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Rare presentation of Ewing sarcoma metastasis to the sella and suprasellar cistern

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Abstract

We present an exceedingly rare case of a Ewing sarcoma metastasis manifesting as a sellar mass mimicking a pituitary adenoma. The differential diagnosis of the young adult with a sellar mass is presented and correlated with a review of available literature, demonstrating this case's unique potential for clinical teaching. More specifically, this case illustrates that in a patient with a clinical history of Ewing sarcoma, a metastasis may involve the sella and suprasellar cistern without apparent osseous involvement.

Keywords

Ewing sarcoma; metastasis; sella; suprasellar; pituitary

1. Introduction

Ewing sarcoma (ES) family of tumors is now considered a spectrum of neoplastic disease which may develop in any bone or soft tissue, including extraskeletal variants, Primitive Neuroectodermal Tumor (PNET) and Askin tumor. Most commonly, ES presents in the first 3 decades of life with osseous involvement of the pelvis or femur. Unfortunately, at least subclinical metastatic involvement must be presumed present in nearly every patient, and chemotherapy is a necessary treatment following local surgical control [1]. Intracranial or CNS metastatic involvement has been appreciated as a significant risk in up to 15% of patients [2]. We present a rare case of a Ewing sarcoma metastasis to the sella in a young adult.

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2. Case Report

A 30 year old woman presented to her primary physician with 3 months of progressively worsening left supraorbital headache. The patient had a history of 3 primary cancers all considered to be in remission. These included Ewing sarcoma of the iliac bone status post chemotherapy (7 years prior), papillary thyroid carcinoma status post thyroidectomy (5 years prior), as well as ovarian mucinous adenocarcinoma status post salpingo-oophorectomy (1 year prior). The patient does not have a known genetic syndrome to account for her three malignancies. No neurologic deficits were identified during initial clinical examination.

She was referred for imaging at our institution, at which time an MRI of the brain revealed a 1.4 cm mass in the sella with suprasellar extension and impingement on the optic chiasm (Fig. 1). Noncontrast imaging demonstrated heterogeneous signal characteristics with confluent regions of both T1 and T2 hypointensity (Fig. 1) as well as trace foci of T1 hyperintensity possibly secondary to subacute blood products. There was no evidence of suspicious or abnormal marrow signal to suggest osseous involvement. Following endocrinologic evaluation (with unrevealing serum functional testing including measurement of prolactin, IGF-1 and TSH levels), the lesion was felt to represent an incidental nonfunctioning pituitary macroadenoma with subacute blood products, although close followup was advised. Her headache improved with steroid taper. Regularly scheduled MRI of the brain/sella obtained 4 weeks later demonstrated interval increase in size of the sellar/suprasellar mass (now 2.0 cm) (Fig. 2). Postcontrast T1 weighted images demonstrated primarily hypoenhancing and nonehancing components (Fig. 3). In addition, there was trace T2 hyperintensity suggestive of new edema in the left optic nerve (Figs. 4). Whole body imaging with follow up PET CT was performed 3 days after the MRI brain/sella and demonstrated mild sellar FDG avidity but no evidence of additional abnormal hypermetabolic focus or osseous destruction on low mAS CT images (Fig. 5). The patient denied visual changes at this time, but endorsed persistent headache.

Given interval growth and past medical history, differential considerations at this time shifted towards a metastatic lesion, with Ewing sarcoma favored over the patient's other treated primaries. Trans-sphenoidal biopsy of the mass was obtained, and histologic examination of the sellar mass disclosed a monomorphous small round blue cell tumor (Fig. 6a). Immunohistochemical studies supported the diagnosis of Ewing sarcoma/PNET by demonstrating strong membranous cytoplasmic labeling for CD 99 (Fig. 6b) and nuclear labeling for FLI-1 (Fig. 6c) in the absence of labeling for CD 45, synaptophysin, chromogranin, TdT, desmin or myogenin. Fluorescence in situ hybridization (FISH) analysis confirmed rearrangement of the EWSR 1 gene. The patient underwent chemoradiation therapy with subsequent improvement in headache related to the tumor mass. A 3 month follow-up MRI demonstrated decreased size of the sellar/suprasellar metastasis and resolution of mass effect upon the optic chiasm.

3. Discussion

The young adult presenting with a sellar mass presents a challenging differential, for which imaging can be helpful in targeting clinical evaluation and treatment. Both true “pediatric”

and “adult” entities must be considered. Pediatric neoplasms located in the sella or suprasellar region are a distinct entity from their much more common parenchymal counterparts. In order of general decreasing incidence, one must consider craniopharyngioma (80-90% of all pediatric sellar tumors), pituitary adenoma, germ cell tumors, suprasellar glioma, and finally metastatic lesions [3]. Pediatric adamantinomatous craniopharyngioma typically presents as a cystic mass with calcifications and enhancing components [4]. In contradistinction to their adult counterparts, chemotherapy and radiation remain useful adjuncts to surgery in treatment of pediatric sellar tumors.

Adult sellar tumors are overwhelmingly pituitary adenomas, the most common sellar mass in patients from the third decade on, and account for 10-15% of all intracranial neoplasms [5]. The pituitary macroadenoma typically presents as a sellar mass inseparable from the pituitary gland with “upward” suprasellar extension [6]. The adult differential includes additional benign and malignant etiologies such as meningioma, lymphoma and chordoma. Metastases to the sellar region account for 1-2% of adult sellar masses, most commonly arising from lung or breast primary [5, 7]. Survival is poor, averaging approximately 6 months in one series [7].

Ewing sarcoma is a rare neoplasm of childhood and young adulthood, typically presenting with an osseous primary and representing approximately 10% of all pediatric osseous primary tumors [2]. Ewing sarcoma of bone, extraskelatal Ewing sarcoma, Askin tumor and PNET are now recognized as a spectrum of neoplasms within the Ewing sarcoma family, which share a common abnormal karyotypic translocation involving chromosomes 11 and 22. Histologically, these lesions demonstrate monotonous sheets of small round blue cells [1]. Various estimates of intracranial or central nervous system involvement vary in the literature, with larger case series reporting 6-8% involvement [8]. Intracranial involvement has been associated with late stage or advanced disease, often centered at the osseous structures of the skull base or calvarium [9]. Our case is unusual in that metastatic disease presented as intracranial sellar/suprasellar mass without discrete osseous involvement on the MRI and PET-CT scans. A potential limitation of the study is that a dedicated high resolution CT scan of the skull base was not performed to more definitively exclude the possibility of the tumor originating in bone.

Ewing sarcoma presenting as sellar mass is a rare presentation and a diagnostic challenge. Past medical history and endocrinologic testing may provide important clues to broaden the differential beyond adenoma in a young adult. Heterogenous imaging appearance may overlap with additional cystic or hemorrhagic masses such as adenoma; however, short term followup as in our case should help focus the differential.

Systematic review of literature yields numerous case reports of calvarial involvement by ES [10, 11]. Examples of predominantly soft tissue sellar or suprasellar involvement were identified in 2 sources [9, 12]. Elias and Uhl report a Ewing sarcoma metastasis centered in the cavernous sinus with an intrasellar component [9]. Weins and Hattab list a single case of sellar/suprasellar involvement of ES in their retrospective review of 20 years of CNS metastases at their institution, although no imaging or further detail (i.e. osseous

involvement) is presented. Our patient therefore likely represents the first reported case of “purely” sellar/suprasellar Ewing sarcoma in the literature to date.

4. Conclusion

We report an extremely rare case of Ewing sarcoma metastasis presenting as a sellar mass. Although rare, it is important to include metastases in the differential diagnosis of an unusual sellar/suprasellar mass in a patient with history of Ewing sarcoma, even if no active osseous involvement is identified. Failure to recognize this appearance could potentially lead to a clinically devastating delay in diagnosis and treatment.

Acknowledgments

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a

b

Fig. 1. Sagittal T1-weighted noncontrast (a) and axial T2-weighted (b) images at presentation through the sella demonstrate an abnormal heterogeneous intensity mass with confluent regions of T1 and T2 hypointensity centered within the sella with mild suprasellar extension. No osseous abnormality of the skull base is appreciated. The mass was presumed to be a pituitary adenoma.

Fig. 2.

Images 1 month after presentation- Sagittal T1-weighted pre contrast image through the sella at the same slice selection as Figure 1 demonstrate marked interval growth of the sellar/suprasellar mass, which now abuts and elevates the optic chiasm (arrow). No osseous involvement or other intracranial abnormality is present.

a

b

Fig. 3. Sagittal (a) and coronal (b) T1-weighted post-contrast dedicated sellar images demonstrate heterogeneous hypoenhancement of the mass in comparison to compressed and displaced normal pituitary tissue (arrowhead). There is mass effect on the optic chiasm, and extension into the intercarotid space of the cavernous sinus on the left (arrow).

a

b

Fig. 4.

Coronal T2-weighted dedicated sella images demonstrating heterogeneous, primarily T2 hypointense signal in comparison to pituitary parenchyma, which is compressed and displaced to the right. (a) The tumor abuts the left cavernous and supraclinoid internal carotid arteries, as well as the bilateral A1 segments of the anterior cerebral arteries. (b) There is new subtle T2 hyperintensity in the left optic chiasm and nerve (arrow).

Fig. 5.
Axial bone window CT from the patient's whole body PET-CT at the level of the sellar/
suprasellar mass demonstrates no evidence of osseous metastatic disease.

a

b

c

Fig. 6.

(a) Hematoxylin and eosin-stained section demonstrating a small round blue cell tumor (40X). (b) Membranous cytoplasmic immunoreactivity of tumor cells for CD 99 (hematoxylin counterstain, 40X). (c) Nuclear immunoreactivity of tumor cells for FLI-1 (hematoxylin counterstain, 40X).