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 and neck. These tumors are usually derived from the testis or ovary. The most common extragonadal 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 2½ year old girl 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 2½ 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 periodic decreasing visual acuity due to repetitively walking in walls. She also had a history of strabismus and was seen by her ophthalmologist who ordered an outpatient MRI scan.

Pediatric otoaryngology, ophthalmology, and neurosurgeical 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 inferriorly without mucosal irregularity. The ears, pharynx and larynx were unremarkable. The eye exam showed evidence of optic chiasm syndrome with loss of light to response to light bilaterally without measurable extraophthalmals. She had full adduction, but limited abduction. Funduscopy 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 3 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 chemotheraphy once the diagnosis was confirmed. She received six rounds of chemotherapy consisting of bleomycin, etoposide and cisplatin.

Radiographic Images

Cancer remains the second most common cause of death in the pediatric population. In Albright’s (2002) 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 by 1-2%, and at a greater rate than childhood cancer in general. The most common malignancies continue to be lymphoblastomas and thyroid malignancies. Also, neural tumors and sarcomas are more common in infants and young children. 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 and are malignant germ cell tumors, derived usually from the ovary or testis that produce alpha-fetoprotein (AFP). Extragonadal lesions occur in approximatly 20% of females with a lower rate of presentation of 18 years old.5,6 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.

Pathology

Serum alpha-fetoprotein (AFP) levels are measured and followed to monitor the tumor response to chemotherapy, as it correlates well with tumor mass on neuroradiographic imaging. Since the extragonadal site presentations are sporadic there are no published survival data for this group. However, in stage III/IV gonad tumors, the overall survival rate is 71% with a 2-year disease free survival of 61-77%.6,9

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 nasopharyngeal and nasopharyngeal malignancies. Early diagnosis and prompt treatment may alleviate many of the presenting symptoms and prolong life.

References