One-year survivor of adult alveolar rhabdomyosarcoma of the maxillary sinus with orbital extension

Case report

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Abstract

Introduction: Rhabdomyosarcoma is uncommon in adults. Adult and maxillary rhabdomyosarcoma with direct orbital extension has been rarely reported. To our knowledge, there is no reported case about adult patient with alveolar maxillary rhabdomyosarcoma and orbital extension survived 1 year with intact ocular function.

Case presentation: A 21-year-old female presented with protrusion of the right eye and an obstructed nasal passage for the past month. Her symptoms were not relieved by oral antibiotic or irrigation. She was referred to our clinic. Computed tomography and magnetic resonance imaging showed a large homogenous well-enhanced mass with surrounding bony erosion and remodeling. The mass extended to the ipsilateral nasal cavity and orbit. Endoscopic biopsy of the nasal cavity confirmed alveolar rhabdomyosarcoma. The maxillary mass was excised using the Caldwell–Luc approach, and the orbital mass was excised using a transconjunctival and transcaruncular approach. A systemic work-up confirmed ipsilateral lymph node metastasis. The patient received 6 cycles of chemotherapy with vincristine, dactinomycin, and cyclophosphamide, as well as 5120 cGy radiotherapy. Her ocular function was intact 1 year after treatment, and magnetic resonance imaging showed complete regression of the tumor.

Conclusion: Rhabdomyosarcoma, which is usually an aggressive malignancy, should be considered in the differential diagnosis of a rapidly growing orbital mass. Aggressive treatment, including surgery, chemotherapy, and radiation therapy, can increase local remission rates and improve the prognosis.

Abbreviations: IRSG = The Intergroup Rhabdomyosarcoma Study Group, MRI = magnetic resonance imaging.

Keywords: adult rhabdomyosarcoma, alveolar variants, one-year survival, orbit tumor

1. Introduction

Rhabdomyosarcoma is the most common primary orbital malignancy in children <15 years of age, accounting for approximately 5% of childhood malignancies.[1] Rhabdomyosarcoma is uncommon in adults. The predilection sites of adult rhabdomyosarcoma are the extremities but the predilection sites in children with rhabdomyosarcoma are the head and neck area. Head and neck sites only account for 24% of adult rhabdomyosarcoma cases.[2] Moreover, cases involving the orbit are extremely rare.

The prognosis of adult rhabdomyosarcoma is poor. More than 60% of adult patients have regional or distant metastasis at the initial diagnosis.[3] It is very difficult or impossible to surgically excise a mass within the maxillary area. When a tumor occurs in this region, it is usually more extensive locally or has metastasized distantly at the time of diagnosis. The 5-year survival rate is ≤8% in cases of head and neck rhabdomyosarcoma.[4] Herein, we report the case of a 1-year survivor of adult alveolar rhabdomyosarcoma of the maxillary sinus with orbital extension.

Written informed consent was obtained from the patient for this case report. Ethical approval was obtained by the Institutional Review Board of Kyung Hee University Hospital at Gangdong (KHU-2010-07-39).

2. Case report

A 21-year-old female without specific medical history presented with a protruding right eye and an obstructed nasal passage of 1-month duration. The patient was diagnosed with sinusitis at another clinic and was prescribed oral antibiotics. Her symptoms were not relieved by the antibiotic treatment or nasal irrigation. She was referred to our clinic, and we performed a diagnostic work-up. Corrected visual acuity was 1.0 in both eyes. Hertel exophthalmometry revealed 3-mm proptosis (Fig. 1A). The extraocular muscle was intact. No specific findings were observed...
in the anterior or posterior segments. Computed tomography and magnetic resonance imaging (MRI) showed a large homogenous well-enhanced mass with surrounding bony erosion and remodeling (Fig. 2). The mass had extended to the nasal cavity and right orbit. Regional neck lymph node involvement was observed. A fiber-optic endoscopic biopsy of the nasal cavity confirmed alveolar rhabdomyosarcoma. The immunohistochemical analysis was positive for desmin, myeloperoxidase, and CD56, consistent with the diagnosis. As neck lymph node metastasis was suspected; surgical debulking, chemotherapy, and radiation therapy were scheduled. The maxillary and nasal cavity mass was excised using the Caldwell–Luc approach, and the orbital mass was excised through a transconjuctival incision in the inferior fornix followed by a caruncular incision. Complete tumor removal was difficult because the tumor contained the orbital wall and was located near the optic canal. The excised mass was pathologically confirmed as alveolar rhabdomyosarcoma. The right eye proptosis was relieved after surgery (Fig. 1B).

Ultrasonography-guided fine needle aspiration of a neck lymph node confirmed malignancy of the tumor. The Intergroup Rhabdomyosarcoma Study Group (IRSG) postsurgical staging was group 3. The patient underwent 6 cycles of VAC (vincristine, dactinomycin, and cyclophosphamide) and radiation therapy (5120 cGy). Her visual acuity and ocular motility were intact 1 year after treatment. MRI revealed complete regression of the previous tumor, mucosal wall thickening, and sinusitis (Fig. 3). A positron emission tomography scan showed no distant metastases. There was no local recurrence of tumors for a total follow-up period of 1.5 years; afterwards, loss of follow-up was occurred.

3. Discussion

Adult rhabdomyosarcoma rarely presents in adults. Orbital involvement is one of the most favorable factors in children, and the 5-year survival rate is >90%. Because of its rarity, no study has reported outcome statistics or an established therapeutic method. The bad prognosis for adults with orbital involvement is the difficulty excising the mass, the histopathological subtype, metastasis, and poor tolerance to intensive chemotherapy. Tumors in the paranasal sinus, nasopharynx, infratemporal palatine fossa, or orbit more often spread into surrounding tissues. These regions have a 35% incidence of meningeal recurrence. The alveolar subtype is more frequent in adults and the embryonic subtype is rare. The alveolar subtype has a higher metastatic rate than that of the embryonic or pleomorphic subtypes. More than 60% of adult patients have regional or distant metastases at the initial diagnosis, and most rhabdomyosarcoma cases are classified as IRSG group 3. Therefore, the prognosis is poor because of impossible complete excision.

The treatment protocol for adults with rhabdomyosarcoma has not been established. Surgery is performed in most cases, and chemotherapy and radiotherapy are used as adjuncts following the pediatric treatment protocol. Surgery is used to debulk, reduce the mass effect on the optic nerve, and improve ocular motility. A combination of chemotherapy and radiation therapy increases the local remission rate and decreases the metastatic rate. The conventional chemotherapeutic agents used are vincristine, dactinomycin, and cyclophosphamide. Darren
et al[10] suggested that 50 to 56 Gy was adequate for patients with completely resected disease and a negative resection margin. We also performed debulking surgery, conventional chemotherapy, and radiation therapy. There have been 7 recent cases similar to our case, but the value of the report is considered sufficient.

In conclusion, adult rhabdomyosarcoma is encountered very rarely in the orbit, where it is usually an aggressive malignancy with a poor prognosis. Rhabdomyosarcoma should be considered in the differential diagnosis of a rapidly growing orbital mass. Aggressive treatment, including surgery, chemotherapy, and radiation therapy can increase local remission rates and improve the prognosis.

**Author contributions**

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**References**

[1] Shieds JA, Shields CL. Rhabdomyosarcoma: review for the ophthalmologist. Surv Ophthalmol 2003;48:39–57.
[2] Hawkins WG, Hoos A, Antonescu CR, et al. Clinicopathologic analysis of patients with adult rhabdomyosarcoma. Cancer 2001;91:794–803.
[3] Wu TH, Huang JS, Wang HM, et al. Long-term survivors of adult rhabdomyosarcoma of maxillary sinus following multimodal therapy: case reports and literature reviews. Chang Cung Med J 2010;33:466–71.
[4] Nayat RC, Purdhomme F, Parise O, et al. Rhabdomyosarcoma of the head and neck in adults: a study of 26 patients. Laryngoscope 1993;103:1362–6.
[5] Christ W, Gehan EA, Ragab AH, et al. The third intergroup rhabdomyosarcoma study. J Clin Oncol 1995;13:610–30.
[6] Christ WM, Anderson JR, Meza JL, et al. Intergroup rhabdomyosarcoma study-IV: results for patients with nonmetastatic disease. J Clin Oncol 2001;19:3091–102.
[7] Su GW, Hong SH. Leiomyosarcoma of the uterus with sphenoid bone and orbital metastases. Ophthal Plastic Reconstr Surg 2007;23:428–30.
[8] Little DJ, Ballo MT, Zagaris GK, et al. Adult rhabdomyosarcoma: outcome following multimodality treatment. Cancer 2002;95:377–88.
[9] Raney RB, Maurer HM, Anderson JR, et al. The intergroup rhabdomyosarcoma group (IRSG): major lessons from the IRS-I through IRS-IV studies as background for the current IRS-V treatment protocols. Sarcoma 2001;5:9–15.
[10] Espanola NF, Rubin BP, Baldini EH, et al. Response to chemotherapy and predictors of survival in adult rhabdomyosarcoma. Ann Surg 2001;234:215–23.
[11] Moon HS, Kwon SW, Lee JH. A case of alveolar rhabdomyosarcoma of the ethmoid sinus invading the orbit in an adult. Kor J Ophthalmol 2006;20:70–5.
[12] Torres-Peña , Ramos Castrillo AI, Mencía-Gutiérrez E, et al. Nasal cavity or alveolar paranasal sinus rhabdomyosarcoma with orbital extension in adults: 2 cases. Plast Reconstr Surg Glob Open 2015;3:e414.
[13] Parikh D, Spindle J, Linden C, et al. Adult rhabdomyosarcoma of the maxillary sinus with orbital extension. Orbit 2014;33:302–4.
[14] Kelly A, Moran M, Primrose W. Alveolar epithelial rhabdomyosarcoma in a young adult male. BMJ Case Rep 2013.
[15] Bagdoniate L, Jeeva L, Chang BY, et al. Multidisciplinary management of adult orbital rhabdomyosarcoma. Orbit 2013;32:208–10.
[16] Sanz-Marco E, España E, Alamar A, et al. Orbital alveolar rhabdomyosarcoma masked by ethmoid sinusitis in a 25-year-old. Arch Soc Esp Oftalmol 2014;89:182–5.
[17] Eshraghi B, Ameli K, Anvar P. Adult rhabdomyosarcoma of ethmoid sinus recurring as an orbital mass. J Clin Diag Res 2016;10:ND06–7.