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Bilateral Cervical Cystic Lymphangioma and Bilateral Plunging Ranula dilemma in differential diagnosis: Or both?

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Abstract

Background / Aim: Cystic lymphangioma occurs in infancy in most cases and occurs usually in the neck. Ranulas are rare pseudocysts that are mucos retantion from an obstructed sublingual gland and located in the floor of the mouth, and it usually occurs congenitally. Plunging ranula as a complicated ranula is herniation of the pseudocyst through the mylohyoid muscle and present as a cervical swelling. Management of plunging ranula is variable and definitive treatmant is usually surgical. In here, this rare case with complicated bilateral cystic lymphangioma and bilateral plunging ranula was presented to review the approach of the differantial diagnosis and treatment.

Case Report: Bleomycin was administered to a 44x22 cm left cervical cystic lymphangioma of a girl neonate at three weeks of age which born with a prenatally diagnosed left cystic lymphangioma. The left ranula, which developed in the third month, was incised and marsupualized. She had a left cervical cystic mass growing at the age of one year and recurrent ranula, and the ranula was re-marsupualized, but the left cystic lymphgangioma shrank and persisted. She presented with bilateral plunging ranula and bilateral cystic lymphangioma, which started suddenly at the age of two and started growing in 15 days. After US, Doppler US and MRI, bilateral cystic lymphangioma and plunging ranula were excised. Histopathological examination was found to be compatible with lymphangioma. Follow ups is normal in the 1st year postoperatively.

Conclusions: Having plunging ranula with cystic lymphangioma and bilateral presence is a very rare condition. It should always be kept in mind that ranula or cervical cystic lymphangioma, which appears simple at first attempt, may recur or develop into plunging ranula. In all cystic lesions in close proximity to all cervical submandibular salivary glands; whether or not cystic lymphangioma or plunging ranula is considered; determination of amylase into the cystic aspiration flued may be helpful in planning follow-up and treatment in cases with a complicated course.

Introduction

Cystic lymphangioma is a congenital hamartamous malformation of the lymphatic system that typically presents during infancy, and has a tendency to grow unless it is complatly extirpated (1) (2). Lymphatic malformations are classified as microcystic (diameter 1 cm), macrocystic (diameter >1 cm) and combination thereof. These classification is also useful therapeutically as size determines whether or not the cystic cavity can be aspirated or compressed (3). Ranula is a rare pseudocyst that is mucos retantion from an obstructed sublingual glandand located in the floor of the mouth. It usually occurs congenitally and diagnosed in early adulthood or rarely result from intra oral trauma (4). Plunging ranula is herniation of the pseudocyst through the mylohyoid muscle, the most

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common presentation is unilateral, painless, progressive cervical swelling (5).

Clinical evaluation and imaging is important for differential diagnosis of cystic lymphangioma and plunging ranula. When ranula and lymphangioma are together, differential diagnosis could be difficult, and treatment of it can be complicated. Treatment options include aspiration, sclerotherapy and surgery.

Here, we aimed to draw attention to the follow-up and treatment of a girl with bilateral cystic lymphangioma and plunging ranula.

Case Report

A 13 days of female baby was admitted to our pediatric surgery department due to cystic submandibular lesion in left side. In history, a prenatal diagnosis of left cervical cystic mass was available on prenatal US. In her physical examination, a cystic mass lesion reaching approximately 2 cm in size, soft and slightly mobile, non-tender, non-pulsatile, without signs of inflammation were found in the left cervicomandibular region. In US / Doppler US, a lobulated dence cystic lesion of 47x22 mm in size with multiple incomplate septa and internal debris without vascularisation was determined in left servical submandibular area, and a lobulated cystic lesion of 17x7 mm without vascularisation was located in the right parotid inferior. Firstly aspiration of cystic fluid were performed, but bleomycin injection was not given because of the hemorrhagic aspiration. The cytology result was benign.

Two months later the left cervical lesion was reach 48x36x59 mm, left submandibular gland was dilated and tortioused, in parotid space was anechoic, avaskuler cyst 14x10 mm in size on US. Bleomycin was given into the cyst after the puncture of the cystic fluid, but fever and whole body rash were seen in first day; patient was hospitalised.

At the third month, a plunging ranula in the left side floor of the mouth that appeared to be associated with the left cervical cyst 15 mm in size and extended to left parotid space and maxiller space were determined on US. Then ranula was incisioned and marsupialised.

At the sixth month, the left cervical lesion size enlarged, and a cystic lesion of 17.8x10.7x9.2 mm in size with thin internal septa which located in the right submandibular gland space on US. Because of intimate associated with vital structures such as nerves and major vessels of the head and neck; the decision of follow up were made.

At the 16th month ultrasound examination, multilocule cystic lesions of 21x39 mm, 16x27 mm, 26x36 mm in size with thick wall in places multiple septa, containing heterogeneous, hypoechoic components, located in the superior of the thyroid gland, extended to medial of right parotid gland to left parotid gland. Left side floor of the mouth was a recurren cyst of ranula of 20x10 mm in size, so ranula incision and marsupialisation were performed as second time (Figure 1).

At the age of 26 months, ultrasound and magnetic resonance imaging (MRI) of the neck were performed as a result of the rapid growth of the submandibular area in the last 15 days (Figures 2 A,B). On ultrasound, a septated dense cystic lesion measuring 65x23 mm was seen in both submandibular regions, related to each other in the midline. MRI showed a bilobe mass measuring 49x56x64 mm, located in the submandibular space, and it passes through

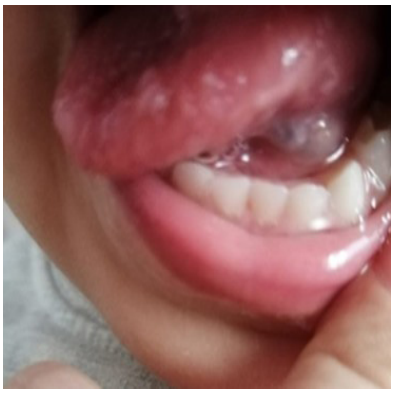

Figure 1: *A cystic mass are seen in sublingulal left side.*

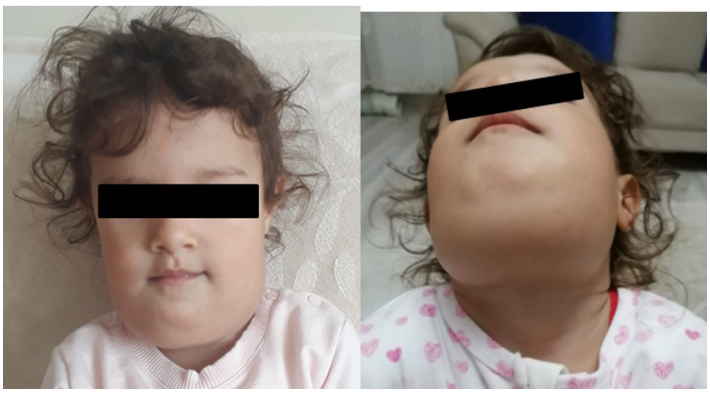

Figure 2 A,B: *Bilateral submandibuler swelling happened in last 15 days.*

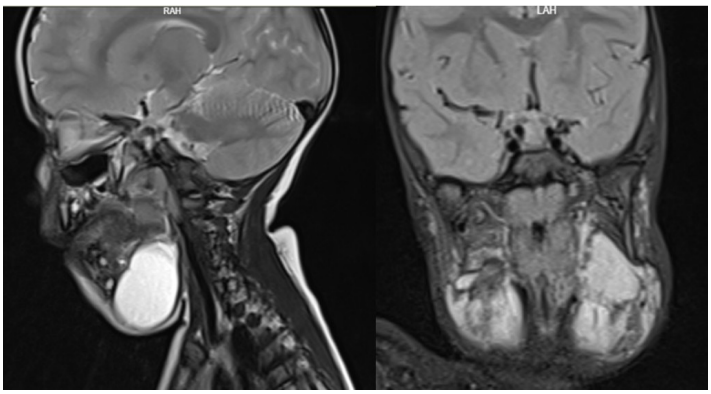

Figure 3 A,B: *MRI showed a bilobe mass measuring 49x56x64 mm, located in the submandibular space, and it passes through the mylohyoid muscle in the neck region and extends into the mouth.*

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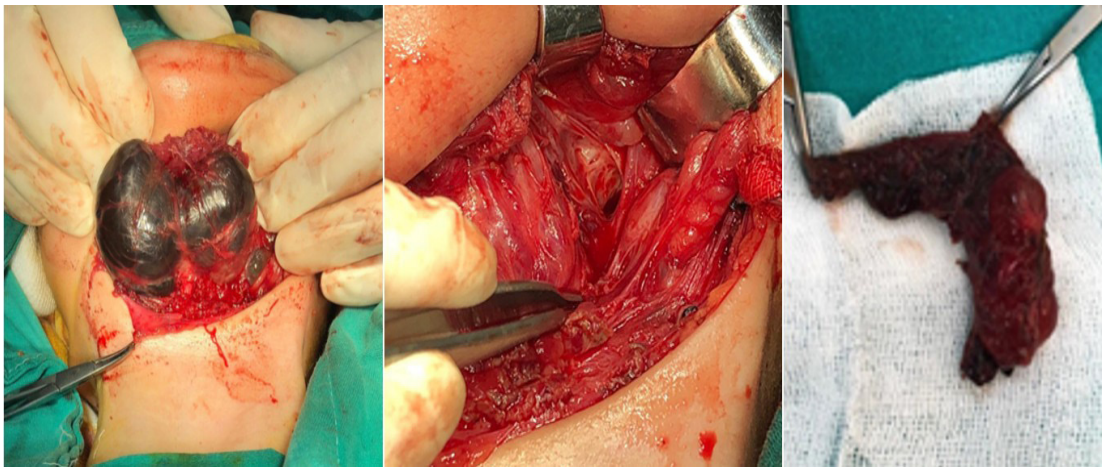

Figure 4 A,B,C: *A: Bilateral submandibular cystic lymphangioma was excised almost completely with the partial excision of the left submandibular salivary gland. B: 1 cm' deep cyst wall could not be completely removed. C: Excised cyst was seen.*

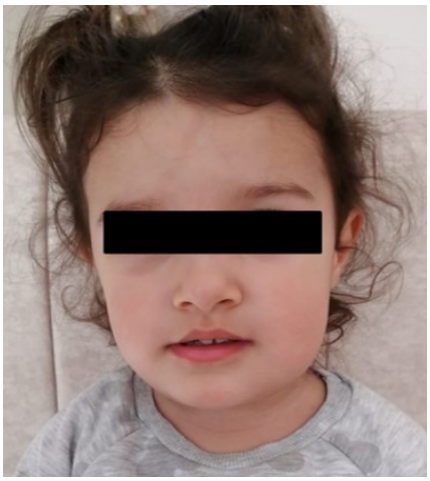

Figure 5: *Postoperative 3th monts a mild palpable mass on the left parotid was detected.*

the mylohyoid muscle in the neck region and extends into the mouth (Figures 3 A, B). There were cystic collections and edema in the left and right parotid glands, except ranula. Tumor markers (b-HCG, CEA, VMA, AFP) were found normal ranges.

The patient was re-evaluated with the otolaryngologist; and it was decided to operate with together. Under endotracheal general anesthesia, a suprahyoid 10 cm collar incision was made, and bilateral submandibular cystic lymphangioma was excised almost completely with the partial excision of the left submandibular salivary gland. Only, 1 cm deep cyst wall, which is tightly adhered to the parotid gland and neurovascular bundle of neck only on the left, could not be completely removed; therefore bleomycin and 30% dextrose were injected into the cyst wall remaining in the deep parotid area on the left (Figures 4 A, B, C). The operation was terminated without complications; The patient was discharged on the fifth day. Histopathological examination was consistent with a lymphangioma.

In the physical examination three months after the operation, a mild palpable mass on the left parotid was detected (Figure 5). On ultrasound reported cystic lesions of 27x9.5 mm in size located in the right submandibular area and 17x10 mm in size with septation loceted in the left submandibular area. Follow ups is normal in the 1st year postoperatively.

Discussion

Cystic lymphangioma is seen in 80-90% at birth, in 30% of infants within one year of birth in 50-60% within two years (6). The most sign or symptom of lymphangiomas is the presence of a mass. Most lesions are recognized early because of their size and associated symptoms of respiratory obstruction and problems with feeding, which are the another common symptoms (7). Cystic lymphangioma of neck usually enlarge rapidly because of an upper respiratory infection (8).

Imaging is very important for clinical diagnosis and for surgical planning of cystic lymphangioma.

Ultrasound is commonly used to investigate a neck mass. Ultrasound enables differentitiation between cystic and solid masses and identifies the anatomic location, association to other structures, and allows accurate needle aspiration or biopsy (9,10). Imaging findings of plunging ranula contain a unilocular large mass, lack of internal septations, a smooth capsuleand location extrinsic to the submandibular gland (11). Cervical US and Doppler US can distinguish between cystic and solid masses and show whether there is vascular circulation. So that the differential diagnosis of cystic lymphangioma from hemangioma can be made. Ultrasound, Doppler US, and ultrasound with fine-needle aspiration cytology, computed tomography or magnetic resonance imaging (MRI) may be used for diagnosis and differential diagnosis all cystic lesions (12,13); however, MRI is the ideal examination in neck cystic lymphangiomas to imaging its association with the adjacent organ and neck vascular nerve bundle (14).

In our case who was followed up for bilateral

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submandibular cystic swelling in the prenatal period, our first diagnosis was left cystic lymphangioma because of the presence of a left submandibular 47x22 mm lesion on the first postnatal US and the absence of a ranula. However, sporadic coexistence of ranula and left cystic lymphangioma was considered in the patient, since the left ranula developed first in the following months and remained uneventful until the age of two after ranula incision and marsupialization. Therefore, until the age of two, follow-up was done with US and Doplerr US.

First line treatment for cystic lymphangioma is complate cystectomy and injection of a sclerotic agent such as bleomycin and or radiotherapy (15,16,17,18). Local recurrence is possible if there component is incomplate. Large and complex lesions are usually related with vital structures such as cranial nerves and major vessels of the neck. Multiple-staged excisions and debulking procedures are often needed (8).

Many methods of treatment for ranula has been suggested and include aspiration of cystic fluid, sclerotherapy, marsupialisation, excision of the ranula only, excision of the ranula and the ipsilateral sublingual gland (19). The lingual nevre and submandibular duct will be at risk for injury particularly in infant patients because of near to the sublingual gland (20). In plunging ranula, the mucus collection is in the submandibular and submental area of the neck with or withouth an connected intraoral collection (20).

Cases which clinical evaluation and MRI can not differentiate a plunging ranula from other lymphatic lesions; fine needle aspiration and amylase measurement are important. Elevated levels of salivary amylase are characteristic to plungingranula fluid (21). Diagnosis is definitely established with fine-needle aspiration and demonstratetion of mucus and high amylase contenent (11). In our case, during the first bleomycin injection for cystic lymphangioma, no characteristic was detected in the intracystic aspiration fluid; however, since there was no ranula in the first application, amylase was not tested in the cyst fluid.

When the sublingual gland is complately excised, recurrence rate is 1% and if complate excision is not achievedit can be higher than 50% (5). Complate cystectomy is difficult in children when the cyst wall is thin and the lesion has infiltrated deeply into areas surrounding important cervical organs or has adhered them; this causes severe complications and recurrence (9,22,23).

In our case, the shrinkage and re-growth of the cyst after the first bleomycin injection suggested that there was a recurrence. When the two-year-old presented with bilateral prominent submandibular swelling and bilateral ranula, it was thought that our patient may have cystic lymphangioma with plunging ranula; this time not only US and Doppler US; at the same time, MRI was performed to show the borders of the cystic lesion, its neighborhoods, salivary glands and its relationship with the neck vascular nerve bundle. Indeed, in our case, it was detected that plunging ranula and cystic lymphangioma were bilateral, and she was operated with a multidisciplinary approach.

In conclusion; having plunging ranula with cystic lymphangioma and bilateral presence is a very rare condition. It should always be kept in mind that ranula or cervical cystic lymphangioma, which appears simple at first attempt, may recur or develop into plunging ranula. In all cystic lesions in close proximity to all cervical submandibular salivary glands; whether or not cystic lymphangioma or plunging ranula is considered; determination of amylase into the cystic aspiration fluid may be helpful in planning follow-up and treatment in cases with a complicated course.

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