Case Report:
NODULAR SCLERITIS WITH PRE-SUSPECTED RETINOBLASTOMA

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ABSTRACT

Nodular scleritis may complicate to uveitis. As we know that retinoblastoma is the most common malignant ocular tumor in childhood and can mimic uveitis, so that to rule out the malignancies in children is a must. Our objective is to report a difficult case of nodular scleritis and posterior uveitis in a child. The patient, a 4 years old boy, complained of red eye and loss of vision. From history revealed painful red eye, leukocoria and proptosis 3 weeks before admission. The patient has been visited to other eye hospital with presumed diagnosed was retinoblastoma. Examination found nodular scleritis and anterior uveitis with anterior synechiae and membrane in the pupil as the complications, and we found multiple dental carries. The USG showed vitreous opacity with echospike 60% and mass was suspected. From the immunology laboratory test found C3c was 114,9 mg/dL (N:90-180) and C4 was 15.5 mg/dL (N: 10-40), but sedimentation rate was increased. We treated with systemic and topical steroid and antibiotic, and performed serial ophthalmic ultrasound to evaluate the inflammation and the mass. After treated with systemic and topical steroid and antibiotic, result showed that the proptosis reduced gradually during two months observation and became phthisis. The nodular scleritis disappeared and the inflammation reduced. In conclusion, with the proper treatment and serial ophthalmic ultrasound give the advantage to save the eye ball in managing patient with nodular scleritis and posterior uveitis. Although blind or phthisical eyes are not at increased risk for malignancy, a tumor can occasionally be the cause of globe degeneration. Ultrasonography is useful in these eyes and planning a proper management.

Keywords : *Nodular scleritis, retinoblastoma, uveitis in child*

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INTRODUCTION

Uveitis in the pediatric age group is relatively uncommon and uveitis in children can present unique challenges to the physicians. (Simon et al. 2006) Nodular scleritis, is a deep red-purple color, immobile, and separated from the overlying episcleral tissue. Nodular scleritis may complicate to uveitis. (Sutphin et al. 2006) As we know that retinoblastoma is the most common malignant ocular tumor in childhood and can mimic uveitis, so that to rule out the malignancies in children is a must. (Simon et al. 2006, Wright & Spiegel 1995)

CASE REPORT

A 4 years old boy visited the Outpatient Clinical of Ophthalmology Department, Dr. Soetomo Hospital, Surabaya, in 19 January 2007 with complaint of red eye and loss of vision. From history revealed painful red eye, leukocoria, loss of vision and proptosis 3 weeks before admission. The patient has been visited to other eye hospital with presumed diagnosed was retinoblastoma because the clinical appearance look-like glaucomatous stage of retinoblastoma and has been planned for enucleation. But the family refused to have that operation.

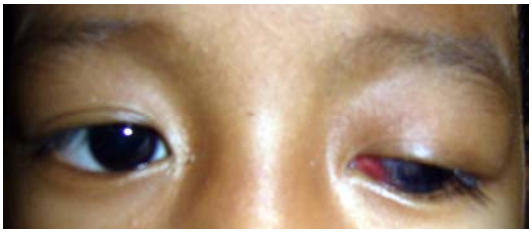

Figure 1a. On first day admission, the appearance looks-like glaucomatous stage of retinoblastoma

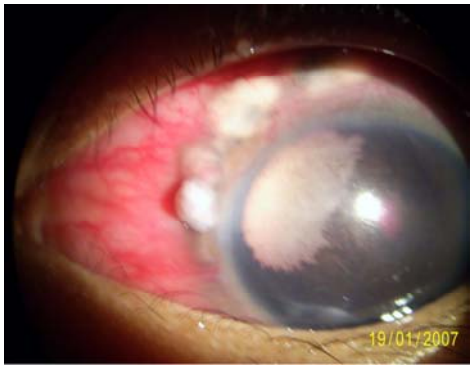

Figure 1b. Nodular scleritis with anterior uveitis

From clinical examination was revealed proptosis with nodular scleritis in the nasal limbus and anterior uveitis, with anterior synechiae and membrane in the pupil (leukocoria) as shown on figure 1a and 1b, we also found multiple dental carries.

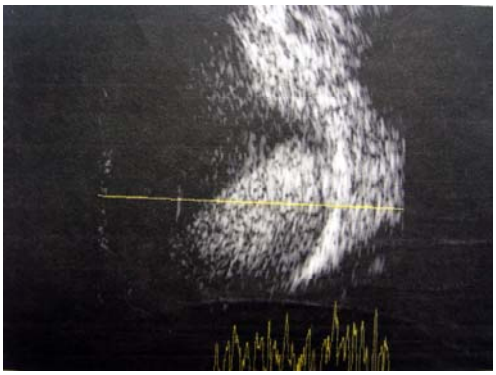

Figure 2.Before treatment. Mass was suspected

On the first ophthalmic ultrasound, showed vitreous opacity with echospike 60% and mass was suspected. There was semisolid mass in the inferonasal, without shadowing or choroidal excavation as shown on figure 2. From the immunologic laboratory test was found C3c was 114.9 mg/dL (N:90-180), C4 was 15.5 mg/dL (N: 10-40), and sedimentation rate was increased.

We didn't perform aqueous humor aspiration to measure the LDH aqueous because the anterior chamber was too shallow. We informed the patient and family about the disease but they still refused to have enucleation, so we treated as inflammation process. We closely observed the patient and if the conditions worsen, the decision to have enucleation must be done. We treated with systemic and topical steroids and antibiotic, and performed serial ophthalmic ultrasound to evaluate the inflammation and the mass.

Figure 3a.The left eye became phthisis and the inflammation reduced

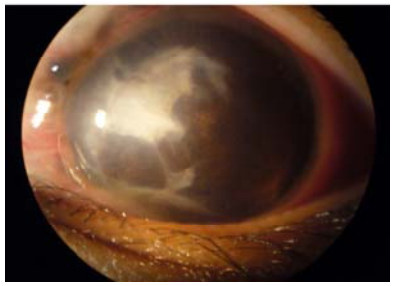

Figure 3b.Nodular scleritis disappeared

After treatment, the proptosis reduced gradually during two months observation and became phthisis. The nodular scleritis disappeared and the inflammation reduced as shown in figure 3a and 3b.

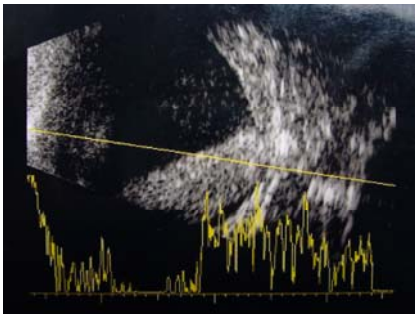

Figure 4.Ten days later. Semi solid mass in the inferonasal without shadowing. Axial length 22,04mm

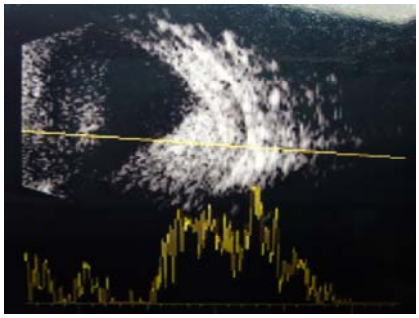

Figure 5.One months later. Mass regression with axial length 18.32 mm

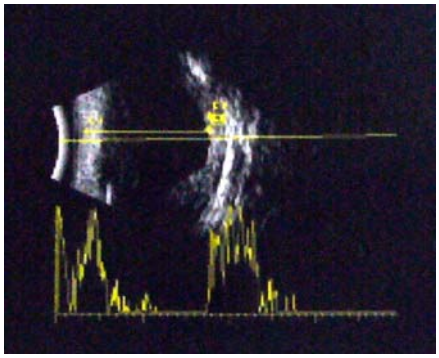

Figure 6. Two months later, vitreous opacity decreased with axial length 14,28mm

Serial ophthalmic ultrasound were performed to closely observe the inflammation and suspected mass. As shown on figure 4, 5, and 6, there were improvement in the inflammation process, in which vitreous opacity decreased and the suspected mass was reduced. The bulbus oculi became phthisis with shorter axial length (from 22,04mm became 14,28mm).

DISCUSSION

Uveitis in the pediatric age group is relatively uncommon, occurring at an annual rate of 6:100,000 and accounting for only 5%-10% of the total cases of uveitis seen in tertiary care centres. Nevertheless, uveitis in children can present unique challenges to the physicians. Children may not verbalize symptoms until disease is advanced. (Simon et al. 2006)

In nodular scleritis, the scleral nodule is a deep red-purple color, immobile, and separated from the overlying episcleral tissue, which is elevated by the nodule. Complications are frequent and include peripheral keratitis (37%), uveitis (30%), cataract (7%), glaucoma (18%), and scleral thinning (33%). Anterior uveitis may occur as a spillover phenomenon in eyes with anterior scleritis. (Sutphin et al. 2006)

In many cases, uveitis in children is idiopathic, but juvenile idiopathic arthritis (JIA) is the most common identifiable underlying systemic disease. Infections including toxoplasmosis, toxocariasis, and herpes viruses are relatively common in children with uveitis. Malignancies that can mimic uveitis in children include retinoblastoma and leukemia. (Simon et al. 2006)

Retinoblastoma is the most common malignant ocular tumor in childhood and one of the most common pediatric solid tumor, with an incidence of 1:14,000-1:20,000 live births. Retinoblastoma is typically diagnosed during the first year of life in familial and bilateral cases and between ages 1 and 3 in sporadic unilateral cases. The most common initial sign is leukocoria (white pupil), which is usually first noticed by the family and describe as a glow, glint, or cat's-eye appearance. Approximately 25% of cases present with strabismus. Less common presentations include vitreous hemorrhage, hyphema, ocular or periocular inflammation, glaucoma, proptosis, and hypopion. (Simon et al. 2006)

Diagnosis of retinoblastoma can be based on its ophthalmoscopic appearance. Intraocular retinoblastoma can exhibit a variety of growth patterns. (Simon et al. 2006) Large tumors often show signs of both endophytic and exophytic growth. Small retinoblastoma lesions appear a grayish mass and are frequently confined between the internal and external limiting membrane.

A third pattern, diffuse infiltrating growth retinoblastoma, is usually unilateral, nonhereditary, and found in children over 5 years old. The tumor presents with conjungtival injection, anterior chamber seeds, pseudohypopion, large clumps of vitreous cells, and retinal infiltration of umor. Because no distinct tumor mass is present, diagnostic confusion with inflammatory conditions is common. (Simon et al. 2006)

Pre-treatment evaluation of a patient with presumed retinoblastoma requires imaging of the head and orbits, which can confirm the diagnosis and can assist in evaluating possible extraocular extension and potential intracranial disease. MRI and ultrasound, which avoid use of radiation, may be preferable to CT, because the risk of secondary tumor is high in many of these patients.

Other, more invasive test are reserved for atypical cases. Aspirations of ocular fluids for diagnostic testing should be performed only under the most unusual circumstances because such procedures can disseminate malignant cells. (Simon et al. 2006) Direct tumor biopsy may performed in highly unusual cases (if there is visual potential), but this is risky due to the chance of seeding the tumor outside the eye. (Rapuano et al. 2000)

TUMOR REGRESSION PATTERN

Three ophthalmoscopically visible patterns of tumor regression have been described. In type I regression, tumor necrosis occurs, resulting in a white, calcify mass resembling cottage cheese. In type 2 regression, the tumor shrinks and takes on a translucent appearence

resembling fish flesh. It is difficult to distinguish such tumors from viable ones, and it is important to carefully document the size and number of each tumor during each examination. Type 3 regression is combination of type 1 and 2. 4

Spontaneous regression of retinoblastoma is also possible and can be asymptomatic, resulting in the development of benign retinocytoma, or it can be associated with inflammation and ultimately, phthisis bulbi. In either case, the genetic implications are the same as for an individual with in active retinoblastoma. (Simon et al. 2006) Anterior segment inflammation is treated with topical steroid and mydriatic/cycloplegic agents.

Because topical steroids do not penetrate well into the vitreous or posterior segment, sub-tenon injection of corticosteroid may be useful in treating older children with intermediate or posterior uveitis. Short courses of oral steroids may be used, but long-term use should be avoided because of significant side effects in young children. (Simon et al. 2006)

Glaucoma and cataract formation are two of the most serious ocular side effect of corticosteroid therapy. In general, the most potent corticosteroids are those most likely to produce an increase in intraocular pressure. Children appear to develop posterior subcortical cataract following oral corticosteroid therapy sooner and at lower doses than adults, but these cataract may be reversible after cessation of treatment. (Simon et al. 2006)

Other risks of long-term systemic steroid use in children include retardation of skeletal maturation and growth, osteoporesis and bone fractures, cushingoid appearance, diabetes, peptic ulcer, myopathy, hypertension, altered mental status, pseudotumor cerebri, and increased mortality from infection. . (Simon et al. 2006)

Blind eyes with opaque media should be suspected of harboring an occult neoplasma unless another cause of ocular disease can be surmised. Although blind or phthisical eyes are not at increased risk for malignancy, a tumor can occasionally be the cause of globe degeneration. Ultrasonography is useful in these eyes and planning a proper management. (Atta 1996)

CONCLUSION

With the proper treatment and serial ophthalmic ultrasound give the advantage to save the eye ball in managing patient with nodular scleritis and posterior uveitis. Although blind or phthisical eyes are not at increased risk for malignancy, a tumor can occasionally be the cause of globe degeneration. Ultrasonography is usefull in these eyes and planning a proper management.

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