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Case Report

A rare case of deep hemangioma presenting as a periorbital swelling and its excellent response to intralesional steroids

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Abstract A hemangioma is a benign and usually self-involuting tumor of the endothelial cells of blood vessels, and is characterized by an increased number of normal or abnormal vessels. It usually appears in the first few weeks of life and grows most rapidly over the first six months. Usually, growth is complete and involution has commenced by twelve months. Half of all infantile hemangiomas involute by age five, 70% by age seven, and most of the remainder by age twelve. In more severe cases hemangiomas may leave residual tissue damage. The dermatologic indications for intervention in a case of hemangioma include maceration and erosion of the epidermis, infection, and cosmetic disfigurement. The treatment options include corticosteroids (oral), interferon, vincristine, propranolol, pulsed dye laser and surgery. The intralesional steroids are usually not recommended in the periocular region because of side effects like eyelid necrosis and central retinal vein occlusion. Here we report the case of a deep hemangioma in periorbital region with no overlying skin changes and its excellent response to intralesional steroids with no side effects.

Introduction

Hemangiomas usually appear within the first month of life although they present as a precursor lesion in the immediate neonatal period. They may be superficial where they appear as bright red in the period of proliferation, deep in which they have a more blue appearance, or a combination of both superficial and deep (25-30%). The deeper lesions are located in the deeper dermis and/or subcutis and do not have any overlying skin changes, which may make diagnosis more difficult or even delay diagnosis. The treatment options include corticosteroids (oral), interferon, vincristine, propranolol, pulsed dye laser and surgery. The topical and intralesional steroids are not usually recommended in the treatment of hemangiomas. The intralesional steroids are usually not recommended in the periocular region because of side effects like eyelid necrosis and central retinal vein occlusion.[1,2,3] Here, we represent the case of a deep hemangioma with no overlying skin changes and its excellent response to intralesional steroids.

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Case Report

We report the case of a 22-year-old young male patient of ethnic Kashmiri origin and a teacher by occupation who presented with periorbital swelling on the right side of 5-year duration. The swelling was initially limited to lower eyelids which progressed gradually to involve the both eyelids. There was no history of pain, itching or bleeding or trauma. There was history of increase in size in the morning hours, however there was no aggravating or

Figure 1 Periorbital swelling involving both upper and lower eyelids on right side.

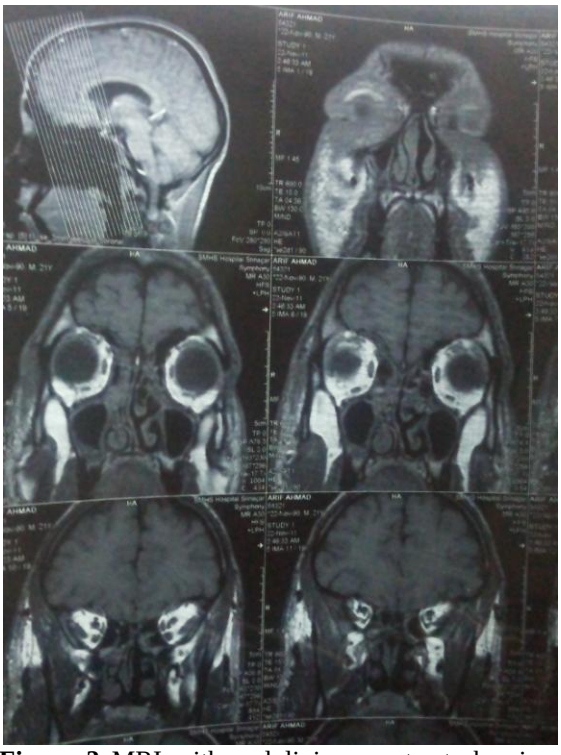

Figure 3 MRI with gadolinium contrast showing homogenous enhancement of subcutaneous tissues of right orbit suggestive of hemangioma. Note there is no intraorbital or intracranial extension of the lesion.

relieving factor. There was no history of diminution of vision, headache or any other systemic complaint.

On examination the swelling was diffuse skin coloured involving both upper and lower eyelids on right side, fluctuant and non-tender. There was no bruit. The patient was suffering

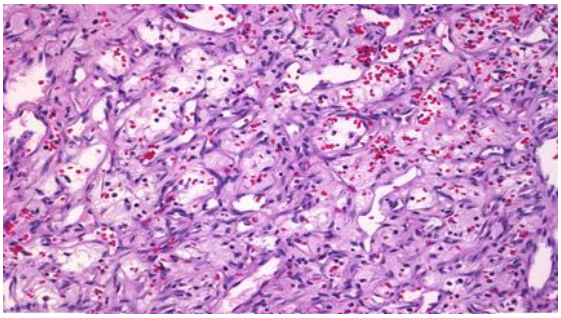

Figure 2 Histopathology of the lesion suggestive of hemangioma showing vessels with intra- and extravascular erythrocytes.

Figure 2 Same patient after receiving intralesional steroids.

from concurrent psoriasis vulgaris which was, however, stable and inactive. The patient was evaluated for all the possible causes of periorbital swelling. The CBC, urinalysis, liver function tests, renal function tests, thyroid function tests were all within normal limits. Serum cortisol, dexamethasone suppression test and serum prolactin levels were also within normal limits. Skin biopsy showed predominantly small vessels and extravasated erythrocytes (suggestive of hemangioma, **Figure 2**). There was no inflammatory infiltrate. Colour Doppler ultrasound studies were suggestive of a vascular swelling (most likely hemangioma). The MRI of periorbital regions was performed which confirmed the vascular nature of the swelling. However there was no intraorbital or intracranial extension of the swelling (**Figure 3**). The patient received

oral steroids and clofazimine with no response. The patient then received four intralesional steroid injections (triamcinolone 10mg/ml) at 3 weekly intervals (after ruling out intraorbital and intracranial extension with imaging) which showed excellent response (the swelling regressed completely) and with no local side effects such as eyelid necrosis (**Figure 4**). The patient is under follow-up from last 3 months and there are no signs of recurrence.

Discussion

Hemangiomas usually appear within the first month of life although they present as a precursor lesion in the immediate neonatal period. This can be very subtle, with a faint telangiectatic patch or even a patch of nevus anemicus like pallor. The hemangiomas may be superficial where they appear as bright red in the period of proliferation, deep in which they have a more blue appearance, or a combination of both superficial and deep (25-30%). The deeper lesions are located in the deeper dermis and/or sucutis and do not have any overlying skin changes, which may make diagnosis more difficult or even delay diagnosis.[4,5] The diagnosis is usually not difficult if the classical signs of a precursor lesion are present followed by a period of growth and involution. However, deep hemangiomas may pose more difficulty with differentiation from vascular malformations. The latter tends to stay fixed and may enlarge during physiological stimuli, for example puberty and pregnancy and does not go through a period of involution. The use of CT and MRI scanning is not particularly helpful in aiding diagnosis, as both hemangiomas and vascular malformations are reported as hemangiomas. There is evidence to support the use of scanning in the delineation of the extent of hemangioma and its response to therapy, for example steroids or interferon.[6,7]

In addition to size the anatomical location is of major importance in determining the potential for complications such as airway obstruction, visual disturbance and ulceration. Other complications which can be seen in case of hemangiomas include heart failure, auditory obstruction, disfigurement and associated abnormalities such as spinal dysraphism and PHACES syndrome. The major goals for the management of hemangiomas include the prevention or reversal of life-threatening complications, prevention of disfigurement from skin changes following involution, treatment of ulceration to minimize scarring and reduction of psychological stress to minimum.[8]

The management options include active non-intervention (observation) and therapeutic intervention. The possible therapeutic interventions include systemic corticosteroids for expanding, function- or life-threatening hemangiomas (topical and intralesional steroids are usually not recommended); interferon (second line for expanding, function- or life-threatening hemangiomas but used in children older than 1 year); vincristine (2^{nd} line for expanding, function- or life-threatening hemangiomas in combination with oral steroids); propranolol, pulsed dye laser (for residual or large plaque hemangiomas); surgery (may lead to scarring) and topical immunomodulators (limited evidence of benefit for small hemangiomas).[9,10]

Systemic corticosteroids especially oral have been used in the treatment of hemangiomas for more than 30 years. Topical and intralesional steroids are not usually recommended. Their precise mechanism of action is not clear, but it is known that corticosteroids reduce angiogenesis, induce vasoconstriction and reduce vascular permeability. More recent evidence shows increased apoptosis. Oral steroids are recommended in a dose of 2-

5mg/kg for 1-3 months followed by gradual tapering.[11]

In our case oral steroids were given but the patient failed to respond. The patient showed excellent response to intralesional steroids with no local and systemic side effects and is under follow-up with no signs of recurrence.

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