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Title: Surgical Treatment Results in Craniofacial Dermoid Cysts: Retrospective Analysis of 29 Cases

Running Head: Surgical Treatment Results in Craniofacial Dermoids

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Abstract

Objective: Dermoid cysts are developmental malformations originating from ectoderm and mesoderm. They are congenital and usually localized on the head and neck region. Their wall is covered with squamous epithelium and can contain different skin patches and tissues (multiple sebaceous glands, hair follicles, sweat glands, oil, nail, eye, dental, cartilage).The purpose of this study was to present an approach to masses in head and neck region, evaluate the relationship between preoperative imaging and ultimate pathology, differential diagnosis, and discuss how treatment planning should be done.

Methods: In this study, between January 2010 and July 2017, 37 patients who admitted to our clinic for masses in head and neck region and underwent surgery after necessary consultations and preoperative evaluation, were scanned retrospectively. Patients were evaluated for age, sex, lesion size, lesion placement, preoperative imaging, intracranial involvement, treatment and complications.

Results: Of the 29 patients included in this study, 15 were male (51.7%) and 14 were female (48.2%). The age of the patients ranged from 1 to 28 and the mean age was 10 years. Of the lesions, 22 were localized in the lateral side of the eyebrows (75.8%), 2 in the glabellas (6.8%), 2 in the temporal region (6.8%), 1 in the occipital region (3.4%), 1 in the forehead (3.4%), and 1 in the medial canthal (3.4%), region. At the time of admission, all patients complained of swelling under the skin at the localization where the lesion was present. At least one image, primarily CT, was requested to assess intracranial involvement in all patients' preoperative examinations. The main reasons for the removal of lesions were seen as cosmetic problems.

Conclusion: Dermoid cysts are operated not only for the removal of bad cosmetic appearance, but also for the prevention of possible leakage and infection, getting precise pathologic diagnosis and prevention of secondary bone changes.

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Keywords: Dermoid cyst, head and neck, intracranial extension

INTRODUCTION

Dermoid cysts are the congenital malformations which are originated from ectoderm and mesoderm. They usually localized on head and neck region and trunk. They are lined by squamous epithelium and involve different kinds of skin-related structures such as multiple sebaceous glands, hair follicles, sweat glands, fat, nail, dental, cartilage or bone structure. Dermoid cysts are often benign as they contain mature tissue. Squamous cell carcinoma in adults and endodermal sinus tumor in infants and children are some rare malignant forms of dermoid cysts. Head and neck dermoid cysts constitute less than 10% of all dermoid cysts. There is no definite information on prevalence, racial selectivity and gender discrimination. However, most cases of dermoid cysts in the literature belong to white race. In infants, they are usually found as subcutaneous masses throughout the embryonic skin fusion lines. The regions where they are commonly seen include periorbital (zygomaticofrontal suture), nasal (frontonasal suture and rhinion), intraoral (floor of the mouth), scalp (anterior fontanel and cranial sutures) and postauricular region (1, 2).

In this study, we aimed to retrospectively review our experience with distribution regions, clinical findings and surgical treatments of dermoid cysts located in head and neck region, to better define the characteristics of each subgroup and to establish a comprehensive treatment algorithm.

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METHODS

In this study, a total of 37 patients who were admitted to our clinic between January 2010 and July 2017 with a mass on head-neck region and operated after required consultations and preoperative evaluations screened retrospectively. Patients were evaluated in terms of age, gender, size and location of lesion, preoperative imaging results, intracranial involvement, treatment and complications. Preoperative radiology reports were examined and postoperative pathology reports were taken from the archives and patients with dermoid cysts were detected as the result of definitive pathology report. Eight patients (22.8%) were excluded from the study due to the fact that the dermoid cyst was one of the initial diagnoses according to preoperative imaging reports, but the definite pathology results were not reported as dermoid cysts. Patients were invited for control every 3 months for at least 1 year to avoid potential complications or recurrence. At the end of this period, no recurrence, pathological scar and additional complications were observed. The details, characteristics on presentation and management of the patients were given in Table 1. In the direct excisional approach, the incision was made in the periorbital masses on the lateral edge of the eyebrows and in the hair for the masses on the scalp. In the unsuitable cases, incision was made in the middle of the masses to generate minimal stress. If the mass was close to a nerve, that nerve was dissected and preserved. After dissection, the mass was excised and controlled for any bone or orbital extension. All excised masses were sent to for pathological examination.

RESULTS

Out of the 29 patients included in this study, 15 were male (51.7%) and 14 were female (48.2%). The age of the patients ranged from 1 to 28 and the mean age was 10 years. Twenty two of the lesions were lateral to eyebrow (75.8%), 2 on glabella (6.8%), 2 of them were on temporal region (6.8%), 1 on occipital region (3.4%), 1 on forehead (3.4%), 1 on medial canthal region (3.4%). The mean size of the lesions was 13.2 * 7.03 mm

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(between 18 and 4 mm). When we examine the localization of lesions, 14 of them (48.2%) were on the right side, 11 (37.9%) were on the left side and 4 (13.7%) were at the midline. On admission, there was swelling under the skin at the localization of the lesion. Pain or discharge on lesion was not reported. On preoperative evaluations, all patients had at least one of the computed tomography (CT), ultrasonography (USG) and magnetic resonance imaging (MRI) examinations. Two patients (6.8%) had both CT and MRI images. Nine patients (31.03%) had only USG image, 12 patients (41.3%) had USG and CT images, 4 patients (13.7%) had USG and MRI images, and 2 patients (6.8%) had USG, MRI and CT images. Also, the comparison between initial diagnoses based on preoperative imaging and postoperative pathology results were shown in Table 2.

The main reason for the removal of the lesions was cosmetic problems. All patients were operated under general anesthesia. The lesions were excised by proper incision through the skin and the defects were closed primarily. Pathological examination of all lesions was performed. No complication was observed. All masses were superficial and none had intradural or intracranial extension. In one patient, the mass was compressing the temporal muscle, but there was no muscle perforation or deeper penetration. In one case, the cyst was fistulated into the orbita from the lateral wall of the orbital cavity (Figure 1, 2). In one patient, the mass caused erosion on occipital bone. In this case, bone was trimmed and bone wax applied. No cutaneous fistula or signs of inflammation were observed. Out of the 37 patients operated, 29 (82.8%) were reported as dermoid cysts. In other 8 patients, definitive pathology results were not reported as dermoid cyst although they had dermoid cyst according to preoperative imaging reports. In six patients (16.2%), epidermoid cyst, in 1 patient (2.7%)

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DISCUSSION

Facial dermoides are generally sporadic but familial linkage is described in the literature. There are 2 theories about the embryological origin of craniofacial dermoids. The first is the superficial sequestration theory described by Bland-Sutton in 1893. The second and more widely accepted theory is the prenasal space theory which was described in 1910 by Grunwald. This theory focused on nasofrontal fontanelle which is formed by intramembranous ossification between frontal and nasal bone (2). Frontozygomatic dermoid cysts are superficial masses in zygomaticofrontal suture. They are usually removed by simple direct excision. Dermoid cysts contain histologically dermal adnexal structures such as intraluminal keratin and pilosebaceous units (3-5). Epidermal cysts are similar to dermoid cysts but do not contain pilosebaceous units in the cyst wall. 7% of dermoid cysts occur in the craniofacial region (5). Dermoids were divided into three subgroups topographically as the frontotemporal region, occipital and nasoglabellar region by Bartlett et al in 1993.Dermoid cysts on the frontotemporal region are the most common subgoup and they are located in the lateral eyebrow region. If they are not infected, they arise as slowly growing asymptomatic masses. Dermoid cysts on this region and on the orbital region are typically superficial lesions, but nasoglabellar dermoid cysts, especially those under the nasofrontal suture line, often exhibit sinus tract and intracranial extension (7). In another study by Pryor et al., which enrolled 49 pediatric patients, it has been observed that dermoid cysts are mostly (61%) seen in the periorbital region (8). In our study, they were located on periorbital region in 75.8% of the cases and on the lateral of eyebrow in 75.8% of the cases and in 3.4% of the cases on medial canthal region. When dermoid cysts around the orbita have been examined in another study; it has been observed that dermoid cysts are located 70% on superotemporal-zygomaticotemporal suture, 20% on superonasal maxilla frontal suture and 5% in nasal soft tissue localizations in series (9). Orbital dermoid cysts may exhibit ocular symptoms such as proptosis and limitation of eye movements. Dermoid cysts create a mass effect that does not cause inflammation, they show slow growth pattern and compress the surrounding tissues. Histologically, rupture is about 50% and low grade inflammation is observed in surrounding tissues (9). Twenty percent of the dermoid cysts on the cranial midline show intracranial extension. Embryologicaly, at 8-9 weeks, they develop as a result of

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continuation of the connection between ectodermal and neuroectodermal structures during closure of the frontal and orbital segments. Bone erosion can be seen especially in dermoid cysts which is located on the periorbital region. In a study by Sathananthan et al., which included 70 patients, bone erosion was found in 87% of the patients and whole layer bone defect was detected in 34% of them (10, 11).

In adult patients, preoperative imaging is more important because the cysts are larger and the probability of complications is higher. In our study, the size of the present mass was less than 2 cm in all cases and no intracranial involvement was observed. In only one case, a dermoid cyst on the lateral of the right eyebrow was fistulated into the orbita through the lateral wall of the orbita. That's why preoperative ultrasonography is sufficient and CT or MRI may not be used for the lesions smaller than 2 cm, localized on the frontotemporal region and especially seen at early ages. Although the initial diagnosis of dermoid cyst was established by clinical examination and USG in our patients and most of the localization was periorbital region; additional imaging studies such as CT and / or MRI were needed for differential diagnosis due to the fact that these masses could not be palpated superficially. According to the treatment algorithm for frontotemporal dermoid cysts in adult patients which is described by Chang et al. and approach to orbitofascial dermoids in pediatric population which is described by Bartlett et al., we evaluated our patient population, size of mass and localization, then we created the examination and treatment algorithm which was shown in Table 3 (3, 6).

Although a congenital intracranial frontotemporal dermoid cyst may be seen clinically as a cutaneous fistula in the first stage, intracranial extension and cutaneous sinus tract formation are rarely seen in these dermoid cysts (12). On scalp, dermoid cysts are usually adhered to the periosteum and the usual diameter of the lesions is 1 - 4 cm.

While dermoid cysts are rare in the head and neck region, they should be included in the differential diagnosis of all nodular and cystic lesions in this region in infants and children. An intraoral nodular lesion or tongue tumor may be a dermoid cyst. A giant dermoid cyst on the neck can mimic the cystic hygroma and MRI is required for differentiation (13).

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In our study, although in 8 patients dermoid cyst was one of the initial diagnoses according to preoperative imaging reports, the final pathology results were not dermoid cysts. Reports of these patients were resulted as epidermoid cyst, juvenile hemangioma and pilomatrixoma. This shows that the gold standard in the diagnosis is definition of lesion by the pathology. Trichilemmal cysts, pilomatrixomas, hemangiomas or epidermal cysts may be similar to dermoid cysts on imaging. Therefore, most reliable way to the definitive diagnosis and understanding of how deeply the lesion extends is showing the lesion during the operation and total excision of the cyst tissue. In addition, in contrast to epidermal inclusion cysts, dermoid cysts in the skin are covered with an epidermis with various epidermal extensions. As a rule, these extensions are fully matured. Hair follicles containing hairs entering the lumen of the cyst are often found. The dermis of the dermoid cysts usually contain sebaceous glands, eccrine glands, and apocrine glands in many patients. Surgical excision is the preferred treatment for cyst in any localization. In order to prevent recurrence, the total removal of the dermoid cyst is necessary so that no residual cyst tissue is left behind. In our study, no recurrence was observed in any patient.

Although the treatment of dermoid cysts is surgical, surgical planning should be performed after the physical examination and radiological examination are completed. Due to the lesion may cause psychological and social problems, especially in children, treatment should not be delayed (14). Treatment is surgical excision before rupture formation. The surgical procedure is determined by the extension of the mass and the age of the patient. There are many surgical approaches described in the literature. Some of these approaches are open septorhinoplasty, bi-orbitofrontal nasal craniotomy, anterior craniotomy with lateral nasal flap and total excision of the lesion with gull wing incisions (15, 16). There are many options described in literature for the repair of defects in the bone, including autogenous bone, cartilage and dermofat graft, alloplastic materials and allogenic grafts (15).

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CONCLUSION

Dermoid cysts are excised not only for elimination of worse cosmetic appearance, but also for prevention of possible discharge and infection, definitive pathologic diagnosis and prevention of secondary bone changes. Since it may cause psychological and social problems in children and may cause bone erosion and defects in later ages, it is important not to delay the treatment.

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Table 1. Demographic characteristics and clinical data summary of patients

Case	Age	Gender	Complaint	Intracranial extension	Operation	Preop scan	Lesion size	Complication
1	3	F	Mass on glabella	Absent	Excision-primary repair	USG	10*7 mm	Not observed
2	16	M	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG MRI	14*9 mm	Not observed
3	13	F	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG MRI	13*8 mm	Not observed

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4	9	M	Mass on right postauricular region	Absent	Excision-primary repair	USG	15*7 mm	Not observed
5	10	F	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG	15*8 mm	Not observed
6	10	M	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG	13*7 mm	Not observed
7	10	F	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG	13*6 mm	Not observed

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8	28	M	Mass on occipital region	Absent	Excision-bone wax-primary repair	USG CT	17*9 mm	Not observed
9	14	M	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG CT	14*6 mm	Not observed
10	17	F	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG CT MRI	15*7 mm	Not observed
11	4	M	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG	10*5 mm	Not observed

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12	22	M	Mass on left postauricular region	Absent	Excision-primary repair	USG CT MRI	16*7 mm	Not observed
13	3	F	Mass on left medial canthal region	Absent	Excision-primary repair	USG	10*4 mm	Not observed
14	4	F	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG	11*5 mm	Not observed
15	9	M	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG CT	14*7 mm	Not observed
16	9	M	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG CT	13*8 mm	Not observed

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17	4	F	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG CT	12*6 mm	Not observed
18	15	F	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG MRI	16*8 mm	Not observed
19	16	M	Mass on forehead	Absent	Excision-primary repair	USG CT	17*8 mm	Not observed
20	3	F	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG CT	10*5 mm	Not observed

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21	1	M	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG MRI	9*7 mm	Not observed
22	13	M	Mass lateral to right eyebrow	Absent	Excision-primary repair	CT MRI	18*9 mm	Not observed
23	2	M	Mass on glabella	Absent	Excision-primary repair	USG CT	11*8 mm	Not observed
24	16	M	Mass lateral to left eyebrow	Absent	Excision-primary repair	MRI CT	15*8 mm	Not observed
25	3	M	Mass lateral to left eyebrow	Absent	Excision-primary repair	USG	12*6 mm	Not observed
26	1.5	F	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG CT	11*6 mm	Not observed

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27	16	F	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG CT	15*9 mm	Not observed
28	16	F	Mass lateral to right eyebrow	Intraorbital involvement	Excision-primary repair	USG CT	11*7 mm	Not observed
29	15	F	Mass lateral to right eyebrow	Absent	Excision-primary repair	USG CT	11*7 mm	Not observed

Table 2. Comparison between inital diagnosis based on preoperative imaging methods and postoperative pathology results

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Number of case	**Percent**	**Inıtial diagnosis on preoperative imaging(USG and MRI)**	**Posoperative pathology result**
1 case	%2.7	Epidermal cyst? Dermoid cyst?	Juvenile Hemangioma
1 case	%2.7	Epidermal cyst? Dermoid cyst?	Pilomatrixoma
6 case	%16.2	Epidermal cyst? Dermoid cyst?	Epidermoid cyst
29 case	%82.8	Epidermal cyst? Dermoid cyst?	Dermoid cyst
37 case	%100		

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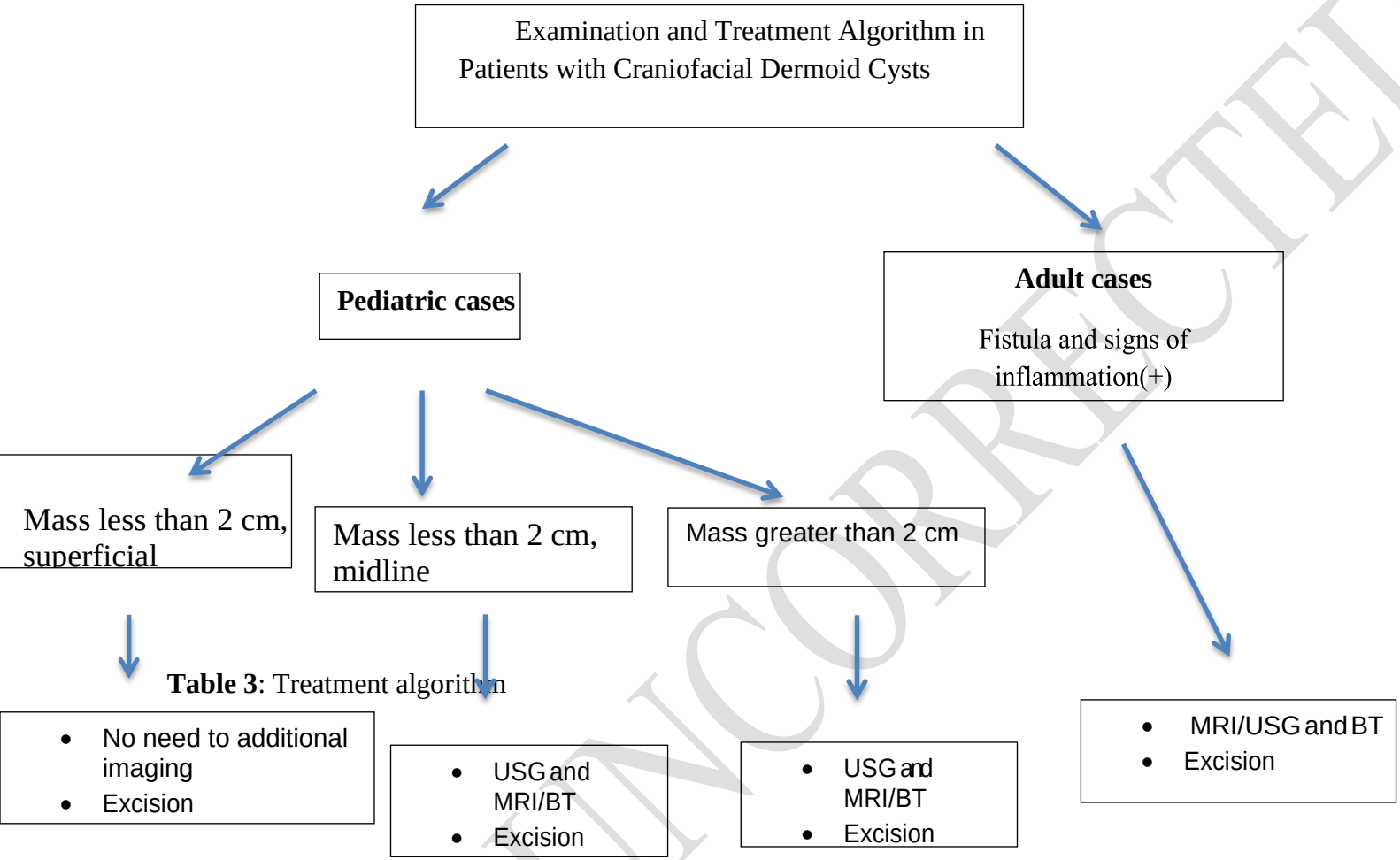

Table 3: Treatment algorithm

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Figure 1a, b. CT image of the dermoid cyst on the lateral right eyebrow in a 16-year-old patient. (a) Coronal section; (b) Axial section; fistula into the orbita from the lateral wall of the right orbita (arrow).

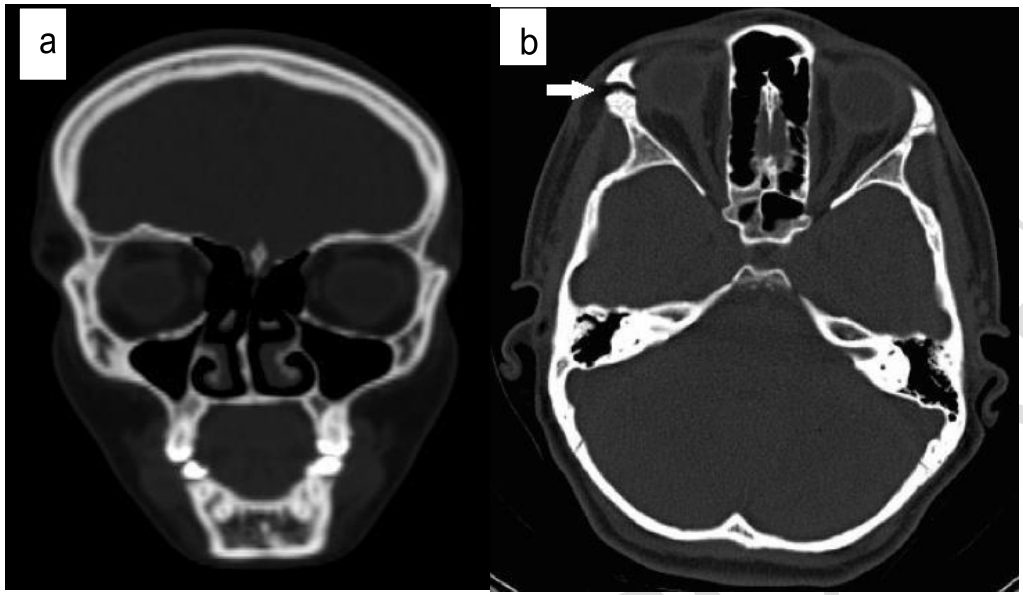

Figure 2a, b. Peroperation view of the same patient in Figure 1. (a) Exploration of the dermoid cyst from the eyebrow incision. (b) Fistula into the orbita from the lateral wall during removal of the dermoid cyst

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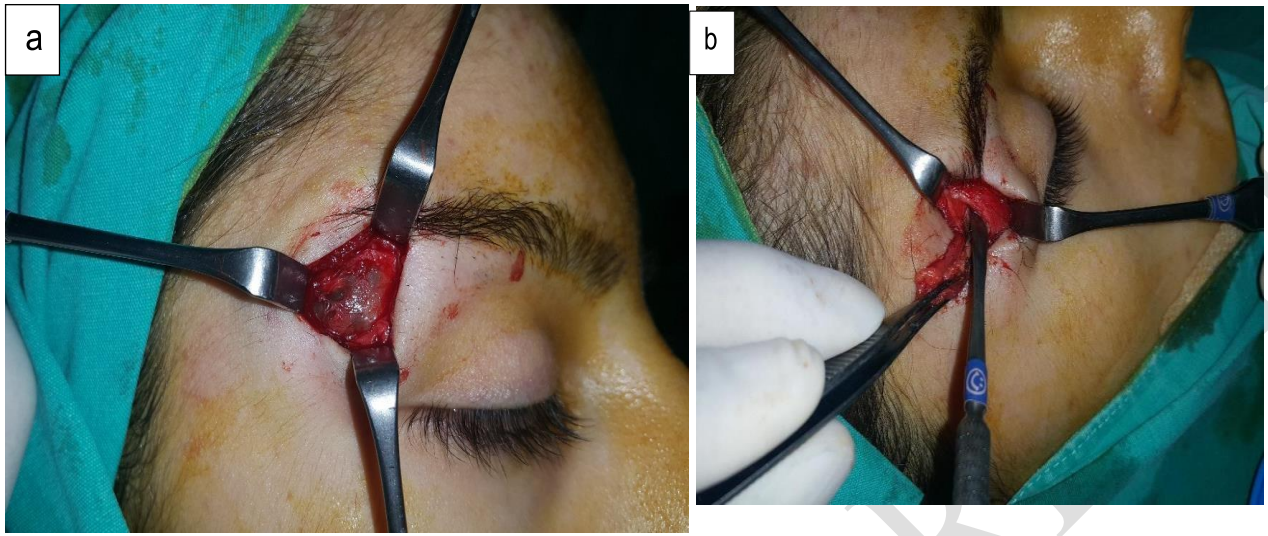

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