Rare Craniofacial Anomaly: Tessier No. 2 Cleft

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Four cases of facial cleft that fit the anatomic description of the rare Tessier no. 2 cleft, with two patients having the no. 12 cleft extending to the cranium as no. 2 clefts, are presented. In all patients, clinical expressions of the anomaly were different. Thus, diverse surgical procedures were used in all cases. These cases and review of the literature help to define the soft-tissue and bony course of these clefts, and also emphasize the role of three-dimensional computed tomography scan imaging to show the bony cleft route. The diagnosis and treatment plan of the no. 2 cleft as well as its cranial counterpart are discussed in this report.

Key Words: Tessier no. 2 cleft, Tessier no. 12 cleft, hypertelorism

The no. 2 craniofacial cleft as described by Tessier[1,2] is an extremely rare cleft of unknown cause with distinct soft-tissue and bony characteristics. Among the 336 rare craniofacial cleft examined by Tessier,[1] only 3 could be classified in this category. Similarly, in 1987, Monasterio and co-workers[3] reported 23 cases of no. 2 cleft among 345 rare clefts in 176 patients. Initially, Tessier (personal communication, 1975) doubted whether this cleft existed as a distinct entity or as a transitional form between clefts no. 1 and 3. Therefore, this cleft was represented by a dotted line in his original drawings.[4] However, the cleft does have unique soft and hard tissue characteristics. When present, the associated cleft of the lip lies in the area of the common cleft of the lip. The most distinguishing soft-tissue characteristic of the no. 2 cleft is the deformity on the middle third of the nostril rim. The defective area is hypoplastic rather than a true notch, which contrasts it with the notched dome of the no. 1 cleft and the undermining of the alar region of the no. 3 cleft.[5] On the affected side, the lateral part of the nose is flattened and the nasal bridge is broad. The skeletal involvement in the no. 2 cleft is also distinct because it traverses the alveolus across the socket of the lateral incisor to encroach on the pyriform aperture. The nasal septum is spared but deviated by the surrounding distortions. The normal separation between the nasal cavity and the maxillary sinus is present. A notch is seen near the junction of the nasal bone with the frontal process of the maxilla. The palpebral fissure is not involved as it is in the no. 3 cleft. However, orbital hypertelorism is seen. Enlargement of the ethmoid labyrinth also contributes to the hypertelorism.[6]

The Tessier no. 12 cleft is the cranial counterpart of the no. 2 cleft on the face. It passes between the frontal process of the maxilla and nasal bone. The medial border of the eyebrow is distorted. The location of the eyebrow disturbance also serves to distinguish the cleft.

Materials and Methods

Four patients with Tessier no. 2 cleft were admitted to the Department of Plastic and Reconstructive Surgery, Ege University Medical School (Izmir, Turkey) during the 8-year period from 1992 to 2000. Two

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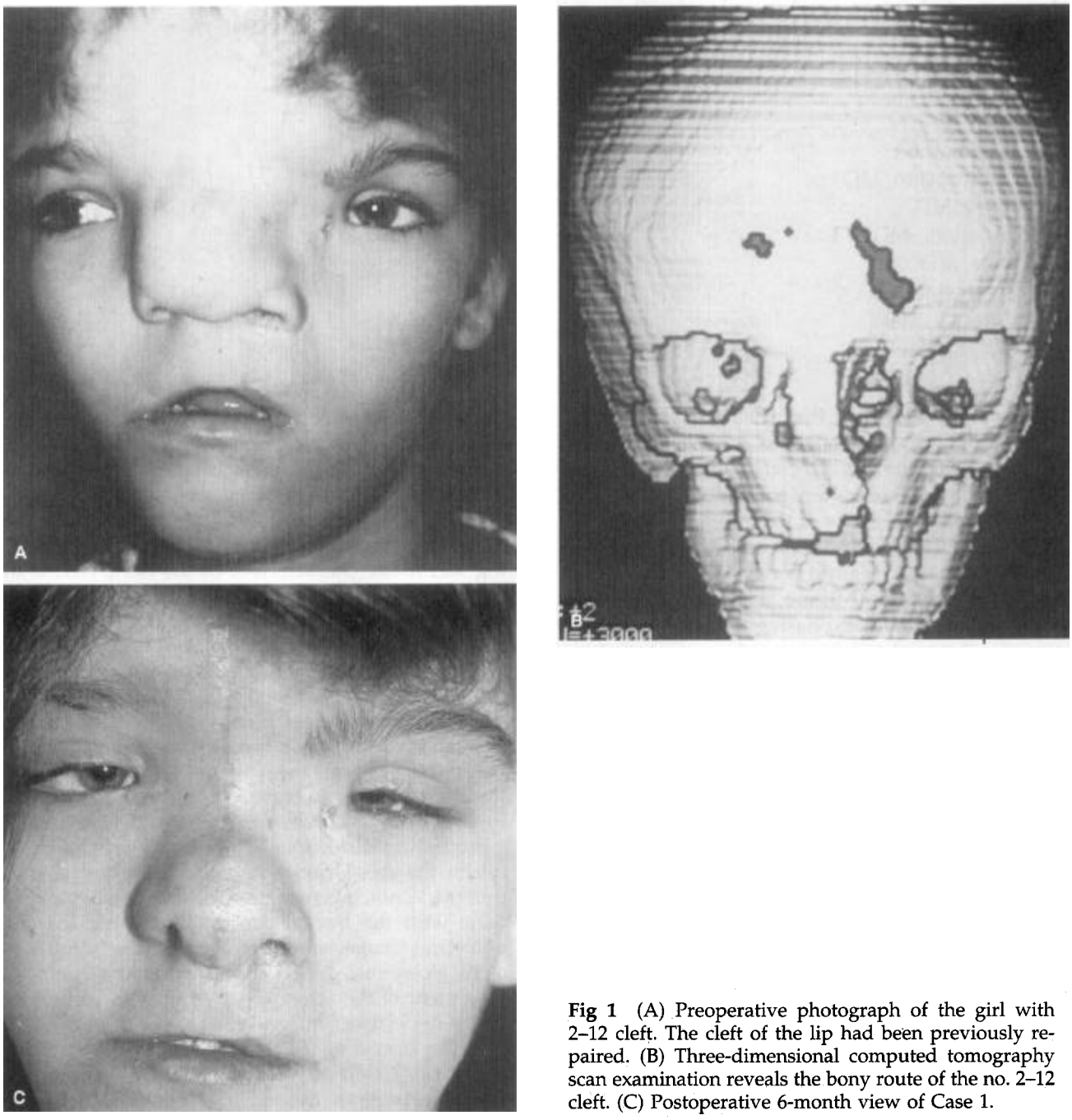

Fig 1 (A) Preoperative photograph of the girl with 2–12 cleft. The cleft of the lip had been previously repaired. (B) Three-dimensional computed tomography scan examination reveals the bony route of the no. 2–12 cleft. (C) Postoperative 6-month view of Case 1.

of these patients were male and two were female (i.e., male-to-female sex ratio: 1:1). The average patient age at time of surgery was 9 years (ranging from 3 months to 17 years of age).

The family history was unremarkable in all patients. There were no reported histories of craniofacial abnormalities in the extended family, and their parents were unrelated. In all patients, there were no known exposures to dangerous drugs, chemicals, or radiation during the gestation. Additionally, there was no evidence (such as ring-constriction deformities of the digits or extremities) of amniotic bands being a source for the facial cleft except in one patient.

The cleft was unilateral in all patients and in two patients of four, the no. 2 cleft extended to the cra-