www.jmscr.igmpublication.org *Impact Factor 1.1147*
ISSN (e)-2347-176x

Journal Of Medical Science And Clinical Research
IGM Publication An Official Publication Of IGM Publication

Hemangiolymphangioma of Neck – A Rare Case with Review Report

Authors

Prof. Dr C.V. Kulkarni[1], Dr Parul Nema[2], Dr Harshul Patidar[3], Dr Swati Soni[4] Dr N P Tiwari[5]

(Department of Pathology MGM Medical College Indore M.P. India)

[1]Professor & Head of Department of Pathology M.Y.Hospital, Indore (MP) 452001 India

[2]Resident Department of Pathology M.Y. Hospital, Indore (MP.) 452001 India

[3]Resident Department of Pathology M.Y. Hospital, Indore (MP.) 452001 India

[4]Resident Department of Pathology M.Y. Hospital, Indore (MP.) 452001 India

[5]Resident Department of Pathology M.Y. Hospital, Indore (MP.) 452001 India

Correspondence Author

Dr. Parul Nema

MGM Medical College Indore (M. P) India 452001

Email: *drparulnema@gmail.com*

ABSTRACT

Hemangiolymphangioma is a very rare benign vascular neoplasm, of both the lymphatic and the blood vessels. We report a case of hemangiolymphangioma in a 3 months old female presented with multiple swelling in postauricular region measuring 2.0 x 1.5 cm in greatest dimension, present since birth, gradually increasing in size. On histological examination of excisional biopsy shows features of hemangiolymphangioma.

Keywords- *Hemangiolymphangioma, rare vascular neoplasm, post auricular swelling*

INTRODUCTION

Lymphangiomas are malformations of the lymphatic system, which is the network of vessels responsible for returning of excess fluid from tissues to the venous system. These malformations can occur at any age and may involve any part of the body, but 90% occur in children less than 2 years of age and involve the head and neck. Lymphangiomas have traditionally been classified into three subtypes: capillary and cavernous lymphangiomas and cystic hygroma. This classification is based on their microscopic characteristics. A fourth subtype, the hemangiolymphangioma is also recognized. Capillary lymphangioma are composed of small, capillary-sized lymphatic vessels and are characteristically located in the epidermis. Cavernous lymphangiomas composed of dilated lymphatic channels, generally present at birth, but may appear later in the child's life. These bulging masses occur deep under the skin, typically on the neck, tongue and lips. They vary widely in size, ranging from as small as a centimeter in diameter to several centimeters wide, usually painless, the patient may feel mild pain when pressure is exerted on the area. Cystic hygromas are large, macrocystic lymphangiomas filled with straw-colored, protein-rich fluid.

Hemangiolymphangioma, as suggested by their name, are lymphangiomas with a vascular component. Although histologically it is a benign disorder, local invasion into the muscle, bone, and underlying tissue can lead to severe deformity. It has a propensity for rapid growth and invasion into the adjacent tissues, and to recur locally. It can occur in a variety of anatomical locations, such as head & neck, axilla, abdominal cavity, extremities and urinary bladder.

Case Report

A 3 month old female with multiple bulging masses in posterior cervical region, below and behind the ear present since birth, gradually increasing in size. Physical examination revealed soft, compressible, multiple lobulated masses, with size varying from 2.5 x 2.0 x 1.0 cm to 08 x 0.5 x0.5 cm. Skin over the mass is pinkish in colour, ulcerated with ulcer measuring x 0.8 cm. The patient underwent excisional biopsy.

Gross Examination of the specimen revealed two globular gray white to gray brown soft tissue pieces measuring 2.5 x 2.0 x1.0 cm in greatest dimension with gray white cut surface.. The section studied show stratified squamous epithelium with elongation of rete ridges and bullous lesion on the skin surface . Just beneath the epithelium multiple dilated channels lined by endothelial cells with empty lumen are seen. Fair number of such cystically dilated spaces show endothelial cell proliferation. The intervening stroma is comprising of myxomatous tissue intercepted by spindeloid cells arranged around capillaries with lumen filled with blood, at places empty depicting capillary hemangioma. Focal dense inflammatory cell infiltrate also noted.

Histopathological examination of the excised specimen confirmed the diagnosis of Hemangiolymphangioma

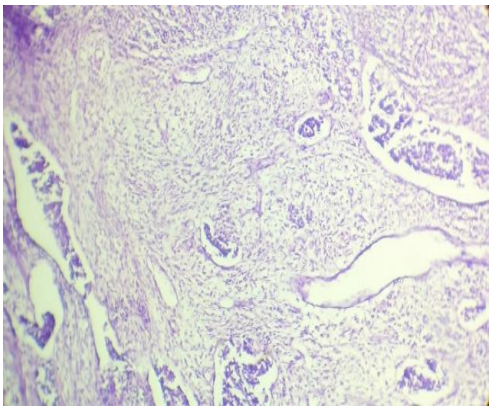

Figure 1- low power view of hemangiolymphangioma

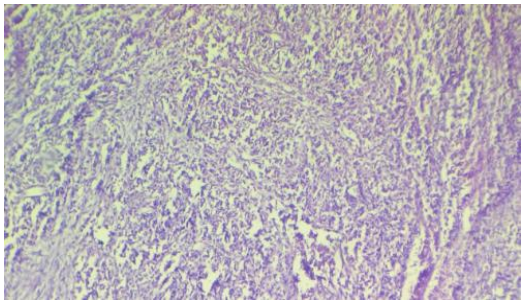

Figure 2- low power view of hemangiolymphangioma

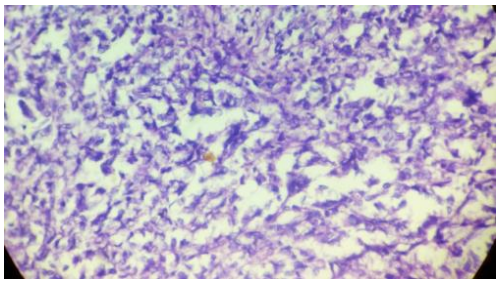

Figure 3- high power view of hemangiolymphangioma

DISCUSSION

Hemangiolymphangioma are very rare benign vascular lesions. They consist in localized centres of abnormal development of the lymphovascular system. Three theories have been proposed to explain the origin of this abnormality. The first suggests that a blockage or arrest of normal growth of the primitive lymph channels occurs during embryogenesis, the second that the primitive lymphatic sac does not reach the venous system, while the third advances the hypothesis that, during embryogenesis, lymphatic tissue lays in the wrong area.

Hemangiolymphangiomas are lymphangiomas with a vascular component. It can occur in a variety of anatomical locations, such as head & neck, axilla, abdominal cavity, extremities and urinary bladder.

Hemangiolymphangioma defined as a proliferation, or network of vascular spaces or vessels of varied nature (lymphatics, capillaries, veins or venules, arteries or arterioles), lined by a benign endothelial lining with intervening connective tissue stroma, and forming a tumor. Hemangiomas and lymphangiomas displayed an interanastomosing network of (usually) small vessels. Some were composed of purely blood vessels (hemangiomas) or purely lymphatics (lymphangiomas), whereas others were composed of an admixture of the two.

The macroscopic appearance of HLA is usually polypoidal. On microscopy, the lesions consisted of a proliferation of varied vessel types (lymphatics, capillaries, veins). The predominant lesional vessel type was lymphatic, capillary or venous. Vascular cavities are dilated and irregular. Diagnosis is based on histopathological demonstration of both vascular component and lymphatic component in the lesion.

REFERENCES

1. Shah KD, Chervenak FA, Marchevsky AM, Rosenberg JC,Berkowitz RL. Fetal giant hemangiolymphangioma: report of a case. Am J Perinatol1987; 4: 212–4
2. Giacalone PL, Boulot P, Marty M, Deschamps F,Laffargue,F,VialaJL. Fetalhemangiolymphangioma: a case report. Fetal Diagn Ther1993; 8: 338–40
3. Goncalves LF, Munoz Rojas MV, Vitorello D, Ternes Pereira E,Pereima M, Saab Neto JA. Klippel–Trenaunay–Weber syndrome presenting as massive lymphangiohemangioma of the thigh: pre-natal diagnosis. Ultrasound Obstet Gynecol2000; 15: 537–41
4. Chandna S, Bhatnagar V, Mitra DK, Upadhyaya P. Hemangiolym-phangioma of the uirnary bladder in a child. J Pediatr Surg1987; 22:1051–2
5. Kaminopetros P, Jauniaux E, Kane P, Weston M, Nicolaides KH,Campbell DJ. Prenatal diagnosis of an extensive fetal lymphangioma using ultrasonograph, magnetic resonance imaging and cytology.