

A RARE CRANIOFACIAL CLEFT: TESSIER NO. 7: A RETROSPECTIVE ANALYSIS

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SUMMARY

In this report we present a retrospective analysis of our craniofacial cleft cases, classified as Tessier no. 7.

Key Words: Rare Craniofacial Clefts, Tessier No. 7

ÖZET

Nadir Görülen Bir Kraniofasial Yarıklık: Tessier No. 7: Retrospektif Bir Analiz

Bu yayında nadir görülen fasiyal yarıklardan, Tessier No. 7 olarak sınıflandırılan vakalarımızın retrospektif bir analizi bildirilmiştir.

Anahtar Kelimeler: Nadir Görülen Fasiyal Yarıklar, Tessier No. 7

The Tessier No. 7 cleft is the least rare seen atypical craniofacial cleft. The incidence of the malformation is reported to be between 1: 3000 and 1: 5642 births (1). Males are more frequently affected than females. Bilateral involvement is rare.

It has been termed as hemifacial microsomia (1), craniofacial microsomia (2), first and second branchial arch syndrome and otomandibular dysostosis (1).

Clinical expression is variable. A preauricular skin tag can be present in microform cases. In its complete form, cleft begins as a macrostomia at the oral commissure and continuous across the cheek toward a microtic ear. All soft tissues may be underdeveloped on affected site. Osseous manifestations also cover a wide range.

MATERIALS AND METHODS

Five patients with Tessier No. 7 treated in our clinic, during the last 17 years, were evaluated. The age of admission was between 4 months to 12 years of age. All patients were female. There was no family history (Table 1).

Table 1: Age and sex distribution of the patients.

Sex	Age	Family History
Female	4 months	(-)
Female	7 months	(-)
Female	11 months	(-)
Female	2 years	(-)
Female	12 years	(-)

There patients had the lateral fascial cleft on the left oral commissure and two had the deformity on the right (Figure 1a, Figure 2a, Figure 3a). Two patients had pretragal skin tag as associated deformity (2a, 3b). One of these two patients had bilateral prominent ear deformity and left, Puzansky type Ia hypoplastic mandibular ramus additionally (Figure 3c). All patients were classified as type Ia, according to Harvold classification. Correction of the macrostomia was done according to Skoog technique (Table 2).

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Table 2: Clinical presentation of the patients

Site of the lateral cleft	Associated Deformities	Classification (According to Harvold)	Operative Technique
R	(-)	Type Ia	Skoog
L	Preauricular skin tag	Type Ia	Skoog
R	(-)	Type Ia	Skoog
L	(-)	Type Ia	Skoog
L	Preauricular skin tag, bilateral prominent ear, left hypoplastic mandibular ramus (Pruzansky type Ia)	Type Ia	Skoog

OPERATIVE TECHNIQUE

After the excision of preauricular skin tag (Figure 3d) oral commissures were reconstructed with the technique described by Skoog (3).

After the proposed oral commissure is marked, a surgical oral commissure is created laterally because of the expected postoperative contraction. A dot is placed opposite on the lower lip (Figure 1b, Figure 3e). After the vermilion turnover flap is prepared oral mucosa is closed (Figure 3f). Upper and lower muscle bundles are skeletonized and divided and upper bundle is sutured over lower one. Skin closure is done according to Z-plasty principles (Figure 3g). In one patient skin is closed primarily (Figure 2b).

RESULTS

During the postoperative follow-up period we didn't see any problem (Figure 1c, Figure 2c, figure 3h).

**Figure 1b:** Preoperative planning**Figure 1a:** Preoperative appearance of the cleft**Figure 1c:** Late postoperative result



Figure 2a: Preoperative appearance of the cleft and the preauricular skin tag



Figure 2b: Intraoperative view



Figure 2c: Late postoperative result

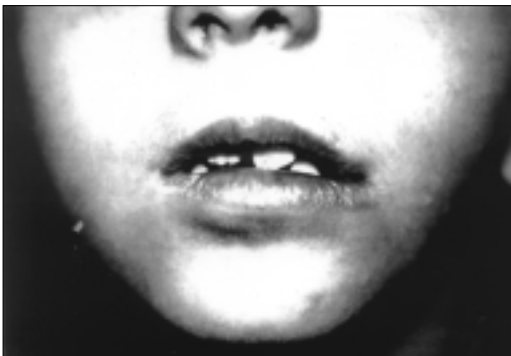


Figure 3a: Preoperative appearance of the cleft



Figure 3b: Preoperative appearance of the preauricular skin tag

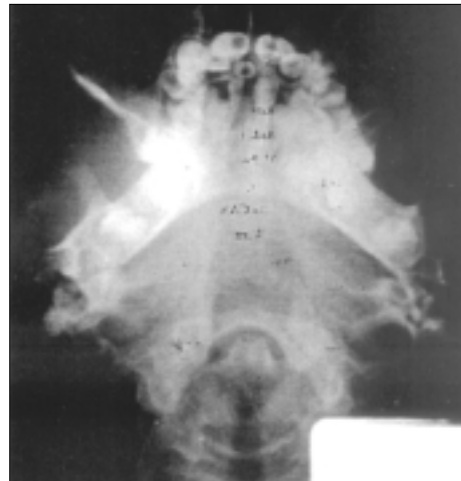


Figure 3c: Radiographic appearance of the hypoplastic ramus mandibula



Figure 3d: Excision of the preauricular skin tag



Figure 3e: Preoperative planning

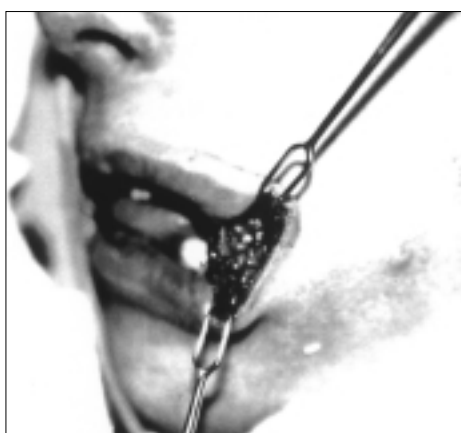


Figure 3f: Closure of the oral mucosa



Figure 3g: Closure of the skin according to z-plasty principles

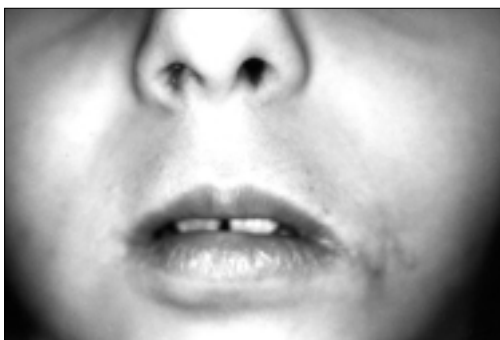


Figure 3h: Late postoperative result

DISCUSSION

Although this deformity is seen more frequently in males, all of our patients were interestingly, female.

As previously mentioned this syndrome shows wide variety in pathologic expression.

According to Pruzansky (4) mandibular deficiency may be classified as;

Type I: Mild hypoplasia of the ramus, and the body of the mandibula is minimally or slightly affected.

Type II: The condyle and ramus are small; the head of the condyle is flattened; the glenoid fossa is absent; the condyle is hinged on a flat, often convex, infratemporal surface; the coronoid process may be absent.

Type III: The ramus is reduced to a thin lamina of bone or is completely absent.

Maxilla, zygomatic complex, the temporal bone and the frontal bone may be hypoplastic. Orbit is often reduced in all dimensions.

One of our patients had Pruzansky Type Ia mandibular deformity.

On the affected site preauricular skin tags are common and the skin, the subcutaneous tissue, tongue, soft palate mimic muscles and muscles of mastication may also be hypoplastic.

Two of our patients had preauricular skin tag.

Absence of the facial nerve function in the distribution of the marginal mandibular branch is seen approximately %25 of patients, with weakness of other components (5). There were no problems associated with facial nerve in our patients. Involvement of the auricle occurs in most of cases and varies from near normalcy to complete absence. One of our patients had prominent ear deformity.

There are some classification systems of the syndrome. Harvold, Vargervik and Chierici proposed following classification (6):

- 1a. Unilateral facial underdevelopment without microphthalmos or ocular dermoids but with or without abnormalities of vertebrae, heart or kidneys.

- Ib. Similar to type I (a) except for the presence of microphthalmos.
- Ic. Bilateral asymmetric type in which one site is more severely involved.
- Id. Complex type that doesn't fit the above but doesn't display limp deficiency, frontonasal phenotype or ocular dermoids.
- II. Limp deficiency type-unilateral or bilateral-with or without ocular abnormalities.
- III. Frontonasal type. Relative unilateral underdevelopment of the face in the absence of hypertelorism with or without ocular dermoids and vertebral cardiac or renal abnormalities.
- IV. (A) Unilateral or (B) Bilateral. Goldenhard type with facial underdevelopment in association with ocular dermoids, with or without under lid coloboma.

All of our patients were classified as Type Ia.

All treatment plans must be customized according to the needs and the age of the individual patient.

Under two years of age:

Excision of preauricular skin tag and correction of macrosomia by commissuroplasty.

Two to six years of age:

In children with severe reduction in the vertical height of mandibular ramus distraction osteogenesis may be performed.

In the patients with a Pruzansky Type III deformity a preliminary costochondral rib graft reconstruction should be performed at the age of four.

In patients with bilateral craniofacial microsomia bilateral mandibular distraction can be performed at the age of two years.

Six to fourteen years of age:

This is the period of orthodontic treatment and facial soft tissue augmentation.

Beyond the fourteen years of age:

Limited autogenous bone grafting of the deficient portions of the craniofacial skeleton, combined LeFort I osteotomy, bilateral mandibular ramisection and genioplasty, bilateral mandibular advancement in patients with mild to moderate mandibular micrognathia and microvascular free flap soft tissue augmentation of the soft tissues may be considered.

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