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Vascular malformation of the iris – a case report

According to the literature vascular malformation of the iris is a rare condition (1–7). It can be a benign stationary state that has no apparent systemic associations and local complications (10), rarely it can cause spontaneous hyphema (7), coexist with orbital lymphangioma (14) or other underlying disorders (12). In our patient besides vessel malformation of the iris in the left eye we observed keratoconus and symmetrical large optic nerve heads bilaterally with no other anomalies.

CASE REPORT

A patient aged 20 was referred to the First Eye Hospital in April 1996 with low visual acuity and irregular astigmatism in both eyes. Best corrected visual acuity with spectacles in the right and left eye was 0.1 and 0.4 on the Snellen chart respectively. The slitlamp examination revealed keratoconus in both eyes and a large vessel of the iris in the left eye (Fig. 1). It passed from the anterior chamber angle toward the pupil, went on the pupil margin of the iris and returned to the anterior chamber angle. It occupied about one fourth of the iris margin. The vascular anomaly could not be easily observed in slitlamp examination because it was partly covered by iris stroma. Besides large diameter of optic nerve heads the optical fundus in both eyes did not demonstrate any arteriovenous anomalies or other significant changes. The patient was generally healthy with no systemic disorders. Corneal topography confirmed keratoconus in the right and left eye, with 62 dioptres and 55 dioptres in the apex respectively. The patient started wearing rigid contact lenses and was examined in the hospital occasionally. In September 2005 the patient was referred again because of worsening visual acuity to counting fingers from 4 meters in her right eye. Refractive error of this eye could not be longer corrected with contact lenses. Visual acuity in the left eye corrected with contact lenses was 0.7 on the Snellen chart. Intraocular pressure was 11 mmHg and 12 mmHg measured by applanation tonometry. In corneal topography keratoconus progressed bilaterally to 64.25 dioptres in the apex in the right eye and 57.20 in the right eye. In slitlam examination the same large blood vessel could be observed with no visible progression. During nine years of follow-up since 1996 none local complications have been reported. Fluorescein angiography of the iris that was proceeded easily visualized in the left eye large blood vessel. There was rapid filling and transit of dye within first 10 sec (Fig. 2). The vessel was uniformly hyperfluorescent and with no leakage even in the late phase of angiography (Fig. 3). Gonioscopy showed the open angle bilaterally, with no vascular malformations. The examination of the visual field in the right eye revealed general reduction of sensitivity due to low vision acuity. Visual field in the left eye was within normal limits. No systemic disorders have appeared. Because

in January 2006 visual acuity in the right eye worsened to counting fingers from 3 meters and could not be any longer corrected with contact lenses the patient was proposed to undergo penetrating keratoplasty in the nearest future.

Fig. 1. Slit-lamp photograph of the left eye, demonstrating the iris vascular malformation

Fig. 2. Early phase of fluorescein angiography of the iris (after 10 sec)

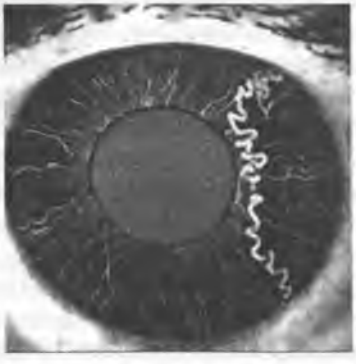

Fig. 3. Late phase of fluorescein angiography of the iris (after 2 min)

DISCUSSION

Vascular malformations described in this case report appear to be arteriovenous anomalies of the iris. The etiology of this anomaly is unknown. According to the literature it can be acquired or congenital (10). Because of similarity to retinal racemose hemangioma which is believed to be congenital malformation (2, 4) and because no other eye disorder as a cause of this abnormality was

observed, we would rather take advantage of congenital origin. It might develop during embryogenesis as a response to a localized failure of iris examination (9). Lately Shields et al. (10) have published 14 case reports of arteriovenous malformations. Clinical state and fluorescein pattern of angiography of our patient are quite similar to those described by them. However, there were no reports about coexistence of keratoconus and vascular malformation of the iris. We do not have an explanation for this state. Probably these disorders are unrelated because keratoconus was present also in the right eye, where it was more severe.

There have been a few other reports of such lesions and their visualization in fluorescein pattern (5, 6, 12). Parodi et al. presented fluorescein angiography and indocyanine green videoangiography (6). They concluded that angiography with indocyanine green is superior in showing iris hypoperfusion surrounding the vascular abnormality, although both techniques are able to precisely delineate the vascular pattern. In different studies angiographic pattern seems to be very similar: a rapid filling of abnormal blood vessel without any evidence of leakage or iris hypoperfusion (5, 6, 8, 9, 10, 12, 13).

Some authors have described additional to iris anomaly episcleral ("sentinel") blood vessel, which can suggest underlying ciliary body melanoma (3). In such cases detailed fundus examination, ultrasonography and transillumination is essential. However, none of the patients described in the literature with iris vascular malformations was diagnosed for melanoma. Authors suggest that epibulbar large vessel is rather continuous to the iris vessel (9, 13). In many patients with iris vascular abnormality previously described there were no other ocular disorders. In a few cases orbital lymphangioma, abnormal retinal vessel and smaller iris diameter and angioma racemosum were observed (14).

Vascular malformations of the iris as in our case report usually do not give any local complications such as increased ocular pressure, iris atrophy or neovascularization. In different studies authors described patients with vascular anomalies which caused spontaneous hyphema (1, 7, 11). In other two additional vascular tufts were observed three months after cataract surgery in the patient with similar iris anomaly (6). In differential diagnosis one should include iris varix, iris cavernous or capillary haemangioma, iris neovascularization and melanoma of ciliary body if large episcleral vessel is observed. It is possible that the same anomalies could be found in anterior chamber angle causing secondary glaucoma or in the retina as angioma racemosum being the cause of vitreous hemorrhage. In some cases Sturge-Weber syndrome or Wybum-Mason syndrome may manifest by iris malformation (10, 12). Usually no treatment is needed for this benign condition. Regular and detailed ophthalmological examination should be performed to detect and prevent progression or local complications (11, 15).

CONCLUSIONS

Vascular malformation of the iris is an unusual, benign condition that usually do not coexist with other ocular disorders. The clinical picture appears to be stable throughout the follow-up period. Local complications such as spontaneous hyphema are observed very rarely. Fluorescein angiography of the iris seems to be a useful technique to visualize vascular anomaly.

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SUMMARY

The aim of our study was to report clinical and fluorescein angiography features in a patient with vascular malformation of the iris and keratoconus. We reviewed the medical records of a patient referred to the First Eye Hospital in Lublin in April 1996. The patient aged 20 was referred to the First Eye Hospital with low visual acuity in the right eye and irregular astigmatism in both eyes. Best corrected visual acuity with spectacles in the right and left eye was 0.1 and 0.4 on the Snellen chart respectively. The slitlamp examination revealed keratoconus in both eyes and a large vessel of the iris in the left eye. The examination of the fundus did not demonstrate any significant changes. Corneal topography confirmed keratoconus in the both eyes, more severe in the right eye. In September 2005 the patient was referred again because of worsening visual acuity to counting fingers from 4 meters in her right eye. Due to severity and progression of keratoconus refractive error could not be longer corrected with contact lenses. Intraocular pressure was 11 mmHg and 12 mmHg measured by aplanatory tonometer. The slitlamp examination revealed again the same blood

vessel of the iris that did not change along the time. Fluorescein angiography of the iris of the left eye easily visualized large blood vessel uniformly hyperfluorescent and with no leakage of dye. Gonioscopy showed open angle in both eyes, with no vascular malformations. In corneal topography keratoconus progressed bilaterally to 64.25 dioptres in the apex in the right eye and 57.20 in the right eye. Vascular malformation of the iris is an unusual, benign condition that usually does not coexist with other ocular disorders. The clinical picture appears to be stable throughout the follow-up period. Local complications are observed very rarely.

Malformacja naczyniowa tęczówki – opis przypadku

Celem pracy było przedstawienie cech klinicznych i obrazu angiografii fluoresceinowej u chorego z malformacją naczyniową tęczówki i stożkiem rogówki. Posłużono się dokumentacją medyczną chorej, skierowanej do I Kliniki Okulistyki w Lublinie w kwietniu 1996 roku. Chora w wieku 20 lat została skierowana do Kliniki z powodu niskiej ostrości wzroku w oku lewym i nieregularnego astygmatyzmu w obu oczach. Ostrość wzroku z korekcją okularową wynosiła odpowiednio w prawym oku 0,1 i lewym oku 0,4 na tablicy Snellena. Badanie w lampie szczelinowej wykazało stożek rogówki w obu oczach i duże naczynie tęczówki w oku lewym. W badaniu dna oka nie zaobserwowano żadnych istotnych zmian. Topografia rogówki potwierdziła obecność stożka w obu oczach, bardziej nasilonego w oku prawym. We wrześniu pacjentkę skierowano ponownie z powodu pogorszenia ostrości wzroku w oku prawym do liczenia palców z 4 metrów. Ze względu na powiększenie się i wielkość stożka wada wzroku nie mogła być dalej korygowana przez soczewki kontaktowe. Ciśnienie wewnątrzgałkowe, mierzone tonometrem aplanacyjnym, wynosiło 11 mmHg i 12mmHg w prawym i lewym oku. W badaniu w lampie szczelinowej ponownie zaobserwowano nieprawidłowe naczynie tęczówki, które nie zmieniło się w ciągu kilku ostatnich lat. W angiografii fluoresceinowej tęczówki oka lewego wykazano duże naczynie krwionośne, z jednolitą hyperfluorescencją i bez przecieku fluoresceiny. W gonioskopii nie stwierdzono żadnych malformacji naczyniowych. Malformacja naczyniowa tęczówki jest rzadkim, łagodnym schorzeniem, które zazwyczaj nie współistnieje z innymi patologiami oka. Na przestrzeni kilku lat obraz kliniczny wydaje się stabilny. Miejscowe powikłania obserwuje się rzadko.