Nasopharyngeal Yolk Sac Tumors: A Rare Pediatric Occurrence

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Introduction

Yolk sac tumors as germinal cell tumors are unusual occurrences in the head neck. These tumors are usually derived from the testis or ovary. The most common extra-gonadal sites include the mediastinum, the pelvis and the abdominal cavity, as well as the liver. Few cases have been reported in the head and neck, and fewer still in the nose and/or nasopharynx of children. We present a 2 ½ year old with acute sinusitis presenting with acute bilateral blindness and complete nasal and nasopharyngeal obstruction by tumor, confirmed as a pure yolk sac tumor by pathology.

Case presentation

A 2 ½ year old female patient was referred to the emergency room by her pediatrician with an acute onset of bilateral blindness of 5 hours. She had a 6 week history of low grade fever, nasal congestion, and clear rhinorrhea. Approximately 2 weeks prior to this admission, the nasal discharge became purulent and she was treated with a course of Amoxicillin for presumptive bacterial rhinosinusitis. Two days prior to this admission to the emergency room, the patient was noted to be somewhat lethargic, with decreasing appetite, and perceived decreasing visual acuity due to repetitively walking into walls. She also had a history of strabismus and was seen by her opthalmologist who ordered an outpatient MRI scan.

Pediatric otolaryngology, opthalmology, and neurosurgical consultations were obtained on admission. Nasal exam showed a large friable erythematous mass occluding the nasal cavities bilaterally extending anteriorly from the nasopharynx. There was evidence of purulent, non-foul smelling mucus as well. There was an obvious protrusion of a well defined mass pushing the soft palate inferiorly without mucosal irregularity. The ears, pharynx and larynx were unremarkable. The eye exam showed evidence of optic chiasm syndrome with lack of response to light bilaterally without measurable exopthalmos. She had full adduction, but limited abduction. Fundoscopic exam confirmed optic nerve edema bilaterally. Her anterior chamber was quiet and her macula was flat.

CT and MR imaging of the brain, orbit and paranasal sinuses were performed: see Figs.1-4. The patient was taken to the operating room and underwent nasal endoscopy with biopsy of the nasal mass which was noted to be extremely vascular as suggested in Fig 2. Histopathologic analysis - see Fig 5 was found to be consistent with a nasopharyngeal pure yolk sac tumor. Orbital decompression was deferred as was any neurosurgical intervention. The patient was started on high dose steroids to relieve the optic neuritis and chemotherapy once the diagnosis was confirmed. She received six rounds of chemotherapy consisting of bleomycin, etoposide, and cisplatin over a 4 month period. Post-treatment imaging studies showed resolution of the large nasopharyngeal mass and its involvement of skull base and orbit. A small amount of persistent enhancement noted in the nasopharynx on post-treatment CT/MR was evaluated in the operating room . Clinically this was consistent with dried mucus and necrotic debris. The underlying mucosa was normal. The patient underwent endoscopic biopsies of the nasopharynx which showed no signs of residual tumor , consistent with her normal AFP level of 3.5(initial 12,174, nml<9).

Four months post-treatment, the patient has regained sight in the right eye with limited vision in the left. She is otherwise neurologically intact and has returned to pre-kindergarten.

Radiographic Images

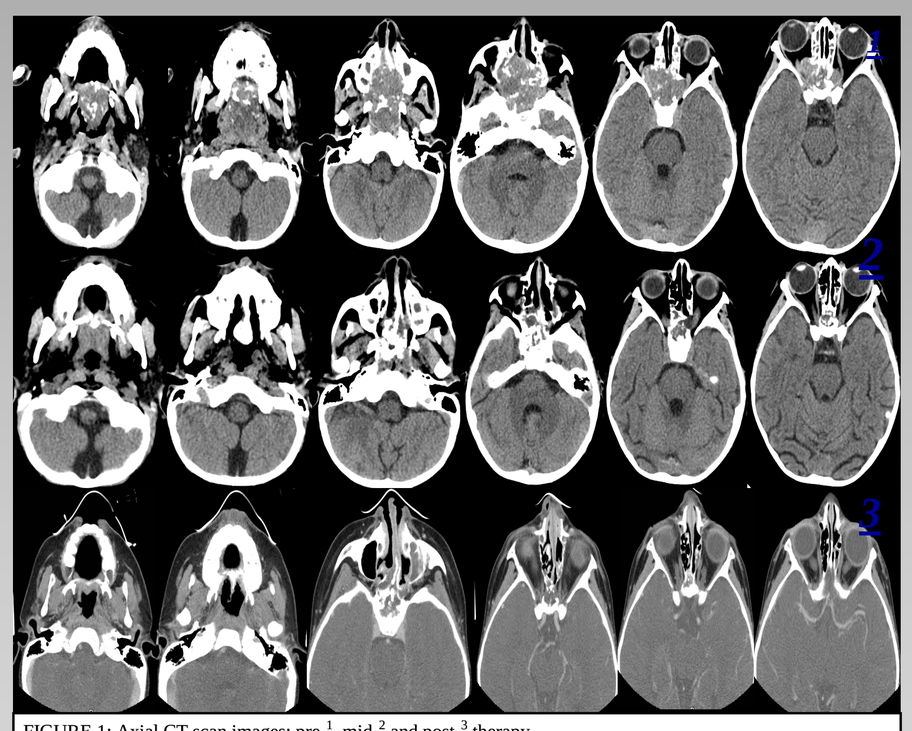

FIGURE 1: Axial CT scan images: pre-[1], mid-[2] and post-[3] therapy.
[1] 4x4cm heterogenous tumor pushes on the palate and is centered on the nasopharynx, involves the orbital apex distorting both orbital nerves. It extends into the pterygopalatine fossa, and thorough the cribiform plate into the anterior cranial fossa. The planum sphenoidale, lamina papyracea and portions of the maxilla are destroyed. AFP level 12,174.
[2] Mass decreased to 3x2.5cm. Decreased palatal, nasal, and intracranial extension with posterior remodeling of the medial orbits has occurred after 3 rounds of chemotherapy. AFP level 78.9
[3] Marked decrease in nasopharyngeal mass with persistent soft tissue density, obvious bony destruction at sphenoid and clivus. AFP level 3.5.

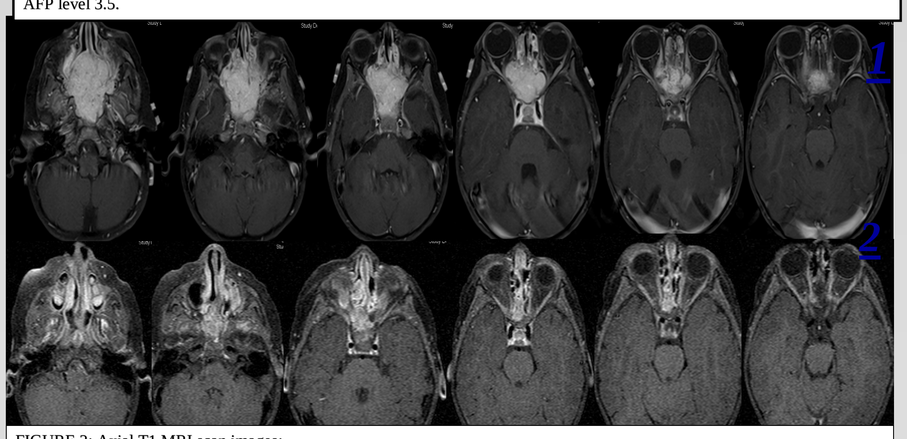

FIGURE 2: Axial T1 MRI scan images:
Pre- therapy[1] The tumor extends anteriorly to the mid-septum, posteriorly to the basi-sphenoid and appears to be highly vascular suggestive of an aggressive malignancy
Post-therapy[2] There is complete resolution of the nasal portion of the tumor, residual maxillary retention cysts and obstructive pattern seen in the ethmoid sinus after 6 rounds of chemotherapy.

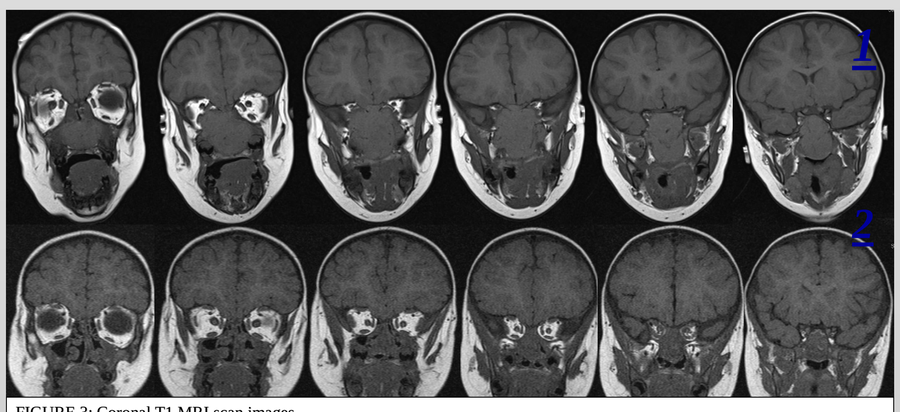

FIGURE 3: Coronal T1 MRI scan images
Pre-therapy[1] Tumor invasion of the orbit – right greater than left is more clearly seen along with destruction of the base of the skull. No dural contrast enhancement is seen.
Post-therapy[2] Complete remodeling of the orbit, - muscles and nerves at the apex are noted with clearance of the base of the skull, though persistent soft tissue density is seen centered on the nasopharynx.

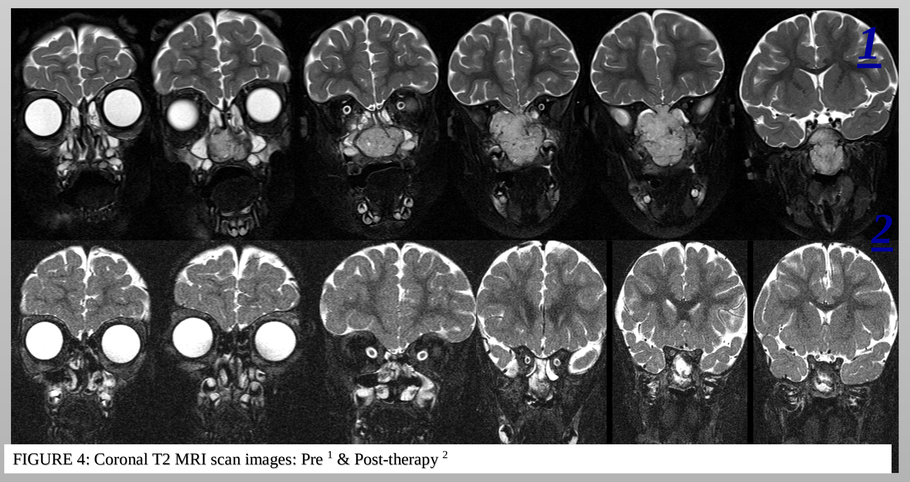

FIGURE 4: Coronal T2 MRI scan images: Pre [1] & Post-therapy [2]

Pathology

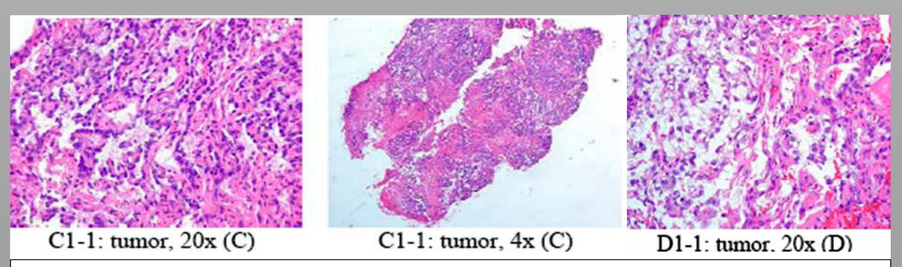

FIGURE 5: H&E stain shows a malignant neoplasm, consisting of highly pleomorphic hyperchromatic tumor cells with scant cytoplasm (C) and epithelioid tumor cells with abundant clear cytoplasm (D). Complex architectural patterns are also noted, with complex microcystic or glandular patterns in C and solid to nested patterns in D.

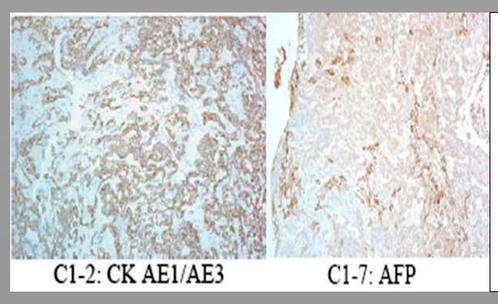

FIGURE 6: Stains show diffuse and strong reactivity for keratin AE1/AE3 and AFP (mostly cytoplasmic and some nuclear stain, in both original and additional sets with diluted antibody concentrations) and alpha-1-antitrypsin. No/little reactivity was observed in tumor cells for myoid (desmin and myoD1); vascular-related markers (CD31 and CD34); neuroendocrine markers (synaptophysin & chromogranin).

Discussion

Cancer remains the second most common cause of death in the pediatric population. In Allbrights' (2002) [2] review of the SEER data, the incidence of head and neck malignancies in the United States from 1973 through 1996 among children younger than 15 years increased by1-2%, and at a greater rate than childhood cancer in general. The most common malignancies continue to be lymphomas, rhabdomyosarcomas and thyroid malignancies. Also, neural tumors and sarcomas are more common in infants and young children. [2] The neck is the most common site.

Yolk sac tumors (YST), also known as endodermal sinus tumors are rare in the head and neck. They represent 3-5% of pediatric malignancies [1] and are malignant germ cell tumors, derived usually from the ovary or testis that produce alpha-fetoprotein (AFP). Extragonadal lesions occur in approximately 20% of females – with an average age of presentation of 18 years old.[3,5] The most common extragonadal site is the vagina. Other extragonadal sites include the mediastinum, pelvis and liver. In the head and neck, exclusive of the intracranial cavity, there have been reports in the nose, orbit, parotid gland, oral cavity and esophagus.

Diagnosis: Since the extragonadal presentations are sporadic, there is no consistent clinical pattern of presentation. Symptoms depend on site affected. The characteristic finding however includes rapid growth over a few weeks. Radiologic imaging with CT and MRI scans are usually required. A biopsy of the lesion along with immunohistochemical staining is usually needed for confirmation of the diagnosis. Histologically, extragonadal lesions behave as in the gonad with strong reactivity for keratin and alpha-fetoprotein, see Figure 6.

Treatment options: Successful treatment of this tumor requires preoperative chemotherapy to reduce the size, followed by completion surgical extirpation of residual disease and postoperative chemotherapy as needed. Cisplatinum remains the mainstay of the therapy, with bleomycin, and either etoposide or vinblastine used in combination therapy. At least four courses have been recommended. [4,6,7] This treatment has a high success rate, low relapse rate, little evidence of late relapse, and less toxic side effects. [4] Radiotherapy is included as part of the armamentarium primarily in non-head and neck sites.

Serum alpha-fetoprotein (AFP) levels are measured and followed to monitor the tumor response to chemotherapy, as it correlates well with tumor mass on neuroimaging. Since the extragonadal site presentations are sporadic there are no published survival data for this group. However, in stage III/IV gonadal tumors, the overall survival rate is 71% with a 2-year disease free survival of 61-77%. [6,7]

Conclusion

Nasopharyngeal germ cell tumors are aggressive lesions, that often present late, as with other extragonadal sites. Recurrent sinusitis and nasal obstruction in children should be evaluated carefully and with a high level of suspicion for nasal and nasopharyngeal malignancies. Early diagnosis and prompt treatment may alleviate many of the presenting symptoms and prolong life.

References

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