

Case Report

Tessier number 30

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ABSTRACT

Group 30 cleft of Tessier classification of craniofacial clefts is supposedly one of the rarest clefts seen. This case presented in a most interesting and bizarre fashion - a neck swelling becoming prominent on coughing. The associated finding was even more amazing and rare - a congenital foregut duplication cyst in the neck. We proceed to discuss the rarity of such a case and the possibility of this being one of first few of its kind.

KEY WORDS

Median mandibular cleft, midline facial cleft, Tessier Group 30 cleft

INTRODUCTION

Group 30 cleft of Tessier, lower midline facial cleft or the median mandibular cleft, as it is otherwise called, is not very commonly seen. Only 66 cases have been recorded worldwide. More so, in this particular case, the patient presented to us in the most interesting and bizarre fashion and the associated findings were even more amazing.

CASE REPORT

A 13-year-old boy presented in our outpatient department with a midline neck swelling of just one year duration. His peculiar complaint was the swelling was increasing in size on coughing. He also gave a history of split mandible since birth and difficulty in neck extension [Figure 1].

His previous history showed that he had a tongue release for ankyloglossia at birth. This was followed by a soft tissue repair of the lip and chin at six years of age and an

attempted bone grafting for mandibular repair at nine years of age.

On examination of the patient, the positive findings were midline scar over lower lip and chin and mobile mandibular segments [Figure 2]. There was a 9 x 7 cm lobulated spherical swelling in front of the midline of neck. There were no signs of inflammation and no skin fixity. The swelling did not move with tongue protrusion or with swallowing. The most interesting finding was that the swelling was becoming more prominent with the Valsalva maneuver.

With this, our provisional diagnosis was median mandibular cleft with dermoid cyst which was possibly infected. Other differential diagnoses for midline neck swelling included the possibility of thyroglossal cyst, tracheomalacia, laryngocoele etc.

His blood and urine examination were normal and we proceeded to do a CT scan of the neck [Figures 3-5]. We found that the thyroid gland, trachea, larynx etc. were all normal. The lobulated swelling had mixed consistency and was situated in front of the thyroid cartilage and had extension to upper part of the neck.



Figure 1: Anterior view



Figure 2: Lateral view

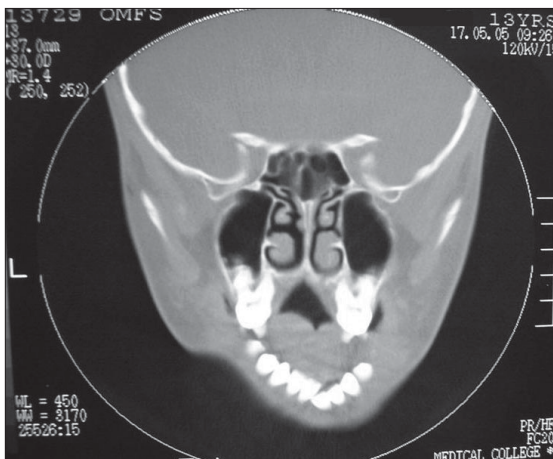


Figure 3: CT scan - split mandible

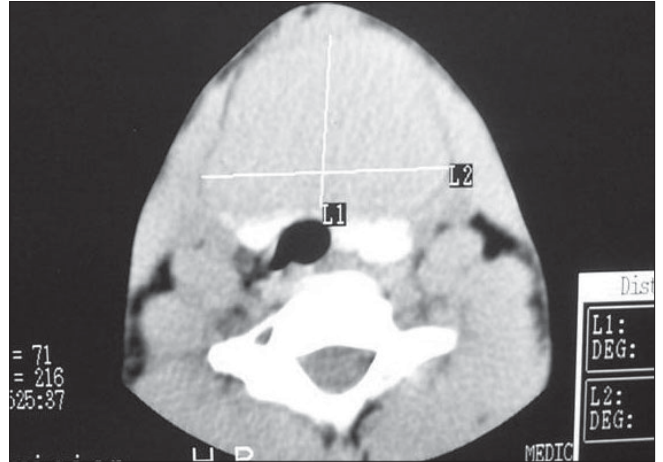


Figure 4: CT scan neck swelling

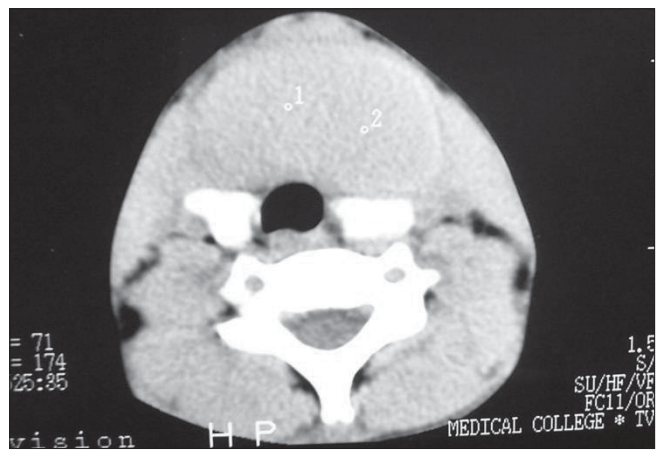


Figure 5: CT scan neck swelling



Figure 6: Specimen

We then proceeded with a formal neck exploration using a Kocher's incision with a vertical midline extension upwards. We were amazed to find that the strap muscles were so thinned out that they were replaced by a fibrous cord in the midline pressing on the swelling giving it a

lobulated appearance. The swelling itself was a smooth white encysted structure which on dissection was found to extend to the base of tongue [Figure 6]. The removed cyst on aspiration gave a copious yellow fluid. The neck was closed with Z plasty of the flaps.

Our diagnosis then was median mandibular cleft with a thyroglossal cyst which was infected.

Postoperatively, as the neck swelling disappeared and patient had sufficient neck extension; and the patient and his parents were happy [Figure 7].

This was not the case with the pathologist, who summoned us to identify the specimen. They found the specimen, on gross examination to be an encysted white structure which when cut open had a thick wall with rugosities on the inner surface [Figure 8].

On closer examination of the section under low power magnification, they got a well-defined four-layered structure with mucosa, submucosa, muscular and serosa walls [Figure 9]. On high power, the submucosa contained gastric glands and lymphoid follicles [Figure 10]. Their diagnosis was this was gastric tissue. Being outside

the normal Gastro Intestinal Tract, they call it a foregut duplication cyst.

So, our final diagnosis was median mandibular cleft with a foregut duplication cyst (stomach in the mouth?!)

DISCUSSION

Tessier No. 30 cleft

It was first described by Couronne in 1819.^[1] It is one of the rarest craniofacial clefts to be seen. Up to 1996, almost two centuries later, only 65 cases have ever been recorded worldwide.^[2,3]

It is believed to be an anomaly in the fusion of the first branchial arch. Since the arches fuse in a sequential manner from above downwards, anomalies can occur in the structures formed from the subsequent arches also. Many of the documented anomalies are: Mandibular cleft,



Figure 7: Postoperative

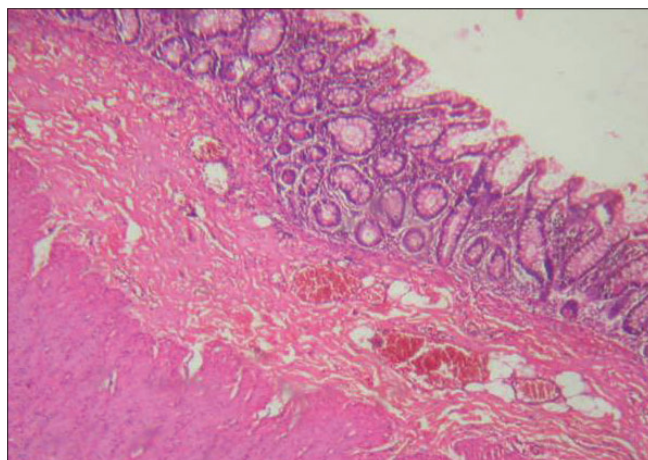


Figure 9: 40x mag: Hemotoxylin and Eosin: 4 layered structure



Figure 8: Cut open specimen

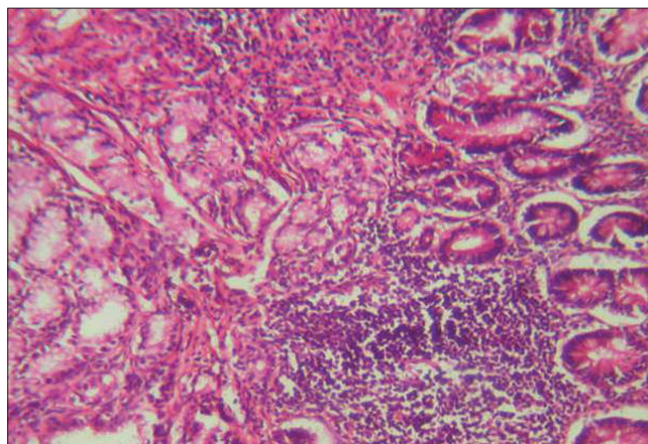


Figure 10: 100x mag: Hemotoxylin and Eosin: submucosa with gastric and mucosal glands

bifid tongue, ankyloglossia and abnormal dentition. Other associated findings are thinned out strap muscles, split - hyoid, sternum, tracheal cartilages, cysts - thyroglossal cyst, dermoid cyst^[4] etc.

The treatment as advocated by Armstrong and Waterhouse,^[5] is to tackle the condition in a staged manner.

- 1st stage - soft tissue correction including Z plasty in chin, lip and neck
- 2nd stage - mandible reconstruction after 10 years of age to minimize damage to the developing tooth buds.

Reconstruction is with bone graft and/or reconstruction plates.

- However, earlier treatment may be indicated when the segments are hypermobile causing respiratory or feeding difficulty.

In this case, the preoperative findings appeared to suggest infection, hence mandibular plating and bone grafting was deferred and is being planned in the next few months as next stage.

Foregut duplication cysts

They are remnants of gastrointestinal tissue that persist after the sixth week of Intra Uterine Life, when the rest of the tissue has canalized and the tract has been formed. It may also be due to a delay in canalization or fusion of the affected tissue.^[6] They can be of the tubular or spherical types and it is the spherical type (as in our case) in which the lumen does not communicate with the gastrointestinal tract lumen.

Foregut duplication cysts are more commonly seen in boys rather than girls. They are most often seen in the posterior mediastinum, the neck being an extremely rare site.^[7,8]

The neck swelling in this case had all the prerequisites for a foregut duplication cyst, namely^[9]

- Focus of gastric or esophageal mucosa
- Well-developed smooth muscle

- Some form of attachment to the gastro intestinal tract

The factors which make this a unique case include:

Rare entity - Tessier Group 30 cleft, is not commonly seen.

Rare presentation - Neck swelling that bulges out on coughing.

Rare association - Rare foregut duplication cyst in the neck, an uncommon site.

Hence, we would probably be justified in calling this case as the "Rarest of the Rare."

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REFERENCES

1. Monroe CW. Midline cleft of lower lip, mandible and tongue with flexion contracture of neck: Case report and review of literature. *Plast reconstr Surg J* 1966;38:312-8.
2. Oostrom CA, Vermeij-Keers C, Gilbert PM, van der Meulen JC. Median cleft of the lower lip and mandible: Case reports, a new embryologic hypothesis and subdivision. *Plast Reconstr Surg* 1996;97:313-20.
3. Ishii M, Ishii Y, Moiyama T, Enomoto S, Ono T, Ohyama K, *et al.* Seventeen year follow up of patient with median cleft of the lower lip, mandible and tongue with flexion contracture: A case report. *Cleft Palate Craniofac J* 2002;39:555-9.
4. Henry K. Kawamoto Jr. Rare craniofacial clefts. *In: McCarthy J. Plastic Surgery.* WB Saunders Company: 1990.
5. Armstrong AP, Waterhouse N. Tessier 30 median mandibular cleft: Case report and literature review. *Br J Plast Surg* 1996;49:536-8.
6. Laraja RD, Rothenberg RE, Chapman J, Imran-ul-Haq, Sabalini MT. Foregut duplication cyst: A report of a case. *Am Surg* 1995;61:840-1.
7. Edwards J, Pearson S, Zalzal G. Foregut duplication cyst of the hypopharynx. *Arch Otolaryngol Head Neck Surg* 2005;131:1112-5.
8. Stringer MD, Spitz L, Abel R, Kiely E, Drake DP, Agrawal M, *et al.* Management of alimentary tract duplication in children. *Br J Surg* 1995;82:74-8.
9. Geller A, Wang KK, DiMango EP. Diagnosis of foregut duplication cysts by endoscopic ultrasonography. *Gastroenterology* 1995;109:838-42.

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