ELSEVIER

Clinical Imaging 37 (2013) 354–357

CLINICAL IMAGING

Case Reports

Suprasellar non-Langerhans cell histiocytosis (Erdheim–Chester disease)—a case report☆

Manas Sharma[a], Beth Vettiyil[b,*], Eric Bartlett[c], Eugene Yu[c]

[a]Head and Neck Imaging, Princess Margaret Hospital, Toronto M5G 2M9
[b]Head and Neck Imaging, University Health Network, Toronto M5G 2M9
[c]University of Toronto, Toronto M5G 2M9

Received 11 April 2012; accepted 11 June 2012

Abstract

Erdheim–Chester disease (ECD) is an uncommon non-Langerhans cell histiocytosis that affects multiple body systems and can present clinically in a myriad of ways. An adult onset is most common with bony involvement and constitutional symptoms. We report the case of a 52-year-old female presenting with diabetes insipidus and a suprasellar mass on imaging, with no evidence of extracerebral involvement. Histopathology was consistent with ECD.

Keywords: Suprasellar; Erdheim–Chester; Diabetes insipidus; Non-Langerhans cell histiocytosis

1. Clinical history and imaging findings

A 52-year-old female was referred for imaging of the sellar region with a history of central diabetes insipidus. Magnetic resonance imaging (MRI) of the brain and sella was performed that revealed an expansile and enhancing intraaxial lesion involving the medial and posterior aspects of the optic chiasm, anteroinferior hypothalamus infundibulum, and neurohypophysis (Fig. 1). A differential diagnosis included sarcoid; lymphoma; optic chiasm glioma; metastatic disease; and, less likely, histiocytosis. Additionally, focal signal alteration within the sphenoid sinus and marginal sphenoid bone was thought to be due to chronic sinusitis with inspissated mucus.

A 1-year follow-up MRI study showed no significant interval change of the suprasellar mass (Fig. 2). However, long-term follow-up MRI studies over a span of about 7 years showed mild gradual increase in size of the lesion (Fig. 3).

A biopsy performed in this period revealed the lesion to be non-Langerhans cell histiocytosis consistent with Erdheim–Chester disease (ECD). Interestingly, there was no clinical evidence of any extracranial involvement by the disease process in this patient.

2. Discussion and review of literature

ECD is a rare condition characterized by xanthomatous infiltration of tissues with foamy histiocytes. It is categorized as a form of non-Langerhans cell histiocytosis [1].

Histiocytosis refers to neoplastic or nonneoplastic proliferation of tissue phagocytes that are involved in processing and presenting antigens to the lymphocytes [2]. Syndromes are divided based on the predominant cell type within the infiltrate [3]. ECD is nonfamilial and typically of adult onset, with a median age at diagnosis of 53 years, and can affect multiple body systems with various clinical

☆ Address of institution where work was performed: Head and Neck Imaging, Joint Department of Medical Imaging, University Health Network, Toronto M5G 2C4.

* Corresponding author. Medical Imaging Research, 700 Bay Street, Toronto M5G 2C4. Tel.: +1 647 459 0233.

E-mail addresses: drmanassharma@gmail.com (M. Sharma), vettiyil@gmail.com (B. Vettiyil), Eric.bartlett@uhn.on.ca (E. Bartlett), Eugene.Yu@uhn.ca (E. Yu).

0899-7071/$ – see front matter
http://dx.doi.org/10.1016/j.clinimag.2012.06.006

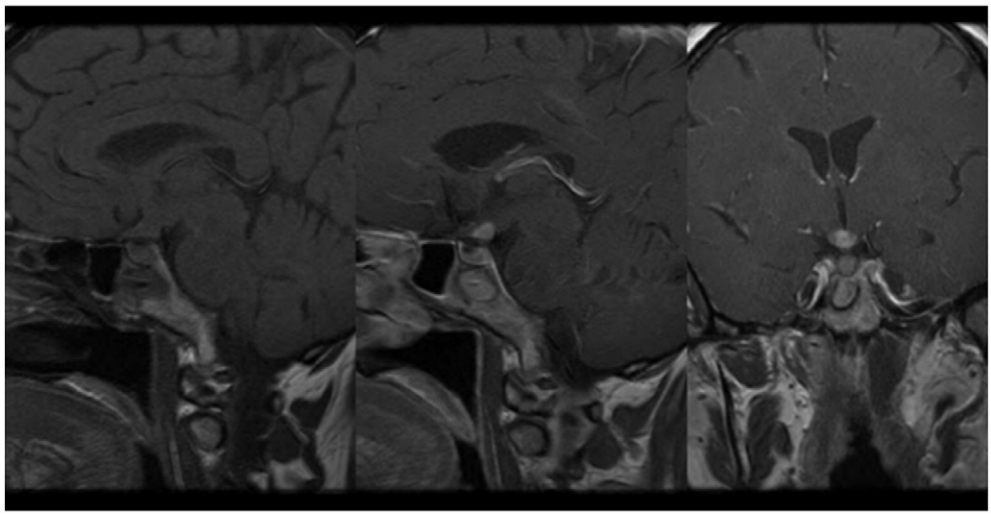

Fig. 1. Sagittal MRI pre- and postgadolinium T1-weighted imaging and coronal postgadolinium image shows an enhancing lesion in the suprasellar region involving the posterior aspect of the optic chiasm, hypothalamus, and infundibulum. Note signal change within the sphenoid sinus and its bony margins.

presentations [4]. A male preponderance was reported in a study of 59 cases [4].

Often asymptomatic, ECD can commonly manifest with bony involvement and may present with prolonged bone pain and osteosclerosis on imaging. Constitutional symptoms including fever, weight loss, and weakness may be seen. Xanthelasma, diffuse interstitial lung fibrosis, xanthogranulomatous changes in the retroperitoneum and kidneys with periaortic fibrosis, and cardiac and neurological involvements have been reported [1]. One third of ECD cases show neurological manifestations [5] such as diabetes insipidus and cerebellar syndromes [6]. Approximately 30 % cases have intra- or extraconal orbital involvement, often leading to clinical exophthalmos [7]. Intracranial lesions may be intraaxial, meningeal, perivascular, and infundibular but are rarely isolated [7].

The diagnosis of ECD is often a challenge for the treating physician because of its complex pattern of presentations. It is often confused with Langerhans cell histiocytosis because of common presenting features such as exophthalmos, diabetes insipidus, and osteolytic lesions on bone survey. However, ECD usually presents at a later age and has a worse prognosis than Langerhans cell histiocytosis [4].

ECD usually has multiorgan involvement, with the bones being the most common site of involvement and with patients often presenting with chronic bone pain. A skeletal survey will typically show osteosclerosis of the long bones. Other clinical features include diabetes insipidus and exophthalmos which is often bilateral, symmetric, and painless. Constitutional symptoms including fever, weight loss, and weakness may be seen. The neurological system, heart, lungs, kidneys, and skin may all be involved. Patients may develop diffuse interstitial fibrosis of the lungs, xanthogranulomatous involvement of the kidneys and retroperitoneum, and skin lesions such as xanthelasma [4].

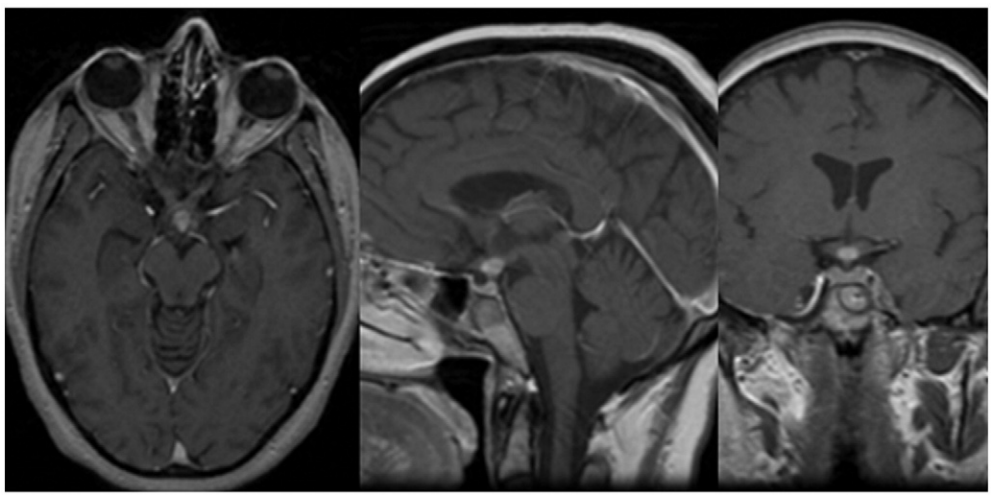

Fig. 2. One-year follow-up study did not reveal significant change in the size or extent of suprasellar mass.