Journal of Clinical Neuroscience 50 (2018) 20–23

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Review article

Sellar plasmacytomas masquerading as pituitary adenomas: A systematic review

Joseph DiDomenico [a], Leonel Ampie [b,c], Winward Choy [d], Jonathan B. Lamano [a], Daniel E. Oyon [a], Kartik Kesavabhotla [a], Orin Bloch [a,*]

[a] Department of Neurological Surgery, Northwestern University, Feinberg School of Medicine, 676 N. St Clair St, Suite 2210, Chicago, IL 60611-2911, USA
[b] Surgical Neurology Branch, NINDS, National Institutes of Health, 10 Center Drive, Room 3D20 MSC 1414, Bethesda, MD 20892-1414, USA
[c] Department of Neurological Surgery, University of Virginia School of Medicine, Charlottesville, VA 22908, USA
[d] Department of Neurological Surgery, University of California, San Francisco, 505 Parnassus Ave., Room 779 M, San Francisco, CA 94143-0112, USA

ARTICLE INFO

Article history:
Received 8 July 2017
Accepted 8 January 2018

Keywords:
Sellar
Plasmacytoma
Pituitary
Intracranial
Multiple myeloma

ABSTRACT

Given the rarity of intracranial plasmacytomas, these lesions are frequently misdiagnosed as pituitary adenomas. We report on the distinguishing characteristics of sellar plasmacytomas from cases in the literature and our experience. A literature search was conducted to collect all documented cases of a plasmacytoma originating in the sellar region. Patient characteristics, medical history, presentation, tumor characteristics, and survival data were collected. An additional case from our institution not previously reported was included. Thirty-one patients with sellar plasmacytomas were studied. Presenting symptoms were most commonly headache (68%), diplopia (65%) and visual field disturbances (10%). Fifteen patients (48%) were initially suspected of having a pituitary adenoma. Pathologic diagnosis of plasmacytoma preceded a finding of multiple myeloma in 14 cases (45%). Thirty patients (90%) had surgical intervention. Adjuvant therapy consisted of radiotherapy for twenty-five patients (81%) and chemotherapy for sixteen (52%). Tumor recurrence was reported for 7 cases (23%). Nine deaths were reported (23%). We demonstrate that cranial nerve involvement is far more common in sellar plasmacytomas than conventional pituitary adenomas. Given the successful management of these tumors with radiotherapy, such deficits, particularly in patients with known multiple myeloma, should impact the diagnostic workup and treatment considerations.

1. Introduction

Intracranial manifestations of plasma cell dyscrasias are exceedingly rare, even amongst patients with a historic diagnosis of a hematogenous disorder. More specifically, plasmacytomas originating in the sellar region are often misdiagnosed as an adenoma due to the rarity of this disease and the limited clinical experience of the entity in this location. Here within, we report our experience in diagnosing a sellar plasmacytoma following a discovery of multiple myeloma. We also present a qualitative review of the literature to further expand on the commonalities in clinical presentation and characteristics of patients diagnosed with sellar plasmacytomas.

2. Report of a case

A previously healthy, 57-year-old man presented with worsening, atraumatic hip pain of three weeks. Physical examination was notable for pain on palpation of the left greater trochanter. No focal neurologic deficits were noted. Laboratory studies revealed an elevated lactate dehydrogenase of 555 units/liter; hematologic values were otherwise within normal limits. A computerized topography (CT) scan of the left lower extremity revealed a pathologic fracture beneath the greater trochanter and a 6.7 × 2.4 cm soft tissue mass extending through the anterior cortex of the proximal femur. A metastatic work-up was initiated and remarkable for bilateral lesions in the lower poles of the kidneys. Magnetic resonance imaging (MRI) of the spine revealed multiple lesions in the bony structures. A core biopsy of the mass along the left femur was performed, with an immunohistochemical profile notable for neoplastic cells positive in CD138, kappa light chains, CD56, and CD45, concerning for a plasma cell neoplasm. Serum plasma electrophoresis and immunofixation demonstrated elevated beta

* Corresponding author.
E-mail address: orin.bloch@northwestern.edu (O. Bloch).

https://doi.org/10.1016/j.jocn.2018.01.022
0967-5868/

globulin (=1.7 g/dL; N = 0.6 to 1.3 g/dL), albumin (=3.2 g/dL; N = 3.5 to 5.7 g/dL), and serum kappa light chains (=654 mg/dL; N = 170–370 mg/dL). Serum lambda light chain levels were decreased (=69 mg/dL; N = 90–210 mg/dL). Serum quantitative IgA was markedly elevated (=1950 mg/dL; N = 70 to 400). Urine studies were notable for elevated total protein (=121 mg; N = 50–80 mg) and immunofixation positive for IgA kappa light chains. A bone marrow aspirate showed a plasma cell dyscrasia with 95% cellular involvement (10% IgA kappa light chain restricted), confirming a diagnosis of multiple myeloma, Durie-Salmon (DS) stage III.

Ten days after operative repair of the pathologic fracture, the patient developed left-sided ptosis and diplopia on awakening. On exam, that patient's left pupil was dilated and minimally reactive, his visual acuity in the left eye had diminished to 20/100, and left third and sixth cranial nerve palsies were discovered. Hypothalamic-pituitary axis hormone studies were within normal limits. Brain MRI revealed a 1.2 × 1.8 × 0.9 cm hypoenhancing, T2 isointense intrasellar mass with mild sellar expansion, left cavernous sinus invasion, and minimal suprasellar extension (Fig. 1A-C). CT imaging demonstrated erosion of the left dorsum sellae and left anterior clivus (Fig. 1D). Positron emission tomography–computed tomography (PET/CT) imaging demonstrated abnormalities throughout the axial and appendicular skeleton consistent with active myeloma, and a prominent focus of increased metabolic activity in the region of the sella suggested a myelomatous process (Fig. 2). The patient underwent gross total resection of the mass by an endonasal, transsphenoidal approach. A biopsy specimen demonstrated sheets of kappa light chain restricted, CD138/kappa/Ki-67 positive plasma cells, confirming the diagnosis of a sellar plasmacytoma (Table 1). Adjuvant treatment included a combination of bortezomib, lenalidomide, and dexamethasone, along with stem cell therapy, for the systemic disease, and radiation therapy to the thoracic spine for a metastatic component with associated cord compression.

3. Literature review

3.1. Methods

A literature search was conducted via OVID (Medline) utilizing the following search terms and boolean operators: {plasmacytoma OR myeloma} AND {sella OR hypophysis OR pituitary OR sellar}. The search was further limited to the English language and to humans only. No date restriction was imposed. A total of 97 articles were identified and screened for relevance. Citations were reviewed for referred cases that may not have been included in the search, leading to 11 additional articles screened for relevance. From the total of 108 publications, 29 articles met inclusion criteria of a plasmacytoma in the sellar/pituitary region. There were 12 articles in which the text was not accessible, and the remaining articles diverged from the inclusion criteria and were deemed irrelevant. Cases were disaggregated and clinical data collected including patient demographics, medical history, symptomatology, tumor characteristics, treatment, recurrence rate, and survival data.

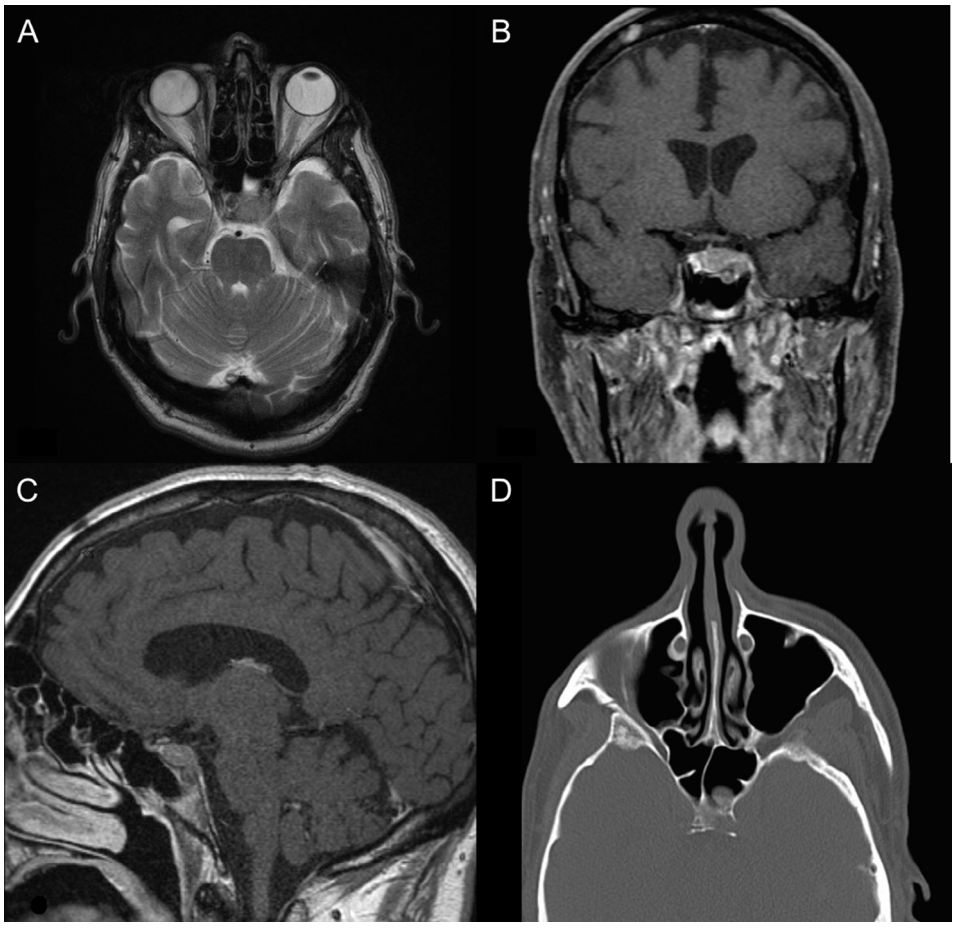

Fig. 1. Pre-operative images of sellar tumor (A) Axial T2-weighted MRI (B) Sagittal T1-weighted post-contrast MRI (C) Coronal T1-weights post-contrast MRI (D) CT of the sinuses demonstrating cavernous invasion.