Title:
Hypertensive Choroidopathy: A Rare Manifestation in a Young Patient with Acute Hypertension

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Abstract:
A 13 year-old Hispanic Female with a history of kidney failure presents with bilateral disc edema and choroidal lesions, consistent with Elschnig spots. A diagnosis of hypertensive choroidopathy appears to be stable without treatment.

Outline

I. Case History
- Patient Demographics: 13 year old Hispanic Female
- Chief Complaint: Doctor directed follow-up for “bone spicule-like” pigment changes in posterior pole and possible papilledema OU
- Ocular History:
  - (+) mild anisometropic amblyopia OS>OD
  - (+) history of bilateral optic disc edema
    - patient’s neurologist attributes ONH edema to current immunosuppressive therapy
- Medical History:
  - End stage renal disease secondary to atypical hemolytic-uremic syndrome
  - Kidney transplant at age 5
  - (+) Trunical obesity, (Height 141 cm, Weight 43.1 kg, BMI 21.5)
  - (-) polydactyly or family history of polydactyly
- Medications:
  - Tacrolimus 1 mg PO
  - Mycophenolate Mofetil 500 mg PO
  - Oxybutynin Chloride 15 mg PO
  - Iron 325 mg PO
  - Calcitriol 0.25 mcg
  - Amlodipine Besylate 2.5 mg
  - Bactrim 40 mg

II. Pertinent Findings:
- VA:
OD: 20/20
OS: 20/30
- Refraction:
  OD: +1.50-1.50x180
  OS: +2.75-1.00x180
- Pupils: Equal, round reactive to light OU, (-)APD
- Color vision (HRR): 6/6 OD + OS
- SLE: Unremarkable OU
- Tonometry (GAT): OD: 16mmHg, OS: 17 mmHg at 9:25 AM
- DFE:
  ONH OU: elevated disc margins 360
  Posterior pole OU: lobular patches of hyperpigmentation surrounded by a margin of hypopigmentation scattered throughout superior and inferior arcades, (-) active inflammation
  Vessels OU: slight vessel tortuosity
  Macula OU: flat & intact; (-) edema
- OCT (Cirrus) through retinal lesions OU: Loss of RPE with resulting visibility of the choroid
- Optos Photos with Fundus Autofluorescence (FAF): Lobule shaped patches of hypo-autofluorescence with an outer border of hyper-autofluorescence OU
- Humphrey Visual Field 30-2: Enlarged Blind spot OU, scattered sup temp and inf temp defects OU
- MRI: Unremarkable OU; (-) space occupying lesions
- Blood Pressure: 96/52 (right arm, sitting)
- Blood Work: Mildly elevated Creatinine and Potassium; otherwise unremarkable

III. Differential Diagnosis:
- Primary/Leading Diagnosis: Hypertensive Choroidopathy
- Bardet-Biedel Syndrome (Truncal Obesity with Retinitis Pigmentosa) (ruled out by DFE and FAF)
- Presumed Ocular Histoplasmosis Syndrome (ruled out by SLE and OCT)
- Multifocal Choroiditis (ruled out by SLE and OCT)

IV. Diagnosis and Discussion
- Hypertensive Choroidopathy (HC)
  - Characterized by the necrosis of choroidal arterioles, with resultant non-perfusion of the overlying choriocapillaris and focal ischemic damage to the retinal pigment epithelium (RPE). This manifests
acutely as yellowish lesions at the level of the RPE, which, over time, become irregularly pigmented with a depigmented halo and are known as Elschnig Spots. (Stryjewski et al., 2016)

- Local RPE detachment and exudative retinal detachment may also occur in the course of HC.
- During the acute phase of HC, spectral-domain optical coherence tomography (SD-OCT) shows subretinal fluid with hyperreflective exudates subretinally. Classic V-shaped adhesions are seen at the location of each Elschnig spot due to fibrous bands extending from the RPE to the external limiting membrane. In the inactive stage, SD-OCT demonstrates total resolution of the subretinal fluid and subtle focal RPE elevation at the location of the Elschnig spots. (Altalbishi, 2015)
- Fluorescein angiography in the area of the Elschnig spots shows focal choroidal hypoperfusion in early phases and multiple subretinal areas of leakage in late phases.

- HC typically occurs in conditions that cause a rapid and severe elevation in systemic pressure, such as preeclampsia and eclampsia in pregnant women, kidney disease in adolescents, renal artery stenosis and pheochromocytoma.
  - The prevalence of ocular complications among pediatric patients following renal transplantation is 17% with 5.7% of patients presenting with optic disc swelling and/or hypertensive retinopathy. (Krause, 2010)
- Treatment of hypertensive choroidopathy is based on lowering blood pressure through the administration of hypertensive drugs
  - Ocular signs such as papilledema or retinal detachment most often subside a few weeks after normalization of systemic BP and normal visual acuity is regained. (Pociej-Marciaik, 2015)

V. Treatment and Management
- Continue taking anti-hypertensive medications as directed by PCP
- FA: not indicated due to stability of retinal lesions and lack of sub-retinal fluid
- Monitor every 6 months with BP, DFE and OCT
- Consider lumbar puncture to rule out papilledema OU

VI. Conclusion
Hypertensive Choroidopathy is a rare finding associated with acute increases in blood pressure. Retinal changes should remain stable as long as blood pressure is controlled with hypertensive medications. Many hypertensive children have underlying renal disorders; therefore screening hypertensive children involves close collaboration between pediatric nephrologists and eye care professionals.
References