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Case Report

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Erdheim-Chester Disease with Calvarial Involvement: A Rare Case of Histiocytosis

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ABSTRACT

Erdheim-Chester Disease is a rare systemic xanthogranulomatous infiltrating disease, characterized by lipid-laden histiocytes accumulating in various organs and almost always in bones. Etiology of the disease is still unknown. It may involve various organs and systems, such as musculoskeletal, cardiac, pulmonary, renal, gastrointestinal and central nervous system (CNS) as well as the skin. The most common systemic manifestations are bone lesions and the specific sign of these are bilateral sclerosis of the diaphysis and metaphysis of long bones. Symptoms and signs can vary related to the organ or system that is involved. In CNS involvement, cerebellar and pyramidal symptoms and signs are the most common, while headache, seizure, cranial nerve paralysis, neuropsychiatric along with cognitive complaints and mood disorders are also reported. Furthermore, there are asymptomatic cases. Histologically lipid-laden foamy histiocytes with small round nucleuses and without nuclear grooves are the characteristic histological features. These histiocytes show positive CD68 and negative S100 and CD1a immunoreaction. Surgery is a reasonable treatment in the patients who have extra- or intracranial lesions with smooth borders when the neurological signs and symptoms are mild. Medical treatment of the disease includes steroid, cytotoxic agents such as cladribin, IFN α-2a, recombinant human interleukin-1 receptor antagonist, tirosine kinase inhibitors, biphosphonate and autologue hematopoetic stem cell transplantation. In this report a 29 years old man was presented with a frontal calvarial lesion who was operated and diagnosed as Erdheim Chester disease.

KEYWORDS: Cranial, Histiocytosis, Erdheim chester disease, Lidip-laden histiocytes

INTRODUCTION

Histiocytosis is a large heterogeneous group of xanthogranulomatous diseases caused by the proliferation or accumulation of reactive or neoplastic histiocytes in tissue. Three categories of histiocytoses have been identified: 1) Langerhans cell histiocytosis (LCH) (including histiocytosis X and Hand–Schüller–Christian disease); 2) Non-Langerhans cell histiocytosis (macrophage-related disorders, including xanthogranuloma and nonmalignant Erdheim–Chester disease (ECD); 3) Malignant histiocytosis (including histiocytic sarcoma, Langerhans cell sarcoma, interdigitating dendritic sarcoma, and follicular dendritic cell sarcoma) (12,14,19).

Xanthogranulomatous diseases are clinically diverse and fall into three major groups. The first of these comprises conditions that primarily affect the skin. The second includes those that affect the skin and also show signs of systemic involvement. The third group consists of those diseases that do not show any symptoms of skin involvement (10,40). Most non-LCH conditions with central nervous system (CNS) involvement fall into the latter two groups. These include juvenile xanthogranuloma (JXG), adult-onset xanthogranuloma (AXG), xanthoma disseminatum (XD), Rosai–Dorfman disease (RDD), and ECD (43). Since these diseases rarely show up in the CNS, diagnosis can be difficult. This is further complicated by the fact that the origin of the pathological cells in all non-LCH is the same (10).

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ECD is a rare type of non-LCH that affects multiple organs. It was first identified and defined in 1930 by Jakop Erdheim and William Chester (8). This was supplemented by the first radiological definition of the disease by Melicow in 1953 (28). Finally, in 1972, Jaffe provided a comprehensive and detailed clinical, radiological, and pathological definition of ECD (20). This led to further studies of the condition. ECD most often manifests in the fifth decade and is more common in men, with a male: female ratio of 2.7: 1 (14).

ECD lesions are defined radiologically as intracerebral or infiltrating masses that manifest as diffuse nodules. In magnetic resonance imaging (MRI), the lesions are hyperintense in T2 sequences and there is little or no edema around them in FLAIR sequences (10). The most common sites for biopsy based on radiological data are sclerotic bone areas and retro-orbital masses. Cerebral biopsies are rare. The typical pathological features of this disease differ from those of LCH. Pathologically, non-LCH xanthogranulomatous lesions, including those in ECD, are composed of lipid-laden histiocytes expressing CD68 but lacking CD1a (a dendritic origin marker), S100 (a calcium-binding protein), and Langerin (a highly selective marker for Langerhans cells) (6,17,18,25,26). This is evidenced by positive CD68 and negative S100 and CD1a immunoreactions in the histiocytes (11,23). Foamy and fatty histiocytes with small round nuclei and no nuclear grooves are typical (41). After extensive diagnostic investigations, a study that repeated a polymerase chain reaction molecular test identified a genetic mutation (B-Raf V600) associated with ECD (28). Once diagnosed, the histological subtype can be identified using electron microscopy. The origin of the histiocytes in ECD has not yet been identified.

Neurological complaints are seen in fewer than 50% of ECD cases with multiple organ involvement and fewer than one-third present with neurological symptoms as the chief complaint (1,33,34). The most common areas of the CNS affected by ECD are the hypothalamo-hypophyseal tract, the cerebral parenchyma, the orbits, the meninges of the brain, and the paranasal sinuses. More rarely, the spinal meninges and vertebral bones may also be involved. Diabetes insipidus and cerebellar symptoms are the most common neurological findings (16).

In this article, a 29-year-old male with chief complaints of headache and nausea underwent surgery for a calvarial mass. Pathological analysis of the mass identified ECD. We describe this case and review the related literature.

CASE REPORT

The study was carried out according to the latest revision of the Helsinki Declaration regarding medical research involving human subjects. Informed written consent was acquired from the patient before the study. No IRC board approval was obtained due to the publication being a case report and anonymity of the patient.

Clinical Summary

A 29-year-old male presented to our outpatient clinic complaining of recurrent headaches over the preceding 6 months and nausea for the last 2 months. His neurological examination revealed a palpable frontal bulge in the midline of his forehead. His routine complete blood count and serum biochemical profiles were normal (hemoglobin, 14.7 g/dl; hematocrit, 45%; white blood cell count, 7.3x10^3/µl; platelet count, 283x10^3/µl; ALT level, 4.1 U/L; AST level, 18 U/L; urea, 37 mg/dl; creatinine, 0.8 mg/dl).

A cranial MRI scan showed a 3.42.3 × 1.5 cm extra-axial lesion that was isointense on T1 and T2 sequences, with homogeneous contrast enhancement in the frontal midline region (Figure 1).

The patient was operated on for a calvarial mass. Perioperatively, the lesion was found to be very soft and yellow and the periosteum appeared to be infiltrated. The mass was completely resected and sent for pathological analysis. A specimen was also taken for culture in case of an infection. The culture results were negative.

Histopathological Findings

The histopathological examination revealed clusters of lymphoid proliferation and histiocytic cells, with fibrotic bands between them. Multinuclear giant cells were detected in both compartments. Differential diagnoses included LCH, ECD, and RDD. Immunohistochemical staining was positive for CD68, negative for CD1a, and focally positive for S100 (Figure 2). These findings were discordant with an LCH diagnosis as there was not enough lymphophagocytosis and hemophagocytosis. RDD was also excluded.

Postoperative whole-body bone scintigraphy showed increased activity in the posterolateral part of the left second costa and right sacroiliac joint (Figure 3). The patient's clinical, pathological, and scintigraphic results all indicated a diagnosis of ECD.

Subsequent thoracic abdominal computed tomography (CT), and cardiac MRI showed no pathology. Postoperative cranial MRI found no residual mass (Figure 4).

Although corticosteroid treatment was started on the recommendation of the Hematology Department, it was gradually stopped because of excessive weight gain after 2 months. At his 3-year follow-up, the patient remained symptom free, and his neurological examination showed no pathological findings. No abnormalities were detected in thoracic, abdominal CT, or cranial MRI (Figure 5). Whole-body scintigraphy showed that the increased activity at the posterior of the left costa and right sacroiliac joint had also diminished. However, there was a significant increase in activity at the right sixth costovertebral joint.

DISCUSSION

Proliferative histiocytic diseases can be defined as tumor-like lesions resulting from the abnormal accumulation of differentiating monocyte/ macrophage-type mononuclear phagocytic system cells. Histiocytic lesions are divided into three major groups: LCH diseases, non-LCH diseases, and malignant histiocytic diseases (11,13,19). Xanthogranulomas are non-LCH diseases resulting from the proliferation of benign dendritic

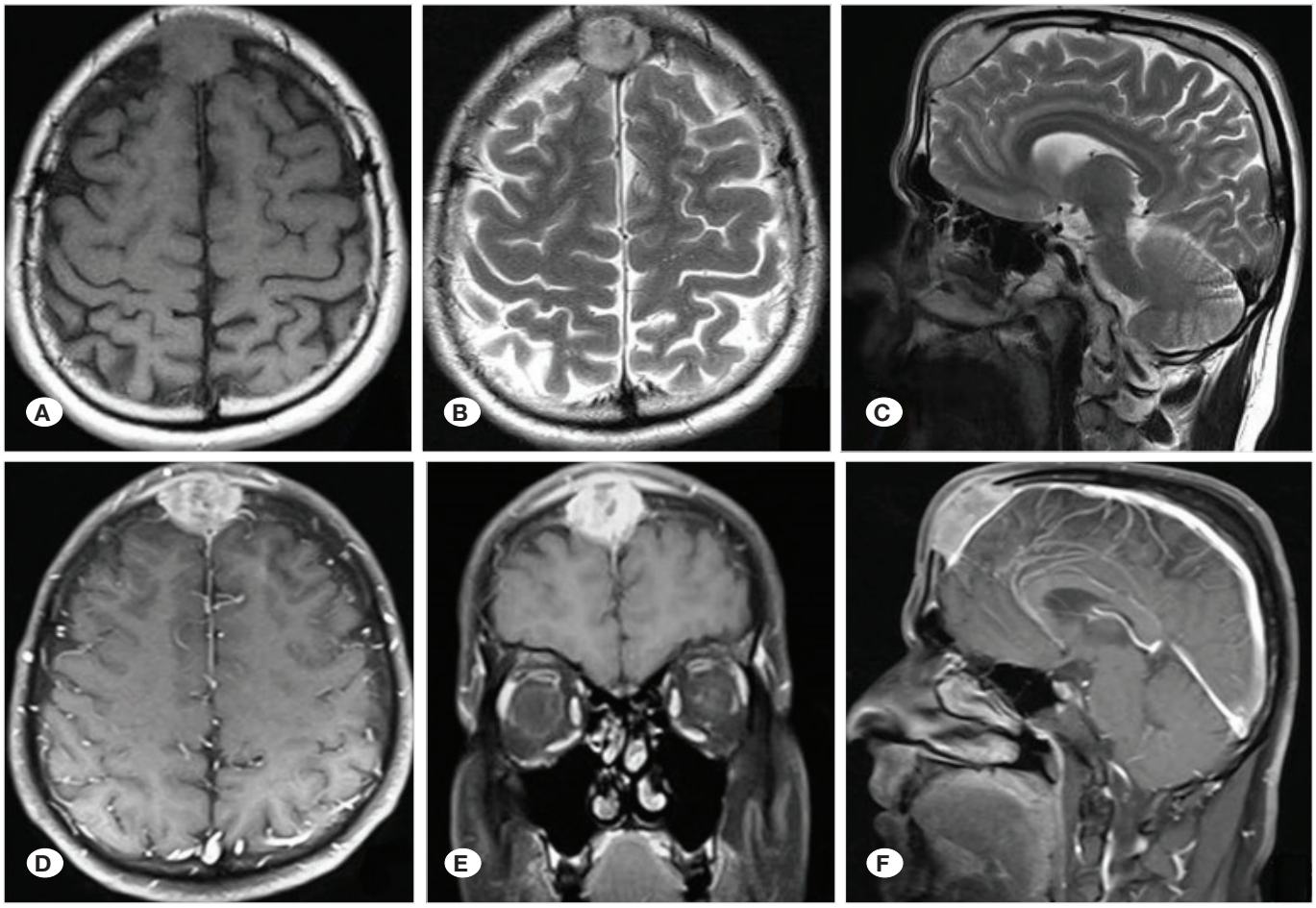

Figure 1: Preoperative MRI sequences. **A)** T1W axial, **B)** T2W axial, **C)** T2W sagittal, **D)** T1W axial+with contrast (Dural tail like appearence extending on both sides mimicking meningioma), **E)** T1W coronal+with contrast, **F)** T1W sagittal+with contrast.

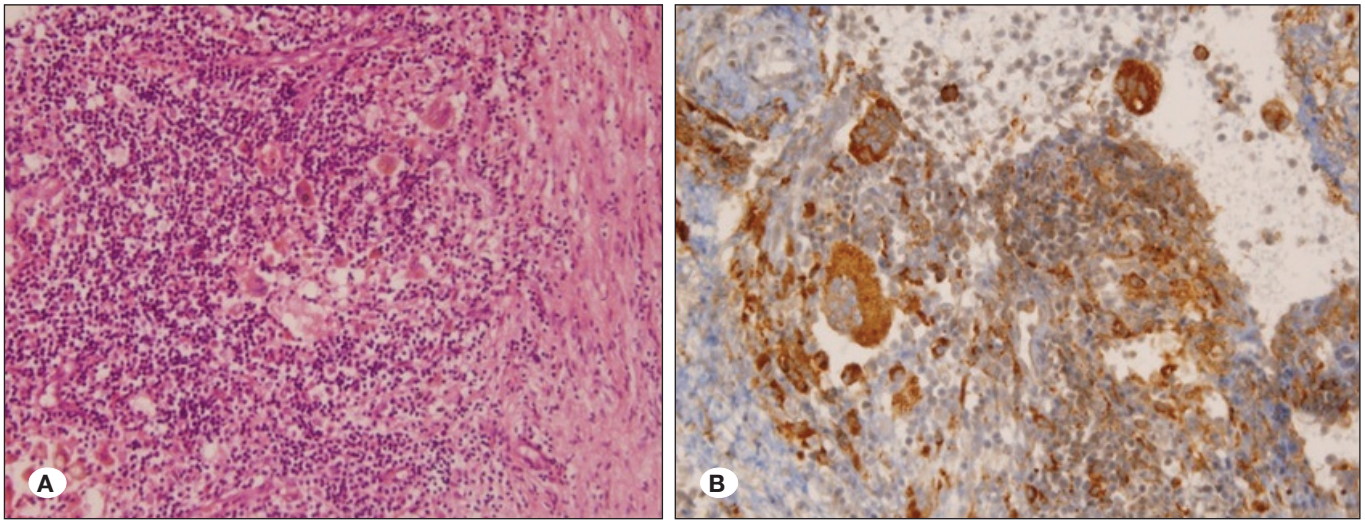

Figure 2: A) Lymphocytic infiltration and multinuclear giant cells (Hematoxylin and eosin, x100), **B)** CD68 immunoreactivity in immunohistochemical evaluation (CD68, x200).

cells and macrophages in the scalp, the skin of the neck, or the trunk. They usually affect young males.

Those xanthogranulomas with systemic effects most often involve the eyes, lungs, pericardium, oral cavity, testicles, and smooth tissue (38). CNS involvement in these diseases is very rare and, when present, usually involves the middle cerebellar peduncle, the brainstem, and the choroidal plexus (42). Cases have also been reported that involve Meckel's cave, the pineal region, the cerebellopontine angle, the sciatic nerve, and the orbit (9,22,31,32,36). Xanthogranulomatous diseases with calvarial involvement, as in the case reported above, are extremely rare. Sedrak et al. found calvarial involvement in only 2 out of 11 patients (34,38,39).

The most commonly reported symptom of these diseases is nonspecific bone pain. This is most often diaphysis or metaphysis of long bones such as the femur, tibia, fibula, and humerus (29, 35). Other manifestations that have been reported include bilateral symmetrical medullary sclerosis and lytic lesions (30%) due to bone involvement, pulmonary fibrosis due to lung involvement, cardiomyopathy, cardiac failure due to cardiac involvement, hypernephrosis, and renal failure due to retroperitoneal fibrosis (24,37). Arnaud et al. analyzed 53 patients who received IFN-α in several medical centers and found involvement of the musculoskeletal system in 96%, the cardiovascular system in 77%, the retroperitoneal system in 68%, the neurological system in 51% and the pulmonary system in 43% (4). The clinical course of the disease is related to the size of the mass and its distribution and extensiveness throughout the body. These masses can occur on a large scale and can range in severity from asymptomatic bone lesions to multisystemic, short-prognoses, life-threatening CNS or cardiovascular lesions (2,4). In our patient, the neurological findings were limited to a palpable swelling in the middle of his forehead (2). Postoperatively, we identified bone lesions at the posterolateral part of the left second costa and right sacroiliac joint with scintigraphy but none were symptomatic.

The patient in this case report presented with the chief complaints of headache and nausea. After clinical and radiological examinations, he underwent surgery for his calvarial mass. Postoperative whole-body bone scintigraphy and the pathology report contributed to a diagnosis of ECD, a rare form of non-LCH. Patients with this type of non-LCH are almost always males aged between 45 and 55 (33).

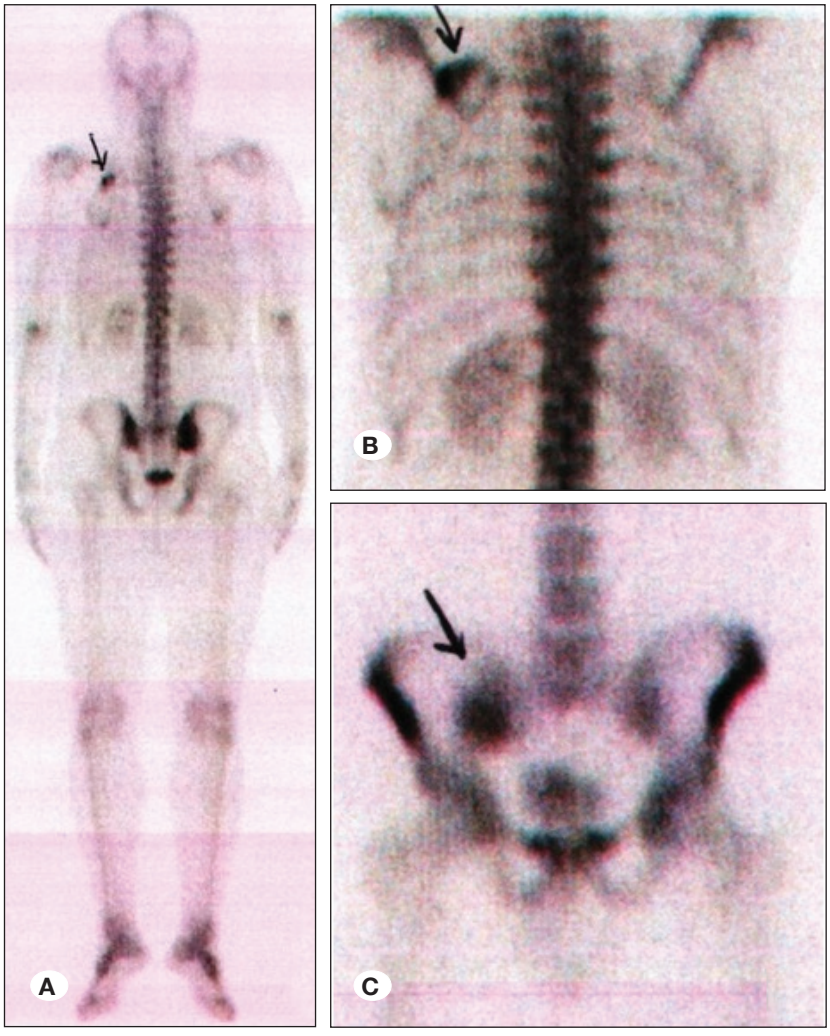

Figure 3: Postoperative whole body scintigraphy. Increased activity at the posterolateral part of the left 2nd costa **(A, B)**, and right sacroiliac joint **(C)**.

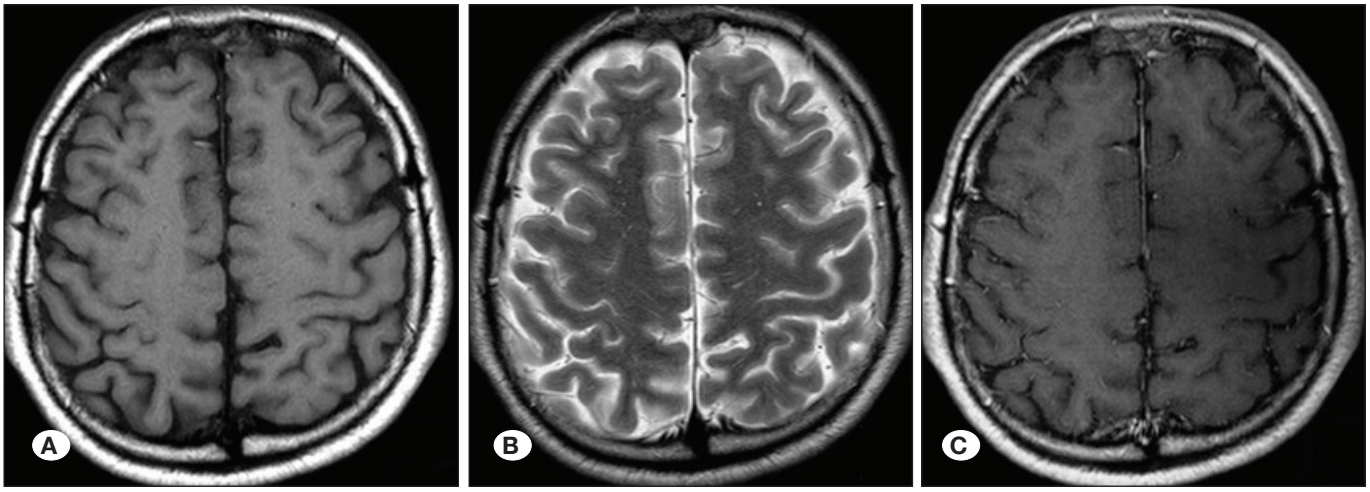

Figure 4: Postoperative Cranial MRI. **A)** T1W axial, **B)** T2W axial, **C)** T1W axial+with contrast.

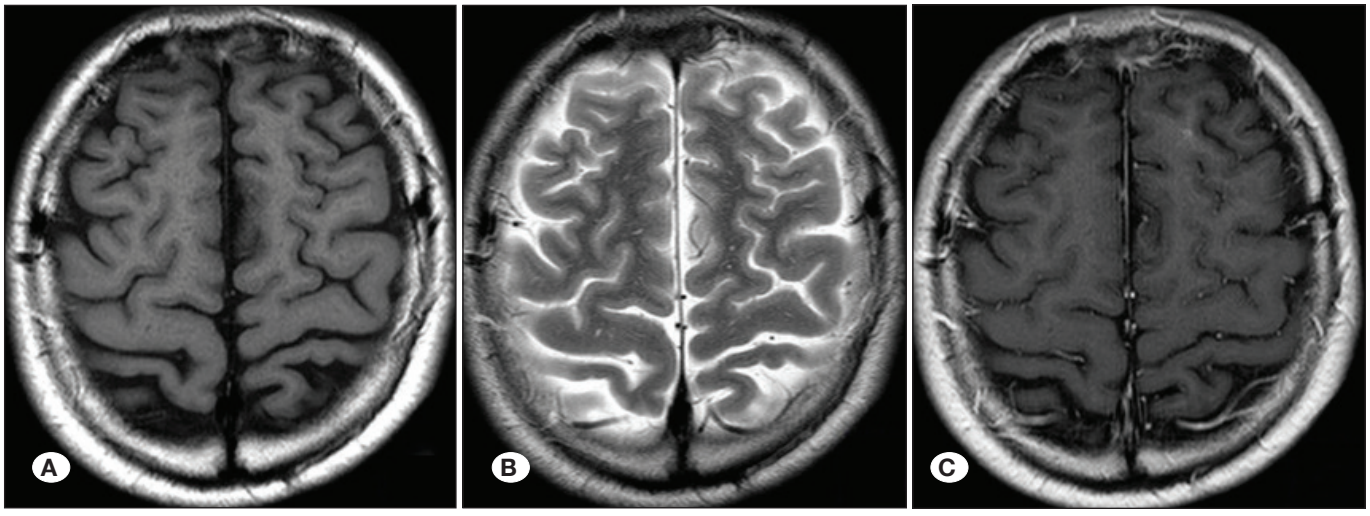

Figure 5: 3rd year control cranial MRI. **A)** T1W axial, **B)** T2W axial, **C)** T1W axial+with contrast. No recurrent tumor was detected.

Lachenal et al. identified three types of neurological involvement in ECD: infiltrative, meningeal, and mixed (27). Mixed-type lesions are isointense or hypointense on T2-weighted MRI and hyperintense on T2 with homogenous holding contrast (7,21,24,37). Our patient's lesion showed homogenous enhanced contrast on T1 sequences. The MRI sequences also detected a dural tail-like appearance, which could mimic meningioma. Nonetheless, the lesion was seen to be infiltrating the bone tissue and extending to the subcutaneous tissue without significant invasion of the superior sagittal sinus, which is not characteristic of meningiomas. The characteristic MRI presentation of the disease is prolonged contrast enhancement, even several days after contrast injection. Kujat et al. presented a case with a lesion that held contrast for 14 days (24). DSA shows venous sinus thrombosis and abnormal venous anastomoses, although hypervascularity cannot be seen (2).

Medical treatment of ECD may include steroids, cytotoxic agents such as cladribine, IFN-α-2a, recombinant human interleukin-1 receptor antagonists, tyrosine kinase inhibitors, bisphosphonate, or autologous hematopoietic stem cell transplantation (3,5,15,30). The recent identification of a proinflammatory oncogene-induced senescence mechanism associated with a BRAFV600E mutation has opened up the possibility of targeted therapy with vemurafenib in some patients. Vemurafenib works by inhibiting the B-Raf/MEK step in the B-Raf/MEK/ERK pathway (17).

Radiotherapy has also been used in the treatment of ECD because the disease was previously considered similar to LCH. In a case series by Arnaud et al., 26% of the patients had died 26.5 months after the diagnosis, and 0.8% died because of CNS involvement. They found 1- and 5-year survival rates of 96% and 68%, respectively (4).

At his three-year follow-up, our patient was symptom free. The only positive finding at this juncture was decreased activity in the right sixth costovertebral joint on bone scintigraphy.

CONCLUSION

ECD rarely involves the calvarium. In patients with involvement of nontypical regions, it can be difficult to differentiate ECD from other systemic granulomatous diseases. Differential diagnoses include RDD, JXG, and XD. CD68, S100, and CD1a immunoreaction studies are required to classify non-LCH into the appropriate subgroup. In xanthogranulomatous diseases, such as ECD, biopsy is essential for diagnosis, along with a combination of clinical, laboratory, radiological, and histopathological data.

Declarations

Funding: No external funding was received for this study.

Availability of data and materials: The datasets used and/or analysed during the current study available from the corresponding author on reasonable request.

Disclosure: The authors declare no competing financial interests and no sources of funding and support, including any for equipment and medications.

AUTHORSHIP CONTRIBUTION

Study conception and design: OB

Data collection: OB

Analysis and interpretation of results: FD

Draft manuscript preparation: FD

Critical revision of the article: OB

Other (study supervision, fundings, materials, etc...): OB

All authors (OB, FD) reviewed the results and approved the final version of the manuscript.

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