Case Report/Case Series

Immature Teratoma of the Maxillary Sinus
A Rare Pediatric Tumor

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Report of a Case

A 10-year-old boy presented with a 1-month history of right upper jaw pain associated with a mild facial weakness on that side. His parents noted a globular swelling over his right cheek that rapidly increased in size (Figure 1). Computed tomographic imaging revealed a soft-tissue heterogeneous mass in the right maxillary sinus measuring approximately 4.4 × 4.6 cm and invading the right ostiomeatal complex, pterygopalatine fossa, and masticator space. It had also eroded the right orbital floor, causing a mass effect on the inferior rectus muscle (Figure 2).

Figure 1. Clinical Photograph Demonstrating Preoperative Appearance

The patient has globular swelling of the right cheek with associated ptosis and superior displacement of the globe.

Figure 2. Preoperative Computed Tomographic Scan

The scan shows a heterogeneous mass in the right maxillary sinus causing bony destruction of the medial wall of the maxilla and right orbital floor. The tumor contains calcifications, and invades the orbit, compressing the inferior rectus muscle and displacing the globe superiorly.
Multiple subcentimeter lymph nodes were seen in the jugulodigastric regions on both sides and the posterior triangles. An incisional biopsy via a sublabial approach revealed a tumor with elements from multiple germ layers, including epithelium (squamous, columnar, and occasional adnexal-like structures) and mesenchyme (adipose, fibroblastic, and cartilaginous features). Neuroectodermal features were present in other areas. All the components displayed multifocal immature or embryonal elements with characteristics consistent with a diagnosis of immature teratoma (Figure 3).

After input from a multidisciplinary tumor board, the patient underwent a partial maxillectomy via a midface degloving approach with the use of calvarial bone grafts for reconstruction of the infraorbital rim and maxillary buttresses and titanium mesh for reconstruction of the right orbital floor. The lesion was resected to the visible margins of the tumor, allowing for the proximity of vital structures. Histopathologic examination of the surgical specimen revealed margins positive for tumor. His postoperative course was uneventful, and he was scheduled for regular follow-up with evaluation and input from the oncology service.

Three months later, the patient developed acute right cheek swelling, and an intranasal biopsy revealed recurrent disease. Despite subsequent chemotherapy with bleomycin, cisplatin, and etoposide, the tumor continued to grow, and a repeated biopsy 3 months later proved positive for persistent teratoma. He underwent revision oncologic resection. Intraoperative and final pathologic margin findings were negative. The prior reconstruction was removed with the specimen, and further reconstruction was deferred to a secondary procedure. He underwent postoperative proton beam radiation therapy to the surgical site in 33 fractions (total dose, 59.4 Gy).

Two years after completion of adjuvant treatment, he developed progressive ipsilateral hypoglossus and midfacial soft-tissue atrophy that was reconstructed with a right orbital floor titanium implant and composite dermal-fat graft. This resulted in improved functional and cosmetic outcome, although a more definitive free-tissue reconstruction is being planned.

Four years after his initial surgical resection, the patient had no evidence of recurrence by serial endoscopic examinations and magnetic resonance imaging studies. Oncology specialists also confirmed by serial measurements that his α-fetoprotein level had not risen since the completion of therapy.

Discussion

Immature teratomas of the head and neck are extremely rare. Congenital teratomas occur in 1 in 4000 births, although those in the head and neck make up less than 2% of cases.1,2 Within the head and neck, most teratomas occur in the neck and nasopharynx, although several other sites have been reported, including the oral cavity, sinonasal cavities, external and middle ear, and mandible.3-5 These benign tumors generally present during infancy, and when they involve the head and neck they can cause neonatal respiratory distress. Surgical excision is the mainstay of treatment. When teratomas occur in adults, they are more likely to be malignant, carrying 3- and 5-year survival rates of roughly 30% and 20%, respectively.6 Some authors describe a bimodal distribution for the presentation of teratomas, with those presenting in infancy more likely to be benign and those in later childhood or adulthood more likely to be malignant.1 The tumor marker α-fetoprotein should be checked every 2 to 3 months after definitive teratoma treatment to monitor for recurrence.

From 20% to 40% of benign teratomas in children contain immature elements. Immature teratomas are a poorly understood subset of teratomas that are not well characterized in children, although they carry a significant risk of decreased event-free survival and overall survival compared with mature teratomas.7 A grading system of 1 to 3 (1 indicating the most mature elements, 3, the least mature elements) has been

Figure 3. Hematoxylin-Eosin-Stained Histopathologic Images

A, Elements from multiple germ layers are seen, including immature fatty and/or fibrous mesodermal and/or mesenchymal tissue. B, Immature ectodermal squamous epithelium and endodermal glandular epithelium (inset) tissue. C, Immature neuroectodermal tissue. Original magnification ×10 for all images.
described for ovarian teratomas. However, the utility of this grading system for nongonadal teratomas is unknown. In some studies of adult ovarian teratomas, grade 3 immature teratomas have been considered malignant, but most authors consider them to be a benign lesion that can be managed by surgical excision alone. If the immature elements are represented primarily by the tumor's neuroectodermal components, the tumors tend to behave benignly but can metastasize. A review of 125 pediatric germ cell tumors noted a correlation between grade 3 immature teratomas and the presence of microscopic foci of yolk sac tumors, emphasizing the importance of careful histologic analysis of these specimens. Malignancy in a teratoma, or somatic malignancy, consists of the concomitant presence of teratocarcinoma cells and malignant nonteratomatous germ cell elements (e.g., yolk sac tumor). The present case involved immature elements from all germ layers, but overtly malignant features (teratocarcinoma, yolk sac tumor, etc.) were lacking.

The present case did not involve a congenital lesion, and to our knowledge, it is the first reported teratoma involving the maxillary sinus in a pediatric patient. Approximately 50 cases of adult maxillary teratocarcinomas have been reported, including 1 case where the patient developed regional and distant metastases and was treated with multiple resections and postoperative irradiation. A report of a series of teratoid carcinomas of the upper respiratory tract describes 8 adults with tumors originating in different areas of the head and neck: the ethmoid sinus (n = 4), unspecified sinus (n = 1), and nasopharynx the maxillary sinus in a pediatric patient. Approximately 50 cases of adult sinonasal teratocarcinosarcoma have been reported, including 1 case where the patient developed regional and distant metastases and was treated with multiple resections and postoperative irradiation. Overall prognosis depends on many factors, including patient age, tumor resectability, and the presence of local or distant metastases. Several chromosomal abnormalities have been observed in the pathogenesis of these extragonadal germ cell tumors, although patients with benign teratomas are often cytogenetically normal. The importance of molecular changes has yet to be clearly elucidated in cases of immature teratomas.

Despite the paucity of reported cases, most authors agree that this disease entity warrants extirpative surgical management, with consideration given to postoperative chemotherapy and radiation therapy in the adult population. In a prospective study of 73 children with extracranial immature teratomas, 22 had tumors in extragonadal sites. Five patients had recurrence of disease 4 to 7 months after surgical excision but were successfully treated with platinum-based therapy. The authors concluded that complete surgical excision is effective treatment in pediatric cases of immature teratomas, even those with histologically malignant elements, although all patients merit close follow-up. The proximity to the orbit, cranial base, and other vital structures in our patient prompted a more cautious initial approach, with excision of all visible disease, but resulted in positive margins. Although this can explain the subsequent recurrence and course, we believe that the rarity of the tumor and the lack of known successful therapies in this location justified the decision.

Conclusions

In the case reported herein, a multidisciplinary team agreed that multimodality therapy was warranted, owing to the aggressive tumor biology and recurrent nature of the disease. Given the paucity of literature on immature teratomas of the head and neck in pediatric patients, our goal was to highlight the challenges faced in treating this rare tumor and to suggest therapeutic and reconstructive options.