XANTHOGRAVLNULOMATOUS DISEASE OF THE ORBIT: CASE REPORT

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SUMMARY - Xanthogranulomatosis is a rare idiopathic histiocytic disorder, histologically characterized by xanthogranulomatous inflammation. It mostly affects bones, heart, lung, liver, kidneys, retroperitoneum, brain, and rarely the orbit. In the literature, it is mentioned as Erdheim-Chester disease. Ophthalmic involvement has been reported in less than 30 cases. We describe a 46-year-old man with unilateral exophthalmos and xanthogranulomatous inflammation of periorbital area, temporal area and face bilaterally, associated with asthma and atopic dermatitis. Dexamethasone was administered retrobulbarly. This therapy resulted in a significant reduction of exophthalmos and edema of the surrounding tissue, which improved the patient’s quality of life. There was no recurrence of inflammation during the one-year follow up. It is concluded that treatment with steroids provides good results in the management of xanthogranulomatous inflammation of the orbit.

Key words: Xanthogranulomatosis; Orbitopathy; Steroids

Introduction

Xanthogranulomatosis or Erdheim-Chester disease is a histiocytic disorder of unknown etiology, characterized by tissue infiltration by lipid-laden histiocytes and pathognomonic Touton giant cells. It was first described by Jacob Erdheim and William Chester in 1930. It is a systemic disease and mostly affects bones, heart, lung, liver, kidneys, retroperitoneum, brain, and rarely the orbit. Orbital xanthogranulomatous lesions are not common and manifest as diffuse or nodular lesions on the eyelids and in the orbit. It may be associated with hyperlipidemia, diabetes mellitus, blood dyscrasias, lymphoproliferative malignancies, atop dermatitis and asthma. Patient observation includes clinical examination, ultrasonography, MSCT or MRI, and definitely biopsy. Histopathology is characteristic and includes proliferation of histiocytes, plasma cells and lymphocytes with Touton giant cells that stain positive for lipids.

Case Report

We report an extremely rare presentation of orbital xanthogranuloma associated with asthma and atopic dermatitis. A 46-year-old man with mild mental retardation had enormous right eye exophthalmos, with lower eyelid retraction and bilateral upper eyelid edema (Fig. 1). He also had periorbital xanthomatous lesions and infiltration of temporal and zygomatic area. On the upper lid there were scars from previous blepharoplasty. Eye movements of the right eye were limited in all directions. He denied double vision. Hertel exophthalmometry was 23-112-5 mm. Bilateral visual acuity with Snellen charts was 0.8, and intraocular pressure was 14 mm Hg bilaterally. The slit lamp examination showed bilateral mild injection of conjunctiva, fluorescein examination was positive on the right eye and indicating filamentous secretion on the left eye. Fundus examination of the right eye was normal, but revealed pale temporal papilla border of the left eye. Laboratory findings showed increased values of erythrocyte sedimentation rate, C-reactive protein, eosinophils, basophils, proteins and immunoglobulin G. Glucose, creatinine and gamma-glutamyltransferease were decreased. Cryoglobulins
as well as ENA and ANCA were negative; lipidogram, CD4 and CD8 were normal. Ultrasonography examination of the right eye showed great enlargement of all extraocular muscles with intercellular edema. MSCT of the brain and orbits confirmed ultrasonography finding showing extraocular muscle enlargement up to 12 mm in diameter. It also revealed slight enlargement of the right optic nerve, reduction of intraorbital fat tissue and wider upper ophthalmic vein. Cortical brain atrophy was present (Fig. 2). Biopsy obtained on previous blepharoplasty showed typical proliferation of histiocytes and lymphocytes, with Touton giant cells (Fig. 3). MSCT of the abdomen and pelvis, and radiography of the lungs, heart and long bones were normal.

The patient was treated with dexamethasone administered retrobulbarly on the right side for 9 days. This treatment resulted in a significant reduction of exophthalmos and periorbital edema, improvement of bulbo-motorics and decrease of conjunctiva injection (Fig. 4). Hertel exophthalmometry was 16-106-7. Upon consultation with an immunologist, the patient was treated with prednisolone 30 mg for one week, then 20 mg to the present. No signs of reactivation of inflammation were noticed during the one-year follow up.

Discussion and Conclusion

Erdheim-Chester disease is a rare non-Langerhans histiocytosis of unknown etiology, characterized by xanthogranuloma present in the tissue. There is proliferation of histiocytes, plasma cells, lymphocytes and Touton giant cells. Touton giant cells are multinucleate cells with the nuclei arranged in a wreath around a ntidus of eosinophilic cytoplasm. This disorder is systemic and
can affect any tissue. Orbital involvement is quite rare.

Our case is one of the few described in the literature, manifested only with orbital and periorbital xanthogranulomatous lesions combined with atopic dermatitis and asthma without any other abnormality. There are many reports on xanthogranulomatosis associated with paraproteinemias, IgG monoclonal gammopathy, plasmacytosis, cryoglobulinemia, complement deficiency and leukopenia. Also, development of multiple myeloma and non-Hodgkin’s lymphoma may occur. Treatment is different from case to case because therapy is modified according to different sequelae of the disease. It is mostly treated with steroids, local or systemic, and by local incision of lesions until decompression of the orbit is achieved. Sometimes, intralungal injection of triamcinolone acetone is an effective and safe treatment for orbital xanthogranuloma in adults. In some reports, patients were treated with retrobulbar radiotherapy (20 Gy) and methotrexate chemotherapy, or therapy with vinblastin and doxorubicin combined with steroids or successful therapy with alfa interferon.

We treated our patient with corticosteroid locally that resulted in significant and fast improvement without any effect on the patient’s general condition. The patient was continuously treated with prednisolone 20 mg/day per os, and there were no signs of recurrence during the one-year follow up. This treatment led to a significant reduction of symptoms, which improved the patient’s quality of life. However, having in mind that xanthogranulomatosis is a systemic disease, long-term follow up is mandatory.

References

Sazetak
KSANTOGRANULOMATOZA ORBITE: PRIKAZ SLUČAJA
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