

Orbital Dermoid and Epidermoid Cysts: Case Study

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SUMMARY

Introduction Dermoid and epidermoid cysts of the orbit belong to choristomas, tumours that originate from the aberrant primordial tissue. Clinically, they manifest as cystic movable formations mostly localized in the upper temporal quadrant of the orbit. They are described as both superficial and deep formations with most frequently slow intermittent growth. Apart from aesthetic effects, during their growth, dermoid and epidermoid cysts can cause disturbances in the eye motility, and in rare cases, also an optical nerve compression syndrome.

Case Outline In this paper, we described a child with a congenital orbital dermoid cyst localized in the upper-nasal quadrant that was showing signs of a gradual enlargement and progression. The computerized tomography revealed a cyst of 1.5-2.0 cm in size. At the Maxillofacial Surgery Hospital in Niš, the dermoid cyst was extirpated in toto after orbitotomy performed by superciliary approach. Postoperative course was uneventful, without inflammation signs, and after two weeks excellent functional and aesthetic effects were achieved.

Conclusion Before the decision to treat the dermoid and epidermoid cysts operatively, a detailed diagnostic procedure was necessary to be done in order to locate the cyst precisely and determine its size and possible propagation into the surrounding periorbital structures. Apart from cosmetic indications, operative procedures are recommended in the case of cysts with constant progressions, which cause the pressure to the eye lobe, lead to motility disturbances and indirectly compress the optical nerve and branches of the cranial nerves III, IV and VI.

Keywords: dermoid cyst; epidermoid cyst; orbit; clinical features; therapy

INTRODUCTION

Dermoid and epidermoid cysts represent congenital cystic benign tumours and belong to choristomas which originate from aberrant primordial tissue. Choristomas are benign tumours that are formed during the embryonic development from the tissues we do not normally expect to be there. Practically, it is normal tissue that is found at the wrong place. During such an embryonic development and formation of the embryonic skull and orbit suture, dermal or epidermal elements remain compressed and form cystic formations with a constant tendency to enlarge and progress [1, 2].

About 50% of these tumours localized in the head appear in the orbit or in its vicinity. Nearly 10% of dermoid head and neck cysts are localized in the orbit, and in this case dermoid and epidermoid cysts of the orbital and periorbital region can be present at different places, which depend on the position of suture, size and degree of growth. More than half of these lesions are localized in the upper-orbital quadrant [3, 4].

Cystic changes are usually discovered early, at birth, but can also be found or even indicate the possibility of growth at any time. The change is usually 1-2 cm in size, soft upon palpation, of an oval shape and mobile under the skin. The inner part of a cyst, which is not accessible to palpa-

ble examination, is usually attached to the bone periosteum near the suture lines. The cyst can very easily be felt in children, while adults have unclear lines and during its growth, the pressure on the bulbous and surrounding bony structures can appear [5, 6, 7].

Clinically, dermoid and epidermoid cysts are divided into superficial (simple) and deep (complicated) cysts. Because of their superficial localization, superficial (simple) cysts rarely develop complications regarding growth and therefore are easily handled surgically. However, they are frequently exposed to injuries and ruptures [8, 9, 10].

Deep orbital dermoid and epidermoid cysts are described as a slow growing mass which spreads during the lifetime. They can form through any bony suture of the orbit, including the upper wall of the orbit. A careful examination is necessary in order to separate superficial from deep lesion formations, because deep lesions can break through the wall of the orbit and propagate into the temporal cavity and sinuses or even into the intracranial area [11, 12].

Differentiation between dermoid and epidermoid cysts is not usually possible to determine during a clinical examination. A pathohistological result shows the difference between these two entities; the basic difference here is the fact that dermoid cysts have squamous epithelium that

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contains extra dermalis and produces keratin. These cysts usually contain blood vessel, fat tissue, collagen, sebaceous glands and hair follicles. The cysts contain material that varies in colour and co-existence. It can be dark yellow, white or yellow colour, with oily, gelatine or relatively solid structure. Since cysts usually have different degrees of inflammation, one should be extremely careful during their removal in order to avoid further inflammation propagation into the surrounding orbital tissue [12, 13, 14].

In our study, a case of a dermoid cyst of the orbit is shown, with a detailed presentation of surgical procedure, as well as discussion of differential diagnosis, necessary diagnosis and therapy.

CASE REPORT

An 8-year old boy was brought for medical examination because of tumorous changes in the inner angle of the right eye, with signs of minor hyperaemia of the involved part. According to his parents, the presence of this change was noticed at birth. The change showed some signs of intermittent growth during the last two years. The boy complained of occasional pains in the area of the tumorous changes and of seeing a double image when looking up and down.

During the ophthalmologic examination normal eye sharpness of both eyes was established (1.0), normal intraocular pressure in both eyes (2.3 kPa), and normal bio-microscopic, as well as ophthalmologic results. Motility of both eyes was also normal. The inspection revealed a possible protrusion of the inner angle of the right eye that covered the inner part of the upper eyelid, which caused narrowing of the inner canthus. A soft and tender cystic structure, clearly limited, which was not fixed to the subcutaneous tissue, could be palpated (Figure 1).

On the axial scanner of computerized tomography of the orbit (CT), on the right side, next to the medial wall, a regular and clearly limited oval hypodensal change corresponding to a cyst was seen. This cystic change pushed the eye aside. A corneal scanner of CT of the right orbit clearly showed a limited soft tissue, a hypodensal cystic formation located between m.obliquus superior and m.rectus medialis,



Figure 1. Tumorous change of the right eye inner angle

in the area of the ophthalmologic artery or v.orbitalis superior, which pushed the bulbus aside, but did not affect the bony structures of the orbit wall (Figure 2).

On 20 Nov, 2005 an operation of the tumorous change was performed in general anaesthesia. After the cut of the inner part of the upper supercillium, wound edges were carefully treated, the target changes were identified (because of the closeness of the bulbomotoral muscles, medial ligament, tear duct, and direct contact between the cyst and the bulbus), with complete preparation and extraction (1-1.5 cm), (Figures 3 and 4).

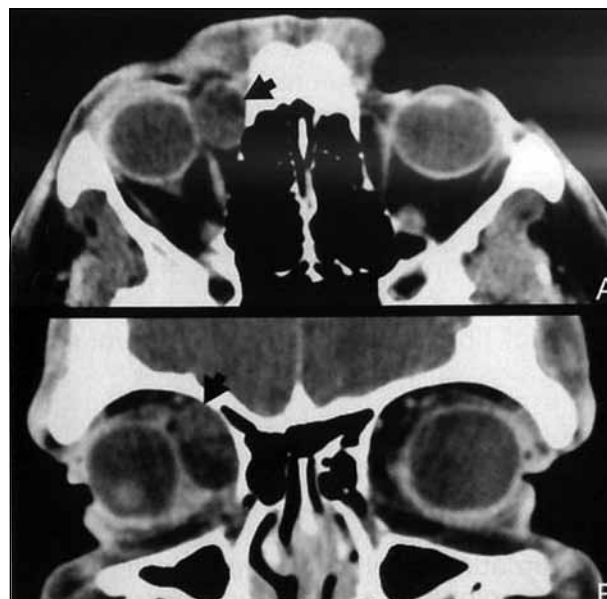


Figure 2. Axial and coronal CT of the orbit



Figure 3. Operative approach

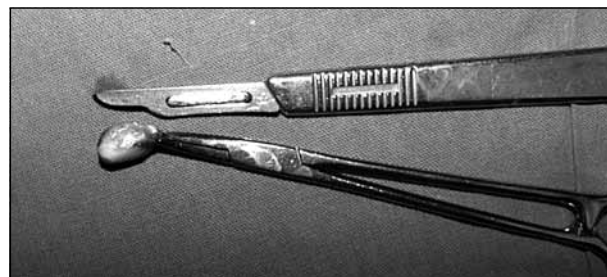


Figure 4. Excised cyst

After the extraction of the cyst, the wound was surgically sutured in layers with adequate haemostasis. Postoperative course was uneventful, without any inflammation signs and with excellent functional and aesthetic results (Figure 5).

The pathohistologic findings confirmed the presence of a dermoid cyst. In Figure 6 the epidermoid cyst which coats the cystic cavity can be seen. The cystic wall, formed of collagen fibres from fibrocyte and infiltrated with rare mononuclear cells is clearly shown (Figure 7).



Figure 5. Postoperative result seven days after surgery

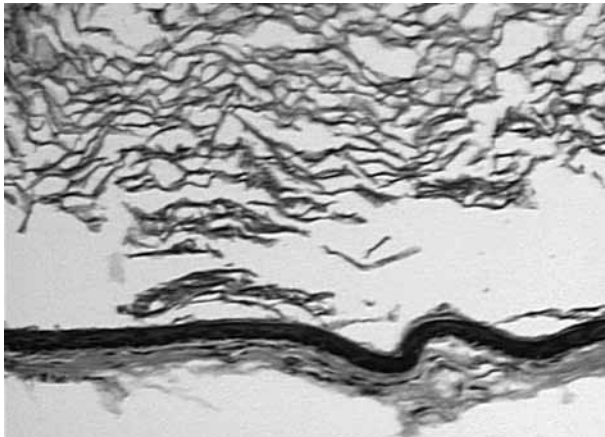


Figure 6. Atrophic epithelium with pronounced parakeratosis and hyperkeratosis

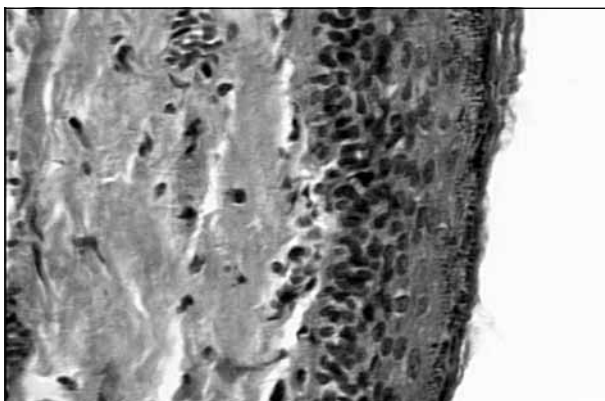


Figure 7. Subepidermal part of the wall with pronounced acellular sclerosis, i.e. hyperplasia of collagen fibers

DISCUSSION

Indications to treat dermoid and epidermoid cysts of the orbit operatively before everything else depend on both the subjective pain caused by them and the prospective estimation that with their progression and enlargement some additional complications can be developed. Considering the fact that they are most frequently formed in the upper temporal quadrant of the orbit, it is in this area that they can cause certain cosmetically aesthetic effects. If it comes to a spontaneous or traumatic rupture of these cysts, an extremely strong inflammation reaction occurs which may present a picture of the orbit phlegmon and demand a proper urgent therapy [15, 16].

With the growth of dermal and epidermal cysts, patients usually register a weak pain in the area where the cyst is localized. They also experience certain bulbous motility disorders. They can be transitory or permanent, and in some cases even signs of proptosis are possible, because of the pressure on the branch nerve, and may cause typical symptoms of compressed optical nerve, with a reduced visual acuity, scotomas in the visual field, disorders of colour perception and the occurrence of a relative pupillary defect [17, 18].

Localization of a dermoid or epidermoid cyst in the upper-nasal quadrant of the orbit is rarer and demands a detailed estimation before any decision about surgery is made. In the case of the boy presented in this paper, the cyst was localized immediately along the bulbus between the upper sidelong and the inner right muscle and near the tear duct on the same side [3, 4].

A decision to perform the surgery due to the child's subjective disturbances caused by both constant progression of the cyst and the pronounced signs of inflammation was made. The CT result was of immense importance since it gave all the detailed information about the size and localization of the cyst. Considering the patient, no signs of bony structure erosion under the tumour compression were found. Some patients might have an obvious bony defect, with clearly defined edges and sclerotic changes. This result made it easier for us to differentiate between superficial and deep cysts; it is extremely important in case of rare, but still present forms of an „iceberg” type with superficial manifestation of the cyst along with simultaneous propagation into the deep orbit structures. Some authors also point out the significance of the ultrasonic examination that can provide us with some useful information concerning orbital tumours [12, 13].

The X-ray, CT and MRI orbit results, as well as ultrasonic examinations are especially relevant in the differential diagnostics of dermoid and epidermoid cysts with other benign and malignant tumours of the orbit. Regarding children, this differential diagnosis is of essential importance in cases of mucocoeles, encephalocoeles, sebaceous gland cysts and echinococcal cysts. In adults, especially if there are signs of progression, in the differential diagnosis thyroid ophthalmopathy should be included, as well as lacrimal gland tumours and other primary and metastatic tumours of the orbit. Clearly limited, a hypodensal oval change with a transparent centre presented in the CT picture of our patient

mostly indicates that it is the case of a benign cystic change [13, 14].

After a precise diagnostic was made, the next step was a correct surgical estimation about the best approach to the cyst, with an aim to avoid all possible disturbances of the surrounding structures, to cause minimal trauma and create the best aesthetic effects. In the case of our patient, we used orbitotomic approach to the tumour in the projection of the lower supercillium and immediately above the tear duct projection. According to the precise estimation about the closeness of the adjacent orbital muscles of the bulb and tear duct, the complete separation of the cyst from the surrounding structures and its extirpation was performed. Since any eventual cyst rupture or any remaining part would cause a serious inflammatory reaction and any possible residua would demand a re-intervention, this operative procedure required extreme preciseness [5]. It should be mentioned that the outer wall of the cyst has a variable thickness. Therefore, in some places, it can be very thin and at the same time the deeper part of the cyst is connected to the periorbital fibrovascular tissue, which implied a special caution while separating the cyst from the surrounding structures.

Operative complications are related to possible disturbances of the surrounding structures, eye and upper eyelid motility disorder, haemorrhage and infections. If an intra-

operative rupture of the cyst occurs, in order to prevent a strong inflammatory reaction, it is recommendable to use antibiotics and corticosteroids.

With surgical treatment of deep cystic orbits, it is often necessary to apply teamwork of an ophthalmologist and a maxillofacial surgeon, and in some rare cases, the presence of a neurosurgeon is needed to surgically treat cyst propagation in the intracranial area [17, 18].

Dermoid and epidermoid cysts beside aesthetic defects, with their constant progression and inflammatory reactions, can cause numerous subjective and objective ophthalmological problems. If a cyst causes dislocation of the bulb, neurological disorders caused by compression on the III, IV and VI intracranial nerves and the optical nerve, or if it demonstrates constant signs of inflammation, these are obvious indications for surgical treatment.

However, if a patient does not complain about subjective disorders and the cyst shows no signs of progression or inflammation, the patient's monitoring is most frequently recommended.

Considering the fact that these kinds of cysts mostly affect children, a special caution and care should be applied regarding the precise diagnostics of the type of tumour and its accurate localization. The cyst should be completely removed, with minimal trauma and a satisfactory aesthetic effect.

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Дермоидне и епидермоидне цисте орбите – приказ болесника

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КРАТАК САДРЖАЈ

Увод Дермоидне и епидермоидне цисте орбите припадају хористомима, туморима који потичу од аберантног примордијалног ткива. Клинички се испољавају као цистичне покретљиве формације најчешће локализоване у горњем темпоралном квадранту орбите. Описују се као површне и дубоке формације с најчешће спорим, интермитентним растом. Посебан дијагностички и терапијски проблем може настати код спонтане или трауматске руптуре цисте, када се јављају изражени знаци запаљења са сликом целулитиса орбите.

Приказ болесника Приказано је дете с конгениталном дермоидном цистом орбите локализованом у горњем назалном квадранту која је показивала знаке постепеног увећања. Компјутеризованом томографијом је откривена циста величине 1,5-2,0 cm. На Клиници за максилофацијалну хирургију у Нишу урађена је екстирпација дермоидне цисте у целини након орбитотомије приступом кроз предео суперцилијума. Операцију су урадили максилофацијални и офталмолошки хирурзи

уз минималну трауму, након које је остао веома мали ожиљак. Постоперациони ток је протекао нормално, без знакова запаљења, а након две недеље је постигнут одличан функционалан и естетски ефекат.

Закључак Пре одлуке о хируршком лечењу дермоидне и епидермоидне цисте неопходна је детаљна дијагностика ради прецизног утврђивања локализације цисте, њене величине и евентуалног продирања у околне периорбиталне структуре. Веома је важно искључити друге бенигне туморе орбите, као што су мукокеле, енцефалокеле и лојне цисте, а у случају израженог запаљења и рабдомиосарком, метастатске туморе орбите и орбитални целулитис. Поред естетских разлога, хируршки захват се препоручује и код цисти које стално напредују, врше притисак на очну јабучицу, доводе до поремећаја мотилитета и непосредно притискају очни живац и гране III, IV и VI кранијалних нерава.

Кључне речи: епидермоидне и дермоидне цисте; орбита; клиничке одлике; хируршко лечење