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CASE REPORT

Congenital Ranula with Caroli's Syndrome in an Infant: A Rare Case Report

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A B S T R A C T

Ranulas are cystic dilatations in the floor of the mouth and a result of obstruction of one of the sublingual salivary glands. It is basically a retention cyst. Ranula may be congenital or acquired. Caroli's disease is a rare communicating segmental or diffuse dilatation of the intrahepatic biliary tree, Cholangitis, liver cirrhosis and cholangio-carcinoma are its potential complications. Congenital ranula in newborn infants is a rarity and thus there is a marked paucity of the literature on the cited subject. This article is an interesting case report of a patient presenting with ranula in a rare syndrome called Caroli's syndrome.

***Key words**: Congenital ranula, Infant, Salivary gland, Caroli's syndrome*

Introduction

The term 'Ranula' is derived from the Latin word 'Rana' and is described as the blue translucent swelling in the floor of the mouth resembling a frog's abdomen.[1-3] A simple ranula can be either a mucus retention cyst or more commonly a mucus extravasation pseudocyst which is confined to the floor of the mouth. A plunging or cervical ranula is a mucus extravasation pseudocyst arising from the sublingual gland and presents as a swelling in the neck. It may appear as a submandibular mass without visible intraoral involvement, in making the diagnosis more difficult. It is commonly a condition of young adults, although the reported age range is 2–61 years.3–5. There is said to be a slight female preponderance of about 1.3:1 (F:M).[2] A plunging ranula usually presents as a painless, fluctuant lateral neck swelling which does not usually alter with swallowing or eating. It is most commonly centered on the submandibular triangle and averages 4–10 cm in size but can extend superiorly in the parapharyngeal space as far as the skull base, inferiorly to the supraclaviculararea, posteriorly into the retropharyngealspace, or

across the midline anteriorly, usually submentally.[2] Mucocele and ranula are mucous extravasation phenomena that affect salivary glands due to malformation or rupture of gland ducts, altering the normal salivary flow and leading to its deposit in the adjacent tissues.. They are clinically characterized by fluctuant soft nodular swellings with normal or slight blue color. Mucoceles can be found in any region of the oral cavity with minor salivary glands, while ranulas are characteristic of the floor of the mouth and usually associated to sublingual glands.[4] Caroli's disease is a rare congenital disease of the liver characterized by cystic dilation of the intrahepatic bile duct. Classic Caroli's disease involves malformations of the biliary tract alone, whereas Caroli's syndrome refers to the presence of associated congenital hepatic fibrosis.[5] Caroli syndrome are rare congenital disorders of the intrahepatic bile ducts. Caroli syndrome is generally inherited in an autosomal recessive manner[6]. Caroli's disease usually presents during childhood and early adult hood between the age group of 5-21 years.[5,7] A more complex form, that is associated with congenital hepatic fibrosis (CHF) and is inherited as an autosomal recessive trait.[8] The clinical features of Caroli's disease include jaundice, right upper abdominal pain, and fever due to the associated complications of hepatolithiasis or bacterial cholangitis. The cause of Carolis disease appears to be genetic, simple form is an autosomal dominant trait and complex form is an autosomal recessive trait. Caroli's disease usually presents with intermittent abdominal pain and hepatomegaly. Cholangitis, cholelithiasis, bilary abscess, septicemia, liver cirrhosis and cholangio carcinoma are all its potential complications.[1]

Case Report

A full term 30 days old female infant was referred to Department of Pedodontics and Preventive Dentistry, College of Dental Sciences, Davangere from a private hospital with a chief complaint of swelling in the sublingual region since birth (Fig1).

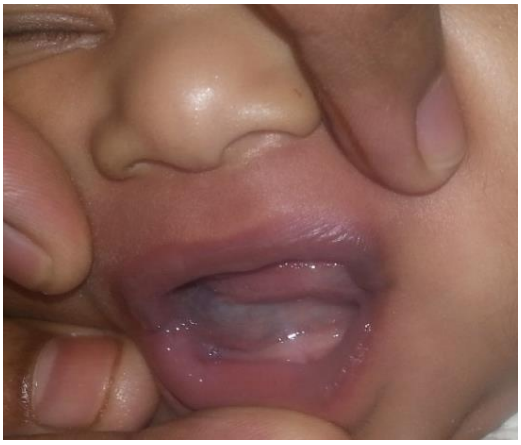

Fig 1. Intra oral ranula on the floor of the mouth

The swelling had increased over a period of time with no difficulty in feeding. Patient was hospitalized in a private hospital due to continuous fever. Patient presented with splenomegly and hepatomegaly and was diagnosed with a rare genetic disorder called Caroli syndrome. Patient was admitted in the hospital and was given treatment for the same .On intra oral examination there was a swelling in the sub lingual region 0.8cm x 0.5 cm in dimension. The swelling was soft fibro elastic, dome shaped slight blue lesion in the floor of the mouth (Fig2).

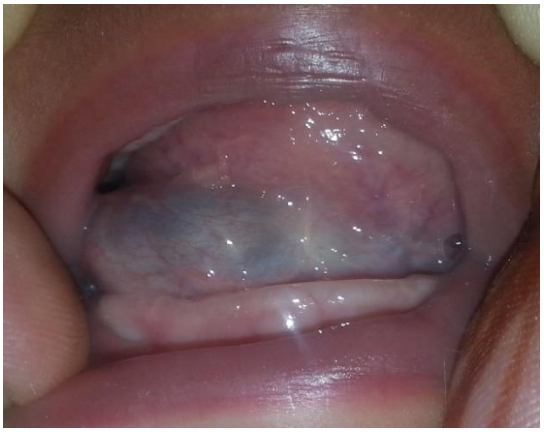

Fig 2. Dome shaped blue lesion at 30 days

Swelling was present since birth and it was a small swelling and gradually it has increased to the present size according to her mother.

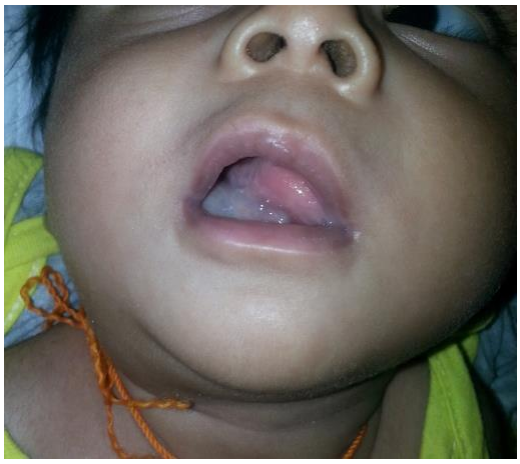

Fig 3. Intra oral swelling at 3 month of age

Provisional diagnosis was made as congenital ranula depending on clinical features. Patient was advised for CT (Computerized tomography) and MRI (Magnetic resonance imaging) for confirmation and to rule out any lymphatic malformation. Parents agreed for investigations, as patient had severe diarrhea due to her systemic problems they couldn't do. Patient reported back to us after two months as the swelling intra orally was increased in size and sometimes patient had difficulty in breast feeding (Fig 3). We observed the swelling had increased in size and it was seen extra orally (Fig 4). Provisionally it was diagnosed as plunging ranula.

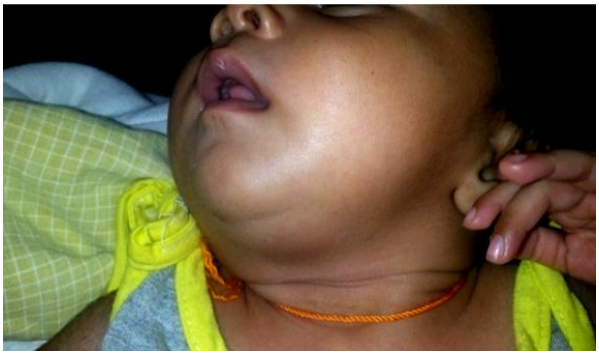

Fig 4. Plunging Ranula

Patient was advised for CT scan and MRI. Treatment plan was explained to parents. As there was more systemic problems to attend for patient, parents dint give the consent for the treatment and patient was kept under observation.

Discussion

Ranula is an extravasation cyst found in the floor of the mouth. They develop from extravasation of mucus after trauma to the sublingual gland or obstruction of the ducts. Ranula is an extravasation cyst found in the floor of the mouth. They develop from extravasation of mucus after trauma to the sublingual gland or obstruction of the ducts.[7] Ranulas are mucus extravasation cyst originating from ` sublingual space but they sometimes extend to the submandibular space and parapharyngeal space, which is defined as a plunging ranula.[10] In plunging ranula the mucus collection is in the sub mandibular and sub mental space of the neck with or without intra oral collection.[11] The diagnosis of ranula is made generally based on the clinical examination. The patient with oral ranula are presented with painless, fluctuant blue translucent color and slowly growing swelling of the floor of the mouth. Ultrsonography , computed tomography scanning , and magnetic resonance imaging can be

helpful in determining the location and size of the lesion. A fine needle aspiration biopsy may detect the mucus with inflammatory cells. Biochemical analysis of aspiration fluid reveals high protein and amylase content.[9] In our case patient presented with congenital ranula and no symptoms were associated with it. After two months the same ranula was increased in size to form a plunging ranula and it was asymptomatic. Patient was diagnosed on clinical signs. Congenital ranulas are usually asymptomatic and are resolved with time.[12-14] Most likely the explanation for resolution would be a rupture as results of feeding .Most of these cases are diagnosed clinically at the time of birth. An MRI scan may be regarded as a gold standard as it not only gives high resolution images, determines precise location and content of the lesion but also enhances the differentiation of ductal atresia from duplication anomalies of ductal system. The treatment protocol for paediatric ranula is still controversial. The medical literature recommends observation for asymptomatic lesions as spontaneous resolution does occur in some cases.[15] Many methods of treatment have been described ,these techniques include aspiration of mucus , incision and drainage , marsupilization, injection of sclerosing gents , excision of ranula with or without excision of ipsilateral sublingual gland , co_2 laser excision , cryo surgery ,and placement of silk suture into the dome of pseudocyst . Spontaneous resolution may be another option for infant ranula. Therefore some investigators suggest that an optimal management of ranula in children my include observation period of 3-6 months for spontaneous recovery. [9] In our case due to systemic condition patient was kept under observation since no immediate treatment could be established. Patient had a very rare genetic disorder diagnosed as carolis syndrome in a private hospital. Systemically patient had recurrent fever, hepatomegaly, splenomegaly, recurrent cholangitis and was frequently admitted to the hospital for these reason. Caroli's disease (CD) was first described by Caroli as a congenital malformation of intrahepatic bile ducts, characterized by segmental cystic dilatation of the intrahepatic ducts; increased incidence of biliary lithiasis, cholangitis and liver abscesses; absence of cirrhosis and portal hypertension; and association of renal tubular ectasia or similar renal cystic disease. Mode of inheritance is still unclear but in majority of cases it is transmitted in autosomal recessive fashion.[16] Endoscopic or percutaneous cholangiography is the traditional method of diagnosis, but magnetic resonance cholangiopancreatography is emerging as the diagnostic modality of choice. The treatment for Caroli's disease includes supportive care with antibiotics for cholangitis and ursodeoxycholic acid for hepatolithiasis.[17]Surgical resection has been used successfully in patients with monolobar disease. For patients with diffuse involvement, the treatment of choice is orthotopic liver transplantation.[5]

Conclusion

Infant ranula is rare. Conservative treatment of infant ranula is advised. It is recommended to observe for asymptomatic lesions, as spontaneous resolution does occur in some cases.[15] Adequate period of observation is 6 months for spontaneous resolution.[9] Rarity of congenital ranula in an infant associated with a very rare genetic disorder made us to concentrate upon systemic well-being and

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just to observe the case for spontaneous resolution unless it becomes symptomatic.

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