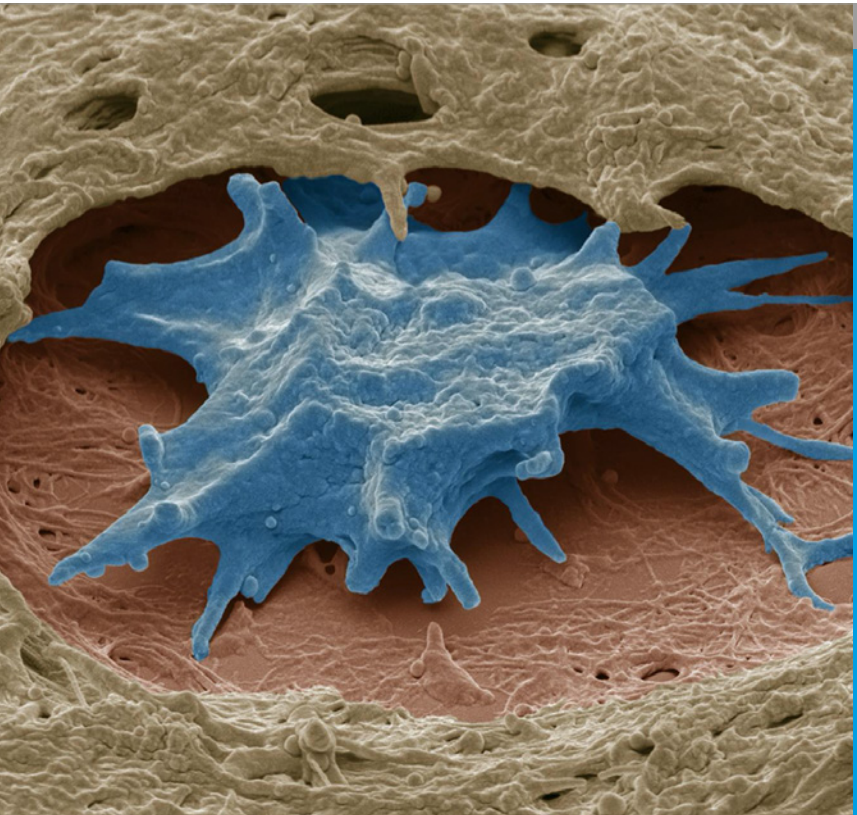
SWISS SOCIETY OF NEONATOLOGY

Atypical localization of a teratoma



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Title figure:
Ovarian teratoma (SEM image by Stephen Gschmeissner, source: theworldcloseup.com)

INTRODUCTION

Derived from the Greek „teratos“, the word teratoma literally means „monster“ referring to the abnormal growth and appearance of these tumors (1). Teratomas are congenital tumors consisting of tissues from all three germ layers: ectoderm, mesoderm, and endoderm. Teratomas are classified as mature or immature teratomas. Mature teratomas require complete resection but are generally benign. Immature ones contain immature neural tissue and have a greater propensity for recurrence or even metastases (2, 3). In newborns, the sacrococcygeal area is the most common anatomic site for teratomas (4). In contrast, orbital teratoma is considered a rare condition (5, 6).

CASE REPORT

This newborn girl was born by an uncomplicated vaginal delivery at 40 2/7 weeks of gestation. Immediately after birth, a huge tumor in the left orbita was apparent (Fig. 1). The baby was the fourth child of non-consanguineous parents with an unremarkable family history. Her birth weight was 3290 g (P26), her birth length was 49 cm (P14), and her head circumference was 35 cm (P41), all appropriate for gestational age. Antenatal care, including 3D-ultrasound at 30 weeks of gestation, had been unremarkable. The girl was transferred to the Neonatal Intensive Care Unit for further investigations following a period of bonding with the mother.

Fig. 1

Baby girl on DOL 1. Marked protrusio bulbi on the left side.

Ophthalmologic exam revealed left-sided proptosis with dislocation of the eyeball out of the orbita in a cranio-lateral direction as well as marked chemosis. Funduscopic findings included swelling of the macula and retinal bleeding. Ultrasound examination demonstrated a cystic mass in the orbita and elongation of the optic nerve with medial displacement.

Magnetic resonance imaging (MRI) (Fig. 2, 3) showed a tumor (3.2 × 3.8 × 3.8 cm) with cystic and solid components without intracranial or periorbital involvement. The orbital volume was enlarged, and the orbital wall was displaced. Differential diagnoses included either a teratoma of unknown dignity or a lymphangioma. Further studies for staging purposes including a thorough physical examination, echocardiography, abdominal ultrasound and chest X-ray were all normal.

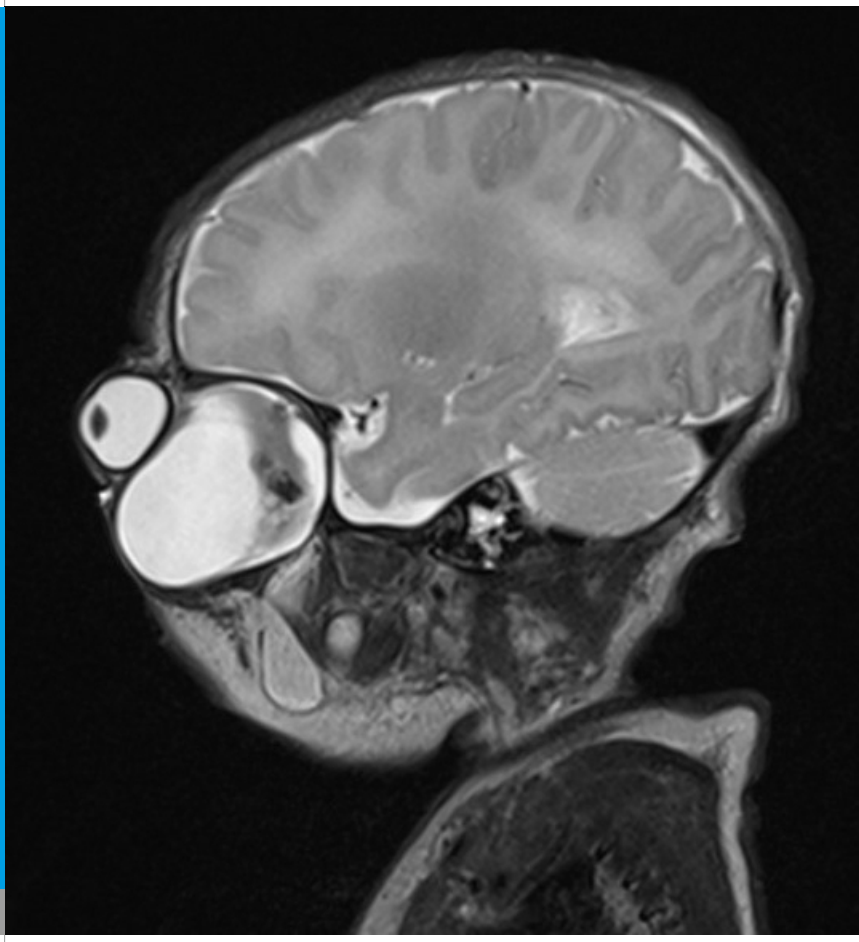

Fig. 2

T2-weighted magnetic resonance images (transverse view): large, left-sided intraorbital mass (3.2 × 3.8 × 3.8 cm) with two large cystic parts separated by a septum, and centrally located, inhomogeneous solid part (1.2 × 1.5 × 2.5 cm).

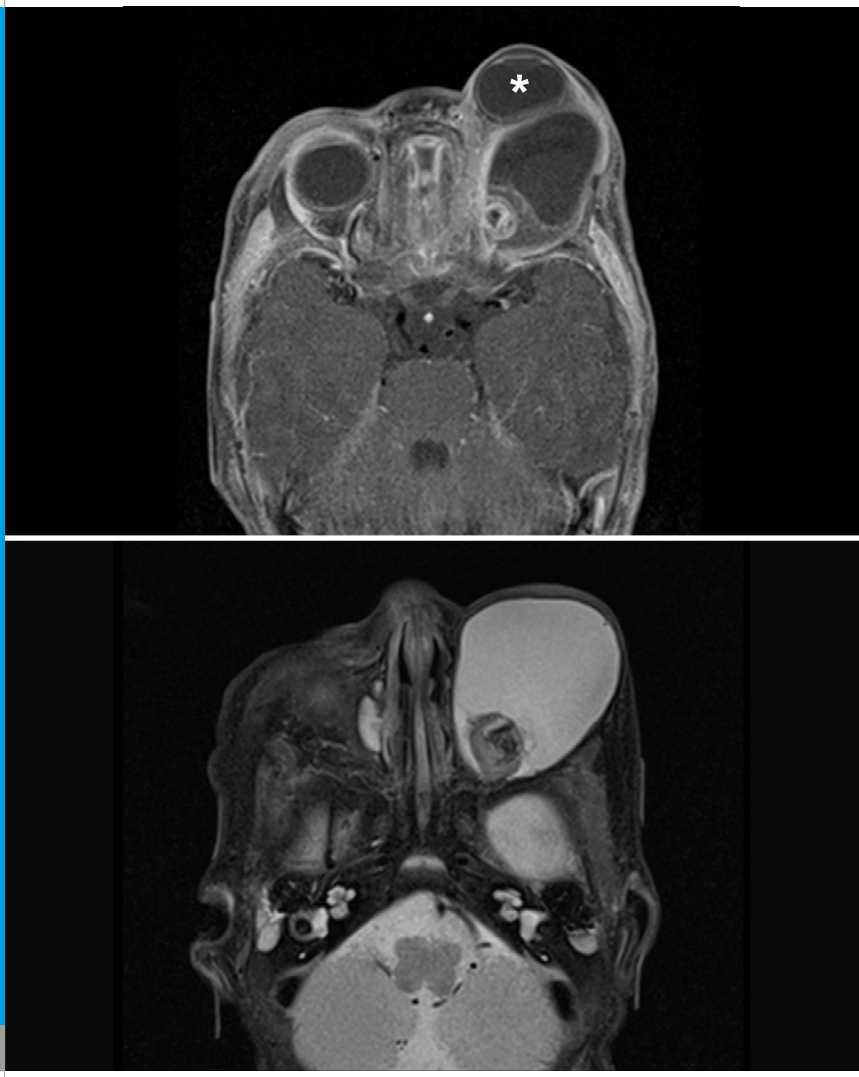

Fig. 3

Magnetic resonance images (coronal views): (top: T1-weighted, bottom: T2-weighted): large, left-sided intraorbital mass with two large cystic parts separated by a septum, and centrally located, inhomogeneous solid part; note impressive displacement of the eyeball (asterisk).

Drainage of the cyst to achieve bulbus decompression was performed on day one of life (DOL 1) with collection of an amber coloured fluid. A few hours after decompression, massive swelling of the cyst reoccurred and was complicated by bilateral retinal hemorrhages. The retinal hemorrhages were most likely caused by ischemic central retinal vein occlusion due to the swelling. Intubation and sedation were required for appropriate pain management.

To preserve visual function, a decision to proceed to immediate tumor resection was taken after interdisciplinary discussions among maxillofacial surgeons, neurosurgeons, ophthalmologists and pediatric oncologists. On DOL 4, eye-saving complete tumor resection was performed by craniotomy via a fronto-zygomatical approach. The orbital floor was remodelled using a fronto-temporal piece of the skullcap (Fig. 4).

Fig. 4

Postoperative result after tumor resection on DOL 4.

Histopathological findings included cysts lined with plexus and squamous epithelium. Moreover, skin appendages, muscle, glial, bone and cartilage tissues were identified (Fig. 5). Therefore, a diagnosis of a mature cystic teratoma, G0 according to Gonzales-Crussi was made (7).

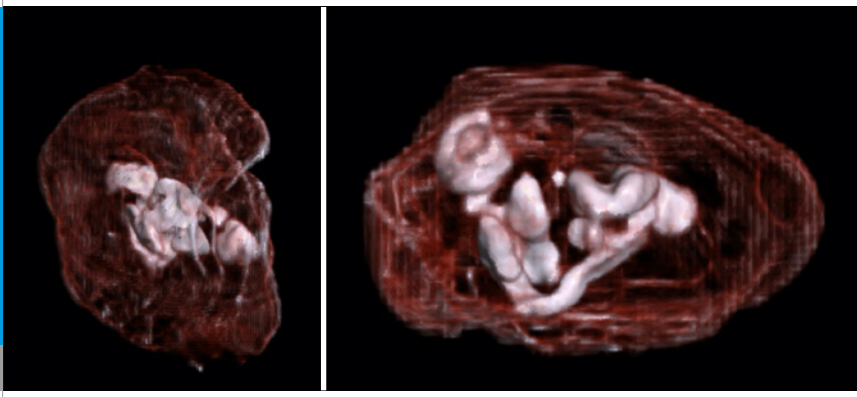

Fig. 5

CT scan reconstruction of the resected teratoma: bone and dental components as well as soft tissue can be identified.

Postoperative recovery was uneventful with regressive swelling under local cortisone therapy. Funduscopic examination was performed at regular intervals and showed regressive retinal hemorrhages. Intermittent occlusion therapy was performed to support visual development of the left eye. The baby was discharged home on DOL 12.

At the age of 10 months (Fig. 6), ophthalmic follow-up documented limitation of lateral and upwards eye movements. These findings are compatible with paresis of the left abducens and oculomotor nerves. Refraction values were borderline. A correction with glasses was not considered necessary at this point and development of binocular vision seemed likely.

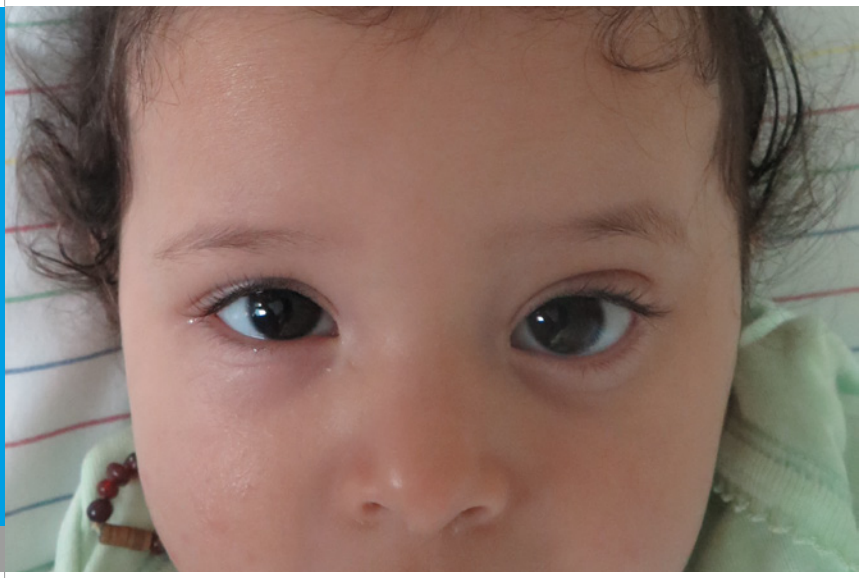

Fig. 6

Clinical appearance at the age of 10 months.

MRI follow-up examinations showed no tumor recurrence. Serum alpha-fetoprotein (AFP) was 114'000 ng/ml (normal value in newborns) in the first week of life and decreased appropriately to 11 ng/ml within the first months of life. Beta-human chorionic gonadowtropin (hCG) was always normal.

DISCUSSION

The differential diagnosis of orbital teratoma is broad and includes microphthalmos with cyst, dermoid cyst, epidermoid inclusion cysts, hemangioma, lymphangioma, cephalocele, neuroblastoma, rhabdomyosarcoma and retinoblastoma (5). The rare occurrence of such lesions coupled with the need for interdisciplinary management often delays adequate therapy.

Early surgical management as a definitive treatment is recommended to avoid further tumor growth which can result in hemorrhage or even rupture of the tumor (8). Early tumor resection has the greatest likelihood to preserve vision in the affected eye (9).

In the presented case, immediate complete tumor resection prevented oculoplastic intervention and lead to likely vision preservation, as well as normal orbitofacial development with satisfactory cosmetic outcome.

Kirvelä and Tarkkanen classified orbital teratomas in a) primary intraocular teratoma b) primary orbital teratoma c) combined orbital and extraorbital teratoma and d) secondary orbital teratoma (9). Our patient had the diagnosis of a primary orbital teratoma. Orbital teratomas are rare and often reported as single cases in the literature. Different grades of complexity in teratomas have been reported, even up to partially or fully developed fetuses (9).

Typically, teratomas contain cystic as well as solid parts. Orbital teratomas are unilateral, more frequently left-sided, and present with moderate to marked congenital proptosis, leading to exposure keratopathy. The globe is normally developed, but eye muscles, and nerves can be elongated, and surrounding bones displaced. Although orbital teratomas can be exceptionally large and extend over the orbita, they have a low tendency to spread intracranially or periorbitally.

Girls are more often affected than boys. Orbital teratomas are usually benign, despite their rapid growth rate. However, malignant forms have also been described (10). The typically rapid enlargement after birth is most likely a result of the accumulation of secretions rather than cell proliferation (9).

Elevated levels of beta-hCG and AFP may occur in malignant teratomas. In benign, mature teratomas, these values are usually within the normal range (11). Physiologically, high AFP values are expected in newborns but decrease rapidly over the first months of life, as seen in our patient.

Diagnostic workup includes ultrasound, MRI or CT scans. The diagnosis is later confirmed by histopathological examination. The goal of treatment is complete tumor resection in an eye-saving procedure.

Despite their benign nature, many cases have been described where the massive size of the tumor necessitated exenteration (9). The risk of recurrence of mature orbital teratoma after complete resection is extremely low. Chemotherapy is not indicated in mature teratomas as no cell division takes place despite potentially rapid volume expansion.

In our patient, MRI follow-up examinations are carried out not only to rule out recurrence, but also to follow orbito-facial development. Observation of the ophthalmological and cosmetic outcomes are crucial in the long-term management of such patients.

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