Case Report

Giant juvenile xanthogranuloma: diffuse and simultaneous palpebral involvement

Xantogranuloma Juvenil Gigante: acometimento palpebral difuso e simultâneo

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■ **ABSTRACT**

Juvenile xanthogranuloma (JXG) is the most common benign tumor of the group of non-Langerhans histiocytic proliferative diseases. Lesions; 2 cm are considered giant JXG, with reports of lesions of up to 18 cm. Oculopalpebral lesions may require surgical treatment to control symptoms. This study reports a case of an 8-year-old boy who had four eyelids and the middle third of the face affected by giant JXG. He underwent three resections, one of which was of great depth that required a full-thickness skin graft directly on the levator palpebrae superioris aponeurosis. Subsequently, three fat-grafting procedures were performed and adequate functional and aesthetic results were achieved without lesion recurrence.

Keywords: Plastic surgery; Eye; Juvenile xanthogranuloma; Myocutaneous flap; Skin transplantation..

■ **RESUMO**

O xantogranuloma juvenil (XGJ) é um tumor benigno e o mais comum do grupo das doenças histiocitárias proliferativas nãoLangerhans. Lesões; 2cm são consideradas XGJ gigantes, com relatos de lesões de até 18cm. Lesões oculopalpebrais podem necessitar de tratamento cirúrgico para controle de sintomas. Esse trabalho relata o caso de um menino de 8 anos que teve as 4 pálpebras acometidas por XGJ gigantes, além do terço médio. Ele foi submetido a 3 ressecções, sendo uma bastante profunda, necessitando enxerto de pele de espessura total diretamente sobre o músculo levantador da pálpebra superior. Posteriormente, 3 procedimentos de lipoenxertia foram realizados, atingindo resultado funcional e estético adequado, sem recorrência lesional.

Descritores: Cirurgia plástica; Olho; Xantogranuloma juvenil; Retalho miocutâneo; Transplante de pele.

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Article received: January 11, 2019.
Article accepted: April 18, 2019.

Conflicts of interest: none.

DOI: 10.5935/2177-1235.2019RBCP0238

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INTRODUCTION

Juvenile xanthogranuloma (JXG) is a benign tumor of unknown etiology formed of histiocytes and included in the group of non-Langerhans histiocytic proliferative diseases[1,2].

It is a rare condition, and although its incidence varies in the literature, it is usually underestimated because it spontaneously regresses in some cases with many lesions not being biopsied for diagnostic definition[3,4,5].

Literature classifies lesions; 2 cm as giant JXG[4]; these occur more frequently in children aged 3–5 years, presenting an almost equal distribution between the sexes. The single skin lesions are the most common, occurring predominantly in the head and neck, followed by the trunk and limbs[6,7].

JXG lesions tend to regress spontaneously within 6 months to 3 years after their first appearance[8]. However, in cases of giant lesions, whose involution is less common, surgical treatment may be indicated when the lesions cause significant functional and/or aesthetic impairment[9,10].

CASE REPORT

An 8-year-old male patient was referred to the service after the excision of a 4 cm × 6 cm occipital skin lesion in which anatomopathological examination revealed JXG. On inspection, yellowish papular lesions were simultaneously affecting the four eyelids in addition to bilateral lesions in the middle third of the face. According to his father, the lesions appeared after 6 months of life and had increased progressively since (Figure 1).

Due to diffuse periorbital involvement, without signs of lesion regression, surgical treatment was chosen with the proposal of lesion excision followed by primary closure or skin grafting, doing also local miocutnaeous flaps when the defects did not allow primary closure. The medical team opted to perform the resections in different surgical stages for each side.

Left upper eyelid

During dissection, this lesion, which apparently was superficial and sized 30 mm × 7 mm, was actually extending to the frontal region, reaching deep planes and invading the orbit, compromising the entire upper orbital cavity except for the aponeurosis of the levator palpebrae superioris muscle.

After complete lesion resection, the frontal region was detached and advanced to cover the upper orbital edge, and a full- thickness skin graft (retroauricular) was placed directly on the aponeurosis of the levator palpebrae superioris muscle, with 100% integration after 5 days (Figure 2).

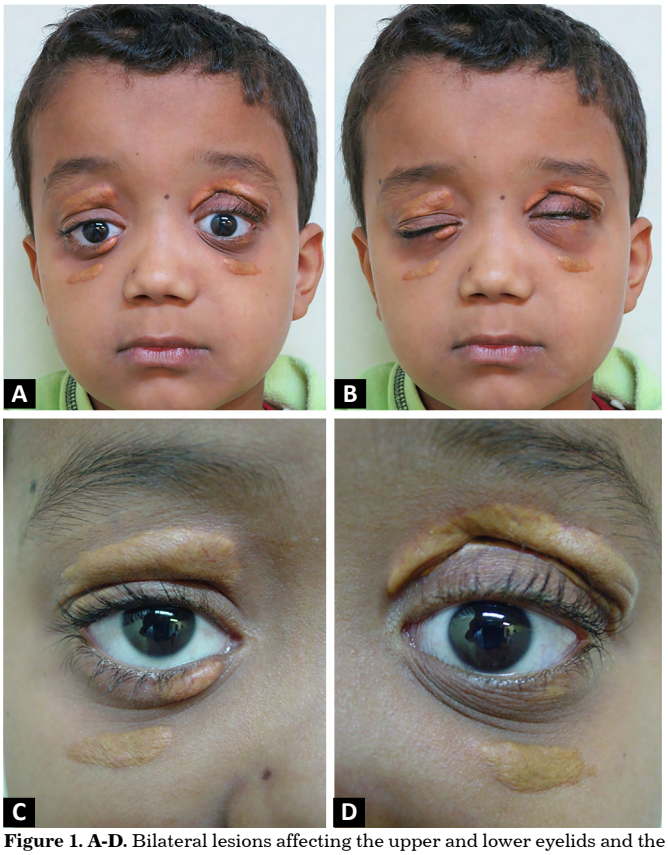

Figure 1. A-D. Bilateral lesions affecting the upper and lower eyelids and the middle third of the face.

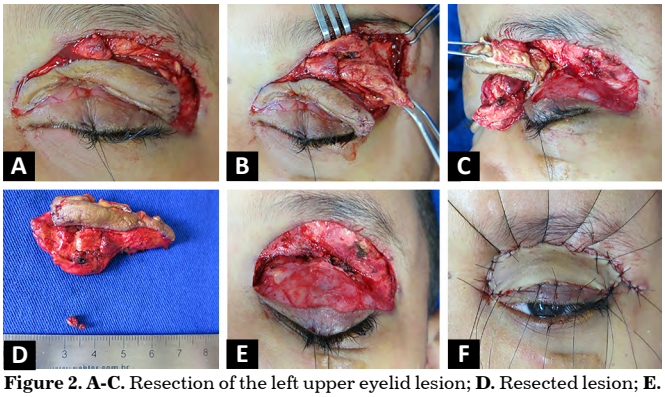

Figure 2. A-C. Resection of the left upper eyelid lesion; **D.** Resected lesion; **E.** Defect created after resection; F. Full-thickness graft.

Left lower eyelid/left middle third

After 3 months, the lesion in the lower eyelid allowed resection and primary closure. On the other hand, the lesion in the left middle third of the face measured 17 mm × 7 mm and extended to deep planes, reaching the periosteum of the maxilla and affecting the infraorbital nerve, which required en bloc resection with the lesion. After detachment of the malar tissue, the middle third was advanced for primary closure (Figure 3).

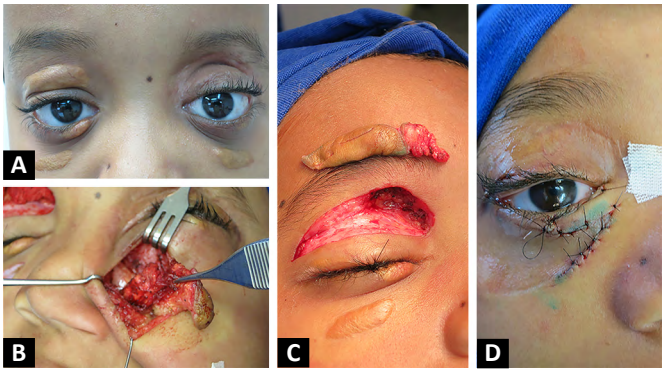

Figure 3. A. Pre-operative view; **B.** Resection of the lesion, left middle third; **C.** Resected right upper eyelid lesion; **D.** Immediate postoperative view of a transposition flap for reconstruction of the right lower eyelid.

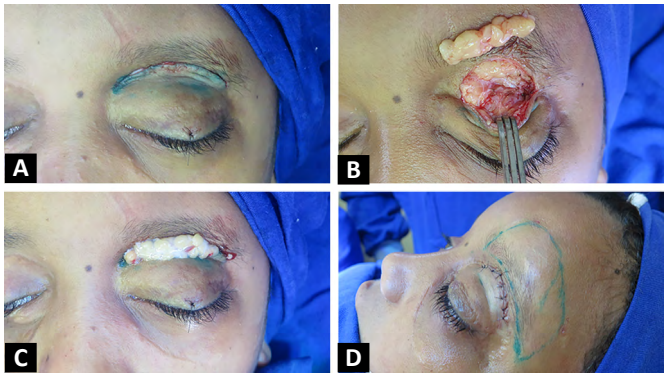

Figure 4. A. Incision below the eyebrow; **B.** Removal of the grafted skin area and the fat strip shown above; **C.** Grafted fat strip; **D.** Fat grafting in the frontal region for volume improvement.

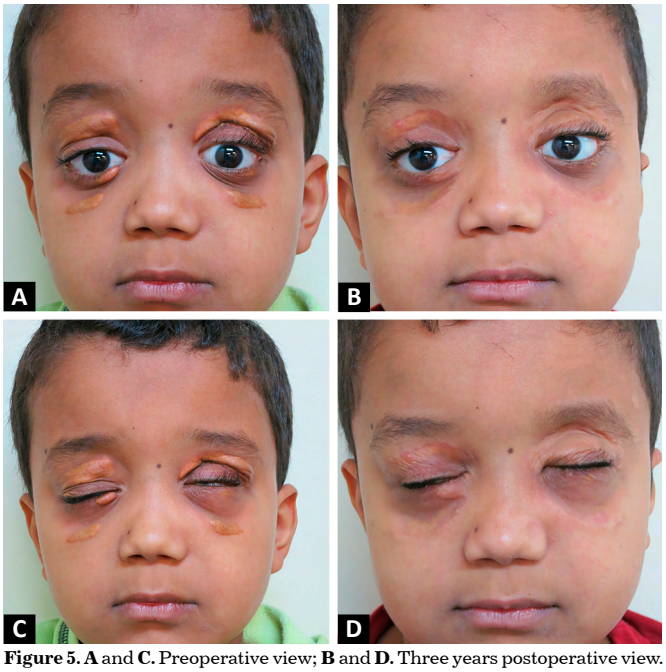

Figure 5. A and **C.** Preoperative view; **B** and **D.** Three years postoperative view.

Right upper eyelid

In the same moment of the left lower eyelid resection, this lesion measured approximately 25 mm × 10 mm and extended to the orbital septum; its strip resection allowed primary closure (Figure 3).

Right lower eyelid

In a 3 rd surgery, after also 3 months, this pretarsal lesion was close to the medial corner, measuring 10 mm × 4 mm, with primary closure impossible after resection. A paranasal transposition flap combined with a superomedial pedicle was used to ensure tension-free closure (Figure 3).

Right middle third

In the same procedure, this superficial lesion, measuring 17 mm × 7 mm, was excised and closed primarily. (Figure 3). Postoperative evolution.

Postoperative evolution

All lesions were sent for anatomopathological examination, which confirmed the diagnosis of JXG. No visual or palpebral motor dysfunction was observed in the postoperative period, but medium-term follow-up showed the need for volume correction of the left upper eyelid. The patient underwent four autogenous fat--grafting procedures in this region (at 6-month intervals), the first using fat strips and the other three using grafted fat, which improved the aesthetic appearance (Figure 4).

The medical team monitored the patient for 3 years from the first procedure to the current day (Figure 5).

DISCUSSION

JXG is a rare benign skin lesion of unknown etiology. Arises as yellow or orange papulonodular single or multiple lesions. Giant JXG, in turn, is characterized by a lesion; 2 cm[3,4,5]. It usually presents isolated or, in the case of multiple lesions, in distant sites[4,5,6], which has drawn attention to this unusual case of diffuse periorbital involvement.

The diagnosis was confirmed histopathologically, with histopathological examination showing dense histiocytic dermal infiltrate and Touton pathognomonic cells (multinucleated giant cells, homogeneous eosinophilic cytoplasm, and peripheral xanthomatization)[6]. In this case, all pieces were analyzed, and the diagnostic hypothesis of JXG was confirmed.

The literature shows that, when lesions similar to the present case do not involute in the first years of life, they tend not to regress despite the use of intralesional steroids[4,5,7,9,10].

Surgical treatment was indicated in this case due to the persistence of the lesions and the possible future

complications (infraorbital growth with compression of local structures and aesthetic impairment).

The surgical challenge, in this case, was the simultaneous involvement of all eyelids and the middle third of the face, which defined the resection strategy for multiple surgical times to ensure patient safety and better reconstruction results.

Resection of the lesion in left upper eyelid led to direct exposure of the aponeurosis of the levator palpebrae superioris muscle. Full-thickness skin grafts placed directly on the aponeurosis of the levator palpebrae superioris proved to be a viable tactical option in cases in which more morbid reconstructive options must be avoided. Although no study reported this maneuver, no impairment of the upper eyelid suspension movement was observed in the medium or long term. Autogenous fat grafts were used to improve the fat volume, although they were small.

It is important to emphasize that the complexity involving the fat grafting in this upper eyelid was due to the thin skin thickness and local fibrosis.

The other lesions were primarily closed or submitted to local miocutaneous flaps. It should be emphasized that the left malar lesion was deeply involved and required en bloc resection of the left infraorbital nerve. .

It is important to be aware that these situations can happen during intraoperative time and surgeons must have tools to solve them, even if they are simple but not common, like the skin grafting on the aponeurosis of the levator palpebrae superioris muscle, as did in this case.

CONCLUSION

The reviewed literature corroborates our finding that apparently superficial lesions may infiltrate deeply into the neighboring tissues, with great adhesion and significant bleeding if the dissection plane is inadequate[5,6,9,10].

The medical team must be prepared for situations that can arise in the intraoperative period that require the use of unusual options such as skin grafting directly on the aponeurosis of the levator palpebrae superioris muscle.

COLLABORATIONS

HF	Final manuscript approval, Project Administration, Supervision, Writing - Original Draft Preparation, Writing - Review & Editing
AHV	Conceptualization, Investigation, Realization of operations and/or trials, Writing - Review & Editing
RCL	Conceptualization, Data Curation, Project Administration, Realization of operations and/or trials, Writing - Original Draft Preparation, Writing - Review & Editing
VAM	Conceptualization, Realization of operations and/or trials, Writing - Review & Editing
MNSJ	Data Curation, Methodology, Realization of operations and/or trials, Writing - Original Draft Preparation
MBS	Conceptualization, Investigation, Realization of operations and/or trials, Writing - Review & Editing
RG	Supervision, Validation, Visualization, Writing - Review & Editing

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