

The solitary sellar plasmacytoma: a diagnostic challenge

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Summary

Solitary sellar plasmacytomas are exceedingly rare and difficult to distinguish from other pituitary tumors. We report a case of a 62-year-old woman presenting with blurred vision of the right eye and tenderness of the right temporal region, which was interpreted as temporal arteritis. MRI revealed a pituitary mass lesion (20 mm × 14 mm × 17 mm) without compression of the optic chiasm and her pituitary function was normal. Pituitary surgery was undertaken due to growth of the lesion, and histopathological examination showed a highly cellular neoplasm composed of mature monoclonal plasma cells. Subsequent examinations revealed no evidence of extrasellar myeloma. The patient received pituitary irradiation and has remained well and free of symptoms apart from iatrogenic central diabetes insipidus. Until now, only eight cases of solitary sellar plasmacytoma have been reported. Most frequent symptoms stem from compression of the cranial nerves in the cavernous sinus (III, IV, V), whereas the anterior pituitary function is mostly intact.

Learning points:

- A solitary plasmacytoma is a rare cause of a sellar mass lesion.
- The radiological and clinical features are nonspecific, but cranial nerve affection and intact pituitary function are usually present.
- The diagnosis is made histologically and has important therapeutic implications.

Background

Plasmacytomas are characterized by monoclonal plasma cell proliferation in a single lesion, whereas multiple myeloma is a multifocal plasma cell neoplasm characterized by elevated serum monoclonal protein levels and/or increased bone marrow clonal cells with related organ or tissue impairment (increased calcium levels, renal insufficiency, anemia, and bone lesions). Solitary plasmacytomas are uncommon and can either arise from bone or soft tissue (1). Solitary sellar plasmacytomas are exceedingly rare and to date there has only been eight reported cases without progression to multiple myeloma

(2, 3, 4, 5, 6, 7). Clinically and radiologically, sellar plasmacytomas mimic benign pituitary tumors, which make the differential diagnosis difficult (3, 6).

We present the ninth case of a solitary intrasellar plasmacytoma, which revealed several emblematic features.

Case presentation

A 62-year-old African American woman presented with blurred vision of the right eye and pain in the

right temporal region. Her previous medical history was unremarkable with no family history of endocrine disorders or malignancies, except for a cousin with multiple myeloma. There were no symptoms of hypopituitarism, hyperprolactinemia, acromegaly, Cushing's disease, or diabetes insipidus. Physical examination revealed a 50% reduction in visual acuity of the right eye and tenderness along the right temporal artery, but there was no diplopia or visual field narrowing. Blood chemistry values were normal apart from a slightly elevated erythrocyte sedimentation rate of 33 mm (normal value <30 mm) and elevated levels of immunoglobulin G (Table 1).

Temporal arteritis was suspected and high-dose prednisolone was administered with transient symptom relief. A biopsy of the temporal artery was normal. Blood and urine profile, X-ray of the thorax, and abdominal ultrasound examination were unremarkable and prednisolone treatment was discontinued. This caused a relapse of the patient's symptoms and prednisolone treatment was restarted with symptomatic effect. To pursue a diagnosis of large vessel arteritis a PET-CT scan was performed, which showed no pathology. The diagnosis of temporal arteritis was re-evaluated, and on the suspicion of multiple sclerosis with optic neuritis, she was referred to the neurologic department. An MRI of the brain was performed, which revealed a homogeneously enhanced 20 mm × 14 mm × 17 mm intrasellar mass lesion encasing the right internal carotid artery and in close proximity to the right optic nerve.

The initial diagnosis was a meningioma. There was no evidence of hypopituitarism. Renewed MRI of the brain performed 5 months later showed tumor growth with encasement of the optic nerves and erosion of the sphenoid bone (Fig. 1A and B). Due to the tumor size and the erosion of planum sphenoidale, a transcranial excision of the tumor was performed. Histopathological examination showed no pituitary tissue, but a highly cellular neoplasm composed of mature plasma cells with round eccentrically placed nuclei (Fig. 2A). Immunohistochemistry was positive for CD79A and MUM-1 (Fig. 2B), while *in situ* hybridization for kappa and lambda light chains showed monoclonality for kappa (Fig. 2C and D), confirming that the tumor was a plasmacytoma. The postoperative course was uneventful except for the development of permanent central diabetes insipidus (Table 1). Extensive investigation for multiple myeloma was performed, including blood and urine profile, skeletal X-rays, lumbar puncture, and a bone marrow biopsy. All results were normal and thus, the diagnosis of a solitary sellar plasmacytoma was made. Postoperative MRI of the brain showed no residual tumor (Fig. 1C and D), but radiation therapy directed to the sellar region (46 Gy over a 5-week period) was conducted.

Investigation

An MRI of the brain was not performed until 6 months after onset of the symptoms. A repeat MRI of the brain

Table 1 Laboratory findings on presentation and posttherapeutically.

	On presentation	Postoperatively	Postradiotherapy	Reference range
ACTH			6.0	7.0–64.0 ng/L
Cortisol	176.0		241.0	171.0–536.0 nmol/L
Cortisol (30 min)			732.0	>500 nmol/L
TSH	0.91	0.26	0.67	0.3–4.5 × 10 ⁻³ IU/L
Total T ₃	1.48	1.31	1.64	1.10–2.50 nmol/L
Total T ₄	78.0	71.0	76.0	60.0–140.0 nmol/L
IGF1			11.3	7.0–26.6 nmol/L
Prolactin	304.0	965.0	697.0	90.0–580.0 × 10 ⁻³ IU/L
FSH	29.0	21.0	19.3	16.0–157.0 IU/L
LH	10.9	6.5	5.0	7.7–59.0 IU/L
Immunoglobulin G	16.0	9.6	7.6	6.1–14.9 g/L
M-protein	Negative	Negative	Negative	
Hemoglobin (whole blood)	7.4	7.5	7.6	7.3–9.5 mmol/L
Calcium	2.26	2.41	2.39	2.20–2.55 mmol/L
Creatinine	76.0	86.0	87.0	45.0–90.0 μmol/L
Sodium	139.0	142.0	138.0	137.0–145.0 mmol/L
Potassium	4.2	4.0	3.9	3.5–4.6 mmol/L

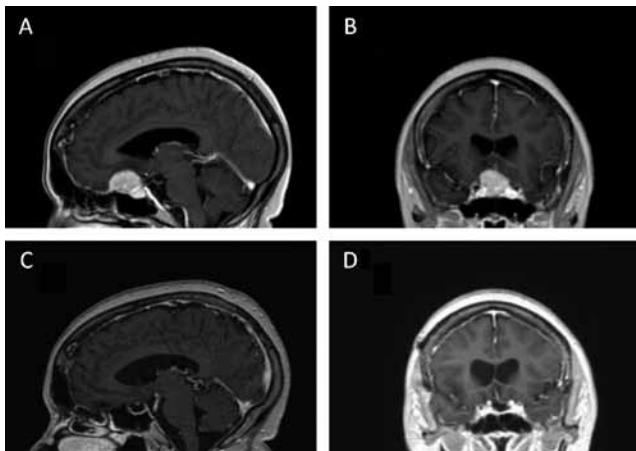


Figure 1
MRI of the brain preoperatively (A and B) and 12 months after surgery (C and D).

was performed 5 months later (Fig. 1A and B) and 1 month after surgery. In addition, postoperative MRI of the brain was performed 6, 12, and 18 months after surgery (Fig. 1C and D).

The histopathological examination of the tumor showed a plasmacytoma with light chain kappa monoclonality (Fig. 2). Investigation for multiple myeloma included blood and urine profile, skeletal X-ray survey, lumbar puncture, and bone marrow biopsy.

Four weeks after cessation of prednisolone treatment, a short synacthen test was performed showing no evidence

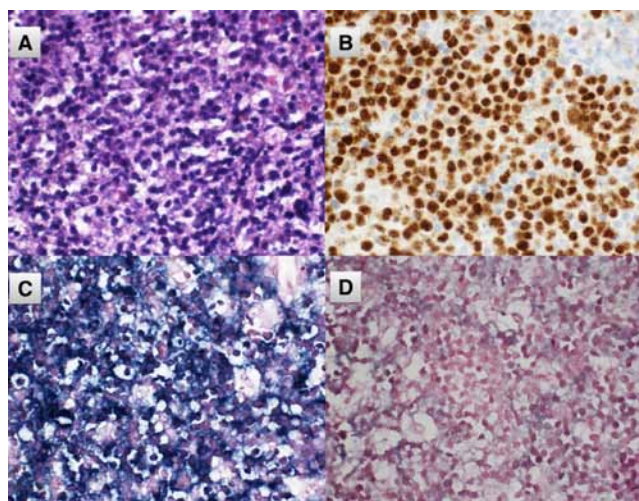


Figure 2
(A) Standard H&E $\times 400$, (B) immunoperoxidase staining (brown nuclear positivity) for MUM-1 $\times 400$, (C) *in situ* hybridization for kappa light chains $\times 400$, (D) *in situ* hybridization for lambda light chains $\times 400$.

of adrenal insufficiency (cortisol levels (mmol/L): 241 (basal) and 732 (30 min)).

Treatment

The patient was treated with transcranial surgery followed by fractionated irradiation of the sellar region (46 Gy over a 5-week period).

Outcome and follow-up

Three and a half years after the first admission to the hospital the patient is doing well. The diabetes insipidus persists, but the plasmacytoma remains in remission and the patient has not developed multiple myeloma. The patient is followed biannually on an outpatient basis by an endocrinologist and a hematologist.

Discussion

We report a rare case of a solitary sellar plasmacytoma, which initially masqueraded as temporal arteritis. So far, 36 cases of sellar plasmacytomas have been reported in the literature (2, 3, 5, 6, 7), but only eight cases were classified as solitary plasmacytomas (4).

Most patients presented with cranial nerve palsies, mainly involving nerves II–VI with symptoms of headache, diplopia, visual loss, eye pain, ptosis, photophobia, facial numbness, and craniofacial pain. The majority of patients presented with intact anterior pituitary function, as in our case.

Sellar plasmacytomas may mimic more common pituitary mass lesions such as adenomas and meningiomas as regards clinical presentation and imaging studies. Additional juxtaseellar mass lesions include craniopharyngioma, chordomas, germ cell tumors, as well as granulomatous lesions and metastases. In most cases, a definitive diagnosis is made histologically, but it is important to recognize that incidental pituitary tumors are frequent and that a histological diagnosis is not considered mandatory in all cases (8). However, rapid tumor growth during follow-up and/or compression of the optic chiasm constitutes an indication for surgical decompression and exploration.

A solitary plasmacytoma involving the pituitary gland without systemic myelomatous dissemination is rare. It is of great importance to continue to follow-up the patient, since more than 50% of the patients will develop multiple myeloma within 10 years (4). Solitary

extramedullary plasmacytomas are less common than solitary bone plasmacytomas. Progressive disease may present as multiple myeloma, solitary bone plasmacytoma, or soft tissue involvement of lymph nodes, skin or subcutaneous tissues. When distant relapse occurs this tends to be within 2–3 years of initial diagnosis (9). Treatment for sellar plasmacytomas is most commonly radiotherapy, whereas chemotherapy is the first-line therapy for systemic myelomatous disease (4, 7).

Interestingly, and illustrated by this present case, distinct ethnic difference exists regarding the epidemiology of multiple myeloma, since the disease is two- to three-fold more common in African Americans compared with Caucasians, and multiple myeloma is the most common hematological malignancy in African Americans (10).

In conclusion, although rare, a solitary plasmacytoma should be considered as a differential diagnosis in a patient presenting a pituitary mass lesion in combination with cranial nerve affection and intact pituitary function.

Declaration of interest

The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of the research reported.

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Patient consent

Written informed consent was obtained from the patient for publication of this case report.

Author contribution statement

A S and U K collected data and wrote the manuscript, S D, G T, and P L P were in charge of the patient treatment, S B was responsible for pituitary tumor histology and the histological perspective of the case report. J O L J was also in charge of the patient treatment and contributed to the manuscript.

References

- 1 The International Myeloma Working Group 2003 Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: a report of the International Myeloma Group. *British Journal of Haematology* **121** 749–757.
- 2 Jiang CZ, Lin QS, Wu XY, Wang CY & Kang DZ 2014 Sellar solitary plasmacytoma progressing to multiple myeloma: a case report and literature review. *Medicine* **93** e58. (doi:10.1097/MD.000000000000058)
- 3 Joukhader R & Chiu K 2012 Sellar plasmacytomas: a concise review. *Pituitary* **15** 146–149. (doi:10.1007/s11102-011-0352-x)
- 4 Yaman E, Benekli M, Coskun U, Sezer K, Ozturk B, Kaya AO, Yildiz R, Uluoglu O & Buyukberber S 2008 Intraseellar plasmacytoma: an unusual presentation of multiple myeloma. *Acta Neurochirurgica* **150** 921–924. (doi:10.1007/s00701-008-0012-8)
- 5 Udiawar M, Bejnariu C & Davies S 2012 Metastatic haematological malignancy presenting as a sellar mass. *BMJ Case Reports* **2012** pii: bcr0320126050. (doi:10.1136/bcr-03-2012-6050)
- 6 Sinnot BP, Hatipoglu B & Sarne DH 2006 Intraseellar plasmacytoma presenting as a non-functional invasive pituitary macro-adenoma: a case report & literature review. *Pituitary* **9** 65–72. (doi:10.1007/s11102-006-8281-9)
- 7 Kahn IS, Javalkar V, Thakur JD & Nanda A 2012 Intraseellar plasmacytoma: an illustrative case and literature review. *Journal of Clinical Neuroscience* **19** 210–213. (doi:10.1016/j.jocn.2011.07.009)
- 8 Levy A 2004 Pituitary disease: presentation, diagnosis and management. *Journal of Neurology, Neurosurgery, and Psychiatry* **75** iii47–iii52. (doi:10.1136/jnnp.2004.045740)
- 9 Soutar R, Lucraft H, Jackson G, Reece A, Bird J, Low E & Samson D 2004 Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extramedullary plasmacytoma. *British Journal of Haematology* **124** 717–726. (doi:10.1111/bjh.2004.124.issue-6)
- 10 Landgren O & Weiss BM 2009 Patterns of monoclonal gammopathy of undetermined significance and multiple myeloma in various ethnic/racial groups: support for genetic factors in pathogenesis. *Leukemia* **23** 1691–1697. (doi:10.1038/leu.2009.134)

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