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CASE STUDY

CHOROIDAL OSTEOMA: REVIEW OF LITERATURE AND FIRST CASE FROM YEMEN

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Abstract

Juxtapapillary choroidal tumors called choroidal osteomas are benign and do not require medical attention. They predominantly occur in young females (median age, 20 years). Ophthalmoscopy and fluorescein angiography can disclose indicative ocular signs, but computed tomography (CT), echography, and plain radiography are particularly crucial for making the right diagnosis. These lesions may be clinically misdiagnosed as a number of dystrophic lesions, as metastatic carcinoma, atypical malignant choroidal melanoma, metastatic carcinoma, leukemic or lymphoma infiltration, choroidal macular scarring, choroidal hemangiomas, or organized subretinal hemorrhage. As therapeutic methods, radiation and enucleation, it is crucial to rule out malignant melanoma from the differential diagnosis. According to an assessment of the literature, all reported instances with CT scans exhibited calcific densities that could be easily seen in the affected eye's posterior pole. The clinical, radiologic, and pathologic findings in our patient are all typical. We emphasize the importance of CT in assessing choroidal osteoma as well as the uniqueness of CT results.

Keywords: Choroid, CT scan, Osteoma, rare episode, Sana'a, Yemen.

INTRODUCTION

The earliest description of choroidal osteoma, a benign choroidal tumor that affected three young ladies and one girl and caused eye symptoms, was made by Gass *et al.*,[1] in 1978. Only 61 cases were seen at a major tertiary center in 26 years, which indicates that the illness is infrequently documented. A tertiary facility, however, is rarely sent to treat asymptomatic choroidal osteoma patients; instead, they are monitored in a community-based approach[2]. We present a case that is the first case reported from Yemen even from Arab countries, paying special attention to the radiological findings of this lesion, which are considered distinctive and useful in establishing the correct diagnosis. A benign ossifying condition called choroidal osteoma causes mature cancellous bone to develop in the choroid. The specific cause of the disease is still unclear, and incidence rates are incredibly low[3-5]. The majorities of these occurrences are unilateral and confined near the optic disc in women during the second and third decade of life[3]. No risk elements have been found. It shows as orange-yellow to yellow-white lesions with a definite edge and blood vessels covering them on the fundus examination. The degree of depigmentation of the underlying retinal pigment epithelium (RPE) determines the color of the lesion[6]. Early on, they usually have an orange-red color, but as the disease progresses, RPE depigmentation gives them a yellowish tint[4]. The diagnosis is primarily clinical and depends on how the lesion appears when the posterior pole is examined, although other diagnostic tests are needed to back up the diagnosis. The most diagnostic methods are ocular ultrasonography and CT, which show the bone character of the tumor[5]. The loss of photoreceptors, choroidal and RPE shrinkage coupled with decalcification, and choroidal

neovascularization (CNV) are the most frequent causes of vision loss in these patients[6,7].

Epidemiology

A rare condition is choroidal osteoma. The biggest case series in the literature consists of 61 patients treated for ocular oncology over a 26-year period at a significant tertiary care facility in the USA[6]. We still don't know its precise frequency. Although it can affect people of all ethnicities, this illness primarily strikes young adults in their early 20s, however it can also strike persons as young as a few months old and as old as late sixties. It favors women and is unilateral in roughly 80% of instances[3,6,8,9]. Although there have been a few familial cases reported[10-13], their overall incidence is low[8]. The majority of cases have been documented in publications from North American, Western European nations or Australia[2,3], and only a small number of instances have been recorded from Arab countries[14].

Etiology

Choroidal osteoma's precise etiology is unknown. Osseous choristoma[15], congenital reasons[11], endocrine abnormalities[3,10] and inflammatory diseases[17,18] are some of the theories about its development. It was not discovered to be connected to any ocular or systemic conditions. A few cases have shown a connection between choroidal osteomas and polypoidal choroidal maculopathy, pregnancy, Stargardt's maculopathy, recurrent inflammatory, pseudotumors of the orbit, intraocular inflammation, and histiocytosis X. Despite early indications linking it to trauma, no defined risk factors could be found[10,19]. The blood chemistries of phosphorus, calcium, alkaline phosphatase, complete blood count, and urinalysis have not been found to be associated with any anomaly in choroidal osteoma, despite prior suggestions[9,10].

Pathology

A branching network of arteries can be seen on the surface of a choroidal osteoma, which presents as a deep orange-yellow lesion with defined geographic or scalloped borders [Figure 1 and Figure 2]. The degree of depigmentation of the retinal pigment epithelium (RPE) is related to the color of the lesion. The hue of choroidal osteomas is typically orange-red in the early stages but takes on a yellowish tinge as they progress due to RPE depigmentation[4]. Histopathology depicts dense bony trabeculae with marrow gaps crossed by pathognomonic dilated thin-walled blood vessels, often known as feeder or spider veins. The choriocapillaris and the bigger choroidal vessels[1,4,15] appear to be connected by these vessels. In 41-64% of cases that were monitored for a total of 10 years[6,8,10,18,20-22], tumor development took place. The majority of choroidal osteomas exhibit sluggish random growth on any of the non-calcified borders, with an increase in mean basal diameter of about 0.37 mm per year[6], with the exception of sporadic cases of fast growth as reported by Mizota *et al.*,[23]. Around 50% of cases of tumor decalcification and resolution, first documented by Trimble in 1988, result in thin, atrophic, yellow-gray regions with overlying RPE and choriocapillaris atrophy. When the decalcification is behind the fovea because of underlying photoreceptor loss, it is linked to poor long-term visual acuity[6,7,24]. Decalcification may occur naturally[25] or may be induced by photodynamic treatment (PDT)[26], which is likely to stimulate osteoclastic activity in the lesion, or laser photocoagulation[7,20].

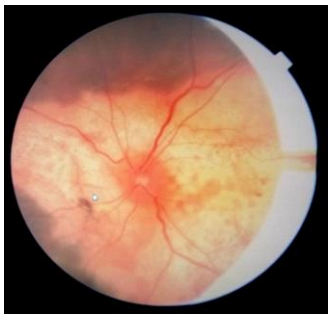

Figure 1: Left fundus photograph showing a posterior pole choroidal osteoma involving the whole posterior pole with overlying atrophy of the retina.

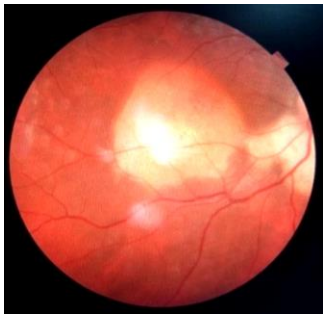

Figure 2: Right fundus photograph showing a posterior pole choroidal osteoma involving the optic disc and the lower part of the fovea.

The apposition of the overlaying retina to the underlying sclera was recently confirmed by optical coherence tomography (OCT)[27]. In choroidal osteomas, choroidal neovascularization (CNV) is highly prevalent. CNV is more likely to form in tumors with underlying bleeding and uneven surfaces[6]. Overall, CNV occurred in 31-47% of patients[6,8] and was associated with decalcification as a result of the RPE and Bruch membrane being disrupted[6]. Shields proposed that the disturbance of the RPE layer promotes the development of new choroidal capillaries under the surface[9]. Foster instead proposed that neovascular membranes are just an expansion of the osteoma[28]. Recent OCT imaging reveals neovascular membranes originating from the center of the osteoma, and osteoclasts were found in a surgically excised neovascular membrane to confirm his theory[27].

Additionally, hemorrhages, serous retinal detachment, and subretinal fluid have all been linked to choroidal osteomas. In the absence of CNV, choroidal osteoma frequently results in serous retinal detachment. In actuality, only about 23% of eyes with subretinal fluid also have CNV[8]. It is hypothesized to be the result of several precise RPE leakage locations seen by fluorescein angiography over the osteoma[29]. Alternately, it is believed that the outer blood-retinal

barrier disruption reduces the ability of the RPE and Bruch membrane to drain subretinal fluid[30].

Treatment

There is no established standard of care for choroidal osteomas, however medications are available to address difficulties brought on by subretinal fluid and choroidal neovascularization (CNV)[2,3].

CASE REPORT

The patient provided written informed consent for publication of her data, and our Institutional Review Board (IRB) approved publication of this case report. The report adhered to the basics of the 1964 Helsinki Declaration and its subsequent revisions. This case concerns a Yemeni female adolescent who was symptomatic and devoid of any clinical features that might indicate the presence of choroidal osteoma. There was no evidence of clinically significant intraocular trauma or inflammation in her ocular history. Her intraocular pressure was 21 mm Hg in the left eye and 19 mm Hg in the right eye, and visual acuity in the left eye was counting fingers at 3 meters and 18/6 in the right. On examination of the dilated fundus, we noted a cupped optic disc. Disc ratio 0.7 in both eyes in addition to bilateral yellowish-white lesions with clearly defined borders in the right eye and irregular borders in the left eye that were on the right on the posterior pole above the optic disc and the lower part of the fovea, measured 3 DD. The lesion in the left eye affects the entire posterior pole including the fovea and optic disc measuring approximately 10 DD. The MRI results: These lesions were both decalcified lesions, with the right one measuring 12 mm by 6 mm by 2 mm and the left one measuring 14 mm by 12 mm by 2 mm. There was no subretinal hemorrhage or CNV seen. A CT scan was advised because osteomas must be diagnosed using multimodal imaging. An irregular hyper-echogenic calcified lesion measuring about 14x12 mm on the left side and 12x6 mm on the right side was found in the posterior eye coating on both sides of the optic discs during a B-scan ultrasound, raising the possibility of choroidal osteoma rather than giant drusen. No aberrant hypo- or hyper-attenuation regions, no peri- or paraventricular calcification were visible in the brain's typical resting CT scan investigation.

DISCUSSION

The present case report refers to bilateral choroidal osteomas in a young female, despite the literature generally reporting a more usual one-sidedness and typical expression of choroidal osteoma in the teenage years. Choroidal osteoma's cause is still a mystery. In 1983, Katz and Gass[31] published the first description of a potential etiology of bilateral osteoma. They demonstrated a case of several osteomas growing in conjunction with bilateral pseudotumors of the orbit, which suggested that inflammation may play a role in the development of secondary ossification[31]. There were no connected triggers in the current instance that were made public. Shields *et al.*,[6] observed that at 10 years, choroidal osteoma showed evidence of decalcification in nearly 50% of eyes and evidence of growth in 51% of eyes in the largest case series on choroidal osteoma. In their series, choroidal osteoma decalcification was typically accompanied by impaired vision[6]. Decalcification frequently coexists with choriocapillaris atrophy and overlaying RPE changes, both of which may cause photoreceptor degradation and impaired visual acuity. In contrast to the calcified component of the osteoma, which had preserved intact outer retina (95%) and intact photoreceptor layer (100%), Shields *et al.*,[3] discovered that the decalcified portion of the osteoma had an overlaying marked thinning to nonexistent outer retina and photoreceptor layers (100%). In our case, no modifications were noted, and our patient exhibits no visual loss.

There aren't many choices for treating choroidal osteoma. In the event of extrafoveal CNV lesions, photodynamic treatment was also mentioned by Shields *et al.*,[26] as a viable option. A caveat was added by the authors at the end of the case report, stating that photodynamic therapy for subfoveal CNV may worsen visual acuity due to decalcification and accompanying RPE loss. Anti-vascular endothelial growth factor medications have recently been successfully utilized off-label to treat CNV secondary to choroidal osteoma[33]. The proper course of treatment is monitoring asymptomatic patients with fundus examinations and multimodal imaging at regular intervals to identify the onset of CNV or the atrophy of retinal layers and prevent further vision loss[33].

The following conditions are included in the differential diagnosis: dysmorphic calcification, metastatic calcification, choroidal metastasis, sclerochoroidal calcifications, amelanotic choroidal melanoma. In our case, the sclerochoroidal calcification fits the patient's clinical characteristics because it is typically bilateral and is seen in young white people who are asymptomatic as multifocal yellowish placoid lesions in the supero-temporal and postequatorial area; no other ocular or systemic signs were present[34,35]. Because it affects otherwise healthy eyes, as in the case of the current patient, choroidal osteoma is unusual in clinics. The case report's highlights include a rare disease condition affecting the choroid bilaterally without noticeable vision impairment that was discovered in a young female patient. The case report's limitations include the lack of follow-up information because our patient didn't have any prior clinic visits or exams. Six-monthly visits will be scheduled for our patient's routine follow-up.

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AUTHOR'S CONTRIBUTIONS

Al-shamahi EY: diagnosis and follow-up. **Al-Shamahi NY:** Writing, review, and editing, supervision. **Al-Shamahi EH:** data analysis. **Al-Shamahy HA:** supervision, review, and editing. All

authors revised the article and approved the final version.

DATA AVAILABILITY

Data will be made available on request.

CONFLICT OF INTEREST

Regarding this project, there is no conflict of interest.

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