Interesting case

Indentification data: a 16-day-old male neonate

Chief complaint: right palpebral mass since he was born.

Present illness: a 16-day-old male infant was born by vaginal delivery with Apgar score 9 and 10, respectively. His birth weight was 2,700 g and his length was 51 cm. Her mother has normal antenatal history. He had a red mass on right upper eye lid since he was born. The doctor told his mother that it was bruise from birth injury and it would be spontaneously regress. Her mother noticed that right palpebral mass became larger by time and totally covered his right eye. He did not had other symptom but mass. His mother brought him back to hospital and he was referred to our institution.

Family history: no other member in his family has the same lesion as him

Physical examination:

GA: A male infant, active, no cyanosis, no dyspnea, BW 3.5 kg, Length 52 cm, OFC 35 cm
Vital signs: BT 36.8°C, HR 155 /min, RR 40/min, BP 65/40 mmHg, O₂sat 100% (room air)
HEENT: no pale conjunctivae, no icteric sclerae, a reddish oval tense cystic mass 3x4 cm in diameter on right upper eyelid without ulcer on top, no tenderness, smooth surface, not fixed to underlying tissue (figure 1), AF 1.5x1.5 cm, no bulging, no cranial bruit, PF finger tip
Heart: regular rhythm, normal S₁ and S₂, no murmur
Chest and lungs: clear and equal breath sound, no adventitious sound, no retraction
Abdomen: no distension, soft, liver is just palpable, spleen is not palpable, no mass
Extremities: no deformity, no rash, no edema
Skin: no red skin nodule, mass or rash on other area
Neurosings: active, pupil 3 mm RTL BE, equal movement and reflex 2+ all extremities, Babinski sign dorsiflexion both sides, clonus negative both sides

Provisional diagnosis: hemangioma right upper eyelid
CT scan of orbit and brain showed an oval shaped homogeneous enhancing hyperdense mass involving right eyelid and preseptal soft tissue (2.9x3.1x3.4 cm) with few calcification within lesion and this mass displaced the globe posteromedially (figure 2). His left eye and brain parenchyma appeared normal. These findings were compatible with hemangioma of right eye lid. He was given propranolol 2 mg/kg/day divided to 3 times a day. However, the mass became larger within three day. Plastic surgeons were consulted and they thought of malignant tumor of eyelid. The patient was undergone debulking mass of right upper eyelid. The pathology showed malignant round cell tumor with eosinophilic cytoplasm involving dermis and subcutaneous tissue. The margin was positive with presence of tumor necrosis and high mitotic rate (figure 3). The immunohistochemical staining was positive for vimentin, cytokeratin, CD99 and cyclin D1 and negative for CD45, desmin and S-100. These results were compatible with malignant rhabdoid tumor.

![Figure 1](image.png)

**Figure 1** A reddish oval tense cystic mass 3x4 cm in diameter on right upper eyelid
Figure 2 An oval shaped homogeneous enhancing hyperdense mass involving right eyelid and preseptal soft tissue (2.9x3.1x3.4 cm) with few calcification within lesion.
Figure 3 Malignant round cell tumor with eosinophilic cytoplasm. These findings is pathognomonic for malignant rhabdoid tumor.

Malignant rhabdoid tumor (MRT) was first recognized by Beckwith et al in 1978. This kind of tumor is rare and very aggressive. It primarily occurs in infants. The kidney and central nervous system are two of most common organ involvement. But it can occur in various organs including skin, soft tissues, liver, mediastinum, retroperitoneum and orbit.

MRT of orbit was originally reported by Rootman et al in 1989. There are few reports in children. This tumor involved in various parts of orbital area but the intraconal area is the most common site of involvement. The differential diagnosis of orbital tumor are rhabdomyosarcoma, neuroblastoma, Ewing sarcoma, granulocytic sarcoma and malignant fibrous histiocytoma.

MRT responds poorly to conventional therapy although multimodalities of treatment including radical surgery, chemotherapy and radiation therapy are used. The prognosis is still very poor with 80% mortality rate.
Bibliography:


