A rare case of complex orbital lymphangiohemangioma

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Background: Complex orbital lymphangiohemangioma is a rare benign vascular lesions. It usually appears as an enlarging mass without specific clinical features and frequently misdiagnosed. This case report highlighted a case of orbital vascular anomalies which presented as intramuscular hemangioma with lymphangioma.

Results: A 12 years old boy with underlying bronchial asthma, presented with painless progressive enlarging swelling over right medial canthal area and right upper lid since age of 6 years old. His best corrected vision was OD 20/50 , OS 20/20. Right eye showed non tender mass at medial canthal area with no skin changes. Anterior chamber and posterior chamber bilateral eye was unremarkable. CT scan showed soft tissue swelling at the medial part of the right orbit involving the medial part of upper and lower eyelid and medial canthal region, measures approximately 2.1cm x 2.4cm x 3.9cm with blocked nasolacrimal duct suggestive of mucocele. Excision biopsy was performed, the intraoperative findings revealed a mass mixed with fibrosis tissue and microcyst with no definite plane with underlying skin and orbicularis oculi muscle. Histopathology examination showed benign vascular lesion likely intramuscular angioma. 3 weeks post operatively, he developed wound breakdown and exploration under GA was done, which intraoperatively showed multiple small slow oozing from remnant of the lesion with multiple cyst surrounding wall of cavity , bluish lesion and small telangiectatic vessels were seen at the upper lid.

Conclusion: Complex orbital lymphangiohemangioma is a rare benign vascular lesion. The recurrence rate is high even after wide surgical excision due to its microscopically infiltrative pattern of diffusion into the surrounding muscular tissue. Long term clinical and radiological follow up are strongly recommended in order to precisely diagnose and treat further recurrences.

Conflicts of interest: The authors report no conflicts of interest.

Keywords: benign vascular lesion, hemangioma, intramuscular hemangioma, lymphangioma, lymphangiohemagioma.

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Background

Vascular anomalies differentiated into two groups based on endothelial chracteristics: hemangiomas and vascular malformations, by Mulliken and Glowacki classifi-

cation 1982. Depending on the type of vessel involved, vascular malformation group was subdivided into high-flow (such as arteriovenous malformation and arteriovenous fistula) and low-flow (such as venous and lymphatic malformation). Intramuscular angioma is a rare special form of hemangioma. It is benign form of tumor in which less than 1% of hemangiomas in the body occur in skeletal muscles, and less than 20% of these occur in the head and neck region arising most frequently in the masseter and trapezius muscle.[1] In contrast to the cutneous hemangiomas of infancy, it never regresses spontaneously.[2] It usually appears as an enlarging mass without specific clinical features and is therefore frequently misdiagnosed.[3] The distinction between lymphangiohemangioma and intramuscular hemangioma is not clear and has been used interchangeably given the overlapping clinical, histologic and imaging features. The recurrent rate following surgical excision in orbital lymphagioma are 52% as reported by Char et al[13], 11% by Gündüz et al[14], while in IMH were reported to be 20% in capillary type, 9% in the cavernous type and 28% in mixed type IMH.[1] This is the reported case of orbital vascular anomalies which presented as intramuscular hemangioma with lymphangioma. To the best of our knowledge, the coexistence of both of hemangioma and lymphangioma is unusual and has been reported only in few cases.

Case history

In 2017, a 12 year old boy with underlying bronchial asthma, was referred for the further management of right orbital tumor which presented with painless progressive, no compressible swelling over right medial canthal area and right upper eyelid since age of 6 years old. Otherwise no history of pus discharge from the swelling, no changing of size during Vasalva maneuver and he has no history of eye trauma. There is no history of malignancy or blindness in his family. On examination, his best-corrected visual acuity was 20/50 OD and 20/20 OS. Right eye showed nontender mass at medial canthal area with no overlying skin changes (figure 1) with subconjunctival multiple cystic lesions medially (figure 2). Otherwise anterior segment and posterior segment bilateral eye was unremarkable.

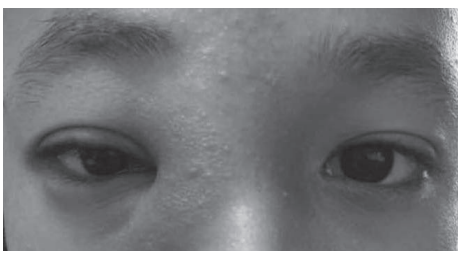

Figure 1: Right medial canthal mass.

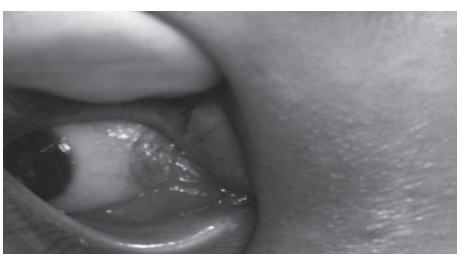

Figure 2: Right eye subconjunctival cystic lesion

Computed tomography (CT) showed soft tissue swelling at medial part of right orbit involving medial part of upper and lower eyelid and medial canthal region, measuring 2.1cm x 2.4cm x 3.9cm with increase density of lateral part of lesion. Right globe and medial rectus muscle are pushed laterally. Streakiness of extraconal fat and slight flattening of nasal bridge with blocked nasolacrimal duct. (figure 3) Right excision biopsy of right medial canthal mass and conjunctival lesion was done. Intraoperatively the tumor size was 2.5cm (width) x 1.0cm (height) and it was mixed with fibrosis tissue and microcyct (specimen A) (figure 4 and 5). It has no definite plane with overlying skin and orbicularis muscle. Multiple cyst of right conjunctival lesion

(specimen B). Nasolacrimal duct was patent. Specimen A and B were given for histopathology examination.

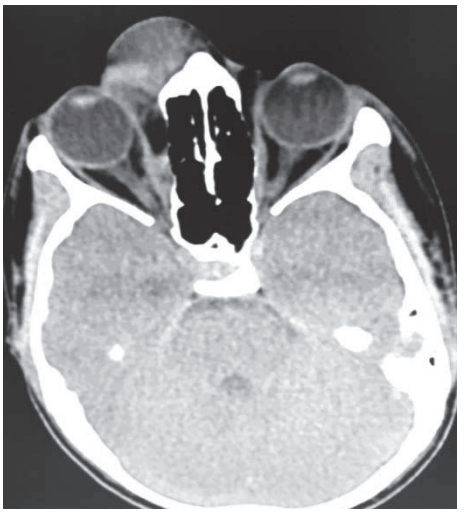

Figure 3: CT findings showed soft tissue swelling at medial part of right orbit involving medial part of upper and lower eyelid and medial canthal region.

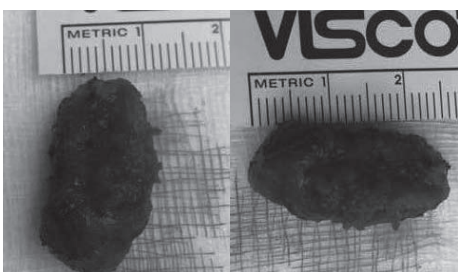

Figure 4 and 5: Macroscopic specimen of the lesion.

Macroscopical examination
A:Specimen consists of a piece of brownish tissue fragments measuring 25x10x10mm. Cut section shows homogenous greyish fragment with areas of hemorrhage. Bisected and submitted entirely in 2 blocks.
B: Specimen consists of 3 fragments of brownish to greyish tissue fragments measuring 2x2x2mm, 4x2x1mm and 5x2x1mm.

Microsopical examination:
A: Sections show fragment of fibrocollageneous tissue interspersed with adipose tissue and skeletal muscles. There are various sized vascular channels present with focal thrombosis. No evidence of malignancy seen. (figure 6)

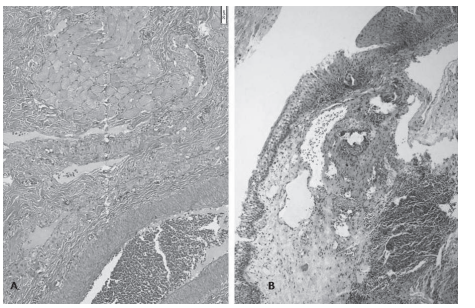

Figure 6: A: Fibrocollageneous tissue interspersed with adipose tissue and skeletal muscles with various sized vascular channels present.
B: Tissue lined by conjuctival epithelium with underlying stroma shows presence of dilated lymphatic spaces and consist of patchy lymphocytic infiltrates is seen between the dilated spaces.

B: Levels show fragments of tissue lined by conjunctival epithelium consists of several layers of columnar cells that contain mucin secreting goblet cells. The underlying stroma shows presence of dilated lymphatic spaces lined by flattened epithelium. Patchy lymphocytic infiltrate is seen in between the dilated spaces. No malignancy seen.

Specimen A was consistent with benign vascular lesion, intramuscular angioma and specimen B was consistent with conjunctival lymphangioma.

Three weeks following the operation, the patient developed wound break down over right medial canthal wound due to the collection of blood underneath the wound (figure 7). Wound exploration and resuturing was done. Intraoperative noted multiple small slow-oozing bleeding from remnant of the lesion with multiple small cystic lesion surrounding of the wall of cavity. A bluish lesion and small telangi-

ectatic vessels at the upper eyelid noted. Postoperatively, there were some residual of the lesions (figure 8).

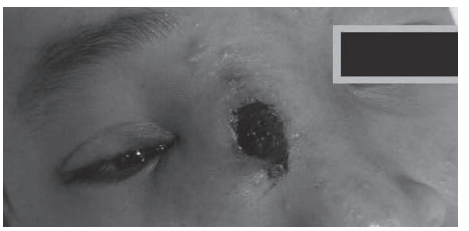

Figure 7:Wound breakdown

Figure 8:After resuturing with minimal residual

During 1 month post operative follow up, MRA and sclerosing therapy was offered to patient and he was further his follow up at different hospital for sclerosing therapy.

Discussion

Intramuscular angioma is the rare special form of hemangioma. It is benign form of tumor in which less than 1% of hemangiomas in the body occur in skeletal muscles, and less than 20% of these occur in the head and neck region arising most frequently in the messeter and trapezius muscle.[1] In contrast to the caverneous hemangiomas of infancy, it never regresses spontaneously.[2] It usually appear as an enlarging mass without specific clinical features and is therefore frequently misdiagnosed.[3]

In the histological classification, intramuscular hemangiomas are subdivided according to their vessel size; capillary, cavernous and mixed form, Beham et al showed in their work that many cases the mixed form prevails[4] as in our case, lesion showed fragment of fibrocollagenous tissue interspersed with adipose tissue and skeletal muscles with various sized vascular channels present with focal thrombosis.

Intramuscular hemangioma often remains undiagnosed preoperatively[5] but its nature may be suggested by MRI, where the tumor will often appear as sharply demarcated, images. This is due to stagnant blood in the larger vessels.[6] Linear areas, isotensive to fat and muscle, are often observed in the lesions representing fibro-fatty septae between vessels. Usually radiological distinction between different types of IMH is not possible.[6,7] Intramuscular hemangioma is poorly defined by CT.[7]

Orbital venous lymphatic malformations, previously known as lymphangiomas, are uncommon and sometimes referred to as no-flow or low-flow vascular malformations. They contain abortive vessels, which spread among normal structures and present as an unencapsulated, primarily thin-walled masses with numerous cystic spaces of different size. They show tendency to spontaneous haemorrhage, resulting in a sudden onset of proptosis combined with periorbital swelling and reduced eye motility, at times leading to optic nerve compression.[8] On imaging they present as an infiltrative, multilobulated mass with poor encapsulated, also intra and extraconal, sometimes harboring calcifications seen on CT. MR imaging is the modality of choice for the evaluation of lymphatic malformations because it best depicts at various components. T1-weighted images best depict lymphatic and proteinaceous fluid, and T1-weighted fat-suppressed images are best for detecting blood or blood products. T2-weighted fat-suppressed images provide improved visibility of component that contain non-hemorrhagic fluid.[9] Fluid-fluid levels produced by hemorrhages of various ages within multiple cysts are almost pathognomonic.[10] In our case, histopathology of conjunctival lesion showed tissue lined by conjunctival epithelium with underlying stroma shows presence

of dilated lymphatic spaces and consist of patchy lymphocytic infiltrates was seen between the dilated spaces.

Treatment of both intramuscular hemangioma and lymphangioma are challenging. Because of the high rate of recurrence, the best treatment for intramuscular hemangioma is total surgical excision[2] as well as for lymphangioma. However the total surgical reception are difficult for lymphangioma. Recurrence rates of IMH were reported to be 20% in capillary type, 9% in the cavernous type and 28% in mixed type IMH.[1] As for the present case, patient developed wound breakdown due to collection of blood underneath the wound and due to incomplete excision of lesion.

A conservative therapy should target the abnormal membranes that make up the lymphangioma, while sparing the adjacent normal tissue through which the lymphangioma infiltrates. Sclerosing therapy has the potential to supply some of these benefits.[11] The idea of using sclerotherapy in the treatment of lymphangioma occurred when it was noted that lymphatic malformations spontaneously involute when they became infected and the infection resolved. Sclerosing agents may have specificity for the abnormal tissues if introduced intralesionally. The first case of lymphangioma treated by sclerotherapy was reported in 1933, using sodium morrhuate. Injection of sclerosing agents has proven efficacy in lymphangiomas in other locations. Some sclerosing agents have been tested in OL, such as sodium tetradecyl sulfate, sodium morrhuate, and OK 432, with different rates of success, although with limited numbers of patient and some local complications, such as pain, swelling, and haemorrhage.[11,15,16] Complete tumour regression was noted in 6 weeks following intralesional injection.[12] There is insufficient evidence demonstrating its efficacy at present. As in our case, patient still has residual lesions after total excision, and he was opted for sclerotherapy.

Conclusion

Complex orbital lymphangiohemangioma is a rare benign vascular lesions. The recurrence rate is high even after wide surgical excision due to its microscopically infiltrative pattern of diffusion into the surrounding muscular tissue. Long term clinical and radiological follow ups are strongly recommended in order to precisely diagnose and treat further recurrences.

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