A case of midline cervical cleft

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ABSTRACT Midline cervical cleft is a rare congenital developmental anomaly of the ventral neck. Less than 100 cases have been reported in published journals to date (Ayache et al., 1997). It is usually found as congenital scar-like skin defect or cord-like contractive abnormality of the skin at the ventral neck. Unlike "median cervical cyst" or "lateral cervical cyst", midline cervical cleft usually has no anatomical association with the hyoid bone. We will present a case of midline cervical cleft without fistula but with very small protuberant tissue. The subject was operated at the age of 5 months. We will discuss the clinical aspect and surgical management of this infrequent anomaly.

Key words: midline cervical cleft, median cervical cyst, lateral cervical cyst, aplasia cutis congenita

Midline cervical cleft, which is usually found as a scar-like skin defect or cord-like contractive abnormality of the skin at the ventral neck, is a rare congenital developmental anomaly (Liu et al., 1994 and Manddalozzo et al., 1993). In the following, we will present a case of midline cervical cleft and discuss its clinical management.

CASE PRESENTATION

A two-months-old baby girl with a skin defect that was 10 mm in width and 80 mm in length at her ventral neck was referred to our department (Plastic and Reconstructive Surgery). The pregnancy was uneventful and the mother had no significant family history of any defects. Since birth, the baby has been treated with several kinds of ointment for suspected "aplasia cutis congenita", a congenital malformation often found on the head in the form of alopecia. After one month of treatment, the defect was covered with thin reddened epidermis although the coverage did not achieve full thickness skin coverage over the cleft. At this point, the baby was referred to our hospital for surgical treatment. A diagnosis of "midline cervical cleft" rather than "aplasia cutis congenita" was made at our department after clinical examination: a spindle shaped defect was observed on the submandibular region down to the upper end of the sternum, the dimensions of which were 10 mm in width and 80 mm in length. The cleft was covered with thin epidermis and scar-like tissue, and there was a very small cutaneous protuberance at the cranial end of the cleft (Fig. 1). The sole associated anomaly was congenital umbilical hernia.

At the age of 5 months, we operated on the girl. The cleft was still covered by the thin epidermis that appeared only slightly thicker than 3 months ago. Under general anesthesia, the total length of the cleft was excised at the margin and the skin defect was directly closed using Z-plasties at 2 locations. The cleft did not have a fistula and only had a 2 mm indention at the caudal end. As we had excised the cleft, there was normal subcutaneous tissue without any other abnormal tissue or organs (Fig. 2).

The pathological diagnosis of the excised tissue was "chronic dermatitis" (Fig. 3). There were dispersed dermal indentations and lymphocyte infiltration. Pathologically, both were categorized within dermopathy as a secondary reaction to chronic dermatitis. Five months have passed since the operation, the cleft has not recurred, and there was no contracture of the neck (Fig. 4).

DISCUSSION

Midline cervical cleft is explained as being one of the group of midline branchiogenic syndromes resulting from abnormal migration of cells derived from the branchial arches (Gargan et al., 1985). Some group this anomaly within the facial cleft category (Karlik, 1966 and Tessier, 1976). It is usually found in Caucasian babies and is found more frequently in females than in males (Eastlack et al., 2000 and Maschka et al., 1995). We have found only 2 Japanese case reports in English language journals (Fujino et al., 1970 and
Ikuzawa et al., 1992). Midline cervical cleft is often associated with hypoplasia of various midline neck structures such as median cleft of the lower lip, cleft mandible and cleft tongue (vanDuyn, 1963), but no associations are made with the hyoid bone, as with "median cervical cyst" or "lateral cervical cyst". Our case had no anatomical association with the hyoid bone or any other structures. We have presented a rare case of midline cervical cleft that did not have fistula and had a very small protuberant tissue. We diagnosed and operated on the case at the age of 5 months and she has shown good post-operative results after 5 months.

Midline cervical cleft could be easily misdiagnosed as a scratch or an abraded wound or overlooked, since it is a rare anomaly in newborns and as the changes in skin appearance are minimal, they may not receive appropriate attention from the clinician, as occurred in this case. Some cases are treated when they are older, or even after reaching adulthood.

We recommend early excisional treatment in order to avoid contracture of the neck and limitation of growth of the mandible. When the cleft is excised, it is also important to recognize that total cleft should be excised, and that the closure should be performed with single or multiple Z-plasties in order to avoid further cutaneous contracture of the neck. The bigger the cleft, the more important the excisional treatment becomes in order to avoid contracture.

REFERENCES
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