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Case Report

Cystic Hygroma of Wrist

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Abstract

Cystic hygroma is the rare congenital lesion. This is a cystic variety of lymphangioma. Cystic hygroma arising outside of the cervicofacial, thoracic, and abdominal areas are extremely rare. Wrist is a very rare site for occurrence of cystic hygroma. Presentation depends on site of location of cystic hygroma. A case of cystic hygroma of wrist in a child is reported. This child presented with painless swelling of left wrist. Excision of cyst was done. This is first case report of literature reporting occurrence of cystic hygroma on wrist.

Introduction

Cystic hygroma is a congenital lesion[1]. These are large painless soft cystic masses with dilatation of associated anatomic areas. Diagnosis is easy in pediatric age group but seldom presents de novo in adulthood. They develop from defective sequestration of lymphatic tissue from lymphatic sacs during the development of lymphatico-venous sacs[2]. The cystic hygroma shows a predilection for the neck (75%) and maxilla (20%), and the remaining 5% arise in rare locations such as the mediastinum, retroperitoneum, bone, kidney, colon, liver, spleen and scrotum.[3] Complete excision is treatment.

The Case

A three years old boy presented with painless swelling of left wrist since birth (Fig1). General physical and systemic examination was apparently normal. Local examination revealed swelling on left lower forearm and encroaching wrist with dimensions being 10×6.8×2.8 cms. The swelling was globular in shape, nontender, soft in consistency, freely mobile along all its directions fixed to overlying skin but free from underlying structures. Wrist movements were free. Transillumination test was positive. X- ray wrist showed soft tissue shadow. Clear colored fluid was aspirated by fine needle aspiration. A diagnosis of cystic hygroma was made. Excision was done and peroperative findings were muticystic swelling originating from subcutaneous planes of wrist. Large cysts lying superficially and small cysts lying in deeper planes and these were containing clear crystal colored fluid (Fig 2).The swelling was adherent, so meticulous dissection was carried out to ensure avoidance of damage of surrounding vital structures and prevention of recurrence. Histopatholgical examination of specimen was consistent with the diagnosis of cystic hygroma (Fig 3).

Discussion

Cystic hygroma is a rare benign tumor of congenital origin and in child it is obvious from birth[4]. Both sexes affected equally and left side of body predominates[5]. It usually involves neck. Other sites reported for cystic hygroma are axilla (20%), mediastinum (5%), abdominal cavity, retroperitonuem, the scrotum and even the skeleton[6,7]. Jugular lymphatic obstruction sequence, abnormal embryonic sequestration of lymphatic tissue, or abnormal budding of the lymphatics results in cystic hygroma formation[6,8,9]. Vascular endothelial growth factor C (VEGF –C) is postulated to play role in lymphatic malformations[10]. About 78% of patients with cystic hygroma are chromosomally abnormal; 58% have a karyotype associated with Turner syndrome phenotype; in the remaining 20% autosomal trisomies and various structural abnormalities are present[11]. Other syndromes associated with cystic hygromas are Noonan syndrome (autosomal dominant), Pena-Shokeir syndrome, Down syndrome, Robert's syndrome (autosomal recessive) and multiple pterigium syndrome (autosomal recessive, X-linked)[12].

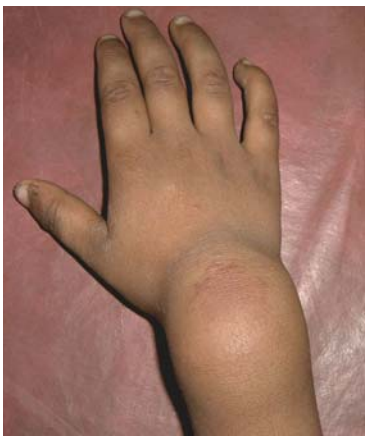

Fig 1: A 10×6.8×2.8 centimeters soft, nontender having fixity to skin, free from underlying structures was arising from dorsum of wrist present right from birth.

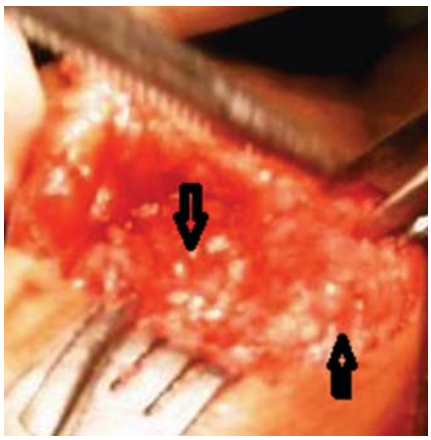

Fig 2: Intraoperative picture with black arrows showing cysts of cystic hygroma.

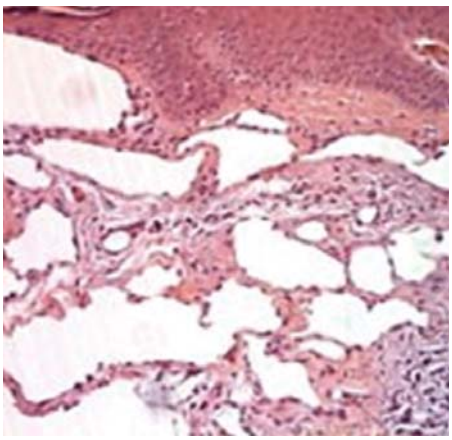

Fig 3: Photomicrograph of cystic hygroma specimen showing dilated lymphatics of different sizes; lined by flat endothelium (HE ×10).

Cystic hygroma consists of an aggregation of cysts resembling a mass of soap bubbles. Cysts range in size from few mm to centimeters and are isolated or freely communicating. Larger cysts are present near the surface, while the smaller ones lie deeper and tend to infiltrate muscle planes[13]. Most cystic hygromas are multicystic with 10% having unilocular cyst; fluid present in these cysts is clear to straw colored being rich in protein and is eosinophilic[14]. Hemorrhage into cyst and nerve compression can presents as parasthesia and pain. Due to rarity of those arising in the extremity, detailed description in literature in regard to extremity is lacking[15].

Diagnosis is usually made by physical examination. Brilliant transilluminiscence aids to differentiate cystic hygroma from other solid masses. Sometimes bleeding into cyst confuses it with hemongioma[16]. In 90%, the diagnosis is usually made before 2 years of age, with aids of FNAC (Fine Needle Aspiration Cytology), ECHO Doppler computed tomography and MRI (Magnetic Resonance Imaging) Scan[17]. On ultrasonography there are classical findings of cyst mass with multiple asymmetric thin walled cysts. Doppler is used to evaluate vascularity of cyst mass and to document relation with adjacent vessels. Computed tomography scan provide information about size, site and density and shows cyst hygroma as non enhancing thin walled masses with near water attenuation value. However it lacks in providing precise diagnosis and its nature.On MRI (Magnetic Resonance Imaging) scan, cystic hygroma has typical solid appearance with low intensity on T1 weighted spin echo images. There is moderate intensity at T2W spin echo image. Contrast enhanced T1 weighted images show a very low central intensity with typical rim enhancement.

Complete surgical resection is the modality of treatment. The characteristics of large size, non invasive infilitration of adjacent tissue planes, close proximity to neurovascular structures and displacement of surroundings sometimes limits its complete surgical excision and attribute to recurrence of certain cysts[18,19]. Complete excision without damage to vital structures is possible in only approximately one third of cases[20]. Other modalities used are OK-432, bleomycin, alcohol, bleomycin and fibrin sealant but are not yielding satisfying results. Injection of hydrocolloid dental impression material is used to remove lymphangioma in adult[21]. Sclerotherapy of lymphangiomas with diluted acetic acid is effective and shorten the treatment period without serious complications[22].

Conclusion

This highlights the importance of clinical history and examination in diagnosing this rare entity which emphasizes that conventional physical signs peculiar for it can substitute unaffordable and unavailable sophisticated tools in countries with limited resources.

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