CASE REPORT

Erdheim-Chester disease: atypical presentation of a rare disease

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► Additional material is published online only. To view please visit the journal online (<http://dx.doi.org/10.1136/bcr-2017-220827>).

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Accepted 26 September 2017

SUMMARY

We report the clinical case of an adult patient referred to our hospital because of trismus due to a tumour in the right infratemporal and pterygomaxillary fossa. He referred hyporexia, weight loss and right trigeminal neuralgia. On physical examination, he had trismus and diplopia. On neuroimaging, the tumour invaded the central nervous system affecting the right temporal lobe and orbit, and the sellar region. Tumour biopsy revealed foamy histiocytes and isolated giant multinuclear cells immunoreactive to CD68 and negative to CD1a and S100. A diagnosis of Erdheim-Chester disease was made. Non-evidence of large bone involvement was found in neither plain radiographs nor Technetium 99 m bone scintigraphy. BRAFV600E mutation analysis was negative. Because of raised intracranial pressure, a debulking surgery of the intracranial histiocytic process was performed. The patient improved his symptoms and remains clinically stable after 12 months of treatment with pegylated interferon- α -2a 180 μ g/weekly.

BACKGROUND

This is an important case because of the atypical clinical presentation with trismus, the absence of large bone involvement and the need of a debulking surgery.

CASE PRESENTATION

A 42-year-old male patient began having pain in the right mandible approximately 3 years before first consultation. Subsequently, he began to notice a progressive difficulty every time he opened his mouth. On February 2014, he was attended at a public Hospital in Buenos Aires. A transoral endoscopic biopsy of a tumefactive lesion located at the pterygomaxillary fossa was performed but the sample obtained was not suitable for anatomic-pathological analysis. He was referred to our hospital on December 2014. He complained of having altered feeding due to marked reduction in mouth opening, hyporexia, weight loss, right hemicranial headache, right trigeminal neuralgia (V1,V2) and diplopia. On physical examination, he had trismus (see online video), right abducens nerve palsy and slight gynecomastia. First imaging studies revealed an infiltrative process of the infratemporal and pterygomaxillary fossa. This lesion, in a matter of weeks to months, rapidly infiltrated the intracranial compartment involving the right temporal lobe and orbit, and the sellar region (see the section investigations). A new biopsy of the

tumour was performed via endoscopic transmaxillary approach. The anatomic-pathological findings were those seen in Erdheim-Chester disease (ECD) (see the section investigations).

INVESTIGATIONS

- Complete blood routine revealed normocytic normochromic anaemia, erythrocyte sedimentation rate (ESR) 66 mm/hour, PCR 43,09 (0–5).
- HIV serology and venereal disease research laboratory (VDRL) negative. Purified protein derivative (PPD) 0 mm.
- Brain MRI on admission revealed an infiltrative lesion of the right mandible, right medial and lateral pterygoid and masseter muscles. The infiltrative lesion progressed into the skull base towards the right sphenoid bone greater wing, right cavernous sinus, right temporal fossa and right orbital fissure with slight temporal lobe oedema. Three months later, another brain and also an orbit MRI was performed which demonstrated that the infiltrative lesion progressed even more into the brain parenchyma with marked right temporal lobe oedema. Involvement of the epiconal and extraconal fat of the right orbit with slight exophthalmos was observed. There was infiltration of the sphenoid ridge, orbital apex, cavernous sinus and orbital fissure. There was pathological gadolinium enhancement of the right pterygoid and masseter muscles, right cavernous sinus and meningeal enhancement at the right temporal and frontal lobe (figure 1).
- Intracranial magnetic resonance angiography was normal.
- Facial paranasal sinuses CT showed the tumour in right pterygomaxillary fossa with sclerotic changes and increased volume of sphenoid, temporal, maxillary and mandible bones.
- C3-C4, antinuclear antibody (ANA), cytoplasmic anti-neutrophil cytoplasmic antibody (ANCAc) and perinuclear anti-neutrophil cytoplasmic antibody (ANCAp) negative.
- Endemic mycoses analysis (aspergillosis, coccidioidomycosis, paracoccidioidomycosis, histoplasmosis) were all negative.
- Cerebro spinal fluid (CSF) cell count, glucose and protein concentrations were normal. PCR for tuberculosis (TBC) is CSF negative. CSF cultures for bacteria, fungus and mycobacteria were negative. No neoplastic cells were present in CSF; flow cytometry of the CSF was normal.
- ACE inhibitor in blood 43.5 U/L (8–52).

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To cite: Calandra CR, Bustos A, Falcon F, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220827

Figure 1 Brain MRI on December 2014. (A) Coronal T1-weighted image with asymmetry of the subarachnoid space at the right temporal pole with marked patchy and leptomeningeal gadolinium enhancement. (B) Axial T2-weighted image with slight hyperintense signal of the right temporal lobe suggestive of brain oedema. Brain MRI on March 2015. (C) Coronal T2-weighted image with marked increase of the hyperintense lesion at the right temporal lobe. (D) Coronal T1-weighted image with marked gadolinium enhancement of the infiltrative lesion at the infratemporal fossa and meninges.

- ▶ 24 hours urine calcium was normal.
- ▶ Prolactin 20.5 (4–15.2). Thyroid stimulating hormone (TSH), follicle stimulating hormone (FSH), luteinizing hormone (LH), testosterone, sex hormone binding globulin (SHBG) and human growth hormone (hGH) were normal.
- ▶ Biopsy of right pterygomaxillary fossa tumour showed foamy histiocytes and isolated giant multinuclear cells which were immunoreactive to CD68, and negative to CD1 and S100 ([figure 2](#)).
- ▶ Cardiac MRI was normal.
- ▶ Thorax, abdominal and pelvis CT with normal findings.
- ▶ Plain radiographs of large bones were normal.
- ▶ Technetium 99m (Tc99m) bone scintigraphy without large bone involvement ([figure 3](#)).
- ▶ Whole body 2-deoxy-2-[F-18]fluoro-D-glucose-positron emission tomography (FDG-PET) showed a hypermetabolic lesion only in the infratemporal fossa (performed after debulking surgery: see below) without large bone involvement ([figure 4](#)).
- ▶ BRAFV600E mutation analysis in sample tissue was negative (Therascreen BRAF RGQ PCR kit for detection of somatic mutations in the BRAF gene using real-time PCR on the Rotor-Gene Q MDx 5plex HRM instrument. Analysis of the following mutations: V600E, V600D, V600K, V600R).

DIFFERENTIAL DIAGNOSIS

Sarcoidosis, extranodal lymphoma, granulomatosis with polyangiitis, other histiocytosis like Rosai-Dorfman disease (RDD).

TREATMENT

After the second biopsy and before having the final anatomic-pathological report, three pulses of intravenous methylprednisolone were administered empirically with reduction of headache severity and oedema of the right temporal lobe previously seen on brain MRI. Treatment was followed by oral meprednisone 60 mg daily and then slowly tapered. Headache and temporal lobe lesion on MRI worsened when oral steroids were tapered below the 40 mg daily. The patient began treatment with pegylated interferon- α -2a (PEG-IFN- α -2a) 180 μ g weekly subcutaneously when the histological diagnosis of ECD was confirmed. He was asymptomatic while on treatment with seven weekly doses of PEG-IFN- α -2a plus meprednisone 40 mg daily. Later on, he complained of severe headache, nausea and vomiting. Signs of raised intracranial pressure were observed in neuroimaging ([figure 5](#)). A debulking surgery was performed with partial resection of the temporal lesion and the dura. A marked relief of the clinical manifestations was obtained right after the surgical procedure.

OUTCOME AND FOLLOW-UP

Oral steroids were slowly tapered. The patient continued monotherapy with PEG-IFN- α 180 μ g once per week. After 12 months of treatment, he has no more headaches, diplopia nor trigeminal neuralgia. Trismus slightly improved and thus feeding and nutritional status. Radiotherapy was in consideration as an adjuvant therapy after debulking surgery but finally it was not administered due to the immediate and sustained relief of the

Figure 2 Histological photomicrographs. (A) Foamy histiocytes. Immunohistochemical staining. (B) Foamy cells immunoreactive for CD68. (C) Cells are negative for CD1a. (D) Cells are negative for S100.

Figure 3 Technetium 99m bone scintigraphy. (B) Abnormally increased tracer uptake of the right mandible, temporal and sphenoid bone. (A) Absence of large bone involvement.

Figure 4 Whole body FDG-PET. (A,B,C) Expansive hypermetabolic lesion in the right infratemporal fossa and extracranial soft tissue with slight to moderate metabolic activity. (D) Absence of systemic pathological FDG uptake.

clinical and radiological manifestations. A new brain MRI, 2 years and 4 months after the first one, showed marked improvement of parenchymal lesion (figure 6). A second Tc99m bone scintigraphy was performed, 13 months after the first one. Still, there is no evidence of large bone involvement and there is less concentration and extension of the tracer in the right maxillary and temporal bone than previously described (figure 7a-b).

DISCUSSION

ECD is a rare disease classified among the non-Langerhans histiocytosis (NLH). It was first described in 1930 by Jakob Erdheim and William Chester, it is slightly more frequent in men and the mean age at diagnosis is 55 ± 14 (range 16–80 years).^{1,2} Recent investigations support the clonal nature of the disease.^{3–6} A very recent and complete review of histiocytoses written by Haroche *et al* has been published addressing the subject of its neoplastic nature. More than half of the patients have mutations in BRAFV600E, and many patients with wild-type BRAF have mutations involving the mitogen-activated protein kinase (MAPK) and Phosphatidylinositol-3-Kinase and Protein Kinase B (PI3K-AKT) pathway. ECD and LH are now being considered as myeloid neoplasia with a marked inflammatory component. But still various aspects of the pathogenesis remain unknown, especially the cells of origin of these disorders.⁷

ECD is multisystemic with involvement of large bone, especially of the lower limbs in almost all cases (96% of the patients in one the largest case series).¹ Large bone involvement is best demonstrated with plain radiographs and/or Tc99m bone scintigraphy. Radiographs typically show bilateral and symmetric cortical osteosclerosis of the diaphyseal and metaphyseal regions in the long bones, mainly the femur, tibia and fibula. Tc99m bone scintigraphy typically shows symmetric and abnormally strong labelling of the distal ends of the long bones. On the other hand, Langerhans cell histiocytosis typically involves axial bones. The mandatory criteria for a definite diagnosis are the histological findings: infiltration by NLH with foamy or eosinophilic cytoplasm and CD68 (+), CD1a (–), S100 (negative/low) immunohistochemistry.⁸ Other sites commonly involved are cardiovascular (circumferential soft-tissue sheathing of the thoracic and abdominal aorta; pericardial infiltration; myocardium infiltration), pulmonary (parenchyma and pleura, commonly asymptomatic), retroperitoneum (infiltration of perinephric tissues—‘hairy kidney’—hydronephrosis and ureteral narrowing), skin (xanthelasma, mainly in the eyelids). CNS involvement may be localised in the intra and/or extra-axial compartment. Pachymeningeal involvement is frequent. Brain parenchyma and spinal cord may be infiltrated. The pituitary gland, stalk and hypothalamus are common sites of infiltration leading to various endocrine abnormalities, mainly diabetes insipidus. The orbits might be involved leading to ophthalmoparesis, visual loss and/or exophthalmos. Cardiac and CNS involvement are poor prognostic factors.⁹

The first drug of choice for medical treatment are INF- α -2a and PEG-INF- α -2a. Vemurafenib, a BRAF inhibitor, is recommended in those cases where the mutation analysis for BRAFV600E is positive and refractory to INF- α -2a.¹⁰ Moreover, vemurafenib is now being considered as first-line therapy in those cases with life-threatening lesions and positive mutation of BRAFV600E.^{7,11} In the same way, cometinib and other mitogen-activated extracellular signal-regulated kinase (MEK) inhibitors are now being considered as first-line therapy in wild-type BRAF patients with life-threatening lesions.⁷ Anticytokine drugs (anakinra, infliximab and tocilizumab) are a second-line

Figure 5 Brain MRI before debulking surgery. (A) Coronal T2-weighted image showing temporal lobe lesion with midline shift deviation to the left. (B) Axial T2-weighted image with midbrain compression. (C) Axial fluid attenuation inversion recovery (FLAIR)-weighted image showing lateral ventricles asymmetry. (D) Axial T1-weighted image with marked gadolinium enhancement of extra-axial infiltrative lesion.

therapy.¹² Surgical treatment is reserved only for those cases with compressing life-threatening infiltrative lesions that are refractory to medical treatment. We think that the delayed onset of treatment with PEG-INF contributed to the requirement of debulking surgery besides the aggressive natural history of ECD involving the CNS. Although surgery was performed while receiving treatment with PEG-INF, we could not ascertain that this case was refractory to medical therapy because of the brief period of treatment so far. Indeed, we decided to maintain PEG-INF- α -2a after surgery. Histiocytes are radiosensitive; thus, radiotherapy may be considered as a palliative treatment in selected cases. The best outcomes have been observed in patients with isolated bone involvement without systemic disease. However, symptom relief is usually temporary and later recurrence of clinical manifestations is common. Variable outcomes have been reported in patients with ECD and CNS and retro-orbital involvement. There is no consensus of doses and fractions of radiotherapy in ECD.^{13–18}

Isolated CNS involvement is highly infrequent. Haroche *et al*² reported a case series of 53 patients of which only 4% of

the patients did not show large bone involvement. There are three more case reports in the literature of ECD with CNS involvement without large bone involvement.^{19–21} Arakaki *et al* reported one case of ECD with CNS involvement. After being published, the authors confirmed the absence of long bone involvement (personal communication by Arakaki), but there is no data about long-term prognosis because the patient did not continue follow-up.²² Munoz *et al* described a series of 14 patients with ECD, with one of them having only skin lesions.¹² Drier *et al* reported a series of 33 patients with cerebral, facial and orbital involvement, all of them with concomitant large bone involvement.²³

Extranodal RDD may present with a clinical picture similar to our case. In RDD, among the most frequent affected extranodal sites are the orbits and CNS. Cranial, facial and tibia are the bones more commonly involved in RDD. However, bone lesions are typically lytic in RDD, meanwhile osteosclerosis of facial and skull bones is frequently observed in ECD (as was seen in our patient). In RDD, intracranial lesions are very similar to meningiomas, attached to the dura and rarely infiltrating the

Figure 6 Brain MRI after 2 years and 4 months from admission. (A) Axial T2-weighted image marked reduction of the oedema in the pole of the temporal lobe (B) Coronal T1-weighted image showing reduction of meningeal gadolinium enhancement (C) Axial T1-weighted image with persistence of gadolinium enhancement of extra-axial histiocytic mass.

Figure 7 Tc99m bone scintigraphy. (A) Reduction in the tracer's extension and concentration at the right maxillary and temporal bones. (B) Absence of pathological tracer uptake of the long bones.

brain parenchyma. In our patient, the lesion clearly infiltrated the brain parenchyma. In both entities, intra-axial lesions (more frequent in ECD) are hypo/isointense in T2-weighted image with hyperintensity due to associated oedema.^{24–26}

Our patient had hyperprolactinemia and gynecomastia, an endocrine abnormality more commonly found in ECD.

The immunohistochemistry of RDD lesions is strongly positive for S100. Positivity for S100 has been observed rarely in ECD samples. Emperipolesis (erythrocytes, lymphocytes and plasma cells engulfed by histiocytes) is typically observed in RDD. However, it is important to mention that this finding may be absent in some cases of RDD (eg, extranodal forms). Many cases of RDD with CNS involvement have CSF with pleocytosis (sometimes with concomitant emperipolesis in CSF). The immunohistochemistry of our patient was S100 (–), emperipolesis was absent and CSF composition was normal.²⁷

We could speculate that our patient could have RDD and ECD simultaneously. However, both tissue samples (pterygomaxillary fossa and temporal lobe) were CD68 (+), CD1a (–) and S100 (–) with none emperipolesis.

Our patient had a good response to the treatment with PEG-IFN- α -2a. There are only anecdotal reports of RDD cases successfully treated with IFN- α .²⁸

Trismus is a person's inability to normally open the mouth. It is commonly seen in stroke, local inflammatory processes, dental procedures, malignant head and neck tumours and due to local radiotherapy. These different aetiologies may lead to trismus if they affect the pterygoid/masseter muscles and/or their innervation, the temporo-mandibular joints and/or other supportive tissues. Trismus is generally overlooked even by the patient itself until it reaches an advanced stage. Trismus affects the oral phase of swallowing, mainly mastication and bolus organisation, leading to dysphagia and an increased risk of aspiration. It can also affect normal speech, oral hygiene and dental care. In cases of severe trismus, surgical intubation cannot be performed normally. Indeed, our patient, during the different surgical procedures, was intubated transnasally and also required tracheostomy. Different definitions of trismus exist. It is classified according to the maximal interincisal distance (MID) from the incisal edge of the maxillary and mandibular incisors. The revised Subjective,

Objective, Management and Analytic (SOMA) scale classifies trismus into four grades: grade 1, MID from 20 to 30 mm; grade 2, MID from 10 to 20 mm; grade 3, MID from 5 to 10 mm and grade 4, MID <5 mm. Grade 3 and 4 are severe trismus.^{29 30}

Our patient is of particular interest because of the scarce reports of ECD with isolated CNS manifestations without large bone involvement, and because of the presence of trismus as the first clinical manifestation. This phenomena was due to infiltration of the right pterygoid/masseter muscles an atypical localisation of histiocytic lesions in ECD.

Learning points

- ▶ Although rare, Erdheim-Chester disease (ECD) might present without large bone involvement.
- ▶ Histological findings are the final and mandatory step to diagnose ECD.
- ▶ Histiocytic infiltration of any tissue can be seen in ECD leading to a wide spectrum of clinical manifestations (ie, trismus).
- ▶ Trismus is a very rare and unusual first clinical manifestation in ECD.

Contributors CRC: substantial contributions to the conception of the work, the acquisition, analysis of data, drafting the work. AB: substantial contributions to the acquisition of data, drafting the work. FF: substantial contributions to the acquisition of data, drafting the work. NA: substantial contributions to the conception of the work, the acquisition, analysis of data, drafting the work. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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