J Oral Maxillofac Surg
70:1891-1895, 2012

Yolk-Sac Tumor of the Postauricular Region: Case Report and Review of the Literature

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The yolk-sac tumor (YST; or endodermal sinus tumor [EST]) is a malignant germ cell tumor that usually occurs in the gonads. This tumor is considered one of the most common tumors in infants and children. The extragonadal presentation of an YST is not common and the mechanism of germ cell misplacement is not well understood. In the head and neck, these tumors usually are found in the central nervous system. A non-midline YST is very rare and most cases have been reported in the neck, nasopharynx, and oral cavity.[1] The treatment of choice is chemotherapy followed by surgical resection or radiotherapy. To the authors' knowledge, there are very few reports of temporal bone and postauricular region YSTs that have been treated solely with surgical excision.

Report of a Case

A 2-year-old girl with known epilepsy caused by West syndrome presented in February 2010 with a mass in the left postauricular region of 2 months' duration. The mass was rapidly increasing in size. She was developmentally delayed and was receiving several antiepileptic drugs because of poorly controlled epilepsy. On examination the ear lobe was pushed forward and its posterior parts were necrotic (Fig 1). The postauricular skin was also necrotic with an ulcer from which necrotic material was draining. The patient had intermittent bleeding from the mass and the laboratory examination showed severe anemia, for which she received 2 U of blood. There was a red hemorrhagic vegetative mass in the left external auditory canal. The facial nerve and other lower cranial nerves were normal. A computed tomographic scan of the temporal bone showed a large solid mass in the postauricular soft tissue with extension to the mastoid and middle ear and erosion of the bony septets (Fig 2). The mass had also extended into the infratemporal space and occupied most of this space. There were several enlarged lymph nodes in the II, III, and V levels of the right side of the neck. Magnetic resonance imaging examination showed a mass with extension to the mastoid and middle ear (Fig 3). An incisional biopsy of the mass disclosed a malignant germ cell tumor. The serum α-fetoprotein (AFP) level was elevated. Surgical resection of the tumor was performed to remove the necrotic tissue. Lateral temporal bone resection, total parotidectomy with dissection of facial nerve, comprehensive resection of the soft tissue of the postauricular and infratemporal regions, and posterolateral neck dissection were performed. After complete gross ablation of the tumor, a plastic surgeon (M.H.) used a temporalis muscle flap, which was rotated over its pedicle, to cover the exposed bone, elements, and vessels. The result of the pathologic evaluation using immunohistochemical studies indicated a YST. The pathologic examination of the mass showed reticular and microscopic areas formed by a loose meshwork lined by flat and cuboidal cells with pleomorphic nuclei, prominent nucleoli, frequent mitotic figures and vacuolated cytoplasm, and a perivascular organoid arrangement of tumoral cells (Schiller-Duval bodies) and solid undifferentiated areas. Intracytoplasmic and extracellular hyaline globules that stained positively with periodic acid-Schiff and multiple hemorrhagic areas were also seen. Immunohistochemistry was strongly positive for AFP and periodic acid-Schiff, focally positive for chromogranin, and positive for keratin (Fig 4). The surgical margin was free of tumor and 0 of 17 lymph nodes was involved. The patient was advised to undergo chemotherapy, but the parents refused. After 9 months of follow-up, there has been no sign of recurrence, the patient is asymptomatic, and the serum AFP level is within the normal range.

Discussion

Germ cell tumors have diverse clinical, pathologic, and prognostic features, with a reported incidence of 3% of pediatric malignancies and with the EST being

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0278-2391/12/7008-0$36.00/0
doi:10.1016/j.joms.2011.08.009

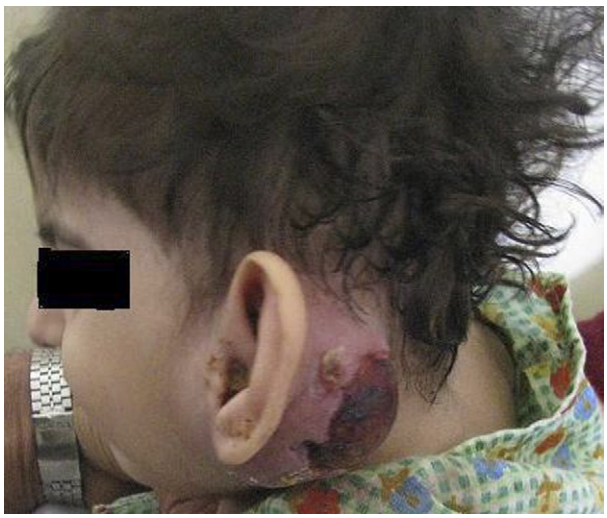

FIGURE 1. A postauricular mass with necrosis and bleeding and the ear lobe pushed forward.

Rozbahany et al. Yolk-Sac Tumor Of The Postauricular Region. J Oral Maxillofac Surg 2012.

the most common type.[2,3] Extragonadal germ cell tumors (EGCTs) of the head and neck account for 5% of all benign and malignant germ cell tumors.[1,4] Teratomas are the most common EGCTs in childhood, which are mostly benign but occasionally have malignant components, eg, embryonal carcinoma or ESTs.[1] Most EGCTs involve the midline[5] and are diagnosed usually before 3 years of age[2].

The EST, also known as an YST, was first reported by Teilum in 1959.[6] The primary origin of such tumors is the testis in the male and the ovary in the female.[7] There are some rare reports of an extragonadal occurrence of ESTs involving the omentum,[7] nasal cavity,[8] and head and neck region;[8] the sacrococcygeal,[9] retroperitoneum,[9] mediastinum,[9] and the paranasal sinuses;[10] the parotid gland;[11] the oral cavity;[12] the liver; the brain; and the vagina.[2] The pathology underlying EGCTs has been suggested to be an abnormal migration of primordial germ cells from the ectoderm of the yolk sac or misplaced totipotential cells of the blastula to the morula stage of embryogenesis.[1] Hoffner et al[13] reported data to support meiosis I nondisjunction, meiosis II nondisjunction, endoreduplication of a haploid ovum, mitotic proliferation of a premeiotic germ cell, and the fusion of 2 haploid ova as the genetic origins for ovarian germ cell tumors.

There are 4 histopathologic patterns for the YST: reticular, polyvesicular vitelline, solid, and pseudopapillary.[1] The reticular form is the most common and the pseudopapillary is the classic pattern, with Schiller-Duval bodies, which were found in the present case.[1] Schiller-Duval bodies are considered pathognomonic for the YST.[14]

These tumors secrete AFP. Immunofluorescent techniques have been used to localize the site of AFP synthesis in YST samples.[15] The serum levels of this protein may be used as a tumor marker to help in the diagnosis and follow-up of YSTs. The serum AFP was elevated in the present patient and decreased after removal of the tumor.

The preoperative level of AFP is high in these tumors; it has been reported to have no prognostic significance.[1]

The prognosis of patients, especially children with ESTs, is unknown, although there have been reports of successful management by surgery, radiotherapy, and adjuvant chemotherapy (cisplatin, bleomycin, and vinblastine).[10,16-18]

Weedon and Musgrave[19] described a female infant who had an YST in the maxillofacial area that was diagnosed at the age of 10 months, and she died 1 month later. Margo et al[14] reported 5 cases of YSTs of the orbit occurring in children 3 months to 4 years old.

Lack[4] reported 3 cases of ESTs; the tumors presented within the oropharynx, nasopharynx, and floor of the mouth. Lack reported a tendency of the cervical teratoma to involve the right side of the neck, but the present patient had the tumor on the left side.

In a series of ESTs reported by Shebib et al,[20] 2 of 11 cases had tumors in the head and neck region. Soft tissues of the facial and temporomastoid regions were the sites of 4 ESTs studied by Dehner et al.[21] The

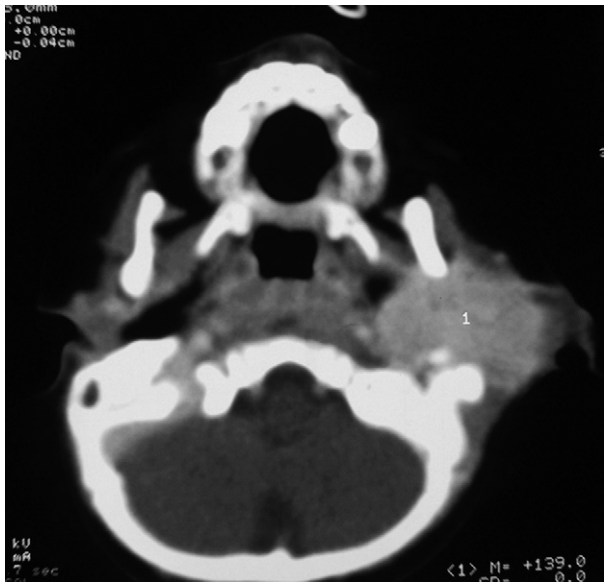

FIGURE 2. Axial contrast-enhanced computed tomogram shows a well-defined, lobulated, avidly enhancing left parotid mass extending to the pre- and poststyloid fossa. Extension to the infratemporal and parapharyngeal fossa is visualized. Invasion to the temporal bone and destruction of the petrous bone are visible.

Rozbahany et al. Yolk-Sac Tumor Of The Postauricular Region. J Oral Maxillofac Surg 2012.

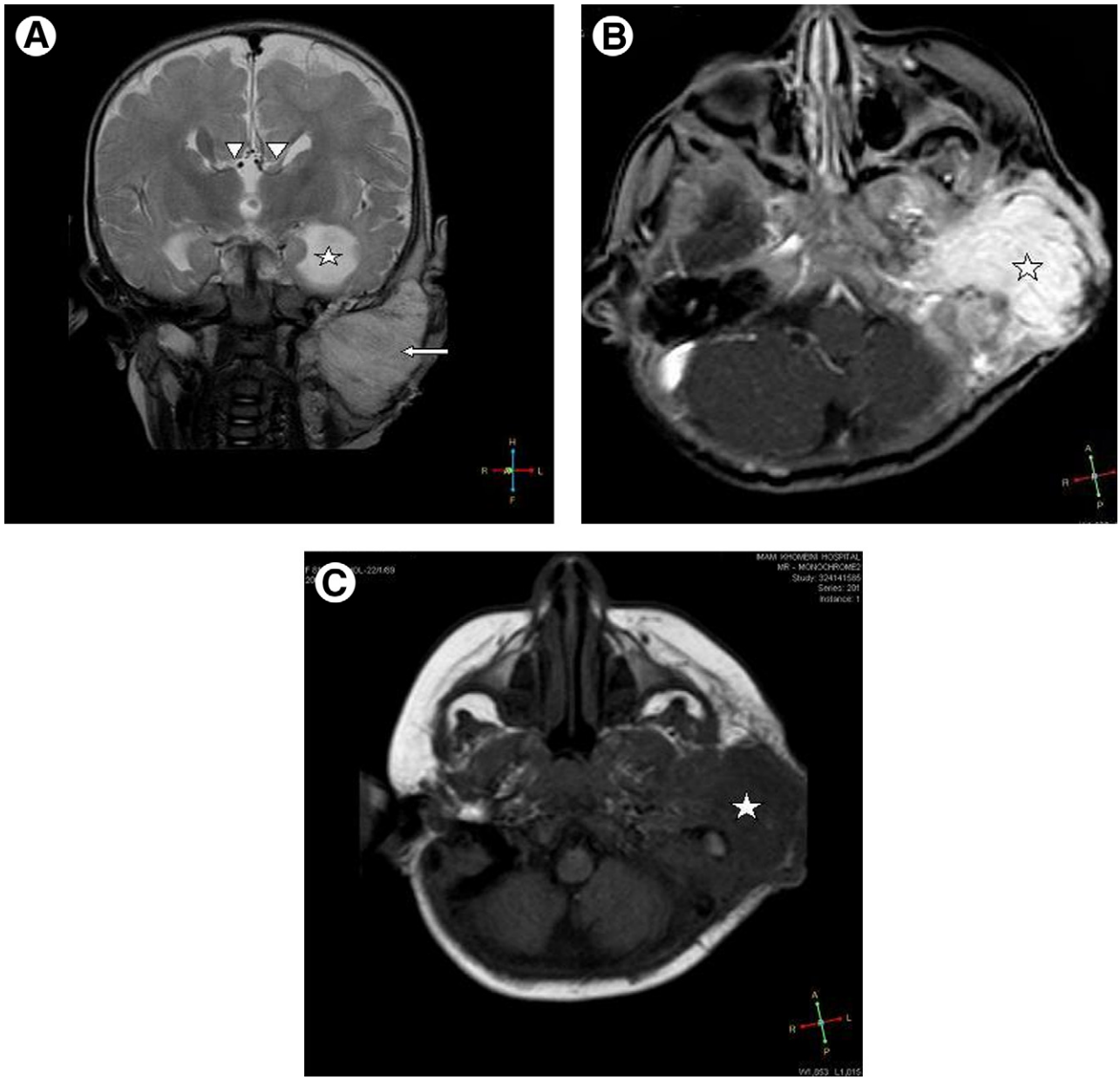

FIGURE 3. Magnetic resonance images show *A*, a strong signal mass on the T2 image (*arrow*), *B*, avid enhancement with gadolinium (*star*) in the left parotid space with extension to the petrous bone and infratemporal fossa, and *C*, a large, well-marginated, lobulated, hypointense mass on the T1 image (*star*). Extension to the parapharyngeal space and the pre- and poststyloid spaces is seen. *A*, Corpus callosum agenesia (*arrowheads*) and hydrocephaly (*star*) are visible.

Rozbahany et al. Yolk-Sac Tumor Of The Postauricular Region. J Oral Maxillofac Surg 2012.

maxillofacial region and temporal region were the primary sites of disease reported by Nair et al[22] and Kebudi et al.[18]

In 1987, Stanley et al[17] presented the first reported case of an EGCT arising from the temporal bone. Their patient was a developmentally delayed patient, as was the present patient, and his tumor was cured by chemotherapy. To the best of the authors' knowledge, there is no known syndrome with mental retardation and a tendency for EGCTs. However, the observation of the occurrence of these 2 conditions together deserves extended studies to discover any possible relations.

The use of AFP in monitoring YSTs has been established in the diagnosis and management of patients.[23] Previous reports of 89 cases of nongerminomatous malignant germ cells showed that testicular and ovarian tumors in children have a better prognosis and that the site of origin is the single most important prognostic factor.[24] In another report, the 4-year disease-free survival rate was 49% in 93 patients treated with chemotherapy (cyclophosphamide, bleomycin,

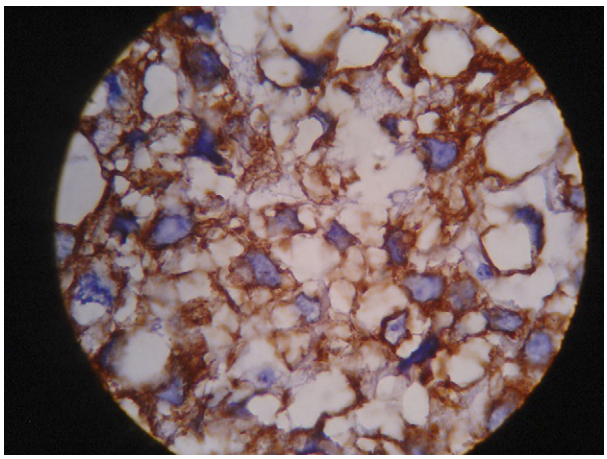

FIGURE 4. Histologic examination of the mass disclosed reticular and microscopic areas formed by a loose meshwork lined by flat and cuboidal cells with pleomorphic nuclei, prominent nucleoli, frequent mitotic figures and vacuolated cytoplasm, a perivascular organoid arrangement of tumoral cells (Schiller-Duval bodies), and solid undifferentiated areas. Intracytoplasmic and extracellular hyaline globules positively stained with periodic acid-Schiff and multiple hemorrhagic areas are displayed.

Rozbahany et al. Yolk-Sac Tumor Of The Postauricular Region. J Oral Maxillofac Surg 2012.

cisplatin, vinblastine, and dactinomycin).[2] The low survival rate is believed to be due to the low doses of cyclophosphamide and cisplatin that were used.[1]

In 2 other studies by St Jude Children's Hospital and the UK Children's Cancer Study Group,[25] the survival rates were 73% and 85%, respectively. There are very few reports of using only surgical excision for the treatment of YSTs and the survival rate is not clear. According to the overall poor response of head and neck EGCTs to primary chemotherapy, it seems that a surgical intervention for potentially resectable tumors is reasonable. In 1997, Kusumakumari et al[1] reported 3 cases of head and neck EGCTs. The primary sites were the orbit, the maxillofacial region, and the retroauricular region. Two patients underwent surgical removal and the only patient who received chemotherapy died of infection after treatment.

In a series reported by Garnick et al,[26] 15 patients with EGCTs were treated with vinblastine, bleomycin, and cisplatin followed by tumor-reductive surgery. Cyclophosphamide and doxorubicin were administered after surgery. Ten patients (67%) achieved complete remission and only 4 patients remained disease free for a median period of 40 months. Thus, these kinds of therapeutic strategies may be inadequate in the treatment of patients with EGCTs. In 1992, Viva et al[11] reported a case of an YST of the parotid gland in a 2-year-old girl. The tumor recurred after chemotherapy. Considering these reports and the special conditions of the present patient, a comprehensive operation was performed.

There are very few reported series of head and neck EGCTs in the literature, so the best choice of treatment has yet to be defined. This in turn necessitates further studies to reach this goal. It is difficult to comment about the present patient's status.

YSTs of the extracranial regions of head and neck are very rare, but there are reports on its occurrence. Patients with tumors that can be resected should undergo surgery followed by adjuvant chemotherapy, which is indicated in all patients. The chemotherapeutic adjuvant should include a combination of cisplatin, etoposide, and bleomycin, which has been reported to be successful in tumor removal. Patients with residual active disease require salvage chemotherapy. The addition of radiotherapy may be beneficial in surgically inaccessible regions.

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