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# LYMHANGIOHEMANGIOMA OF FACE: A CASE REPORT

## ABSTRACT

Lymphangiohemangiomas are combined deformities of blood and lymphatic vessels. Lymphangiomas are congenital malformations of lymphatic vessels filled with clear protein rich fluid containing few lymph cells. Malformations may be seen in different combinations of vascular and lymphatic elements. Histologically cystic channels may be filled with blood and named lymphangiohemangioma or hemangiolymphangioma according to the dominant tissue structure present. Lymphangiohemangioma clinically tend to behave as lymphangioma. lymphangiohemangioma is a rare developmental anomaly. Although there are many therapeutic options; the definitive treatment is surgical. Here we present case of a five-year-old child with a lymphangiohemangioma located in the face.

Keywords: Hemangioma, Lymphangiohemangioma, Lymphangioma, Vascular anomalies

## INTRODUCTION

Lymphangiohemangiomas or hemangiolymphangiomas are combined deformities of blood and lymphatic vessels. Vascular anomalies are mainly classified under two headings: vascular tumors and vascular malformations according to the cellular turnover, histology and clinical findings.[1] Lymphangioma are congenital malformations of lymphatic vessels filled with a clear protein-rich fluid containing few lymph cells. It can also occur in association with hemangioma and the combined lesion is called lymphangiohaemangioma. Here we present a case of facial lymphangiohaemangioma leading to facial disfiguration in a 5-years-old girl. Lymphangioma is considered as a benign hamartomatous tumor of lymphatic vessels. It is widely considered as a developmental and congenital lesion rather than a true neoplasia.[1] Malformations may be seen in different combinations of vascular elements such as lymphatic and venous endothelium and cannot be identified as purely one or the other entities. Histologically if these vessels are filled with blood, these mixed vascular malformations are named as lymphangiohemangioma or hemangiolymphangioma according to the dominant tissue structure present.[2]

## CASE REPORT

A 5-year-old girl presented to the Department of Otorhinolaryngology & Head and Neck surgery, Bir Hospital, Kathmandu with a chief complain of swelling of left side of face with disfiguration. The lesion was present since birth and gradually increasing in size. Past medical and family history was non contributory. Intra-oral examination revealed no abnormality. Examination of swelling revealed 10cm x 7cm sized globular swelling on left side of the face over the malar bone and cheek. It was extending up to forehead with facial asymmetry on the left side (Figure I). Facial muscle movements and facial nerve were normal. The surface of the lesion was regular, not attached to the skin, soft in consistency, slightly tender, non-indurated, non-pulsatile with normal appearing surrounding skin.There was no palpable lymphadenopathy. A provisional diagnosis of lymphangioma was made clinically with differential diagnosis of hemangioma. The USG of swelling showed a complex cystic lesion. Blood mixed with fluid was aspirated with wide bore needle from the lesion. FNAC was inconclusive as it contained blood elements only. The CT scan revealed a large heterogeneous mass of 10cm x7cm x 5.0 cm with cystic areas inside. Few enhancing tortuous channels due to enhancement of vascular elements were also seen. Computed tomography scan was helpful in diagnosis because enhancement of the vascular elements within the lesion was demonstrated on the post-contrast images (Figure II and III).

After taking consent, surgical excision was planned. The patient underwent an uneventful excision under general anesthesia.No obvious

feeder vessel was identified during surgery. The surgical site was closed primarily and the excised specimen was sent for histopathological examination. Gross specimen was soft in consistency with reddish brown in color and approximately of size 7.5 cm × 5 cm x3 cm in dimension (Figure IV). Cut section showed multiple cystic areas with variable dimensions. Sections from the received specimen revealed cystic structures with attenuated lining.The cyst wall was composed of fibro-collagenous tissue infiltrated with inflammatory cells like lymphocytes, aggregates of lymphocytes and few macrophages.

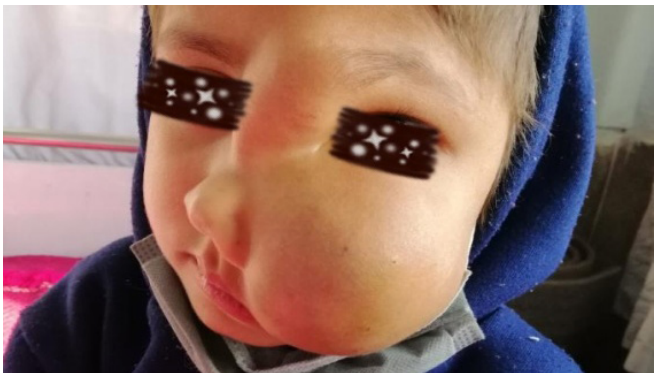

Figure I. Patient profile and clinical presentation

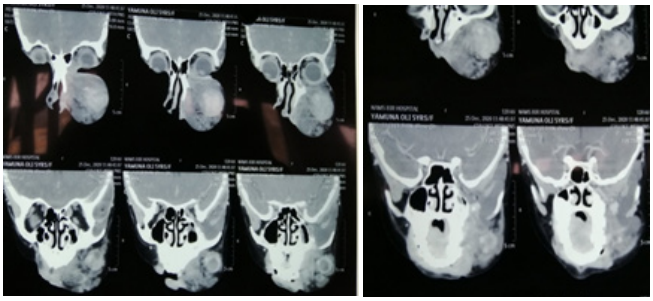

Figure II: CT scan showing large heterogeneous mass with cystic areas. Few enhancing tortuous channels seen due to enhancement of vascular elements (blood vessels inside the mass)

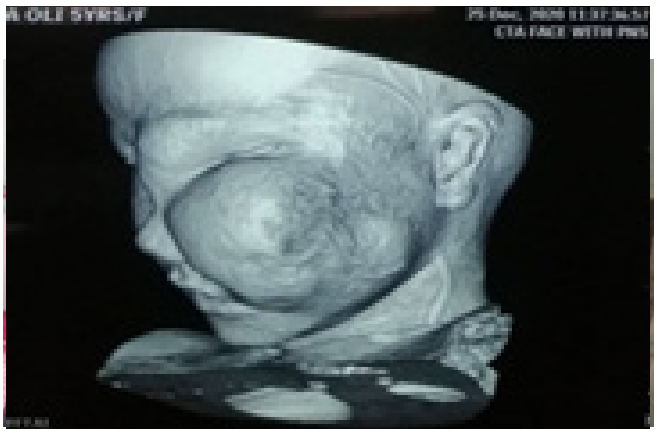

Figure III. 3D reconstruction of the lesion and vascular supply

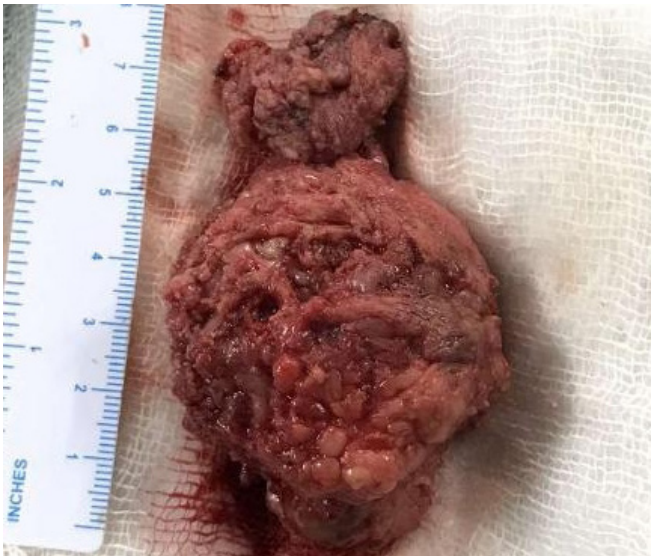


Figure IV. Gross specimen after surgical excision

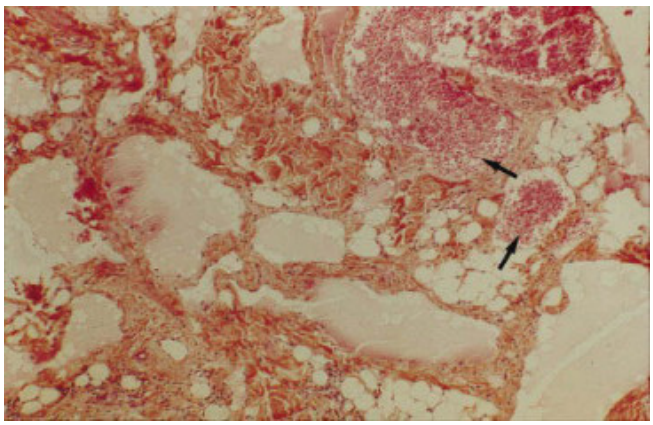

Figure V. Histology: Showing tumor composed of dilated endothelial spaces and smooth muscle cells. Note the combination of lymph-filled and blood-filled spaces (arrow) (Hematoxylin and eosin ×100).

The focal areas showed numerous cystic dilated spaces containing lymph. Many channels were also found with blood cells. Muscles fibers were seen deeper in the connective tissue stroma (Figure V). This histological picture suggested a benign cystic lesion, lymphangiohemangioma. The patient is on regular follow-up without recurrence till date.

## DISCUSSION

Lymphangiomas are usually noted at birth or within 2 years of life. Most commonly occur in the cervicofacial region. The overlying skin is usually normal or may have a bluish hue.[3] Hemangioma is usually seen during infancy and childhood, occurring in 4%–10% of Caucasian infants. Hemangiomas are generally noted within the first 2 weeks of postnatal life. However, they are wide variability in this timing.[3] Hemangiomas mostly

occur in the female. The majority of hemangiomas involve the head and neck.[4] Lymphangioma is classified by the diameter of the vessels into capillary lymphangioma, cavernous lymphangioma and cystic lymphangioma. Structurally, blood vessels are incomplete in hemangiomas, and they are surrounded by hyperplastic cells.[5] Vascular malformations do not show active proliferation, but they consist of ecstatic vessels of either one of the vein, capillaries, arteries, lymphatic vessels or combinations.[5] Lymphangiomas are an abnormality of the lymphatic system, which developed from sequestration of the primitive lymphatic cells. Vessels are capable of accumulating fluids. On the other hand, vessels do not anastomose with bigger lymphatic vessels; therefore, the lymphatic blockage may lead to the cystic appearance of lymphangioma.[6] In our case, looking at the appearance, progression and age of the patient,the diagnosis could be lymphngioma but aspiraton of the lesion revealed frank blood even on ultrasound guidance and created a diagnostic dilemma.

## CONCLUSION

Hemangiolymhangioma is a rare developmental anomaly. Histopathological examination gives definitive diagnosis. Surgical excision is the mainstay of treatment. An early diagnosis and intervention will help inreducing functional, psychological disturbances and cosmetic disfigurement. The patient should be followed up long term to rule out recurrence.

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