



Hemolymphangioma of Neck– A Diagnostic Dilemma

Authors

Dr N N Dutta¹, Dr D K Das², Dr J Talukdar³, Dr Praneeth kumar Koduru⁴

¹MS (ENT) FICS, Chairman & Managing Director, Downtown Hospital, Guwahati, Assam, India

²Senior Consultant, Department of Head & Neck Oncology, Downtown Hospital, Guwahati, Assam, India

³Senior Consultant, Department of ENT, Downtown Hospital, Guwahati, Assam, India

⁴DNB Postgraduate Trainee, Department of ENT, Downtown Hospital, Guwahati, Assam, India

Abstract

Lymphangiomas are rare congenital or acquired lesions that occur mostly in the head and neck region. Some part of lymphatic system gets sequestered during embryonic development and mostly gets resorbed but in some cases, they dilate and form sac like structures. These are of 3 types mostly – capillary, cavernous and cystic hygroma. There exists a 4th variant also called hemolymphangioma where the sac like dilatations will be filled with blood which may be due to infection or trauma. Also a sudden increase in their size may occur due to infection or trauma. Here, we present the rare variety of lymphangioma i.e., hemolymphangioma in a 7 year old girl where a diagnostic dilemma existed during clinical evaluation.

Keywords: *Lymphangioma, lymphatic system.*

Introduction

Lymphangiomas are rare congenital benign lesions. They occur mainly in the head and neck region. Embryonic sequestration of lymphatics is the main congenital cause.¹ Acquired lesions are caused by traumatic or infective obstruction of the lymphatic system.²

90% of cases present before 2 years of age.³ Only some cases do present at a later stage of life. There are 3 variants of lymphangioma normally – capillary, cavernous and cystic hygroma. Here we present a rare 4th variant called hemolymphangioma in a 7 year old girl where there was a diagnostic dilemma during clinical evaluation which got confirmed only after histopathology.

Case Report

A 7 year old female presented to ENT OPD, downtown hospital, Guwahati with chief complaint of swelling in the right side of the neck since last 5 months. Initially, it was of the size of a peanut, not clearly noticeable but started increasing in size to the present size since last 15 days. No history of trauma, recent sore throat, cough, cold or fever was given.

On examination, a single, non-tender 5cm x4cm (approximate) sized oval swelling is present in the upper one-third of right side of the neck, occupying both anterior and posterior triangles, with well-defined edges (figure 1). Skin appeared normal. Surface was bosselated (grape-like multi loculated feel) and soft to firm in consistency, fluctuant, compressible, non-transilluminant.

Transmitted pulsations were present. Skin over the swelling was not fixed. No tracheal shift noted. On examination of oropharynx, right tonsil was found bulging medially.

A clinical possibility of a branchial cyst or a lymphangioma or a tubercular lymphadenopathy was suspected and the patient was further investigated.

Ultrasonography neck showed multi-loculated cysts with homogenous internal debris suggesting a possibility of lymphangioma or venous malformation. MRI (figure 2) showed 6x5x3.5cm³ multi-loculated cystic lesion in right side of neck deep to sternocleidomastoid involving both

anterior and posterior triangles and extending to right parapharyngeal space pushing right tonsil medially and also displacing carotid sheath medially. Haemorrhagic and thrombotic areas were seen. Findings were suggestive of venous malformation.

So, a possibility of a vascular tumour was suspected and patient was posted for excision under GA (Figure 3). Post-operative HPE report showed it to be a hemangio-lymphangiomatous mass with chronic lymphadenitis (Figure 4). Hence a final diagnosis of a “Hemo-lymphangioma” was made.



Figure 1 Patient presentation

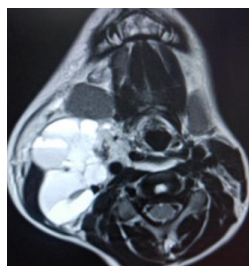


Figure 2 – MRI Neck



Figure 3 – Almost final specimen sent for HPE

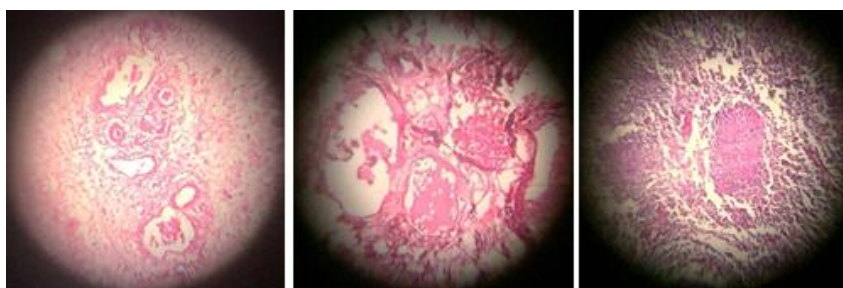


Figure 4 HPE showing collagenous tissue, loose vascular and lymphatic spaces, lymphoid tissue with follicular hyperplasia and infiltration of lymphocytes into fibro-collagenous tissue and septa in the hilum of lymph nodes

Discussion

Lymphangiomas are abnormal cystic dilatations of lymphatic system originating due to congenital sequestration and endothelial fibrillar membrane

proliferation.⁴ Secondary is due to surgery and trauma causing lymphatic damage and poor lymph drainage.

Incidence of hemolymphangioma ranges from 1.2 to 2.8 per 1000 newborns.⁵ According to Landing and Farber⁶, there are 4 types of lymphatic swellings. They are capillary, cavernous, cystic hygroma, and mixed (hemolymphangioma). Hemolymphangioma consists of dilated lymphatic spaces, extravasation of red blood cells, hemosiderin deposition and fibrosis.⁷

Hemolymphangioma of neck is very rare. We could not find any similar case in the pubmed database. Occurrence at other sites such as chest wall, oral cavity, oesophagus, tongue and orbit were found. In the present case of a 7 year old girl, it was in the right lateral part of the neck beneath the sternocleidomastoid occupying both anterior and posterior triangles. Other tumors of common occurrence in this region include branchial cyst, carotid body tumor, cystic hygroma and tubercular lymphadenopathy. Branchial cyst mostly presents in the second decade and moreover rapid growth of tumor in our case did not go in favour of it. Carotid body tumors mostly present in 5th to 6th decade and their rate of growth is very slow, hence could not be considered. Cystic hygroma is a possibility but it was non-transilluminant which went against to it. Tubercular lymphadenopathy was considered a possibility but patient did not have any clinical features of tuberculosis. So, there existed a diagnostic dilemma. But all the investigations suggested it as a vascular tumor. Hence, a surgical excision was done carefully under general anesthesia and the specimen was sent for histopathology which confirmed it as a hemolymphangioma.

Growth of hemolymphangioma can be slow over a period of years or can be aggressive enlarging tumor without any invasive property.⁸ Clinically, they are soft and compressible, loculated in pattern as seen in our case. Complications include hemorrhage, rupture and infection.⁸ MRI shows the extent of the lesion which can help in preoperative planning. Diagnosis is mainly by histopathology. Surgical resection forms the mainstay of treatment. Prognosis is good provided

no tissue is left over during surgical resection. Recurrence rates were found to be 10-27%.⁷ In our case, there was no recurrence at 3 months follow-up. Other treatment modalities include cryotherapy, laser excision and radiotherapy. Sclerotherapy use with OK-432 (picibanil) is becoming most popular these days.⁹

Conclusion

While evaluating neck masses, a rare possibility of having a hemolymphangioma should be kept in mind and can be added as a differential diagnosis.

Conflicts of Interest: None

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