Congenital Melanocytoma Manifesting as Proptosis With Multiple Cutaneous Melanocytic Nevi and Oculodermal Melanosis

Melanocytomas are benign, pigmented tumors usually seen in adults. They are normally asymptomatic, localized, and unassociated with any systemic features.1,3 We report a case of congenital melanocytoma with diffuse ocular involvement, extending into the orbit and causing proptosis, along with oculodermal melanocytosis and multiple congenital melanocytic nevi on the body.

Report of a Case. A 6-month-old boy had progressive protrusion of the right eye since birth. He had a proptosis measuring 34 mm with a pigmented subconjunctival mass in the upper fornices and cutaneous pigmentation on the upper eyelid (Figure 1A). Multiple pigmented cutaneous lesions were seen throughout the body, ranging from 1 to 10 mm (Figure 1B).

A computed tomographic scan showed a large lesion infiltrating the right eyeball and posterior orbit, causing scalloped expansion of the orbit (Figure 1C). An incisional biopsy specimen taken through the superior fornix revealed a melanocytoma. Because melanocytomas of such infiltrative nature and progression can become malignant, an exenteration was done. A nevus was also biopsied. Results of a complete metastatic workup were normal. Gross examination showed a pigmented mass measuring 35 × 25 × 25 mm, filling up the globe and extending superoposteriorly into the orbit (Figure 1D). Microscopical analysis showed heavily pigmented tumor cells, of which bleached preparations revealed plump polyhedral nevus cells with abundant cytoplasm and small, uniform nuclei (Figure 2A and B). The tumor had infiltrated the sclera posteriorly and replaced the entire optic nerve. There was no evidence of mitosis or vascular invasion. Staining for S-100 protein and HMB-45 were positive. There was no evidence of mitosis or vascular invasion. Staining for S-100 protein and HMB-45 were positive. Staining for Ki-67 showed a proliferative index less than 1%. Features of melanosis oculi in the form of increased dendritic melanocytes were seen in the episclera, sclera, and optic nerve sheath (Figure 2C).

Results of the microscopical analysis of the cutaneous lesion were consistent with a congenital melanocytic nevus (Figure 2D).

Three years later, there was no recurrence.

Comment. Melanocytoma, although previously often confused with malignant melanoma, is now distinctly recognized by its typical clinical and benign histological features.1,2 The tumor in this case was diffuse and infl-

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Melanocytoma can manifest with vision loss, glaucoma, scleral pigmentation, or an iridochoroidal mass. Our patient had a large proptosis. A literature search revealed only 1 report of a congenital melanocytoma with proptosis. Unlike our case, it was mainly orbital and surrounded the optic nerve.

Ocular melanocytosis is characterized by hyperpigmentation of the uvea, sclera, episclera, and optic nerve. Shields et al have pointed out the association of melanocytoma with oculodermal melanocytosis and suggested that although they are considered to be different entities, they may be different expressions of the same disease. Our patient also had melanocytoma with oculodermal melanocytosis.

Melanocytomas usually have no systemic associations. To our knowledge, this is the first report of melanocytoma associated with multiple congenital melanocytic nevi. Cases of congenital malignant melanoma of the choroid with multiple congenital melanocytic nevi have been reported. Our case had benign histological features, showing only a locally aggressive infiltration, making it distinct from the other cases reported.

Several reports have cited growth in melanocytoma. A history of rapid progression as in our case is unusual. Few reports have also shown extrascleral extension, but to our knowledge this never occurs with complete globe involvement and extensive orbital infiltration. The overall prognosis of melanocytoma is good. Slight growth may not signify malignancy. However, more progressive growth suggests malignant transformation.

Our case is unusual because it manifested congenitally with diffuse ocular involvement and orbital extension, causing proptosis. It also suggests an association...
between melanocytoma, oculodermal melanocytosis, and multiple congenital melanocytic nevi.

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Evaluating Patient Discomfort, Anxiety, and Fear Before and After Ranibizumab Intravitreous Injection for Wet Age-Related Macular Degeneration

Ranibizumab (Lucentis) intravitreous injection (IVI) has emerged as a common treatment for wet age-related macular degeneration after 2 international, multicenter, controlled clinical trials, MARINA1 and ANCHOR,2 reported positive visual outcome compared with placebo and photodynamic therapy. Despite the therapeutic benefits of ranibizumab, repeated injections are required as often as every 4 weeks. We aimed to study the patients’ perspective of treatment with ranibizumab, specifically pain, anxiety, and discomfort related to the ranibizumab IVI procedure. To