Case Report: Infantile Orbital Lymphangioma with a Rare ENT Presentation

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Abstract: We are reporting a rare case of 22-year-old Malay female with history of left orbital swelling which was noted since the age of 2 month and treated conservatively. Four years later noted the swelling increase gradually in size involving the bridge of the nose associated with proptosis and left intermittent epistaxis. Examination revealed a soft and boggy swelling at the medial canthus of the left orbit extending to the nasal bridge with mild proptosis. Left intranasal endoscopic examination revealed hemorrhagic spots at the septum with bulging at the lateral wall of the nose and intact mucosa. The radiological examinations with biopsy were consistent with lymphangioma of the left orbit. Orbital Lymphangiomas are rare and it is not reported to present with recurrent epistaxis.

Keywords: Infantile orbital lymphangioma, recurrent epistaxis.

INTRODUCTION

Lymphangiomas are malformations of the lymphatic system. These malformations can occur at any age and may involve any part of the body. Ninety percent occur in children less than 2 years of age and involve the head and neck. Most lymphangiomas are benign lesions that result only in a soft, slow-growing mass. Lymphangiomas are usually treated for cosmetic reasons as it has a rare chance of becoming malignant. Lymphangiomas are rare, accounting for 4% of all vascular tumors in children [1]. The tumor may affect the conjunctiva, the lids and the orbit. Associated similar extra orbital lesions include facial and palatal cystic lesions and intracranial vascular anomalies have been reported. The orbital lymphangioma is located behind the orbital septum and involves the intraconal or/and extraconal space. Usually they will present with proptosis and occasionally with pain due to spontaneous intralesional haemorrhage or to lymphoid proliferation in the connective tissue of the tumour during an upper respiratory infection but it is not reported to present with recurrent epistaxis as reported in our case.

CASE REPORT

S.S.S is a 22 year old Malay female who presented with history of left orbital swelling which was noted since the age of 2 month and treated conservatively at that time. Four years later noted the swelling increase gradually in size involving the bridge of the nose associated with left intermittent nasal bleed, which was spontaneously stopped by conservative therapy i.e. pinching of the nose and ice packing. There was no history of nasal trauma, forceful sneezing, nasal blockage, rhinorhea, facial pain, facial numbness, or reduced vision. There were no other medical illnesses with any known allergies. Clinical examination revealed a soft and boggy swelling at the medial aspect of the left orbit extending to the nasal bridge with mild proptosis. There was evidence of telangiectasic spots at the medial side of the cornea with minimal upper and lower lid swelling. Intranasal examination revealed hemorrhagic spots at left septum with bulge at the lateral wall of the nose with intact mucosa (Figure **1**). There were no polyps seen and the fossa of Rosenmüller was free of mass. Other ENT examination was unremarkable.

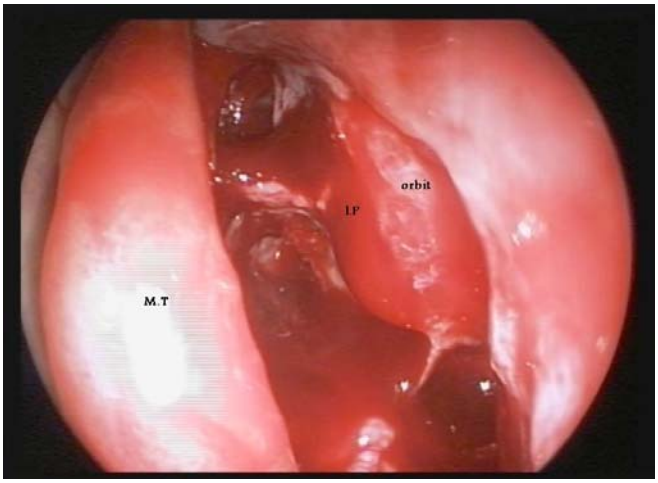

Figure 1: An endoscopic view of the left lateral wall of the nasal cavity.

CT scan revealed a poorly defined multilobulated heterogeneous extraconal mass with intraconal extension involving the left orbit and the upper eyelid

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with multiple cysts like areas with rim enhancement. There was no evidence of calcifications or hemorrhage. There was mild proptosis with the left globe displaced infero-laterally. The rectus muscles were normal in caliber and the left optic nerve showed evidence of enlargement compared to the right with surrounding streakiness (Figures **2**, **3**).

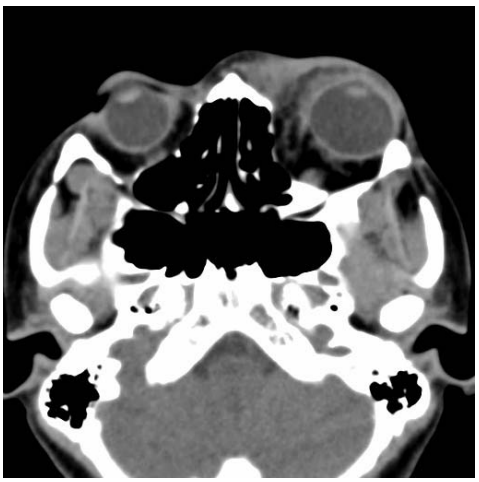

Figure 2: CT scan axial section showing poorly defined heterogeneous extraconal mass with intraconal extension of the left orbit.

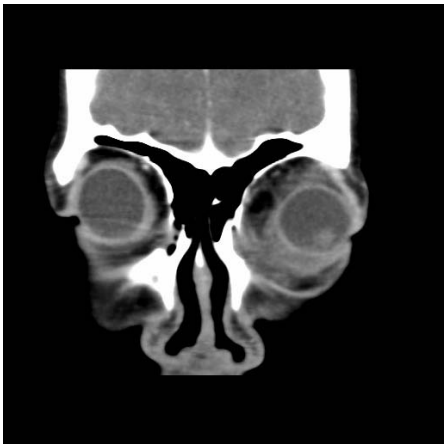

Figure 3: CT scan coronal section showing non inhanced heterogeneous extraconal mass with intraconal extension of the left orbit with displacement of left globe infero-laterally.

MRI revealed poorly defined left lobulated inhomogeneous mass involving almost entire intra and extraconal of the left orbit. The mass is isointense to the extraocular muscle on T1 FAT SAT and hyperintense on T2 and showed evidence of bright inhomogeneous enhancement with contrast. Cystic areas are seen within the mass. The mass insinuates between tissue planes, which makes distinction between the mass and normal extra ocular structures difficult. The left optic nerve is enclosed by the mass (Figure **4**).

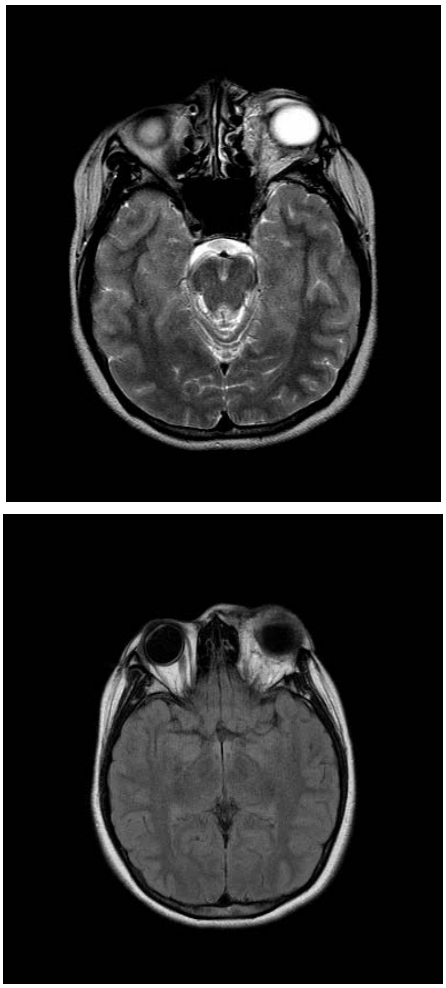

Figure 4: MRI axial section showing poorly defined lobulated inhomogeneous mass involving almost entire intra and extraconal aspect of the left orbit. The lesion is isointense to the extraocular muscle on T1 FAT SAT (bottom) and hyperintense on T2 (top).

All the blood investigation was within normal range. She had an excisional biopsy of the swelling in which the histopathalogical study revealed lymphangioma of the left orbit. Due to the recurrent episodes of epistaxis she had multiple hospital admissions for symptomatic anemia with blood transfusion. A successful Angio-embolization revealed a patchy mucosal telagiectasic vessels from the distal branches of the left internal maxillary artery in which renegade microcatheter was used to cannulate the distal part of the left internal maxillary artery. Polyvinyl alcohol (PVA) 700 microunit were used and injected under intermittent screening until reduction of flow was achieved. The telangiectatic vessels were not seen in the post embolization run.

One month later, she had severe left epistaxis, and underwent examination under anesthesia in which the bleeding point was identified from the medial and inferior wall of the orbit and secured using surgicel and

cautery and no packing was required. Postoperatively the patient is followed up regularly for the past four years in ENT and Ophthalmology clinics with yearly CT scans. Up to date there were no clinical and radiological evidence of tumor extension

DISCUSSION

Lymphangioma of the orbit can be congenital or infantile. It is benign vascular tumor, slowly growing and may not become clinically apparent for months and for years. Associated similar extra orbital lesions include facial and palatal cystic lesions. Lymphangioma are usually diagnosed in the first or second decade of life [2]. Orbital involvement may include the lids, conjunctiva or orbit [2]. There has been no consensus regarding the morphologic nature of lymphangioma. It is believed that the lesions are a variant of venous malformations [3]. According to their haemodynamic relationship, division was made into three groups: no flow, venous flow, and arterial flow lesions [4]. This case is unilateral as in most reported cases. There is no significant race or sex predilection [3]. The prevalence of orbital lymphangioma has been reported to be between 1-3 % of all orbital mass and 10 % of all vasculogenic tumours. Ipsilateral extraorbital localization was observed in our case, which is very rare. In a retrospective study, palatal lesions are found in 13.8 % of cases and maxillofacial vascular lesions have been reported to be 11.8 % of cases [3]. Associated intracranial vascular venous anomaly has recently been reported in 28 % of cases [5] and up to date there is no reported cases for nasal extension with recurrent epistaxis. To establish the diagnosis, CT-scan and MRI of the orbit are obligatory. CT-scan usually shows a multilobulated cystic mass within the orbit (Figures **2**, **3**) and intralesional calcifications may be demonstrated which was absent in our case. MRI is the modality of choice to identify the haemorrhagic cyst as well as the lymphatic component (Figure **4**). Haemorrhagic cyst (acute and sub acute) shows hyper signal intensity on both MRI-T1 and -T2-weighted images whereas lymphatic cysts are hypo-intense on T1-weighted images and hyper intense on T2-weighted images. MRI study is most helpful in documenting fluid level within the multicystic lesion. Pathological examination of a lymphangioma reveals a non-encapsulated lesion with variable sized cystic spaces, lined by flattened endothelial cells. Pericytes and smooth muscles are absent in the vessel wall.

Treatment options for orbital lymphangioma include conservative management, partial surgical resection of the major cyst, needle aspiration, intralesional injection of sclerosing agents and local radiotherapy [5]. Because lymphangioma is a non-encapsulated tumour, which forms a labyrinthic network of interconnected channels with arborization in the orbital tissue, complete removal is not possible without orbital exenteration [1]. The indication for surgery reported in retrospective studies includes acute orbital haemorrhage with compressive neuropathy, severe pain secondary to intraorbital hypertension, cosmetic problem, amblyopia, and diagnostic surgery [6].

CONCLUSION

In orbital proptosis, orbital lymphangioma must be considered as one of the differential diagnosis. Associated extraorbital localizations, particularly intracranial vascular venous anomaly, have to be ruled out. Orbital lymphangiomas rarely present as recurrent epistaxis as shown in our case. A multidisciplinary approach is needed and the treatment has to be as conservative as possible, if vision is not at risk and cosmetically acceptable.

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