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Concurrent Swellings of the Scrotum and the Left Knee; an Uncommon Presentation of Cystic Hygroma – Case Report

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Abstract:
Introduction: Cystic hygroma is a benign congenital lymphatic malformation, usually present in infancy and rare cases in adults. It can pose challenges in diagnosis and management.
Case Report: This is a case of cystic hygroma in a 4-year-old boy with painless swellings of the left knee and scrotum from birth, which have been increasing in size. The patient presented with his mother to an outreach program organized by the Ghana section of the International College of Surgeons (ICS) at Worawora Regional Hospital in the Oti region of Ghana. Examination revealed soft, fluctuant, non-tender and transilluminating swellings in the scrotum, the medial aspect and the suprapatellar region of the left knee. Bedside ultrasonography assessment revealed multiloculated cystic masses in the regions mentioned above. Our surgical team performed an excision biopsy of the swellings under general anaesthesia. We found multiple lobulated cystic masses in the scrotum, the medial aspect, and the suprapatellar regions of the left knee. Histopathology examination of the excised masses microscopically showed cystic spaces filled with eosinophilic material with areas of fibrous and fat lobules and also some blood vessels containing erythrocytes. A diagnosis of cystic hygroma was made. Excision was complete, and all wounds healed well over 6 'weeks' follow-up.
Conclusion: The occurrence of cystic hygroma found at multiple sites makes it an interesting case. The creation of awareness can help raise an index of suspicion among clinicians, especially when patients are seen with multiple swellings on any part of the body from childhood.

Keywords: Cystic hygroma, lymphangioma, congenital anomaly, surgical excision, multidisciplinary approach

Introduction: Cystic hygroma is a benign congenital malformation of the lymphatic system due to blockage of developing lymphatic vessels [1], and it is characterized by cystic growths typically found in the head and neck region. Despite being diagnosed prenatally through ultrasound screening, the management of cystic hygroma can pose challenges due to its variable clinical presentations.

Case Presentation: We report a case of a 4-year-old male who presented with his mother at an outreach at the Worawora Regional Hospital of the Oti region of Ghana with progressively enlarging painless swellings of the scrotum, the medial aspect and the supra-patella regions of left knee since birth. There were no local symptoms associated with the swellings, and there were no other significant medical problems or a history of trauma to the left leg and scrotum.

Upon physical examination, we found that the left hemi-scrotum was enlarged, crossing the midline and pushing the right testis peripherally (Figure 1). The swelling was soft, fluctuant, and the light was brilliantly transilluminated. It was neither warm to touch nor tender and measured 10cm x 6cm in size.

There were two similar masses on the medial aspect of the left knee and the suprapatellar region measuring 4cm x 3cm and 6cmx 4cm, respectively (Figure 2). No signs of other congenital anomalies were found.

Diagnostic Evaluation: To support the clinical diagnosis, bedside ultrasonography of the masses confirmed the presence of cystic lesions with thin walls and septations, suggestive of cystic hygroma.

Management: Given the size and location of the swellings, surgical excision biopsies were deemed necessary. The patient underwent surgical excision of the masses under general anaesthesia. Intraoperatively, the multiloculated scrotal swelling was found arising from the scrotal wall with normal left and right processus vaginalis and testes. The swelling extended to the left groin but was completely separate from the cord structures. Some cysts contained clear fluid while others contained sero-sanguinous fluid (Figure 3).

Meticulous dissection was performed to preserve vital structures and minimize the risk of complications. The scrotal mass was first excised, haemostasis was secured, and a drain was left in-situ. The masses on the left knee's medial aspect and suprapatellar region were similarly excised with separate skin incisions and hemostasis secured (Figure 4). All skin incisions were closed with Vicryl 3-0, and dressings were applied.

Histopathological Results: **Histopathological examination of the specimens confirmed** the diagnosis of cystic hygroma, characterized by dilated lymphatic channels lined by endothelial cells containing eosinophilic material. No evidence of malignancy or other pathological features were identified.

Postoperative Course: The patient's postoperative course over six weeks was uneventful (Figure 5). Regular follow-up assessments have been scheduled with the district physician to monitor for recurrence and assess for any functional or cosmetic sequelae. At 12 'weeks' follow-up visit, the patient exhibited normal growth and development, with no evidence of a new swelling.

Figure 1: Enlarged left hemiscrotum crossing the midline and displacing the right testis

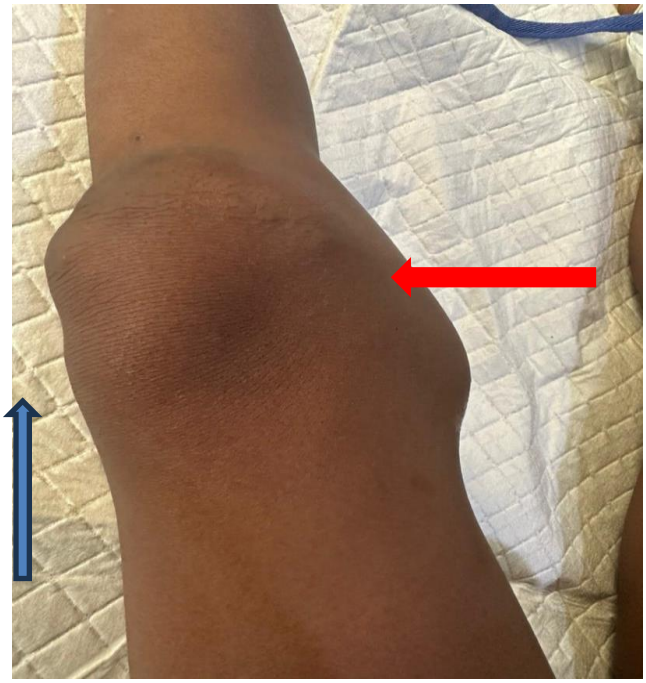

Figure 2: Shows one swelling at the medial aspect of the left knee (red arrow) and one at the suprapatellar region (blue arrow)

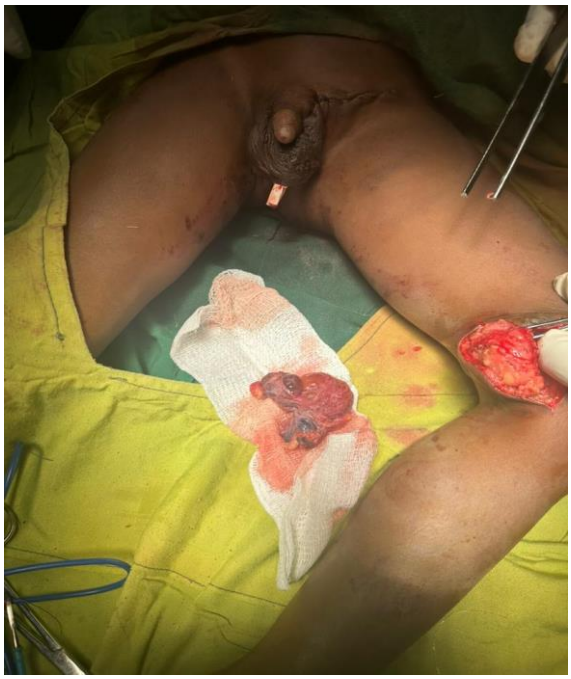

Figure 3: Intraoperative picture after excision of multiloculated scrotal swelling

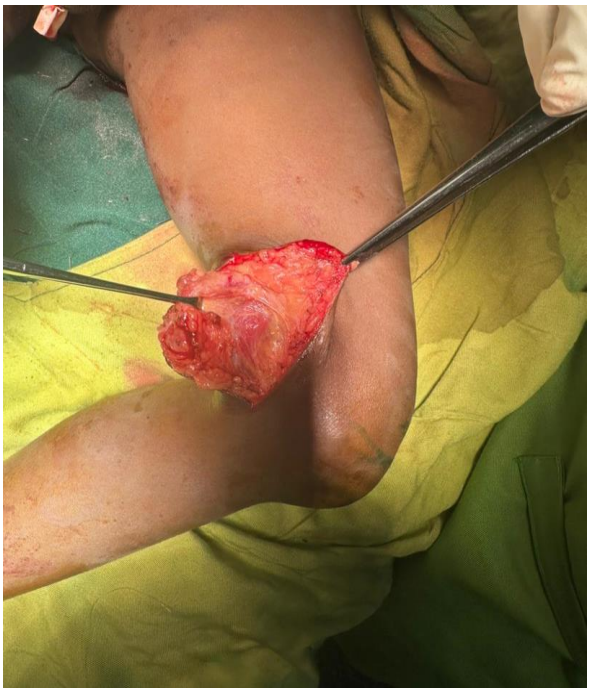

Figure 4: Excision of swellings at the left knee

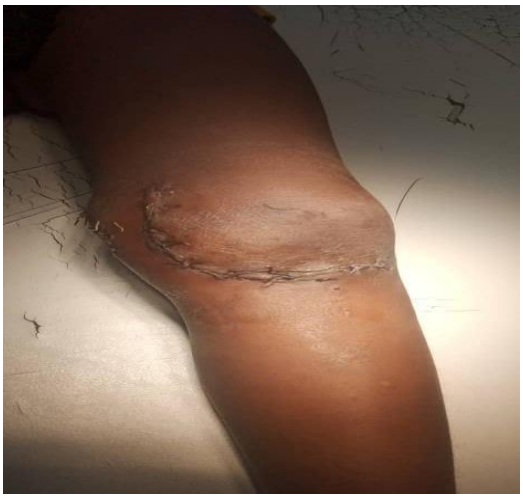

Figure 5 : Healed wound at the left knee seen two weeks post operatively.

Discussion:

Cystic hygromas are congenital malformations of the lymphatic system. They can occur in any region of the body, commonly in the cervico-facial region (75-80%), mediastinum [2], axilla [3] and groin [4]. Occasionally, they may develop in organs such as the upper extremity [5], liver [6], spleen [7], retroperitoneum [8], and mesentery [9, 10]. Singla et al. reported a case of cystic hygroma in the gluteal region [11]. Approximately 50-65% of cystic hygromas are seen at birth and 80-90% by the second year of life. However, cystic hygroma can present in adults, but it is pretty rare, although Al-Ameer et al. reported on rare cystic hygroma of the axilla and breast in adults [3], and Gorai et al. reported a case of cystic hygroma of the left neck in an adult [12].

Cystic hygroma can be diagnosed by ultrasonography at ten weeks' gestation, and the transvaginal route provides superior detail for cervical lesions. Fast-spin magnetic resonance imaging (MRI) can also be used to determine the extent of fetal cystic hygroma. Elevated alpha-fetoprotein levels in amniocentesis fluid have been detected in pregnancies with cystic hygroma. The finding of cystic hygroma is, nevertheless, rare in adulthood. No differences in prevalence between males and females have been reported.

Several mechanisms have been proposed to explain the occurrence of cystic hygroma. Embryologically, these malformations are thought

to arise from the sequestration of lymphatic tissue during the formation of lymphatic-venous sacs. These sequestered tissues fail to connect with the rest of the lymphatic or venous system. Over time, these isolated lymphatic tissues expand, resulting in the cystic appearance of these malformations.

Lymphangiomas are commonly categorized as capillary, cavernous, or cystic. Cystic hygroma can further be classified according to the cysts' size as microcystic, macrocystic, or mixed type. Microcystic lymphangiomas comprise cysts smaller than 2 cm, while macrocystic lymphangiomas consist of cysts larger than 2 cm. The mixed type is characterized by cysts of different sizes [9, 13]. The cyst may be unilocular or multilocular and could be of varying sizes but transilluminates brilliantly.

Typically, cystic hygromas present as painless masses apparent at birth, often prompting concern from parents. Other presentations, such as respiratory difficulties, feeding problems, fever, sudden enlargement of the mass, or infection, may arise due to complications, mostly at late presentation.

Upon clinical examination, cystic hygromas are characterized by their soft, compressible, non-tender, translucent nature and ill-defined. Symptoms may arise when the cyst enlarges to compress surrounding tissue or organs when it may show obstructive symptoms such as dysphagia, dysphonia, and airway obstruction. They do not typically exhibit any vascular sounds.

Ultrasonography commonly reveals a multicystic structure with internal partitions; colour Doppler ultrasound does not detect blood flow within the lesion. CT and MRI scans can give more delineation of the lesion, particularly regarding its extent and relationship with nearby nerves and blood vessels, which is valuable when considering surgical intervention. Fluid aspirated from these cysts may vary in colour and consistency, ranging from milky to serous or serosanguinous. Studies have suggested that fluorescent in-situ hybridization (FISH) can be used to assess lymphatic malformation (LM) in antenatal chromosomal analysis (chromosomes 13, 18, 21, X, and Y). FNAC of cystic hygroma is clear in the majority of cases and milky to haemorrhagic in a few. Histologically, the fluid contains cholesterol crystals, lymphoid cells and endothelial cells. [14]

Diagnosis of cystic hygroma via ultrasound during pregnancy is extensively documented in the literature. This condition is commonly seen in the nuchal region, with about 20% occurring in the axilla and the remaining appearing in other part of the body, such as the mediastinum, retroperitoneum, abdomen, groin, scrotum and bones. Its sonographic appearance is characterized by a multi-septate, thin-walled cystic mass, occasionally showing a more intricate echo texture with both cystic and solid components. Fetuses affected by cystic hygroma can present with associated anomalies, accounting for approximately 62% of cases, including conditions like Turner's syndrome, Down syndrome, Trisomy 18, Trisomy 13, and Noonan syndrome [1, 13, 15]. Biopsy correlation might be necessary for precise diagnosis, mainly when lesions occur in unusual sites, such as the larynx, intraoral cavity, or orbit.

Indications for urgent medical or surgical interventions are respiratory distress, recurrent infection dysphagia, bleeding within the cyst, sudden increase in the size, lymph discharging sinus and obvious disfigurement.

The primary mode of treatment for cystic hygroma is complete surgical excision [16], although sclerosant agents have shown promising results in recent studies. Other treatment modalities include drainage, aspiration, radiation, laser excision, radio-frequency ablation, and cauterization. Surgical excision can be challenging, with potential complications including damage to surrounding structures and incomplete excision leading to recurrence. Aspiration can be performed as a temporary measure to alleviate pressure effects on the respiratory tract and the oesophagus. Sclerotherapy, using bleomycin or OK432, has shown efficacy in reducing the size of cystic hygroma, with minimal side effects, especially in unilocular cysts [9].

Complications of cystic hygroma

Complications of cystic hygroma include potential infection, resulting in increased size, redness,

warmth, tenderness, and fever. Infection covered with intravenous antibiotics may necessitate drainage if it progresses to abscess formation. Other complications include recurrence, spontaneous bleeding within the cyst, respiratory difficulties, dysphagia, and sinuses with lymphatic discharge due to infection or trauma [16].

Conclusion: Cystic hygroma poses diagnostic and management challenges due to its variable presentation and potential complications, including airway obstruction. Prenatal screening and diagnosis allow for early intervention and appropriate counselling of parents. Surgical excision remains the mainstay of treatment, with the goal of complete resection while minimizing morbidity and preserving adjacent structures, although other treatment modalities such as drainage, radiation, laser, radio-frequency ablation and cauterization can also be used in selected patients. Long-term follow-up is needed to monitor for recurrence and assess for any functional or cosmetic sequelae.

Clinical message

Swollen soft tissue in various body regions, notably the scrotum and knee, is an unusual manifestation of cystic hygroma, particularly when it presents without symptoms, as in this case. Nevertheless, conducting a thorough clinical assessment is crucial for clinicians to maintain a heightened awareness of the possibility of encountering a less common occurrence of a typical condition despite contradictory findings on sonography. Consequently, cystic hygroma can be considered a differential diagnosis of swellings in the scrotum and knee with no history of trauma in a child, as illustrated in this case.

Declaration of patient consent: The authors certify they have obtained all appropriate patient consent forms. In the form, the patient's mother has given the permission for his images and other clinical information to be reported in the journal. The patient's mother understands that his name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil

Ethical approval: Based on our institutional policy, ethical approval is not required for case reports.

Source of support: None

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