Development of Chronic Conjunctival Swelling in Association with Congenital Lymphedema

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Abstract:

Although the association between the conjunctival swelling & lymphedema is rare, but this may occur, most of the cases described are part of the lymphedema distichiasis syndrome, this is the first case reported in North America where patient developed many years after birth (when the patient is 9 years old) an isolated chronic conjunctival swelling (not associated with any syndrome affecting the eyes) in association with congenital lymphedema of the feet.

Key words: Conjunctival swelling, Lymphedema, Childhood diseases.

Introduction:

Lymphedema describes a clinical condition characterized by the following:

1- Painless edema of one or both lower limbs usually involves the dorsum of the foot & toes primarily in young women.
2- Initially pitting edema, which becomes brawny & often non-pitting with time.
3- Ulceration, varicosities & stasis pigmentation do not occur, but there may be episodes of lymphangitis and cellulitis.

The underlying mechanism in lymphedema is impairment of the flow of the lymph from an extremity, when due to congenital developmental abnormalities consisting of hypo- or hyperplastic involvement of the proximal or distal lymphatic's, it is referred to as the primary form. The obstruction may be in the pelvic or lumber lymph channels and nodes when the disease is extensive and progressive, the secondary form results when an inflammatory or mechanical obstruction of the lymphatic's which occurs from trauma , regional lymph node resection or irradiation or extensive involvement of the regional lymph nodes by malignant disease or filariasis , secondary dilation of lymphatic's that occur in both forms leads to incompetence of the valve system , disrupting the orderly flow along the lymph vessels & result in progressive stasis of protein rich fluid with secondary fibrosis , episodes of acute & chronic inflammation may be superimposed with further stasis & fibrosis , hypertrophy of the limb results with markedly thickened & fibrotic skin & subcutaneous tissue with decrease in fatty tissue [(1)].

Although the association between the conjunctival swelling & lymphedema is rare, but this may occur, most of the cases described are part of the lymphedema distichiasis syndrome [(2)],

this is the first case reported in north America (as to our knowledge) where patient developed after birth in her childhood a chronic conjunctival swelling in association with congenital lymphedema of the feet .

Case report:

A young girl (DOB: March 19 , 2003) with a history of congenital chronic lymphedema at her feet (figure 1) referred to our office from River valley Pediatrics Clinic complaining from strabismus on September 10, 2007, a thorough examination performed & the following observed : the best corrected visual acuity is 20/30 +2 OD & 20/60 +2 OS , the cycloplagic refraction was +8.00 / -0.75 *8 OD & +9.25/-1.00 *170 OS (anisometropic hypermetropic astigmatism), left accommodative esotropia for distance & near with left amblyopia, the reminder of her slit lamp examination revealed normal structures & dilated fundus examination was within normal limits. Atropine penalization as a form of occlusion therapy was prescribed once daily in her right eye for one month with a full time spectacles were prescribed, a follow up in 2 months were suggested.

The patient continued regular visits to the office with continuous checkup and improvement noticed in her strabismus and amblyopia with time, but on her visit to the office at Jan. 18, 2011, the development of bilateral conjunctival nasal side chemosis worse on the right eye noticed, this associated with her accommodative esotropia and high hyperopic astigmatism, fortunately her left amblyopic eye improved with best corrected visual acuity 20/20 OD, 20/40 OS, her cycloplagic refraction was +6.00 OD & +7.75 / -0.75 *180 OS, full time spectacles required, a small angle esotropia was noted at distance and near vision which becomes better controlled with spectacle correction, otherwise the slit lamp and fundus examination were normal, Alrex eye drops were prescribed in her right eye three times daily for two weeks and part time occlusion therapy of her more dominant right eye was recommended during non-school hours, follow up after one month suggested.

Later with subsequent visits there were no improvement of the conjunctival swelling (figure 2), although all allergic and other causes were rolled out (conjunctival biopsy was refused by the mother), this lead as to the conclusion that the development of this conjunctival chemosis in the patient may be associated with her lymphedema.

Figure (1-a): Bilateral foot & leg edema-frontal view.

Figure (1-b): Bilateral foot &leg edema-frontal view.

Figure (1-c): Bilateral foot & leg edema from back.

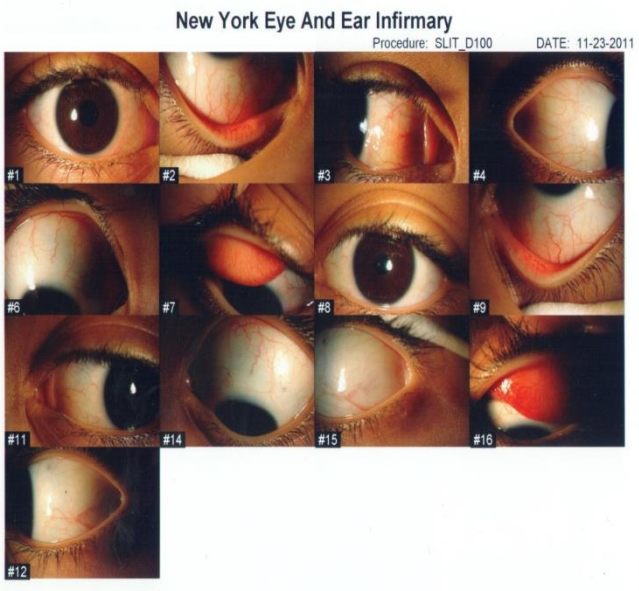

Figure (2): Chronic conjunctival swelling (name removed for patient privacy).

Discussion:

The association between conjunctival swelling in conjunction with chronic lymphedema is rare, most of the cases reported are part from the so called (lymphedema distichiasis syndrome) [(2)], few other cases reported to have such association, one reported by Perry and Cossary, they described a three and half year old female with Turners syndrome and strabismus having bilateral conjunctival chemosis persistent for years in association with lymphedema [(3)], another case reported by Bourcier, Baudrimont, Mayaud and Laroche , they described a patient with a yellow nail syndrome (a rare syndrome characterized by slow growing yellow nails associated with peripheral lymphedema) who developed a chronic conjunctival chemosis [(4)], there is only one case which reported by Tabbara and Baghdassarian, where they described a patient having isolated congenital lymphedema of extremities (not associated with any other syndrome) in association with conjunctival edema ,but their case developed conjunctival swelling earlier (when the patient was 7 years old) [(5)], our case is the first case reported to have isolated congenital chronic lymphedema of the feet who developed later in her childhood (when she became nearly 9 years old) a chronic conjunctival swelling.

Conclusion:

Isolated congenital chronic lymphedema may be associated with the development of chronic conjunctival swelling later in childhood many years after birth.

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