

Delleman syndrome or Haberland syndrome?

Sir,

We read the article by Nocito *et al.*^[1] with interest and wish to make the following observations:

1. The authors have described a dermoid cyst over the iris impairing patient's vision in the left eye. We disagree as the location of dermoid is on the cornea and limbus, rather than iris. Iris is the intraocular pigmented structure of eye, which regulates the entry of light into the eye. Authors also state that patient is having a blue-gray dermoid cyst over the right eye sclera. We disagree with the diagnosis in the right eye since the color of dermoid ranges from yellow to gray to pink rather than blue-gray.^[2] It seems

to be an anterior staphyloma because the color of staphylomata is almost always blue-gray due to underlying uveal tissue. The visible blood vessels over the surface of the lesion also favor the diagnosis of staphyloma since they are known to be present in staphyloma and are always absent in dermoid cyst. Furthermore, dermoid cysts usually have hair on their surface or in rare circumstances, a bunch of hair may be visible through thinned out cyst wall. Anterior and intercalary staphyloma may develop after chronic scleritis, trauma and limbal dermoid excision.^[2] Congenital anterior staphyloma has also been reported in the literature.

2. The minimal diagnostic criteria for Delleman-Oorthuys syndrome include a central nervous system cyst or hydrocephalus, microphthalmos with orbital cyst and focal dermal hypoplasia or aplasia.^[3] As two of these components, that are microphthalmos with orbital cyst and focal dermal hypoplasia or aplasia were missing from the case report of Nocito *et al.*,^[1] it does not appear to be a typical case of Delleman-Oorthuys syndrome. The constellation of findings in this case, i.e., limbal dermoid, frontal alopecia (? nevus psiloliparus), upper eyelid coloboma, papular lesion on the eyelid and arachnoid cyst is suggestive of encephalocraniocutaneous lipomatosis or Haberland syndrome or Fishman syndrome.^[4]
3. Authors did not report the status of visual acuity and intraocular pressure in their case and have stated that enucleation of eyeball is the only possible treatment for cosmetic purposes. We think that the present case is a potential candidate for limbal dermoid excision with keratoplasty in his left eye and staphyloma repair with donor scleral grafting with or without keratoplasty in his right eye. Surgical correction of limbal dermoids should be performed as early as possible to prevent amblyopia. Even if the patient has no perception of light in either eye, enucleation is never a procedure of choice. In such cases (absent light perception or blind eye), evisceration with orbital implant may be the procedure of choice, because it provides better cosmesis in term of greater ocular prosthesis motility and less chances of orbital implant extrusion.^[5] Enucleation alone can lead to post enucleation socket syndrome, which is characterized by

enophthalmos, deep upper eyelid sulcus, ptosis and laxity of the lower lid.^[6]

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REFERENCES

1. Nocito MJ, Luna PC, Contardi ML, Mazzini MA. Delleman syndrome: Report of a case in an adolescent boy. *Indian J Dermatol Venereol Leprol* 2012;78:229.
2. Mansour AM, Barber JC, Reinecke RD, Wang FM. Ocular choristomas. *Surv Ophthalmol* 1989;33:339-58.
3. McCandless SE, Robin NH. Severe oculocerebrocutaneous (Delleman) syndrome: Overlap with Goldenhar anomaly. *Am J Med Genet* 1998;78:282-5.
4. Hunter AG. Oculocerebrocutaneous and encephalocraniocutaneous lipomatosis syndromes: Blind men and an elephant or separate syndromes? *Am J Med Genet A* 2006;140:709-26.
5. Nakra T, Simon GJ, Douglas RS, Schwarcz RM, McCann JD, Goldberg RA. Comparing outcomes of enucleation and evisceration. *Ophthalmology* 2006;113:2270-5.
6. Steinkogler FJ. The treatment of the post-enucleation socket syndrome. *J Craniomaxillofac Surg* 1987;15:31-3.

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