Blepharoptosis and hypertrophic osteoarthropathy: A case report

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A 52-year-old male patient presented to our hospital with a history of secondary hypertrophic osteoarthropathy (HOA) associated with an abdominal neoplasia and blepharoptosis. He had finger clubbing, hyperhidrosis, and hypertrichosis. He also had a recent history of extensive abdominal surgery with a pathology report of myelolipoma. Routine blood work was unremarkable. Upper eyelid reconstruction with blepharoplasty, upper eyelid wedge resection, and brow suspension was performed to address his eyelid concerns. By this case report, we would like to attract notice that the eyelid involvement may be a part of HOA and to emphasize the importance of systemic and pathologic evaluation in failed blepharoptosis surgery.

Key words: Blepharoptosis, hypertrophic osteoarthropathy, myelolipoma

Hypertrophic osteoarthropathy (HOA) is a rare disorder characterized by new bone formation, clubbing of fingers and toes, swelling or pain of the large joints, excessive sweating of the palm, and thickening of the skin.[1] However, its manifestation with eyelid involvement is less common than the other features.[2,3] Here, we present an HOA case with bilateral blepharoptosis which was treated successfully by surgery. The tenets of the Helsinki Declaration were followed. Informed consent for surgical intervention and case report was obtained from the patient.

Case Report

A 52-year-old man presented with bilateral blepharoptosis [Fig. 1]. He had a history of bilateral brow suspension surgery 10 years ago at another institution. The patient had a 30 cm × 20 cm pelvic mass located in the presacral area with boundaries between both parailiac regions displacing the rectum and bladder. It was completely excised with a pathology report of myelolipoma. In physical examination, he had hypertrichosis, finger clubbing, and radiologically cortical thickening of the bones [Fig. 2]. He had no family history of HOA. The laboratory testing was unremarkable.

His best-corrected visual acuity was 7/10 in the right eye (RE) and 5/10 in the left eye (LE). His ophthalmologic examination was normal except corneal astigmatism with corneal peripheral opacifications. There was ptosis of both eyelids. Horizontal length of the upper lids of RE and LE was 46 mm and 41 mm, respectively. Vertical central tarsus lengths of RE and LE were 20 mm and 21 mm. Vertical fissure heights were 6 mm and 4 mm with margin-reflex distances of 0.5 mm and −0.5 mm, respectively. Vertical eyelid contour of the RE showed an inverse "V" shape for the RE and a smooth curve for the LE.

Anterior segment and fundus examinations were normal. He had long eyelashes with trichiasis. Thick mucoid secretion was revealed on everting upper eyelids. The eyelids were floppy and easily everted. To correct these clinical findings, a surgery which combines vertical tarsal shortening accompanied with horizontal whole eyelid wedge resection and brown suspension was performed. The excessive skin tissue was removed by blepharoplasty [Fig. 3]. At the end of the operation, 10 mg/0.25 mL triamcinolone acetate was injected into each supratarsal space to decrease the thickness of the tarsus. Horizontal wedge resection which is not a part of routine ptosis surgery was performed in this operation with the aim of normalizing the obvious horizontal length of the lids. Pathological evaluation revealed epidermal hyperplasia, severe inflammatory changes in subepidermal level, hyperplastic sebaceous glands, and collagen tissue derangement [Fig. 4].

The postoperative course was uneventful and patient satisfaction is good within the 1st year of follow-up [Fig. 5].

Discussion

Primary HOA is a rare disease with a prevalence of <0.2%.[2] It is characterized by skin thickening, clubbing, hyperhidrosis, and periosteal reaction in the long bones.[4,5] Primary HOA is predominantly a male disease that at least 90% of patients are men.[5] Although symptoms may be seen in childhood, it manifests mostly during the fifth decade of life. It has an autosomal dominant inheritance with variable penetrance, and Supradeeptha *et al*. described the largest number of primary

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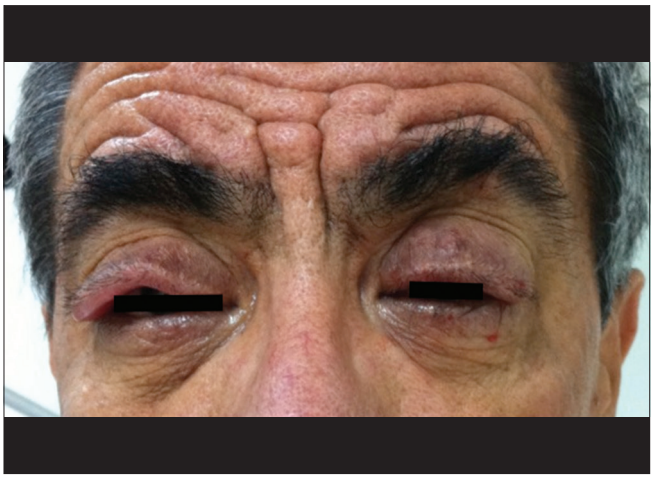

Figure 1: The patient presented with bilateral blepharoptosis with coarse skin folds

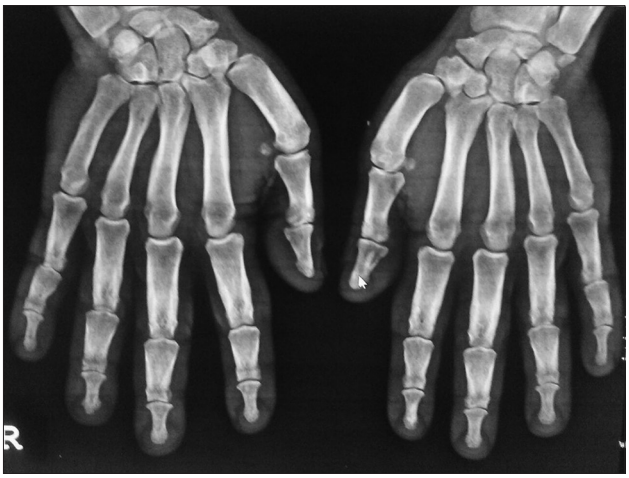

Figure 2: The patient had cortical thickening of the bones and clubbing

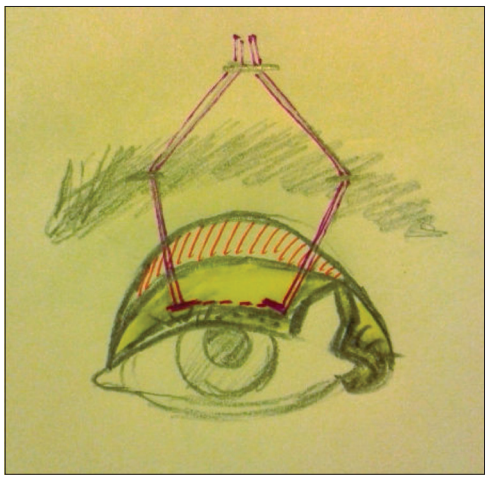

Figure 3: An illustration showing the surgical intervention. Reconstruction of each eyelid was achieved by vertical tarsectomy, horizontal full thickness lid resection, and supratarsal steroid injection. Ptosis was assessed with silicone rod frontalis suspension, and resection of excessive skin was performed as in blepharoplasty

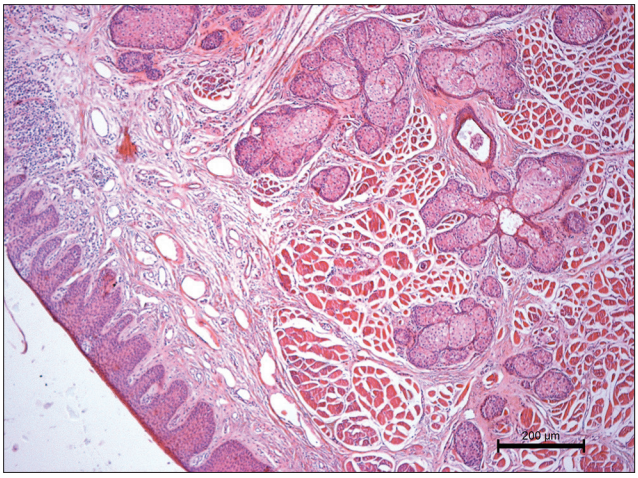

Figure 4: Skin biopsy showing thickening of the dermis with increased collagen content and lymphocytic infiltration (H and E, ×10)

HOA series in the literature that the authors reported that family history was positive in all studied patients.[5] The pathogenesis of HOA is unclear and some studies explained the role of several growth factors in the evolution of the disease.[6] Another study described a mutation in 15-hydroxyprostaglandin dehydrogenase.[7]

Although clinical findings are similar, secondary HOA differs from primary HOA with absence of family history. Secondary HOA is an acquired form that is associated with usually lung disease but also heart, liver, and intestines.[8] The presented patient had a huge pelvic mass which was completely excised with a pathology report of myelolipoma. The last eyelid surgery was performed 3 months after the abdominal surgery. Hence, our diagnosis was secondary HOA due to the paraneoplastic manifestations of pelvic malignancy which causes secretion of many growth factors. Blepharoptosis may develop secondary to sebaceous gland hyperplasia, thickening of the dermis with increased collagen content, mucin deposition, and lymphocytic infiltration or due to additionally marked scarring.[9] Our patient was a 52-years-old man with negative family history which let us exclude the diagnosis of primary HOA. He had a history of failed eyelid surgery. The recurrence may be explained by the continuing proliferative process.

The differential diagnosis includes several diseases as acromegaly, syphilitic periostitis, and thyroid acropachy.[5] Acromegaly is characterized by enlargement of facial bones that are not present in our case. Serology, radiology, and clinical findings did not support a diagnosis of syphilis or thyroid disease. HOA may develop secondary to pulmonary or congenital cyanotic cardiac diseases those were not present in our patient.[3,5] The remaining possible etiological factor for secondary HOA in our case was the huge pelvic malignancy which was excised previously.

The surgery for these kinds of pathologies may be planned as single[9] or staged procedures.[10] We performed multiple corrections in a single session under local anesthesia. Blepharoplasty with excessive skin excision is usually required both for better cosmesis and to reduce the tissue bulk. No complications were encountered during surgery, but bleeding was more than any other lid surgery. The profound inflammatory

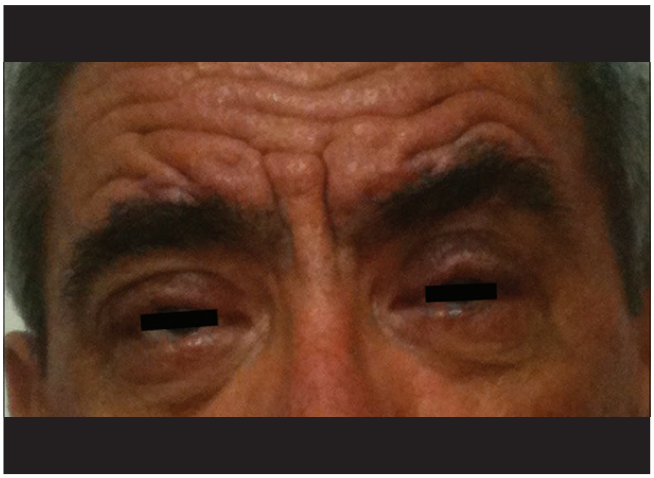

Figure 5: Increased palpebral aperture with a satisfactory cosmesis

reaction in the tissue might be the cause of this excessive bleeding.

Eyelid involvement of HOA is a rare entity though may cause ptosis. HOA may not be always primary, particularly in patients with negative family history. In cases of findings with abnormal fibroproliferative and inflammatory changes, detailed systemic examination and investigations both using serologic and imaging modalities should be performed for possible causes of secondary HOA.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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