• Rarely, cardiopulmonary and CNS complications like cardiopulmonary failure, aseptic meningitis, etc. may occur. These are associated with outbreaks caused by Enterovirus 71.

Prognosis:

Complete recovery is the norm in HFMD

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CASE REPORT

An Adolescent With An Axillary Mass

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Abstract

A 13-year-old boy presented with a progressively enlarging right anterior axillary and chest wall mass. He first noted the mass three days after injuring his right shoulder while playing football. When the mass persisted, the possibility of hemorrhage into a soft tissue sarcoma in an adolescent was raised. Computed tomography and magnetic resonance imaging were used to evaluate the mass. Imaging revealed a multi-loculated cystic mass with an internal fluid-fluid level, and enhancement characteristics consistent with cystic lymphangioma and recent traumatic hemorrhage. This diagnosis was confirmed after histologic examination of the resected mass. This case illustrates an atypical adolescent presentation of cystic lymphangioma, manifest after trauma and masquerading as a soft tissue tumor. High resolution diagnostic imaging provided an accurate diagnosis and guided resection.

Introduction

Cystic lymphangiomas, once referred to as cystic hygromas, are congenital lymphatic malformations constituting 6% of all benign lesions of infancy and childhood. (1) Their origin is not completely understood, but it is thought that they occur if the primary lymphatic sacs fail to join the central venous system during embryologic development. (2) The sequestrations of lymphatic tissue form cysts that are composed of dilated endothelium lined channels. Most masses are identified by two years of age with approximately 50% noted at birth. (3) Approximately 75-90% are found in the head and neck, a region with a complex lymphatic system, with lesions also occurring in the axilla, mediastinum, chest wall, abdomen, inguinal region, and extremities.(3) Rapid or intermittent enlargement can occur secondary to infection or, as in the case presented here, injury related hemorrhage.

Case Report

A 13-year-old boy with no significant past medical history presented with a progressively enlarging right axillary mass extending to the right chest wall. He had been playing football and fell onto his right shoulder. Three days later he noted the presence of a small pea-sized mass. Within five days the mass had increased in size to approximately 5 x 9cm. He reported having intermittent numbness and slight weakness in the right hand with use.

On initial physical exam, the patient was noted to have a soft, mobile, anterior axillary mass that was approximately 6 x 10cm and was nontender to palpation. His upper extremity strength was equal bilaterally and his sensation was intact. Bilateral radial pulses were normal.

The mass was evaluated with computed tomography (CT) revealing a 9.6 (craniocaudad) x 4.4 (transverse) x 5.7 (anterior-posterior) cm multiloculated cystic mass within the upper right lateral chest wall and axilla, with very faint peripheral and septal enhancement most consistent with a large lymphatic malformation or cystic hygroma. (Figure 1a). The adjacent ribs, muscles, neurovascular structures and pleura were normal in appearance. Axial magnetic resonance imaging (MRI) revealed a multiloculated, multiseptated cystic mass with a distinct fluid-fluid level indicative of recent bleeding into the cyst. (Figure 1b). On post-gadolinium images, the mass demonstrated peripheral rim enhancement as well as enhancement of internal septations. (Figure 2). The MRI assisted in identifying the relationship of the mass to the subclavian vein and long thoracic and thoracodorsal nerves. A diagnosis of cystic lymphangioma was made based on imaging. Due to parents' and referring providers' concerns for a hidden malignancy and risk of future infection, elective resection after resolution of acute swelling was recommended.

During the two weeks between presentation and elective operative resection, the mass decreased slightly in size and became more firm. The mass was entirely resected sparing the neurovasculature, and the patient did well postoperatively. The gross specimen was a large multiloculated cyst with serosanguinous fluid. (Figure 3). Histopathology was diagnostic for a lymphangioma. (Figure 4).

Figure 1: 1a: Coronal reformation from a contrast enhanced computed tomography scan demonstrates a cystic mass in the right axilla with faint rim and septal enhancement. 1b: Axial fat saturated fast spin echo T2 weighted magnetic resonance image demonstrates a multi-loculated cystic mass in the right lateral chest wall with internal fluid-fluid levels.

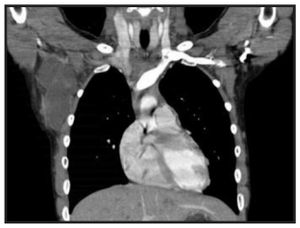


Fig 1a

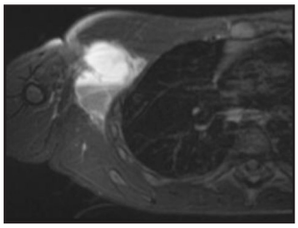


Fig 1b

Figure 2: Axial (A) and coronal (B) fat saturated post-gadolinium T1 weighted images demonstrate circumferential rim enhancement of the mass, and enhancement of the internal septations of the mass. There is no enhancement of the internal cystic fluid.

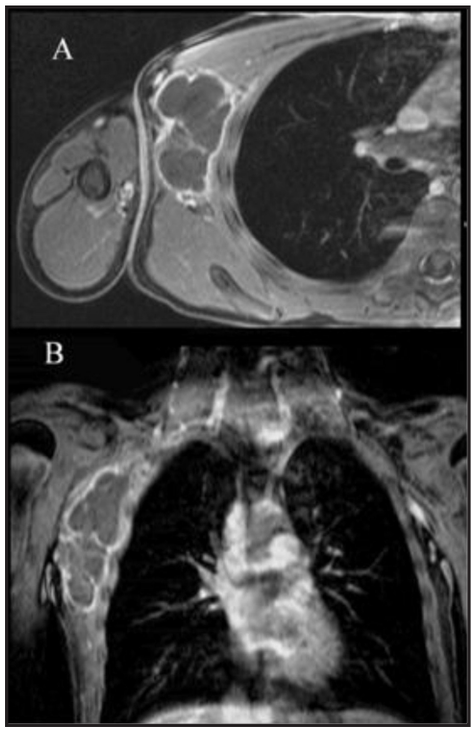


Figure 3: Resected specimen, axillary cystic lymphangioma (A) Cut surface of cystic lymphangioma (B)

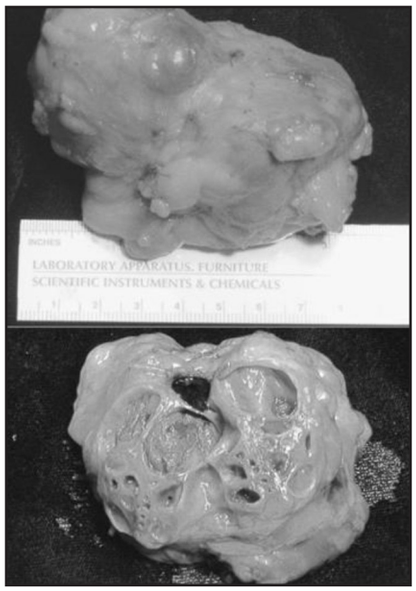
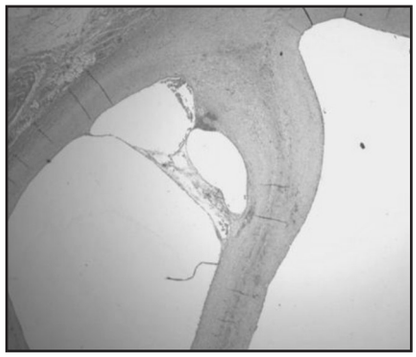


Figure 4: Histologic sections showed large thin-walled cysts most of which were devoid of luminal contents and which were lined by variably attenuated endothelial cells.



Discussion

Cystic lymphangiomas are benign lesions, but often involve extensive invasion of tissue planes by daughter cysts at the periphery of the primary cyst. (1) Treatment options include aspiration; injection with sclerosing agents, radiation and observation, but the most favored management option is surgical resection. Lymphangiomas recur after resection in as many as 10% of cases, and the recurrence rate after infection or repeat excision is even higher. (2) Because the most common cause of recurrence is inadequate tumor removal, a detailed delineation of the size and extent of the mass for successful surgical management is useful. (1) However, vital neurovascular structures should be spared since the lesion is not malignant. Sclerotherapy with OK-432 has given good results, but the highest success rate is still obtained with surgical excision. (5)

Typical MRI findings include high signal intensity

and multiple cysts with well demarcated margins on T2-weighted images and low signal intensity on T1-weighted images. (1) Although the T1-weighted images usually demonstrate low signal intensity, the cyst may demonstrate mixed or even hyperintense signal depending on the age of the hemorrhage. (4) In this case, the MRI did reveal a heterogeneously hyperintense mass on T2-weighted images and a significant fluid-fluid level contained within the cyst with relatively more hypointense fluid layering posteriorly in the supine patient. On post-gadolinium T1-weighted imaging, cystic lymphangiomas typically show peripheral rim enhancement as well as enhancement of internal septations. The mass in this case was determined to be a large lymphatic malformation with associated internal hemorrhage.

As discussed, these lesions typically present prior to age 2 years. This child's presentation at age 13-years-old was atypical, but enlargement after injury in older patients has been reported. (6) Fung, et. al. described in their study that MRI produced highly detailed images that proved to be diagnostic and predictive of intraoperative findings, as was evident in this case. (1) Hemorrhage into the lymphatic malformation and the distinct appearance on MRI led to high confidence in the diagnosis and strategy of resection.

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CASE REPORT

Congenital Fusion Of Maxilla And Mandible (Bony Syngnathia)

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Abstract

Congenital fusion of maxilla and mandible as an isolated occurrence is very rare. Syngnathia is often associated with other intra oral and maxillofacial anomalies. We present a rare condition of isolated complete bony fusion of maxilla and mandible in a neonate without any other associated anomaly.

Introduction

Congenital craniofacial defect comprise 20% of congenital birth defects. (1) The fusion defects can present in a wide range of severity from single mucosal band (synechiae) to complete bony fusion (Syngnathia). Only 25 cases of syngnathia have been reported in literature. (2) We present a rare condition of isolated complete bony fusion of maxilla and mandible in a neonate without any other associated anomaly.

Case Report

A full term neonate 16 hours of age, weighing 3770 grams was referred to our medical newborn unit, for management of birth asphyxia. This was the first-born baby of consanguineous parents. Mother had a healthy gestation period and no occurrence of any illness, trauma or drug use. Baby was delivered by caesarean section and had not cried since birth. On admission baby was lethargic, tachypneic with no cyanosis or grunting. Physical examination revealed hypoplasia of mandible. There was severe trismus and mouth could not be opened even to pass an oropharyngeal tube. There were no other obvious external anomalies. Cardiovascular and respiratory systems were normal. Blood sugar, calcium and renal parameters were normal. Chest X ray was normal. On the second day of admission baby developed seizures which was controlled with Inj. Phenobarbitone. Baby's respiratory distress worsened and at 56 hours of life oxygen saturation could not be maintained with oxygen hood. Baby was started on nasopharyngeal CPAP with 5 cm H2O pressure and there was a brief improvement in oxygen saturation.

Radiography of the skull and facial bones revealed bilateral bony fusion of maxilla and mandible from anterior to posterior ends. In view poor respiratory efforts and declining saturation blind nasal endotracheal intubation was done to maintain oxygen saturation. ENT surgeon secured the airway with an emergency tracheostomy and baby was mechanically ventilated. Saturation was maintained between 85-87% for 24 hours. Repeat X-ray chest revealed bilateral infiltrates on the third day of hospitalization. Baby developed progressive desaturation and succumbed at 78 hours of hospitalization. Postmortem examination revealed