

on the lens surface. (b) Light microscopy of the cross section of the IOLs analyzed with von Kossa stain showed numerous fine, granular, crystalline-like deposits below the surface of the IOL. (c) Image showing deposits in higher magnification.

**Supplementary Fig. 2—(a, b)** The modulation transfer function curves showed a significant decrease in optical quality compared with ISO standard.

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## Footnotes and Disclosure

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## Rosai-Dorfman disease with corneal anaesthesia: case report and review of literature



Rosai-Dorfman disease is a rare systemic histioproliferative disease characterized clinically by cervical lymphadenopathy and pathologically by lymph node sinuses containing histiocytes with intact phagocytosed lymphocytes (emperipolesis).<sup>1</sup>

Extranodal sites may be involved by the disease.<sup>1</sup> Ocular involvement is uncommon and manifests most frequently as eyelid or orbital masses; other ocular manifestations are extremely rare.<sup>2–5</sup> We report a novel case of a patient with unilateral corneal anaesthesia and neurotrophic keratitis as presenting features of Rosai-Dorfman disease.

A 16-year-old male presented with a corneal ulcer in the left eye. He had a history of global developmental delay with seizure disorder, making clinical examination challenging. On examination, there was an absence of corneal sensation in the left eye with a small (1.5 mm × 1.5 mm) inferior corneal ulcer. Corneal sensation was normal in the right eye. There was no lagophthalmos. The rest of the anterior segment was unremarkable. He also had a painless left temporal orbital mass that had been gradually increasing in size for the previous few months. There was no proptosis. Extraocular movements were normal. The ulcer was managed with artificial tear drops, antibiotic ointment, and lid taping; and it healed with corneal vascularization (Fig. 1). A magnetic resonance imaging (MRI) of brain and orbits, examination

under anaesthesia (EUA), biopsy of the orbital mass, and left tarsorrhaphy were scheduled.

The MRI (Supplementary Figs. 1A and 1B, available online) showed an ill-defined mass in the superior-temporal aspect of the left orbit, involving the left lacrimal gland. It measured 17 mm × 34 mm × 27 mm. It had mild mass effect on the left globe as well as the lateral rectus, superior oblique, and superior rectus muscles. No proptosis was seen. The extraocular muscles and optic nerve were normal. Expansile soft-tissue masses involving the nasal cavity, ethmoids, and right osteomeatal complex were seen.



**Fig. 1—Left vascularized corneal opacity. The superior-temporal orbital mass is also partly visualized.**

At EUA, there was left corneal stromal scarring and vascularization. Cycloplegic refraction showed a refractive error of  $-15.00$  D OD and  $-14.00$  D OS. Dilated fundus examination showed myopic changes in both eyes. The mass was firm and nodular on palpation. Enlarged lymph nodes in bilateral anterior and posterior clavicular chains were found.

A small skin incision was used for biopsy. The mass appeared pale pink with a capsule (Supplementary Fig. 2, available online). An incisional biopsy and left tarsorrhaphy were then performed.

Histopathology of the specimen showed an encapsulated mass reminiscent of an obliterated lymph node with no definite residual nodal structure. Sheets of foamy histiocytes separated by fibrous bands were seen. Lymphoid aggregates with follicles were mixed with the histiocytes. Intact lymphocytes and plasma cells were seen within the cytoplasm of histiocytes (emperipolesis) (Supplementary Fig. 3, available online). There was no evidence of malignancy. Lesional histiocytes were positive for S100, CD68, and CD163 by immunohistochemistry (Supplementary Figs. 4A and 4B, available online). Immunostains for CD1a and langerin were negative.

The findings of histopathology and imaging were consistent with Rosai-Dorfman disease. Imaging of the chest and abdomen was done to exclude other sites of involvement, and it was normal. After discussion with the patient's family, it was decided not to start any treatment owing to the patient's general condition and neurological delay. His ocular surface is now stable.

Rosai-Dorfman disease was initially known as "sinus histiocytosis with massive lymphadenopathy."<sup>1</sup> The exact etiology is unknown. An underlying immune dysfunction as well as possible infection (with human herpesvirus 6, Epstein-Barr virus, parvovirus, or *Klebsiella*) have been postulated.<sup>1,4</sup>

Painless cervical lymphadenopathy is the most frequent presenting symptom.<sup>1,2,4</sup> Extranodal disease is reported in up to 43% of patients, the most common sites being skin, nasal cavity, and paranasal sinus.<sup>1,2,4,5</sup>

Ophthalmic involvement is rare (up to 11%).<sup>5</sup> Only 0.03%–0.09% of all ocular specimens received for pathological examination are reported with this diagnosis.<sup>4</sup> Reported ophthalmic manifestations include orbital and eyelid involvement (most common), lacrimal gland involvement, subconjunctival nodules, scleritis, uveitis, and compressive optic neuropathy.<sup>2–5</sup> Associated proptosis has been reported to cause corneal exposure and ulceration,<sup>3</sup> but corneal anaesthesia and neurotrophic keratitis as seen in our patient have not been reported. The orbital mass, cervical lymph nodes, and nasal and sinus masses were the more typical manifestations of the disease in our patient. Corneal anaesthesia was likely owing to compression of the branches of the left trigeminal nerve by the mass. This is extremely rare and has been previously reported to occur only in isolated case reports with some other tumours in the region.<sup>6,7</sup> Alternatively, another cause for corneal anaesthesia such as

herpes simplex or zoster infection cannot be ruled out; however, our patient had no history of skin rash or recurrent eye redness. Rosai-Dorfman disease has been postulated to be associated with other herpesvirus infections (specifically Epstein-Barr virus and human herpesvirus 6), which are not normally associated with neurotrophic keratopathy, but there is a possibility that a coexisting infection led to both corneal anaesthesia and the systemic presentation.<sup>4</sup>

Histologically, the hallmark of the disease is lymphophagocytosis or emperipolesis. The involved histiocytes express S-100, CD68, and CD163, but are negative for CD1a.<sup>1,2,4,5</sup>

Only half of patients with Rosai-Dorfman disease need some form of treatment. The aims of treatments for ophthalmic manifestations are to control functional (e.g., vision, ocular motility) and cosmetic abnormalities. Management options include surgical excision or debulking for lesions in accessible locations, and systemic corticosteroids, chemotherapy, or radiotherapy in patients with severe symptoms.<sup>1,2,4,5</sup>

Discussion with the patient and family is important to make management decisions. In our patient, the lack of significant treatable functional or cosmetic abnormalities as well as the fact that he was developmentally delayed and that his quality of life may further deteriorate from side effects of aggressive treatments were important in the decision to only observe and monitor.

To conclude, Rosai-Dorfman disease, though extremely rare, must be considered in young individuals with slowly progressive orbital masses, especially with associated lymphadenopathy. Care should be taken to check for sight-threatening manifestations like compressive optic neuropathy or neurotrophic keratitis, as in this case. Also, orbital masses should be considered in the differential diagnosis of corneal anaesthesia.

## Supplementary Materials

Supplementary material associated with this article can be found in the online version at doi:[10.1016/j.jcjo.2020.06.003](https://doi.org/10.1016/j.jcjo.2020.06.003).

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### Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

## A case of complete spontaneous regression of extensive Merkel cell carcinoma involving the orbit



Merkel cell carcinoma (MCC) is a rare and aggressive neuroendocrine cutaneous carcinoma with an annual incidence of 0.18–0.42 cases per 100 000.<sup>1</sup> MCC has a propensity for local recurrence, lymphatic, and distant metastasis, and most commonly presents in the head and neck region of elderly and immunocompromised patients.<sup>1,2</sup> Despite its

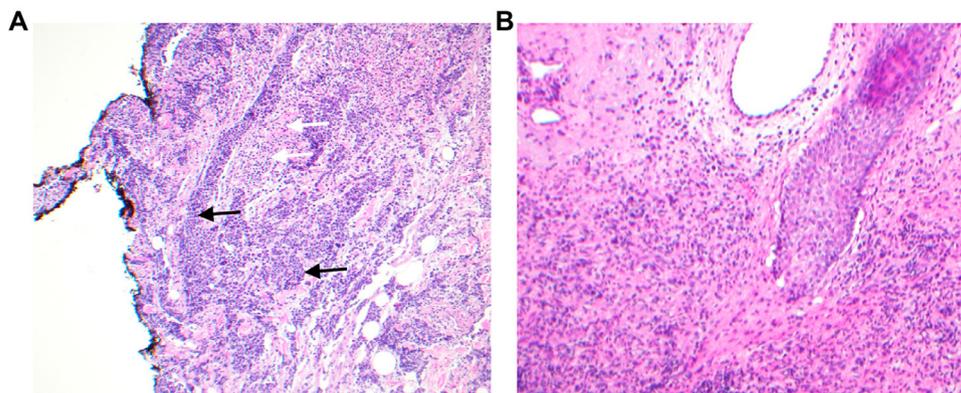


**Fig. 1**—Clinical photograph of a large, nodular, exophytic solid and reddish mass with rapid growth over 4 months at the left lower eyelid–cheek junction. Histopathological studies revealed a diagnosis of Merkel cell carcinoma.

aggressive nature and poor 5-year survival rate (60%),<sup>1</sup> complete spontaneous regression (CSR) has been documented in approximately 1.67% of cases.<sup>3</sup> In all reported cases of CSR, resolution occurred within 18 months of diagnosis of the primary tumour, and the regression was rapid, occurring over 1–3 months.<sup>3</sup> In most cases, CSR followed diagnostic biopsy or incomplete excision of a primary cutaneous lesion.<sup>3</sup> In a few, CSR occurred in locally recurrent disease or nodal metastasis without prior intervention.<sup>3</sup>

To our knowledge, there has not been a previously documented case of spontaneous regression of MCC invading the orbit. Herein, we report a case of CSR following diagnostic incisional biopsy of an extensive MCC involving the left lower eyelid, cheek, and anterior orbit.

A 71-year-old woman presented to an outside institution in March 2016 with a 4-month history of a fast-growing, protruding mass on the left lower eyelid and cheek (Fig. 1). Computed tomography revealed a 4 cm × 3 cm × 3 cm rounded isodense homogenous cutaneous/subcutaneous mass at the level of the left lower eyelid and inferior orbital rim with invasion into the intraorbital fat. There was abutment of the globe with no globe deformation, extraocular muscle involvement, or lymphadenopathy. An incisional biopsy was performed, and histology showed small blue cells with sparse cytoplasm and irregular hyperchromatic nuclei (Fig. 2A). Tumour cells stained positively for



**Fig. 2**—(A) Initial biopsy. Hematoxylin & eosin stain demonstrating small, round malignant cells with little cytoplasm and irregular hyperchromatic nuclei (black arrows) with associated lymphocytic infiltration (white arrows). Immunohistochemical staining was diagnostic of Merkel cell carcinoma. (B) Repeat biopsy after clinical signs of regression. Hematoxylin & eosin stain demonstrating granulomatous inflammation consisting of many histiocytes and lymphocytes, but the absence of malignant cells.