

The treatment for the first branchial cleft anomalies in children

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Abstract The objective of this study is to make a clinical analysis for first branchial cleft anomalies (FBCAs), especially introduce the relationship between the Type I/II FBCA with the facial nerve and to demonstrate the importance of using intraoperative microscope and facial nerve monitoring. This is a retrospective review of patients with FBCAs treated in Beijing Children's Hospital, from Jan 2013 to Dec 2015. Clinical data of patients, including sex, age, chief complains, history of surgery, incision and drainage, the relationship with the facial nerve, pre and post-operative facial paralysis, recurrent rate and complications were recorded. FBCAs were divided into two subtypes according to Work's Classification. All patients had an MRI examination for diagnosis and to locate the lesions before surgery. Both microscope and facial nerve monitors were routinely used for detecting and protecting the facial nerve. The study cohort included 42 patients with ages ranging from 10 months to 14 years. The chief complaint was recurrent swelling or abscess near the ear or angle of mandibular. 76.2% of them (32/42) had incision and drainage histories. Two of them had suffered from facial palsy during the infectious period. Seven patients had undergone surgeries once or twice in other hospitals. Four patients with Type II FBCAs developed temporary facial palsy. We had follow-ups for each patient from 3 months

to 3 years. 4 (9.5%) patients with Type I FBCA had got postoperative external auditory canal (EAC) stenosis. There were no cases of recurrence. Type II FBCAs has closer relationship with facial nerve than Type I FBCAs. The facial nerve of Type II FBCAs may lie more superficially and ascending to the fistula. Intraoperative microscope and facial nerve monitoring were necessary for protecting the facial nerve and decreasing the recurrent rate.

Keywords First branchial cleft anomalies · Facial palsy · Surgery

Introduction

First branchial cleft anomalies (FBCAs) are rare congenital malformations of the head and neck, accounting for less than 8% of branchial cleft anomalies. They develop as a result of incomplete fusion of the cleft between the first and second branchial arches. There are several types of classifications for FBCAs. Olser et al. has divided these anomalies into cyst, sinus and fistula [1]. Whether the defect is a fistula (a tract with two openings), sinus (a tract with a single opening) or cyst (a tract with no opening) depends on the degree of closure [2]. Work [3] has divided FBCAs into two types based on anatomical and histological features. Work has described Type I as being a cystic mass in the postauricular area extending medially and anteriorly along the EAC. Type II FBCAs frequently present with fistula below the earlobe in the upper neck which communicated with a sinus tract which extend medial or inferior to the EAC. Type I lesions are of only ectodermal origin, while Type II lesions arise from both ectodermal and mesodermal. As we know, the first cleft and pouch

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form the external auditory canal, middle ear cavity, Eustachian tube and mastoid air cells [4]. The opening of the sinus or fistula could occur anywhere including EAC, middle ear cleft, the post-auricular region and even within the neck near the angle of the mandible [5]. Surgical excision of the anomalous tract is considered to be the best choice of treatment. Both classification systems do not refer anatomy as group standard, but they are very important for excision. Because of the close relationship between the lesion and the facial nerve, parotid gland and EAC, facial palsy, postoperative leakage of parotid gland and stenosis of EAC are the most frequent complications. These complications increase the difficulty of the surgery. In this article we will review patients with FBCAs in our hospital from 2013 to 2015. Preoperative magnetic resonance imaging (MRI) examination, microscope and facial nerve monitoring during the operation are routinely used for these series of cases. The purpose of this article is to introduce the relationship between the Type I/II FBCA with the facial nerve and to demonstrate the importance of using intraoperative microscope and facial nerve monitoring. We also want to introduce an operative technique in our hospital, which is different from classical facial-nerve-exposure.

Patients and methods

We performed a retrospective analysis of 42 patients with FBCAs treated in Beijing Children's Hospital (BCH) from 2013 to 2015 after obtaining approval from the institution's research ethics board. We made classifications according to Work's standards (Fig. 1). All the patients accepted MRI examination to see the extent of the lesions. Operations were performed under general anesthesia on all the patients in BCH. We made records of the patient's sex, age, chief complain, side of cyst/sinus/fistula, the history of surgery, incision and drainage, the relationship with the facial nerve, post-operative facial palsy, pathology, recurrent rates, postoperative EAC stenosis and so on.

Surgical procedures

Patients with Type I (work's classification)

A fusiform incision was made include the infected area in the post aurum. Another straight incision was made from the top of the fusiform incision to the top wall level of EAC (Fig. 2). Then the subcutaneous tissue was separated and we could see the mass which was usually located between skin and cartilage of the EAC. The cartilage of the auricle should be split routinely (Fig. 3). Then we could see the



Fig. 1 Work's classification. *Black* FBCA Type I; *Brown* FBCA Type II



Fig. 2 The incision of Type I FBCA

mass more clearly. The cartilage should be protected carefully, because it was very important for avoiding EAC stenosis. EAC was the anterior border of the FBCA. The inferior border was the superior of parotid gland. The posterior and upper border was the mastoid. Then we could resect the mass totally.

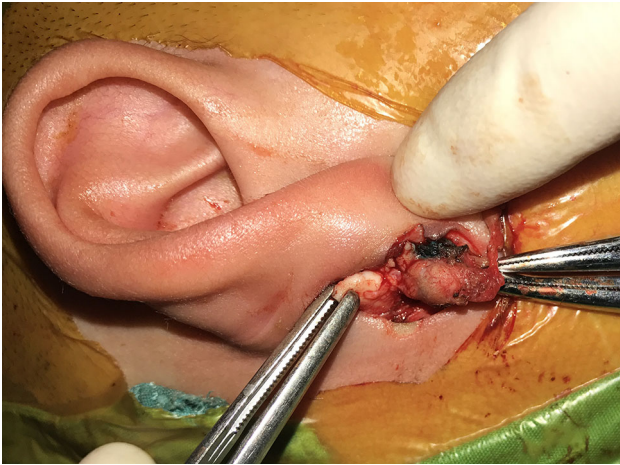


Fig. 3 After splitting the cartilage, we could see the FBCA clearly

Patients with Type II (work's classification)

First, we injected methylene blue to the orificium fistula. A fusiform incision was made include the opening of fistula or sinus. We made another straight incision from the top of the fusiform incision to the inferior wall of the EAC (Fig. 4). We gently separated the subcutaneous tissue because the variant facial nerve might be very superficial. After the lesion was exposed, we did surgery under an operative microscope. With the help of microscope and facial nerve monitoring, the facial nerve could be detected and preserved easier. Once the facial nerve was identified then the cyst, fistula or sinus could be incised easily.

If the opening of the fistula was far away from the EAC and located below the mandibular angle, we chose to make a “stepwise” incision. The first incision was a fusiform incision includes the opening of fistula or sinus. We separated the fistula carefully. When it was close to the EAC, we made another fusiform incision in the inferior wall of



Fig. 4 The incision of Type II FBCA

the EAC. During this step, we should pay more attention to the facial nerve, because it was very closer to the second incision. Facial nerve monitor should be used routinely. After separation of the fistula with the facial nerve, we could make the two incisions connection and cut down the lesion.

Results

The study cohort included 42 patients with ages ranging from 10 months to 14 years. The median age was 4 years and 6 months. Nineteen of the patients were male and the others were female. All of them complained of recurrent swelling or abscess near the fistula, sinus or cyst for 0.16–8 years. 21 of the patient's lesions were located on the left side, 20 of them were on the right side and only one was located bilaterally. 76.2% of them (32/42) had incision and drainage histories. Two of them had facial palsy during the infection. Seven patients had accepted surgeries for once or twice in other hospitals. Three patients also suffered from microtia. Hypointense or isointense signal in T1-weighted, and hyperintense signal in T2-weighted MRI characterized the mass. Some of the lesions infiltrate the parotid and some lesions located around the EAC, which suggested the subtype of FBCAs. In 88% (37/42) of our cases we could see the distribution and extent of the lesions from the MRI. We divided patients into two types (Work's classification) (Table 1). We found that lesions with Type II were more likely to have close relationship to the facial nerve. It has more risk of a complication as facial palsy. Four patients with Type II FBCAs had temporary facial palsy. Their lesions were all located deeper than the facial nerves. They all had the history of incision and drainage. During the surgery we could see the lesions were adhesive with the surrounding tissues. We gave the patients glucocorticoid therapy. Three of them healed within 1 week. One patient healed in 3 months postoperatively. Patients were followed-up from 3 months to 3 years. No recurrence was found. EAC stenosis formed in 4 (9.5%) patients with subtype I.

Discussion

FBCAs were rare and difficult to be treated. The diagnosis of FBCAs was based on symptoms. Clinical signs and an MRI examination helped us choosing the surgical approach. The patient's chief complaints were a recurrent mass or draining sinus in the post-auricular region (Type I) or in the neck over the angle of the mandible (Type II). We used MRI instead of CT scan because of the high radiation damage of CT. MRI could show the soft tissue more clearly

Table 1 The difference between Type I and II

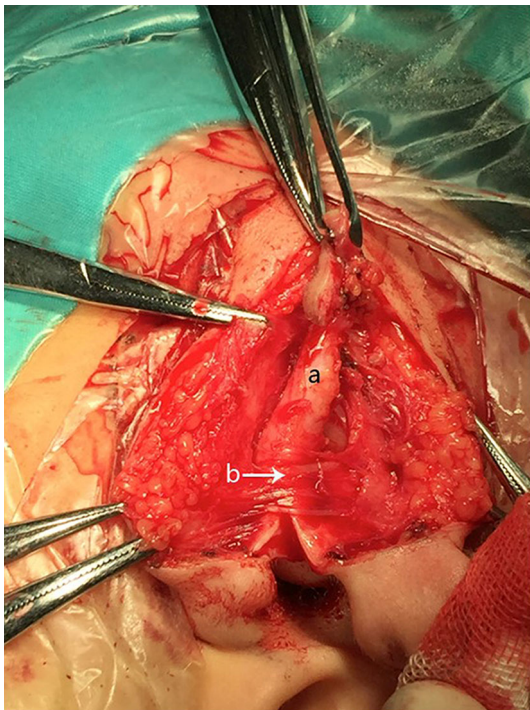
| Work's classification | Type I | | | Type II | | |
|---|---------------------------|-------|-----------|---|-------|------------|
| Number (%) | 24 (57.1%) | | | 18 (42.9%) | | |
| Sex | Male | | Female | Male | | Female |
| | 12 (50%) | | 12 (50%) | 7 (38.9%) | | 11 (61.1%) |
| Lateral | Left | Right | Bilateral | Left | Right | Bilateral |
| | 13 | 11 | 0 | 8 | 14 | 1 |
| | Preoperative facial palsy | | | 2 | | |
| Relation to the facial nerve | No close relationship | | | 38.9% (7/18) adhesive to the facial nerve | | |
| Concomitant with double external auditory canal | 21 | | | 9 | | |
| Postoperative facial palsy | 0 | | | 4 temporary facial palsy | | |
| Recurrent rate | 0 | | | 0 | | |
| Postoperative EAC stenosis | 4 | | | 0 | | |
| Olser classification | Cyst 15 | | | Cyst 5 | | |
| | Sinus 5 | | | Sinus 6 | | |
| | Fistula 4 | | | Fistula 7 | | |

than CT. MRI might help identifying the extent of the lesion. It also helped us recognize the relationship between the lesion, parotid gland and EAC, which might contribute to planning the surgery (Fig. 5). But the facial nerve could not be located with MRI before surgery. In a T1-weighted MRI, the lesion was hypointensive or isointensive and hyperintensive in T2-weighted MRI.

Surgical excision was the only choice of treatment. Because of the lesion's proximity to the facial nerve, the

postoperative recurrence and facial palsy rate was high. Recurrence happened if one considered more about avoidance of facial nerve damage. On the other hand, facial palsy happened if the surgeon concerned more with no residual lesions. The facial nerve and its muscles migrated upward between the sixth and eighth weeks of development. It could be inferred from the embryological patterns that a FBCA might be found superficial to, deep to, or within the branches of the facial nerve [7]. Liston [11] proposed that the developing facial muscles were responsible for this variation. If the facial musculature migrated inferior to the first cleft, the facial nerve would be deep to the lesion and versa. Souza et al. [6] showed the incidence of this complication, which ranged from 21 to 41%, including temporary facial nerve palsy and permanent facial palsy. Recurrence after previous surgical procedures or infection in this region might lead to inflammatory adhesion in the surrounding tissues. It might increase the risk of injury to the facial nerve, negatively impacting the patient's quality of life [8]. Lower recurrence rates were reported (3%) compared with as high as 20% when the excision was performed in the acutely inflamed stage [10]. So we suggested making diagnosis and treatment as soon as possible because the repeated drainage might result in fibrotic adhesion. In our study, one patient developed facial palsy postoperatively and healed 3 months later. We found that this patient's surgery was performed during the acutely inflamed stage. We could not see the structure clearly because of bleeding of the granulation. So we suggest not performing this surgery during this stage.

The authors have argued that as a result of the anomaly development of the first branchial cleft some patients have a double extent auditory canal. A double extent auditory

**Fig. 5** a fistula b facial nerve

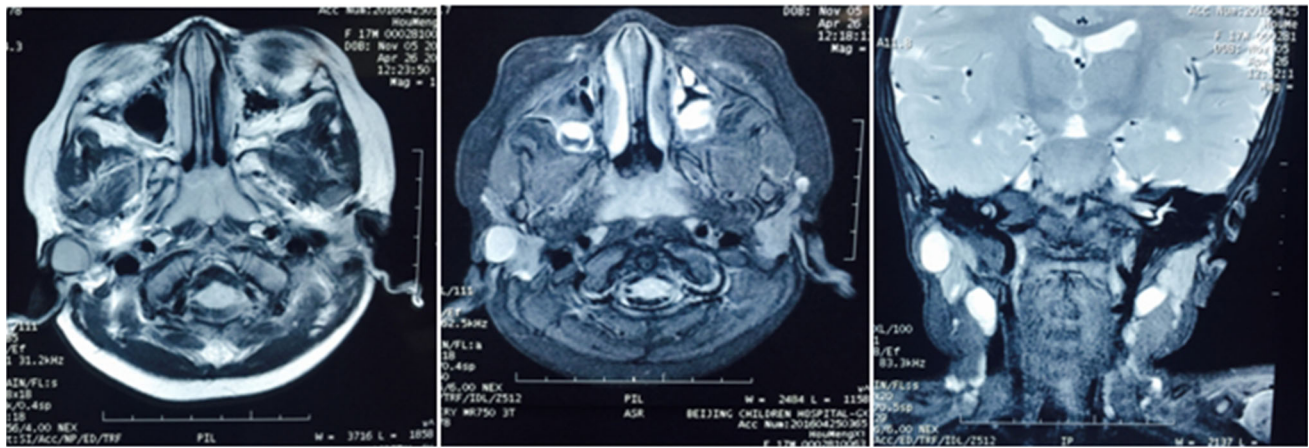


Fig. 6 MRI examination

canal was defined as a second, more or less rudimentary, external auditory canal coexisting with a usually normal external ear canal [9]. In our study, there were 90.2% (37/41) FBCAs patients concomitant with double external auditory canal. So it was very important to see the EAC when doing physical examination. Three patients (two with Type I and one with Type II) also had microtia.

Because of the origin of the FBCA was from the EAC, so we split the cartilage of the auricle routinely. After we split the auricle cartilage, we could see the FBCA more clearly. Then we could find the anterior border of the mass and remove it completely. It was very important for avoiding recurrence. In our opinion, the FBCA with Type I did not have close relationship with the facial nerve, so the rate of postoperative facial palsy was low. However, it has high rate of EAC stenosis than Type II. In our hospital, the postoperative rate of EAC stenosis was 4 (9.5%). The four patients were all Type I FBCA. Too much skin or cartilage of the EAC was resected after we split the auricle cartilage and it lead to the EAC stenosis. Actually, there was no difference of postoperative EAC stenosis rate between Type I and II. No matter Type I or II FBCA, if you resected too much EAC cartilage or skin, the postoperative EAC stenosis would happen.

Due to the relationship between the facial nerve and cleft lesion, the traditional standard treatment was superficial parotidectomy with facial nerve exposure. A sufficient length of facial nerve needed to be dissected. However, over-managing the nerve would result in edema, which could also lead to facial nerve palsy. According to our experience, the facial nerve of FBCA patients might be more superficial than normal anatomy, which means the variation in facial nerve was extended in patients with FBCA and could be damaged by traditional excision. A previous study showed a high (22%) incidence of facial nerve palsy even with prior identification

of the facial nerve [11]. For patients with Type I FBCA, a fusiform incision was made include the infected area in the post aurum. Another straight incision was made from the top of the fusiform incision to the level of EAC. The length of the incision was about 3 cm accordingly. For the patients with Type II FBCA, we made a fusiform incision along the fistula or sinus and made another straight incision from the top of the first incision to the inferior wall of the external auditory wall. If the opening of the fistula was far away from the EAC and located below the mandibular angle, we chose to make a “stepwise” incision. This kind of incision was relatively invasive. We considered the stepwise incision to be enough to expose the lesion and the adjacent facial nerve. However, the intraoperative facial nerve monitoring was essential because the facial nerve was always variant and we could not recognize it easily.

In our study, there were four patients with Type II FBCAs that had fistula openings below the mandibular angle. All four patient’s facial nerves were ascending to the fistulas and the facial nerves were abnormal (Fig. 6). We used intraoperative facial nerve monitoring to confirm that the abnormal tissue was facial nerve. Our study has shown that if the FBCA patient’s classification was Type II and he or she had fistula openings below the mandibular angle, we needed to pay more attention because the facial nerve may lay more superficially and ascending to the fistula. If we do not pay attention, then the facial nerve may be cut off when we separate the subcutaneous tissue. The conventional management of FBCAs does not use a microscope. However, the lesions have always been repeatedly infected. The structure was difficult to be identified. With the use of an operative microscope we could see the nerves and vessels more clearly. When a part of the lesion was exposed, we did the surgery under microscope. It could help decreasing the rate of facial nerve injury.

Conclusion

The management of FBCAs was difficult. Type II FBCAs has closer relationship with facial nerve than Type I FBCAs. During the surgery we should have paid more attention to the Type II FBCAs patients whose fistula openings were below the mandibular angle. Because the facial nerve may lay more superficially and ascending to the fistula. Intraoperative microscope and facial nerve monitoring were indispensable for protecting the facial nerve and decreasing the recurrent rate.

Author contributions Each of the authors has contributed to read and approved this manuscript.

Compliance with ethical standards

Conflict of interest None of the authors has any conflict of interest or otherwise.

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