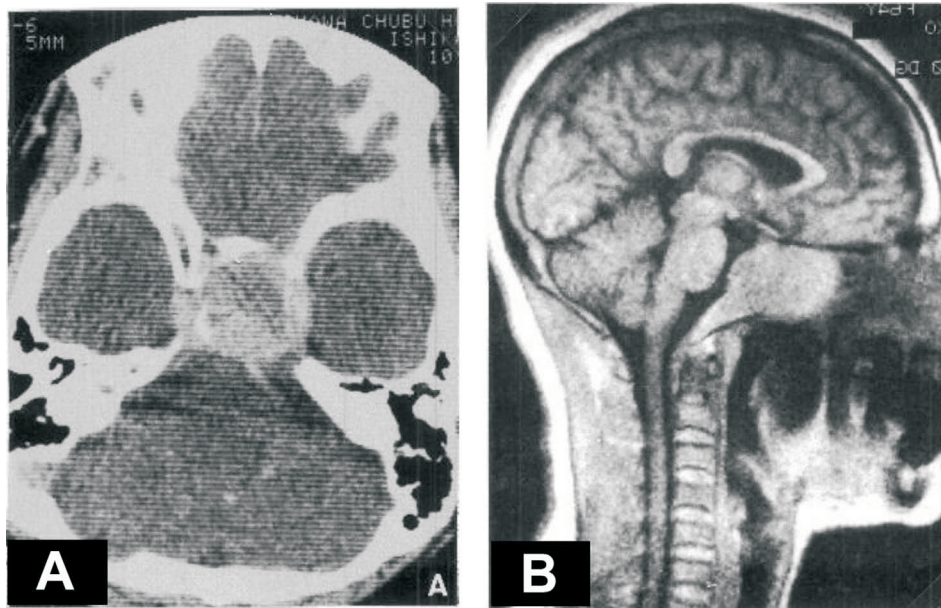
However, the scintigram of radiotracer showed a normal distribution over the skeletal system, and the roentgenogram of the whole skeletal system was also normal. There was no evidence of plasmacytosis in two studies of bone marrow aspirate. Skull roentgenogram and tomogram showed an enlarged turcica sella and eroded floor. The mass shadow measured 5 by 5 cm, extending downward into the sphenoid sinus. The computerized tomography revealed an intra- and supra-sellar isodense mass. The mass was homogeneously enhanced after intravenous injection of contrast medium (Fig. 1A). The MRI image depicted a sphenoid sinus tumor extended and invaded the sellar region. The whole outline of the tumor was clearly delineated by magnetic resonance imaging (Fig. 1B). The carotid angiograms showed that both internal carotid arteries were compressed laterally and the anterior cerebral arteries elevated. The tumor was slightly hypervascular, which was supplied mainly from both internal carotid arteries.

A normal skeletal survey with an additional solitary lesion found in the MRI of this studied subjects. Based on the findings above, plasma cell myeloma (PCM) was highly suspected in this patient.

#### 2.1.1. Surgical intervention

The transcranial approach was performed to evacuate this space occupying lesion. Grossly, the tumor was dark-reddish, encapsulated and well-demarcated. The tumor tissue was soft and easily removed by vacuum. The intraoperative findings confirmed that the thin pituitary gland-like organ was compressed posteriorly by the lesion.

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**Fig. 1.** A Computerized tomogram. The intrasellar tumor is homogeneously enhanced by contrast medium. B The sagittal T1 figure of magnetic resonance imaging reveals that the tumor is clearly demarcated.

## 2.2. Immunohistochemistry and pathological examinations

Light microscopy findings revealed a hypercellular tumor with fine fibrous septa. The tumor cells were round with eccentric nuclei and cartwheel arrangement of chromatin (Fig. 2A). The immunocytochemical stain by the method of peroxidase-antiperoxidase indicated that the tumor was positive for IgG (Fig. 2B), but negative for other immunoglobulins, prolactin, LH, FSH, GH and ACTH. The electron microscopical examinations of the thin sections which were refixed from the formalin-fixed specimen revealed a prominent nucleolus in an eccentric cartwheel nucleus. The rough endoplasmic reticulum and mitochondria were well developed, and the secretory granule was absent (Fig. 3). The tumor was then diagnosed as plasmacytoma based on these findings.

## 2.3. Postoperative studies

The laboratory examinations demonstrated a remarkable increase of IgG: 2610 mg/dl (normal 800–1800 mg/dl). The Immunoelectrophoresis of serum showed reactions to anti-IgG and anti-kappa serum. Urine was positive with Bence-Jones protein of kappa type.

## 2.4. Postoperative course

The postoperative course was uneventful. Visual acuity of the

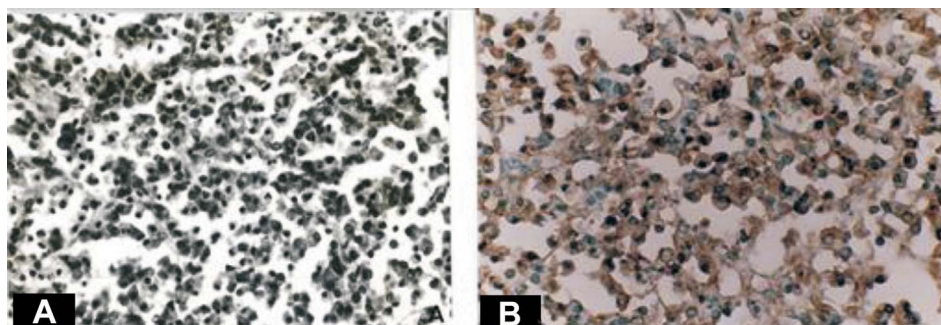
left eye improved to n.d./30 cm after 50 Gy of cobalt irradiation locally.

3 months after operation, the pituitary hormones and IgG decreased to normal level, and the proteinuria diminished. The systemic skeletal roentgenogram revealed no osteolytic change. The patient remained well until one year after operation, when she complained of right femoral pain. She was hospitalized again for the osteolytic lesion of right femur. Laboratory data showed high level of IgG (7300 mg/dl) and Bence-Jones protein. The patient developed overt multiple myeloma in spite of irradiation and chemotherapy. Lasmacytoma usually has good prognosis but 15% can progress to PCM. Whether it is originally PCM or progresses to PCM. It will be better clarified by normal skeletal survey and MRI or CT.

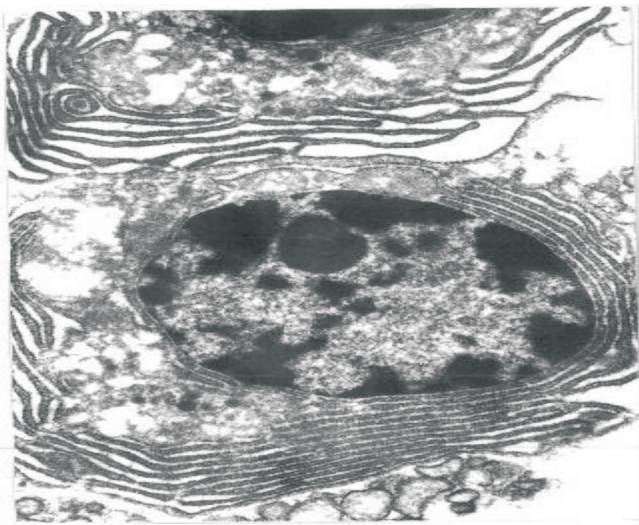
## 3. Discussion

Intrasellar plasmacytoma. Intracranial plasmacytomas are rare tumors that constitute less than 1% of intracranial neoplasms. They may present as a solitary plasmacytoma or may be part of a systemic malignant plasmacytosis, as in multiple myeloma. Isolated cranial mononeuropathy (e.g., sixth nerve palsy) leading to diplopia is a common presentation.<sup>7,8</sup> We describe a case of Intrasellar plasmacytoma presenting with abducens nerve palsies, bilateral visual disturbance, hyposphagma.

This tumor may occur at any age, but is mostly seen in patients



**Fig. 2.** A Light microphotogram. The tumor is abundant of round cells with eccentric nucleus and cartwheel arrangement of chromatin HE  $\times$  400. B Immunohistochemical stain shows positive stain with Ig G.  $\times$ 400.



**Fig. 3.** Electron microscopical findings. The eccentric cartwheel and well developed rough endoplasmic reticulum are seen. The mitochondria are artificially swollen for incomplete fixation.  $\times 6000$ .

in their fifties or sixties. Pituitary masses can be of various etiologies. The most common is still pituitary adenoma. Other, less common entities include gliomas, meningiomas, craniopharyngiomas, Rathke's cleft cysts, epidermoids, chordomas, germ cell tumors, metastatic tumors, vascular lesions and granulomatous, infectious or inflammatory processes.<sup>9–11</sup>

The clinical and neuroradiological findings are generally non-specific, so they are often misdiagnosed or masqueraded pre-operatively. On both CT and MRI scans, there may be mild to significant enhancement as shown in our case. Plasmacytomas involving sella turcica are extremely rare. All patients complained of diplopia or visual loss for a period of 5 days–6 months before hospitalization. Since it is difficult to differentiate from pituitary adenoma, some cases were operated under the diagnosis of pituitary adenoma.<sup>12</sup> Additionally, the tumor invades suprasellarly, the hormonal alterations owing to the blockage of the releasing or inhibiting factors from hypothalamus<sup>13</sup> make this tumor mimic the non-functioning pituitary adenoma.

Consequently, the final diagnosis should be made by pathological findings, especially by immunocytochemical stain of immunoglobulins and electron microscopy proof of the plasma cells are lacking secretory granules.

Plasmacytomas are malignant tumors that are essentially osseous. The cervicocephalic region is rarely afflicted (1%). Extension to the intrasellar region seldom occurs. Fewer than 30 cases have been documented in the literature simulating the other numerous neoplastic intrasellar lesions.

The role of surgery in solitary plasmacytoma treatment remains

controversial due to the incongruent findings reported by a limited number of studies. Based on our large population study, we conclude that therapy of combined RT with surgery is beneficial in axial plasmacytoma. Although increased survival rates were observed for patients treated with r or surgery, disease progression to myeloma was still evident.<sup>14</sup>

The patient remained well after operation. She was rehospitalized for femoral pain, which developed to overt multiple myeloma despite irradiation and chemotherapy treatment. It coincides with the literature that tumors that measured at 5 cm or more in diameter at diagnosis had a higher risk of treatment failure when managed with local radiotherapy alone.<sup>15</sup>

### Conflicts of interest

All authors declare no conflict of interest.

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