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CASE REPORT

The importance of a lifeboat-median artery forearm flap in Goldenhar Syndrome

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Abstract
We present a case of a 32-year-old male with left-sided Goldenhar Syndrome and delta phalanx of the thumb, who was offered free tissue transfer from the forearm to address an intra-oral soft tissue deficiency. Despite the presence of appropriately developed right radial artery, used in previous facial reconstruction, the left radial artery occurred to be hypoplastic. He ultimately underwent free flap transfer based on the anomalous persistent left median artery. We suggest that in face of an unusual hand anatomy, flexible flap creation techniques that allow a lifeboat strategy of adjusting flap design should be considered preoperatively. Level of evidence: Level V, therapeutic study.

Keywords Median artery forearm flap · Vascular anomalies · Anomalous thumb · Goldenhar Syndrome

Introduction

The radial forearm flap is typically supplied by the radial artery and its perforators to the overlying skin [1]. Although the radial arterial system is rather consistent, rare anomalies have been reported. The radial artery has been noted to vary in site of origin [2, 3], course [2], number and location of perforators [4, 5]. Additionally, radial hypoplasia and aplasia have been previously described [6]. Typically, the median artery forms part of the main arterial supply to the hand within the first trimester [7–11]. Although the median artery regresses in 90% of population, it can sometimes be found running parallel to the median nerve into the carpal tunnel and contributing to the vascularization of the hand [7–12]. In case of the dominant median artery, the skin of the forearm can also be supported by the perforators stemming from this vessel, which is of great importance when designing a forearm-based flap [5].

Goldenhar Syndrome, also called oculoauriculovertebral dysplasia, refers to patients with the spectrum of anomalies including unilateral microtia, blepharoptosis, mandibular hypoplasia, microstomia, epibulbar tumors, and vertebral deformities [13]. Very rare limb anomalies occurring in Goldenhar Syndrome have never been described as arterial system anomalies or ipsilateral delta phalanx.

Case report

Our patient was a 32-year-old male who was born with left Goldenhar Syndrome. As a child, he underwent multiple reconstructive procedures of the facial skeleton. Six years prior to presentation, he sustained a self-inflicted gunshot wound to the mandible, which necessitated right osteocutaneous fibula and right radial forearm free flap transfers to reconstruct the defects.

He presented to our practice with an intra-oral soft tissue deficiency and drooling (Fig. 1). To address the shallow buccal sulcus and provide a lower lip sling, he was offered free tissue transfer from the left forearm, including the tendon of the palmaris longus to improve oral competence. The patient was found to have normal anatomy of the right forearm;

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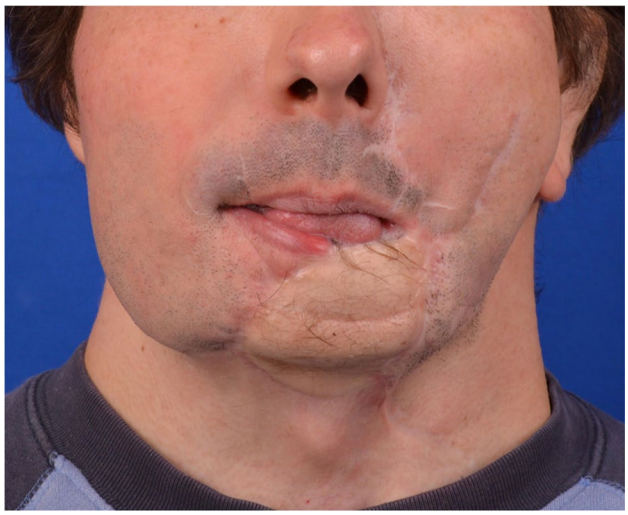

Fig. 1 Preoperative photograph of 32-year's old patient with Goldenhar Syndrome, depicting soft tissue deficiency and tightness of the lower lip

however, the left thumb appeared to be distorted harboring a delta phalanx (Fig. 2).

Pre-operative Allen's test using Doppler machine revealed adequate hand perfusion via the ulnar artery and questionable deep palmar arch perfusion from the radial artery. Intra-operatively, Doppler signal over the radial artery appeared weak and followed a linear vessel pattern in a usual location. The skin paddle of the flap was designed over the marked radial artery and tendon of the palmaris longus. The dissection revealed multiple perforators arising from the area above the median nerve. To allow the dissection to proceed radially, some of these perforators were ligated. Surprisingly, instead of the expected anatomy, a sizeable vascular bundle overlying the median nerve was identified. The median artery was found to originate from the proximal ulnar artery, running directly above the median nerve into the carpal tunnel, 2 mm in diameter (Figs. 3 and 4). Thus, the remaining perforators connecting the median artery and the skin paddle were meticulously preserved. A vestigial radial artery originated from the distal brachial artery and continued only to the midportion of the forearm, without any perforators. After the vessels were identified and the viability of the hand was assured, the median artery was ligated distally, and the flap was raised from distal to proximal liberating the artery together with median veins from the intact median nerve. The flap was then transferred to the recipient site

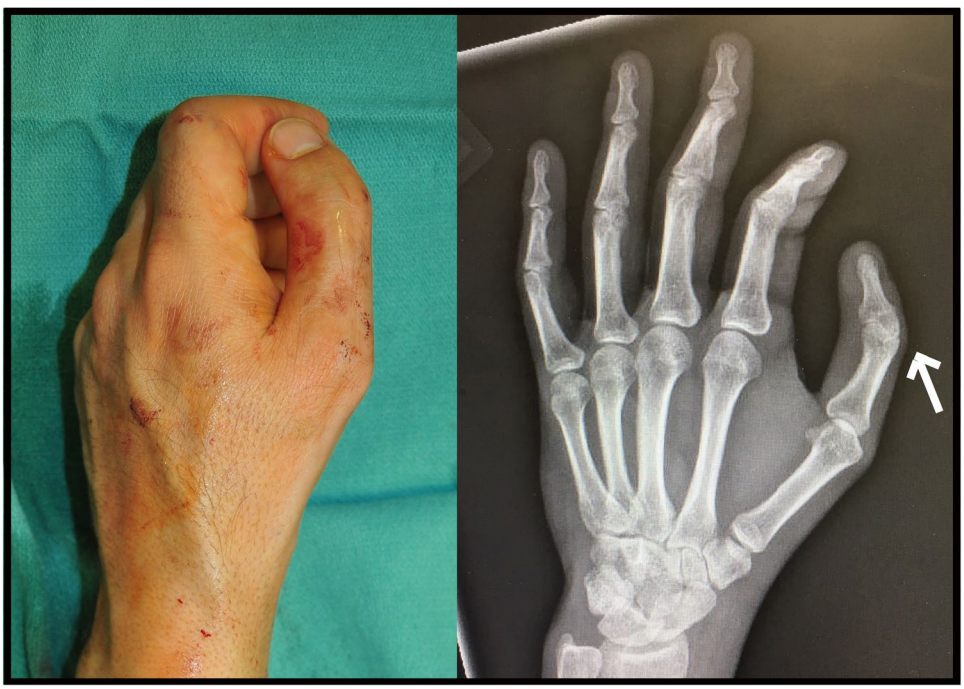

Fig. 2 Figure shows 32-year's old patient's left hand with distorted thumb and an X-ray revealing harbored delta-phalanx. The arrow points at the trapezoidal distal phalanx

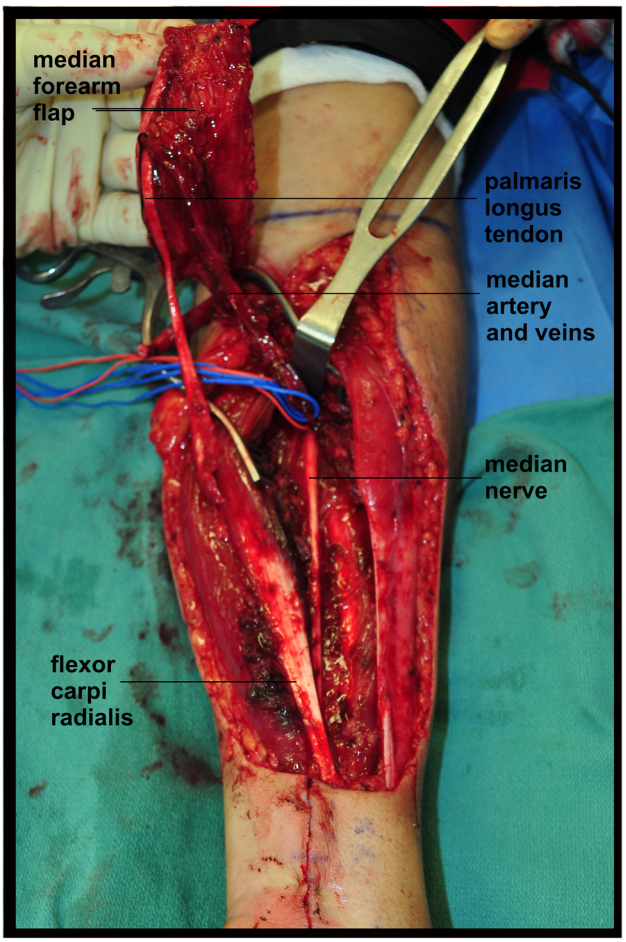

Fig. 3 Intraoperative appearance of the patient's left forearm during median free flap harvest

and the median vessels were gently tunneled and anastomosed to the right facial vasculature. The flap inset was completed without complications. The patient's postoperative course was uneventful. The improvement of oral competence was successfully achieved (Fig. 5).

Discussion

It has been reported that various hand deformities can coincide with at least 127 different syndromes, such as Apert's, Pfeiffer or Nager Syndromes [14]. Therefore, finding an atypical thumb can be a predictor of other congenital anomalies [15, 16]. The oculo-auriculo-vertebral spectrum patients including Goldenhar Syndrome may also present with limb deformities [17]. In our case with multiple facial deformities and delta phalanx of the thumb, we found a hypoplastic radial artery, arising from brachial bifurcation. Hypoplasia of the radial artery has been associated with congenital malformations such as Klippel-Feil, VATER, and Down's syndromes [18]. It also occurred in patients with isolated triphalangeal thumb [16]. It is well documented that certain vascular anomalies are linked with musculoskeletal deformities [18], although an association of a delta phalanx with radial hypoplasia and/or persistent median artery occurrence has not been reported. It has also never been reported in Goldenhar Syndrome.

Our patient presented with the persistent median artery, whose incidence is more common among the African Americans (up to 53%) than Caucasians (2.5% Polish, 4.3% British) [7–11, 19]. The median artery in our case took origin from the most common location, the ulnar artery, although it has also been reported to arise from interosseous or radial arterial system [20]. It did not terminate, as in most cases, before entering the carpal tunnel [7]. As no peri-operative angiography was conducted, we are not able to determine whether the artery was contributing to the palmar arch formation or continuing as the palmar digital artery as described in the literature [7–11, 18]. Interestingly, 80% of patients with one persistent median artery, display the same anomaly on the other side [12]. In case of our patient, the anatomy of the contralateral non-syndromic right upper extremity was typical.

Based on our experience, we suggest that the vascular architecture must be meticulously studied preoperatively in case of congenital limb deformity, especially in the absence of contralateral flap. We propose that, when presence of the persistent median artery is suspected, the modified Allen's test should be performed first [5]. During modified Allen's test, compression of the hypothetical median vessel instead of the radial is applied [20]. If any suspicions arise, additional studies involving duplex Doppler ultrasound or magnetic resonance angiography are advised [20]. However, even normal pre-operative angiogram does not guarantee typical intra-operative findings [5].

When considering options for an alternative flap design, adequate hand perfusion should be assured first. Clearly, the surgeon should be well familiar with the anatomy of the forearm to avoid damage to neuromuscular structures while executing alternate dissection techniques.

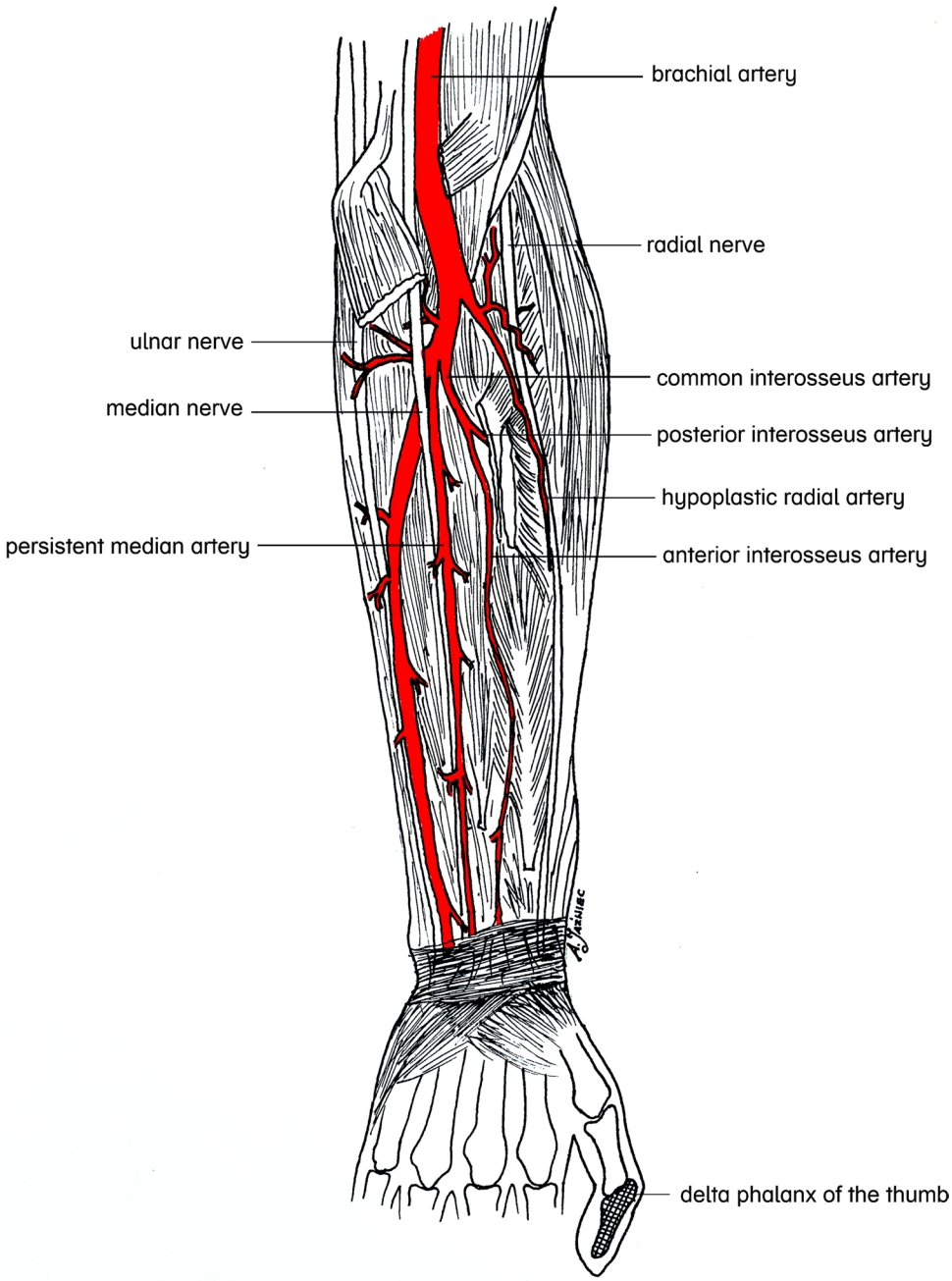

Fig. 4 Scheme of the patient's left forearm vasculature and delta phalanx of the left thumb

In our case, while approaching the presumed radial artery, we inadvertently ligated some of the perforators not realizing their abnormal origin. But after revealing the atypical anatomy, the remaining vessels, connecting the median artery and the skin paddle, were able to be preserved. Also, the septum was intact, so the intraoperative switch from radial to median artery flap was possible.

The possibility of similar adjustment from radial to ulnar artery forearm flap harvest, using the same skin island, has been previously described [5]. We advocate that in face of an unusual hand anatomy, flexible flap creation techniques that allow a lifeboat strategy of redesigning the flap should be considered.

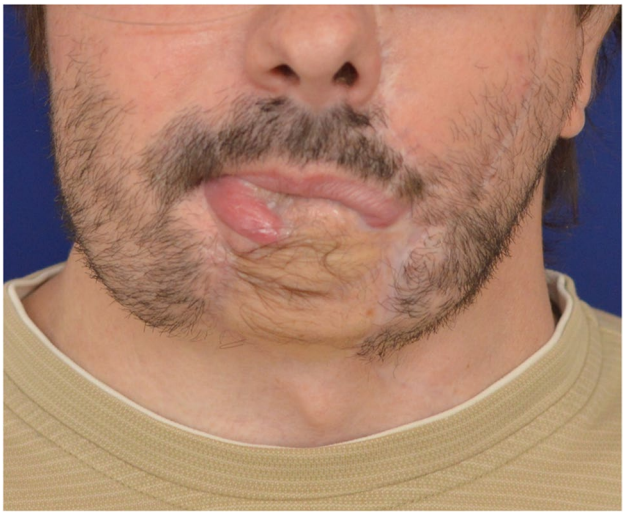

Fig. 5 Postoperative photograph of 32-year's old patient with Goldenhar Syndrome demonstrating improved lip closure and support

Conclusions

The surgeon seeking to use a skin flap from the forearm should always examine the upper extremity and employ pre-operative assessment techniques to delineate the anatomy of the forearm vessels if anatomic abnormalities are noted. Potentially "modifiable" flap design should be implemented in cases of congenital anomalies even if pre-operative studies appear unremarkable.

Declarations

Ethical approval This case report was conducted complying with the ethics requirements of the institutional research committee and with the 1964 Helsinski Declaration and its later amendments. No ethical approval was required for this case report.

Informed consent Informed consent was obtained from the patient included in this case study.

Patient consent Patient signed informed consent regarding publishing his data and photographs.

Conflict of interest Anna M. Jaźwiec and Ewa D. Komorowska Timek declare no conflict of interest.

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