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Case Report

Langerhans cell histiocytosis of the orbit and frontal sinus of the adult woman: A first case report in Poland

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ABSTRACT

Background: Langerhans cell histiocytosis (LCH) is a term describing a clonal proliferation of pathologic Langerhans cells (histiocytes), which may manifest as unisystem (unifocal or multifocal) or multisystem disease. LCH is a rare cause of the orbital tumor with the predilection to its lateral wall which is particularly common in children.

Case Description: We report an unusual case of a 33-year-old woman, 6 months after childbirth, who presented with the edema of the right orbit and upper eyelid with headaches. On physical examination, the patient had a right superior and lateral swelling of the eyelid and the orbit and right enophthalmos, without blurred vision. Magnetic resonance imaging showed well-defined, expansile, intensely homogeneously enhancing mass lesion in the right superolateral orbital rim with the destruction of the upper wall of the orbit, growing into the frontal sinus and frontal part of the cranium with the bold of the dura mater in this region. Radical excision of the tumor was achieved through a right fronto-temporo-orbito-zygomatic craniotomy. Histopathological examination had confirmed the diagnosis of the LCH. The patient was discharged home with a modified Rankin Scale score of 0.

Conclusion: The main purpose of this case report is that LCH should be considered as one of the possible causes of quickly appearing tumor of the orbit in adults.

Keywords: Histiocytosis X, Langerhans cell histiocytosis, Orbital tumor

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a term describing a clonal proliferation of pathologic Langerhans cells (histiocytes), which may manifest as unisystem (unifocal or multifocal) or multisystem disease. LCH is a rare cause of the orbital tumor with the predilection to its lateral wall which is particularly common in children.

CASE DESCRIPTION

We report an unusual case of 33-year-old woman, without chronic diseases, 6 months after childbirth, who was admitted to our clinic because of the edema of the right orbit and upper eyelid with headaches of 1-month duration. On physical examination, the patient had a right superior and lateral swelling of the eyelid and the orbit and right enophthalmos, without blurred vision.

On admission, magnetic resonance imaging (MRI) showed well-defined, expansile, intensely homogeneously enhancing mass lesion in the right superolateral orbital rim with destruction of the upper wall of the orbit, growing into the frontal part of the cranium with the bold of the dura mater in this region [Figure 1a-d].

Radical excision of the tumor was achieved through a right fronto-temporo-orbito-zygomatic craniotomy.

On histopathology, sheets of histiocytic cells with indented pale nuclei with nuclear grooves suggestive of LCH were seen. Positive immunohistochemical staining for S-100, CD1a, and CD68 confirmed the diagnosis. Ki67 was about 30%.

The whole-body scintigraphy showed the increased collection of the tag in the region of the right orbit, but the postoperative MRI showed no features of the recurrence. We do not decide to use the radio- or chemotherapy in this case. One year after treatment, there is still no recurrence, yet.

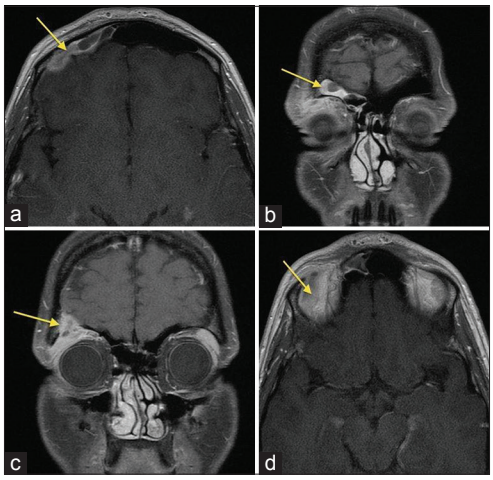

Figure 1: Tumor of the frontal sinus. (a) Infiltration of the tumor into frontal sinus, (b) Infiltration of the tumor into frontal sinus; coronal view, (c) Infiltration of the tumor into the right orbit; coronal view, (d) Infiltration of the tumor into the right orbit; axial view.

DISCUSSION

Langerhans cell histiocytosis (LCH), previously called histiocytosis X, is a term describing a clonal proliferation of pathologic Langerhans cells that may manifest as unisystem (unifocal or multifocal) or multisystem disease.[19,20,23] Although LCH may be diagnosed at any age, LCH usually occurs in young children with a peak age of 1–4 years.[6,10] In adults, the diagnosis of LCH is made at a mean age of 35 ± 14 years, with a peak age ranging from 20 to 30 years.[1] The pathogenesis of LCH is unclear. Although a clonal proliferation of Langerhans cells has been identified,[19,20,23] LCH is not probably a true neoplasm and may represent an atypical immunoreaction.[4] Accumulation of Langerhans cells in LCH results from survival rather than uncontrolled proliferation and is associated with the expansion of regulatory T cells.[16] There are also some evidence for an aberrant immune interaction between the clonal proliferation of dendritic cells and T-cells leading to a "cytokine storm."[10] Uncontrolled cytokine production induced by several factors including viral infections, minor trauma, may also contribute to the transformation of precursor cells into pathologic Langerhans cells.[22,25] However, as the cell cycle is obviously not blocked in LCH lesions and chromosomal instability was found in LCH as well, a neoplastic process cannot be completely excluded.

Histopathologically, the histiocytic infiltrate consists – despite its relatively benign-looking histologic features – predominantly of a clonal proliferation of pathologic Langerhans cells that resemble tissue macrophages rather than the typical dendritic shape of Langerhans cells in the skin.[13] The Langerhans cell infiltrate is accompanied by a varying amount of giant cells and eosinophils which are not mandatory for the diagnosis of LCH. Lymphocytes, plasma cells, polymorphonuclear leukocytes, macrophages (histiocytes), and varying amount of necrosis can be also present. The lesions are very often vascular, and erythrocytes are frequently observed due to hemorrhage. Mitotic figures are occasionally found. Larger studies have shown that there is no correlation between the histologic features and disease severity.[15] The combination of immunopositivity for the neuronal marker S100 and CD1a, which is specific for Langerhans cells (and thymocytes) and is not expressed by macrophages, helps to confirm the diagnosis. Other immunohistochemical markers that may be positive in LCH include adenosine triphosphatase, peanut lectin binding, alpha-mannosidase, CD207 (langerin), and fascin.[11,13,14]

LCH is classified into four groups based on a clinical staging system, i.e., Group A – bone only or bone and contiguous soft-tissue involvement, Group B – skin or other squamous mucous membranes only or with involvement of related superficial lymph nodes, Group C – soft tissue and viscera only, and Group D – multisystem disease.[3]

The ocular manifestation of the LCH usually occurs in children but may also affect adults.[24] LCH of the orbit usually presents as an isolated bone lesion with an associated soft-tissue mass, although it can also be associated with multifocal or multisystemic disease.[5,7] It occurs predominantly in the superior or superolateral orbital roof and exhibits other features depending on the location of the Langerhans cell infiltrate such as a visible mass with ptosis and/or (erythematous) swelling if located in the anterior orbit. This may be misinterpreted as an infectious process. Proptosis is predominantly seen in lesions that involve the posterior orbit. This may be accompanied by diabetes insipidus and skull lesions forming the triad formerly called Hand–Schüller–

Christian disease. Depending on the location and the size of the lesion as well as the infiltrated structures such as extraocular muscles, ocular movement may be impaired resulting in diplopia, and nerve palsies may occur. Visual acuity may be somewhat affected in young patients due to ptosis and the development of amblyopia. Fundus abnormalities, such as optic disc edema, dilated venous channels, and macular edema, have been reported in eyes with orbital LCH.[5]

Diagnostic imaging, including computed tomography and magnetic resonance tomography, shows well-defined bony lesions with a classic "punched-out" lytic appearance that are often accompanied by soft-tissue involvement in the orbit.[17] Lesions without bone erosions have also been reported.[5]

There is still no recurrence in our case, but it can appear in patients with localized orbital lesions within even 13 to 16 years after the initial treatment (most common within 12–18 months).[2,9]

As the pathogenesis of LCH is still unknown, treatment is empirical and depends on the disease severity and degree of systemic involvement. In general, the diagnosis of LCH should be proven by a biopsy. Incisional and excisional biopsies are preferred over fine-needle aspiration biopsy because the latter might not provide enough material for a sufficient histologic diagnosis.[8] Single bone lesions have a far better outcome than multiple bone lesions and multiorgan disease, so most physicians recommend biopsy and curettage for solitary orbital lesions. Careful curettage/excision in combination with intralesional steroids may be another effective treatment for primary unifocal lesions of the orbit and recurrent lesions.[21] One study group observed the absence of recurrence in twenty patients with unifocal, unisystem bone lesions of the orbit treated with excision and curettage alone, but recurrences occurred in three patients treated with excision and adjuvant chemotherapy (n = 2) and excision and intralesional triamcinolone (n = 1), respectively.[12] Although different doses of steroids have been used, 125 mg methylprednisolone is recommended as it might have an inhibitory effect on osteolysis.[8] The favorable outcome of excision and curettage alone may be attributed to changes in the microenvironment leading to the disruption of the pathological cascade.[8] Because of the possibility of the recurrence, close follow-up is recommended.[18] Radiation is predominantly used to treat recurrences.[8] Chemotherapy is regarded as a treatment for multifocal/multisystem disease or recurrences.[8] The most common chemotherapeutic agents are vinblastine, prednisone, etoposide, and methotrexate in various combinations.

CONCLUSION

The main purpose of this case report is that LCH should be considered as one of the possible causes of quickly appearing tumor of the orbit in adults. Moreover, an early diagnosis and multidisciplinary approach are required for proper staging of the disease to plan the best management of each case as treatment varies from case to case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Conflicts of interest

There are no conflicts of interest.

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