Case Study

UNUSUAL SWELLING IN HAND – A LYMPHO VASCULAR MALFORMATION

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Lymphovenous malformations are benign vascular anomalies which arise from defective embryological development of lymphatic system. They are frequently seen in head and neck areas. Here is the case report of a child with a swelling in the hand, which was evaluated and diagnosed as lymphovenous malformation, which was successfully excised and treated. This is presented here because of its rare location and successful treatment.

Keywords: Vascular, lymphatic malformation, children, hand

INTRODUCTION

Congenital vascular malformations are a group of vascular anomalies occurring due to abnormal development of vascular system during the period of embryogenesis. They involve either venous, arterial, venous, lymphatic or capillary systems. These can be defined as disorganised vascular development without new cell growth. They may also refer it as hemangio-lymphangiomas. Incidence is 5 to 12% of all vascular anomalies. There is no sex predominance noted. (1)

Over three fourths of the vascular malformations are seen in cervico facial region. The other observed sites are axilla, chest, gluteus, perineum, retro peritoneum and also seen in mediastinum (2).

The exact etiopathology is not known. There are several theories proposed like connection failure between abnormal endothelial buds and the venous system which it originates from, loss of connection between the buds and the central lymph channels, or pinching out of a proportion of lymphatic channels from the main lymphatic system [3, 4].

The classification of Lymphatic malformations, on the basis of the size of the cysts in the lesion are microcystic, microcystic and combined. Macrocystic lesions measure more than 2 cm and microcystic lesions are those less than 2 cm in size.

CASE REPORT:

5-year-old boy E admitted with history of swelling in the ulnar aspect of right hand noted for 5 months of age. It gradually increased in size and has associated pain for the past 20 days. No history of trauma or fever.

The child was second born to non-consanguineous parents who is completely immunised and developmentally appropriate admitted in neonatal period for respiratory distress. He was evaluated to have Patent Ductus Arteriosus with Coarctation of Aorta. Child had undergone subclavian flap aortoplasty with PDA ligation in a tertiary cardiac centre. Post operative period was uneventful.

The swelling is soft, cystic, warm, tender, fluctuant, not transilluminant in the hypothenar aspect of right hand which measures 5cm*3cm. Distal pulses are well felt and with no bruit. The finger movements are normal. No sensory abnormalities noted. (Fig 1)
Systemic examination revealed a cardiac murmur. Vitals are within normal limits. Other systems are normal.

He was evaluated with imaging. Ultrasonogram wrist and hand showed an ill-defined heterogeneous mass lesion with cystic spaces interspersed between hyperechoic solid areas within the subcutaneous plane on the medial aspect of right hand, suggestive of slow flow lympho vascular malformation.

MRI showed well defined T2 weighted hyperintense lesion with multiple cystic components noted around the little finger in palmar aspect involving subcutaneous and inter muscular plane extending up to the middle finger in between flexor tendons. The lesion measures 3.4 *3.3 cm. The lesion receives feeders from branches of ulnar artery. These suggestive of a possible- slow flow lymphovenous malformation.

The child was assessed by vascular surgeon and planned for excision and biopsy. The lesion was excised and covered with a flap under general anaesthesia. Post operative recovery was uneventful. Wound is healthy. Finger movements are normal. Biopsy confirmed vascular malformation. There were fibrofatty tissue admixed with many congested blood vessels and dilated lymphatics.
He is in regular follow up. There is no residual deformity or vascular deficit. (Fig 2)

Figure 1 showing the swelling of right hand

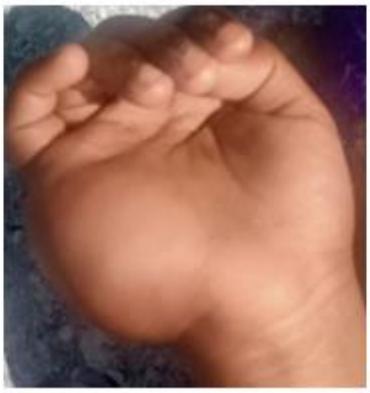

Figure 2 showing post operative picture of right hand

DISCUSSION:

Vascular malformations are inherited anomalies affecting arteries, veins, lymphatics or capillaries, accounting ≈1 to 5% of the population. (1) Majority of the vascular lesions may be sporadic. Inherited malformations may also be a part of complex congenital diseases. The presentation may range from a simple birthmark to even severe life-threatening lesions. Symptomatology may be bleeding, disfigurement or functional deficits. (2)

Rudolf Virchow in 1863 classified vascular malformations into 3 categories like angioma cavernosum, angioma simplex, angioma racemosum.(3) These malformations may be classified as low fow and high flow based on the blood flow characters and type of vessel affected. Low flow lesions may be lymphatic, venous, capillary or combination. High flow lesions are arterial or arteriovenous. This classification can be made radiologically or by physical examination. High flow lesions have warmth and palpable thrill. Flow characteristics helps in planning interventions.

Over 50% of the lesions are venous malformations. The second common lesions affecting 35% are arterial and arterio-venous malformations. Other types are lymphatic malformations (≈10%), and mixed combined malformations (≈5%). (3)
There are two hypotheses proposed for the pathogenesis. First is malformation of lymphatic vascular pathway and second is by cellular endothelial hyperplasia. Mutations of VEGR3 and TIE2/TEK genes are associated with vascular malformations mainly lymphangiomas. (4)

Evaluation of the lesion done with ultrasonogram, MRI, angiography. They reveal a multiloculated cystic lesion with low intensity. It helps to determine the extent of the lesion and to delineate anatomy.
On microscopy, these lesions demonstrate lymphatic and venous channels. Immune histochemistry with CD31 highlights the vascular and lymphatic channels, whereas D2-40 is exclusive for lymphatics.

Management of lymphovenous malformations may not be easy and it depends on the symptoms, size and the extent. Treatment options are observation, aspiration, sclerotherapy and surgical excision. Still, Complete surgical lesion is the standard line of management. (5) Laser coagulation, radiofrequency are other approaches.

CONCLUSION:

Lymphovenous malformations are rare vascular anomalies which need high degree of suspicion. It is a completely treatable condition. Early diagnosis and prompt treatment is necessary for functional recovery.

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