Case Report
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Anterior Cystic Hygroma and its Effects on the Dentofacial Structures

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Abstract

Cystic hygroma is a benign growth that is often congenital and probably results from tissue malformation during the early development of the lymphatic system. The cervical cystic hygroma involving the entire craniofacial skeleton is attributed to both local and distant effects on the dentofacial structure. Here we are reporting a case report on cystic hygroma having a secondary effect on the dentoalveolar structure, providing information on potential complications of cystic hygroma and the treatment needed. A 6-year-old female patient with cystic hygroma of the anterior triangle of the neck at birth with post-surgical recurrence and had marked dentofacial malformation.

Keywords: Cervical Cystic Hygroma, Dentofacial Malformation, Pediatric Patient

1. Background Section

Cystic Hygroma (CH) or cystic type of lymphangioma is a heterogeneous group of benign vascular malformations of the lymphatic system composed of cystically dilated lymphatics[1]. It has also been called a malformation, a hamartoma, and an embryonic tumour as 65% to 70% of lesions are present since birth. About 90% of these develop before the age of 2 years, their occurrence in an adult is very rarely observed[1,3,8,9]. The cystic hygroma incidence rate is 1.2 to 2.8/1000 new born[9]. There are three theories proposed to explain its growth, first theory says that blockage or arrest of normal growth of primitive lymph channels occurs during embryogenesis. The second theory says that the primitive lymphatic sac doesn't reach the venous system. The third theory says that during embryogenesis lymphatic tissue lies in the wrong place[2]. It affects the tongue, anterior triangle of the neck, and posterior triangle of the neck, and rarely it affects the parotid gland. The clinical features of head and neck cystic hygroma along with dentofacial abnormalities (clinical and radiological) are described according to the site of involvement as shown in Table 1[2–6,8,10,11,13].

Radiologic evaluation of cystic hygromas includes conventional radiography, sonography, CT, and MR imaging. On CT scan cystic hygroma is seen as a hypoattenuating ill-defined neck cystic mass. On MRI T1 phase shows a predominantly low signal unless there are haemorrhagic components, in T2 phase shows a predominantly high signal, in the T1C+(Gd) phase shows no enhancement on any component except the occasional faint enhancement of the rim[7].

As such literature on cystic hygroma showed that it is not much uncommon entity but very few reports are available on dental-facial malformation in cystic hygroma. Here we are reporting the case of recurring cystic hygroma of the right anterior triangle of the neck with a dentofacial malformation in a 6-year-old pediatric patient.

2. Case Presentation

A 6-year-old female patient was diagnosed with cystic hygroma of the anterior triangle of the neck at birth. The patient came to our OPD for dentofacial malformation and

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Table 1. Clinical features of head and neck cystic hygroma

	Tongue (Cavernous lymphangioma)	**Anterior triangle of neck (cystic hygroma)**	**Posterior triangle of neck (cystic hygroma)**
Percentage of occurrence	40.17%	44%	56%
Develops in	Dense tissue	Loose fascia	Loose fascia
Clinical features	• Enlarged tongue • (macroglossia) • reddish tongue with numerous granular lesions. • translucent in colour • many fine, prominent radiating blood vessels	• Soft, compressible, loculated, non-tender, mobile, pulsatile, fluctuant, transilluminate • Arising from submandibular and submental spaces • Extension in floor of mouth, base of tongue, and epiglottis.	• Soft, compressible, loculated, non-tender, mobile, pulsatile, fluctuant, transilluminate. • swelling Located beneath the platysmal muscle. • Extension into the anterior compartment of neck, may cross the midline, may reach into the cheek or down into the mediastinum and axilla.
Associated symptoms	• eating problem or dysphasia • airway obstruction • malocclusion • difficulty in mastication • secondary infection of respiratory tract	• Bony overgrowth of mandible • airway and feeding difficulty	• May involve the vital structure, such as sympathetic chain, carotid sheath content, branches of hypoglossal, lingual, and facial nerve • obstructive sleep apnea • dysphagia • airway obstruction
Effect on dentialveolar structure (Clinical examination)	• Prognathism or enlargement of mandible • anterior open bite • posterior cross bite.	• Skeletal distortion and hypertrophy, mandibular prognathism • class3 malocclusion • anterior open bite • lateral cross bite. • Retroclination of mandibular anterior teeth • lip incompetence	• Till now there has not been a single case reported because it does not affect the dentofacial structure.
Effect on dentialveolar structure (radiographic examination)	• transverse pressure on body of mandible • marked decrease in the anterior-posterior width of the mandibular ramus • decrease in posterior facial height • increase in posterior facial height	• reduction in width of ramus • increase in length of body • produce an anteriorly direction force on mandible • Increase in the anterior and posterior lower facial height • increase in frankurt mandibular plane angle	• Does not affect the dentofacial structure.
Recurrence rate	• 29.23%	• Cervical involvement 8.33%	

an opinion regarding extraoral swelling in the right facial area. The patient was operated on for cystic hygroma at the age of 2 months. Within 1 year the extraoral swelling reoccurred which was larger than the previous one. The patient had been operated second time at another hospital at the age of 1 year. Facial swelling again reoccurred of the same size and got operated 3rd time at the age of 6 years but still swelling still persisted except it regressed in size up to 4*5 cm at the time of reporting to us.

The familial background excluded the existence of similar clinical manifestations in any other family member. Normal full-term birth without any complication. The patient was not systemically compromised or syndromic with normal intellect and a moderately built physique.

On extra oral examination, unilateral, painless, diffuse swelling was present on the right jaw as well as the submandibular region approximately 4*5 cm in size extending anteroposteriorly from the parasymphysis



Figure 1. **(A)** Depicts extra oral diffuse swelling present on the right side of the face involving the submandibular area at approximately 1 year of age. **(B)** shows recurrent and chronic extraoral swelling after the third surgery at the age of 6 years of age. **(C)** slight vestibular obliteration on right depth of vestibule with midline shift towards left side. **(D)** lingually placed right side mandibular arch.

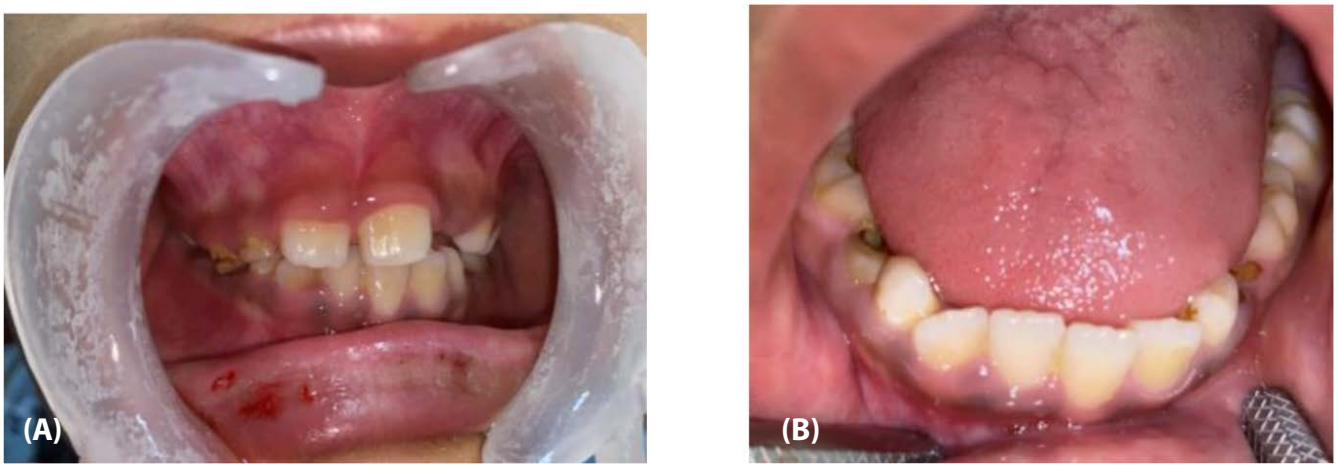


Figure 2. Dento-alevolar changes due to cystic hygroma A) shows vestibular obliteration of right side; B) Lingually tilted mandibular molar

region to the angle of the mandible and superio-inferiorly from ala of the nose to 1 cm below the inferior border of mandible with the normal overlying surface, without any pus discharge or sinus tract (Figure 1-A). While opening deviation of the mouth towards the left side and jerky movements of the temporomandibular joint were noticed. The swelling was soft in consistency, cystic in nature, fluctuant, compressible, and non-tender (Figure 1-B) as classical features of cystic hygroma. Submandibular lymph nodes were of normal size, non-palpable, and non-tender.

Slight vestibular obliteration was seen on the right side of the posterior mandibular region (Figure 1-C). The right mandibular first molar was lingually tilted (Figure 1-D). There was no evident tooth mobility. The tongue deviated towards the right side during protrusion.

Radiographic examination by Orthopantomogram (OPG) revealed a mixed dentition stage with comparable developmental age on right and left sides. Mandible was hypoplastic on the right side extending from the right parasymphysis region to the condylar neck level. No evidence of any impacted, supernumerary, or dental abnormality was present. The inferior border of the mandible was irregular in shape with no break in the continuity on the right side (Figure 2). Above mentioned inferior border of the mandible was distorted due to the pressure effect of CH.

Magnetic Resonance Imaging (MRI) showed a multilocular cystic lesion extending from the base of the skull to the neck on the right side (up to the C4 vertebral level). The lesion bilaterally involved the floor of the mouth, bilateral submandibular, right lingual, right master, right parotid, right parapharngeal and right temporal fossa. The lesion appears hyperintense on T2/STIR images and hypointense on T1W1 (Figure 3).

3. Discussion

Cystic hygroma is a benign, congenital malformation of the lymphatic system with a marked predilection to the head and neck region with 90% of the reported cases involving the head and neck area[2]. The majority of cervical hygromas are located in the posterior triangle of the neck, but if the lesion is sited in the anterior triangle of the neck it may extend into the submandibular and submental spaces, resulting in a swelling of the floor of the mouth[8]. Small cervical CH either involving the anterior or posterior triangle is frequently superficial and freely moveable from deeper structures and the skin. Extensive cervical CH may invade adjacent structures, spreading into the axilla or mediastinum or into the facial structures. Massive lesions of this size account for only 3% of all cases[11].

Cystic hygroma of the anterior cervical triangle of the neck has oral and mandibulofacial involvement extending into the submandibular, submental, and sublingual spaces. They produce an anteriorly directed force on the mandible that causes marked skeletal and postural



Figure 3. OPG shows hypoplasia of the right side of the mandible, Inferior border of mandible on the right side is irregular; the Developmental staging of teeth in right and left sides are same.

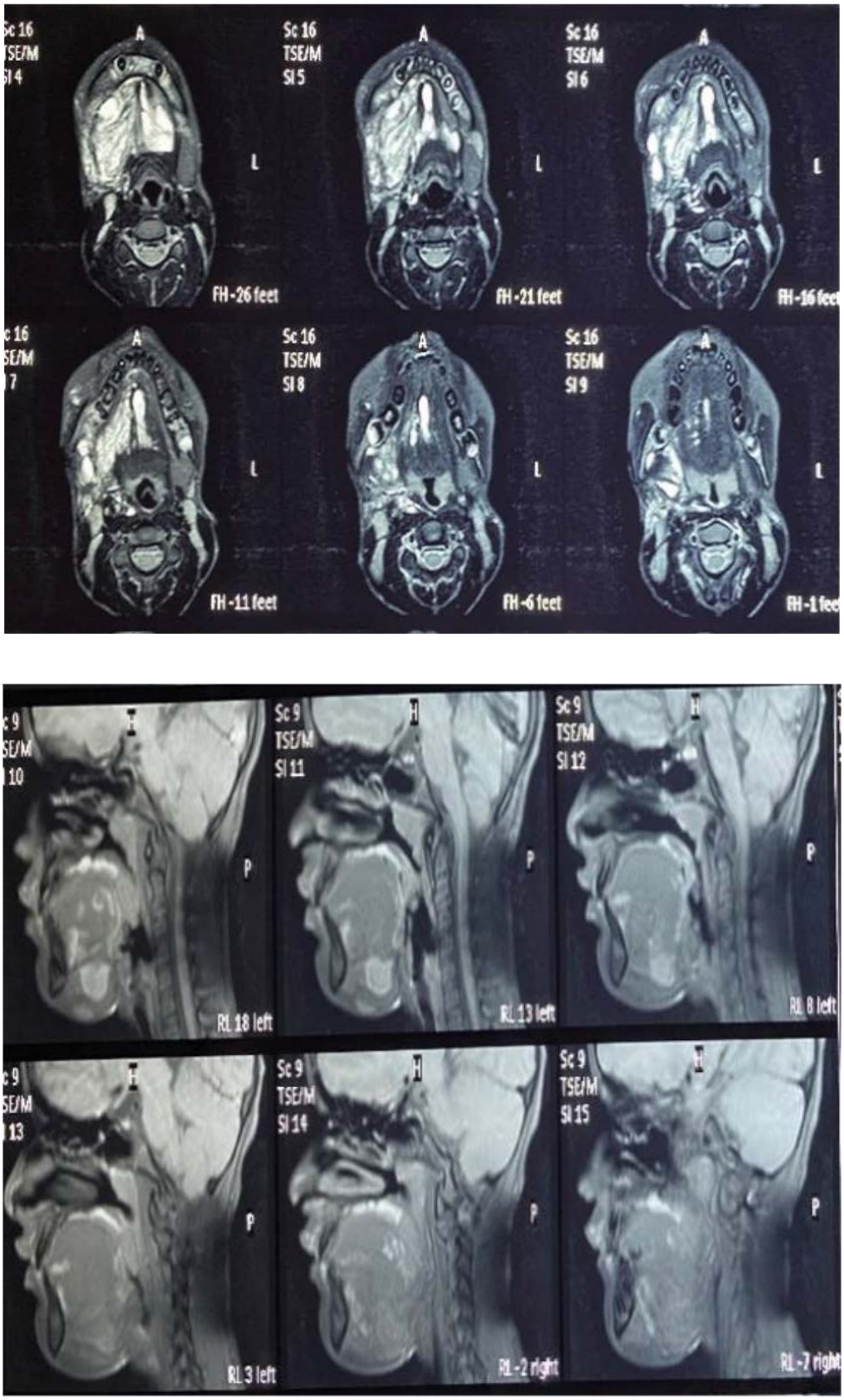


Figure 4. MRI shows a multiloculated cystic lesion extending from the base of skull to neck on right side (up to C4 vertebral level). The lesion bilateral involved floor of mouth, bilateral submandibular, right lingual, right master, right parotid, right parapharngeal, and tight temporal fossa. The lesion appears hyperintense on T2/STIR images and hypointense on T1W1.

mandibular prognathism. There is often engorgement and marked protrusion of the tongue from the mouth that contributes to the anterior force and also causes transverse pressure on the body of the mandible[11]. In our patient, diffuse swelling was present on the right side of the face. While opening deviation of the mouth towards the left side and jerky movements of the temporomandibular joint were noticed. The tongue deviated towards the right side during protrusion.

Very Few cases reported CH involving the anterior triangle of the neck, stating pressure effects on the dentoalveolar structure like slight irregularities in the lower border of the mandible, especially in the angle region[6,8]. In our patient also irregularity in the inferior border and hypoplasia of the right-side mandible was noted.

Etiopathogenesis of bony overgrowth association with lymphatic malformation includes pressure effects, increased blood flow, and direct bony involvement. Another theory suggests that it is due to forward subluxation of the mandible secondary to the mass of soft tissue or Gravitational forces act on the formed tissue mass and distort the functional matrix or due to presence of the lymphatic abnormalities in the submandibular, submental and sublingual spaces causing anteriorly directed force on the mandible that in turn causes skeletal and postural mandibular prognathism[11,12].

Till today there have been fewer than 150 reports of adult cervicofacial cystic hygroma reported, their effect on the dentofacial structure may vary its treatment according to its site and location. When it affects the anterior triangle of the neck it majorly affects the mandible. In cervical CH involving the tongue where mandible prognathism occurs, the chin strap is designed so that pressure is exerted upward and backwards and if the patient has an open bite, orthodontic treatment is recommended.

There is considerable evidence to support the benefits of early orthodontic treatment to correct malocclusions and parafunctional habits as well as to improve oral function in a growing child, thus contributing to better skeletal and occlusal development. It consists of the first phase of dentofacial orthopaedics to try to induce a more harmonious growth pattern and improve function and facial aesthetics. The second phase of treatment would consist of fixed orthodontic appliances to correct the remaining dentoalveolar discrepancies. Remaining skeletal malocclusion can be camouflaged with fixed orthodontic appliances after the completion of her craniofacial growth[14].

4. Conclusion

Cystic hygroma of the head and neck affects the tongue, anterior triangle of the neck and posterior triangle of the neck. Dentofacial malformation occurring in cystic hygroma involving the anterior triangle of the neck should be thoroughly evaluated periodically. Hence timely diagnosis and treatment of dentofacial malformation in patients with cystic hygroma will not affect the malfunctioning of dental structure.

List of abbreviation

CH: Cystic Hygroma
MRI: Magnetic Resonance Imaging
OPG: Orthopantamogram

5. References

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